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Pediatric Neurosurgery

Theoretical Principles – Art of Surgical Techniques

Updating of the Neuroimaging by G. Trasimeni
The Chapter on Epilepsy Written with F. Cardinale

Second Revised and Enlarged Edition
with 567 Figures in 1514 Separate Illustrations, 355 in Color
and 47 Tables

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To my wife, Lucia
and my son, Marco Antonio

Preface to the Second Edition

"Old men that knowen the grounde
well yenoughe
Call it the battell of Otterburn:
At the Otterburn began this spurne
Upon a monnyn day.
Ther was the daugghte Doglas slean:
The Perse never went away."

ANONYMOUS*

"Thys fraye bygan at the Otterborn
Bytwene the nyghte and the day;
Ther the Douglas lost his lyfe,
and the Percy was lede away."

ANONYMOUS*

There are three compelling reasons for an Author to undertake a second edition of a text book: correction of unacceptable errors or inexactitudes, up-dating (putting into perspective) new concepts and techniques, the very personal wish to express more completely and graphically his messages. My goal for this Edition is – within the limits imposed by true differences of opinion – to present a corrected text reshaped by time, and enrichened by an exhaustive personal reading of the literature pertaining to pediatric neurosurgery during the 10 year period 1987-1997. At a time-distance of 13 years from the date the First Edition was given to the publisher, the Second Edition left my desk for Heidelberg, holding reviews of papers heard, journals and articles read, thoughts clarified.

The methodology consisted of stacking clinical notes and publications according to themes, highlighting messages to discuss or convey, writing these then as critical reviews, and lastly re-reading the text *of Pediatric Neurosurgery: Theory and Art of Surgical Techniques* with Dictaphone in hand to re-elaborate the new text. Hence, some sections of this work are verbatim reprintings from the First Edition, others are new, and others still are re-evaluations of the subject or the context of their expression. The narrative style chosen remains that of writing in the first person (so as to avoid the implication that the information given is the eternal truth), with concepts and conclusions developed consequentially. Thus, surgical attitudes and/or principles along with clinical theory are structured upon anatomy, pathology, clinical problems, operative methodology, interpretation of results... and developed as considerations, perspectives, approaches.

To simplify, enrichen, communicate visually the narrative message, extensive use is made of MRI, line and half-tone and color drawings, tables. Every effort was spent to bring these into spatial proximity to the texts, though at times we did not succeed. Color was chosen when it was necessary to distinguish clearly between adjacent anatomical structures, half-tones for perspective, line drawings for focus or concept. The tables used are either new elaborations, transcriptions from other writings, or reworkings of the two, and the design for their use is didactic and documentary.

As for the First Edition, this text has all the advantages and disadvantages of a single-author treatise, though I have painstakingly – throughout the literature review and multiple re-readings over time – attempted to incorporate other perspectives into the work. Nevertheless, one sees with his own eyes, thinks with his own mind. There is an absolute and pervading difference between an Editor and an Author. Thus, the

*RELICS OF ANCIENT ENGLISH POETRY. Two ballads sung by two *different* anonymous minstrels, relating a "historic" single event which occured within the living memory of the listeners... with two very different issues.

aseptic and telegraphic, at times disjointed, conglomerates of information and attitudes of a multi-authored book are counterbalanced against the defined visual arc of a single author. As an information source for those who wish to practice the theory and art of pediatric neurosurgery, I think a single author treatise is preferable.

Let us see now what this book holds, and let us outline its methodology and goals:

The imaging studies, by and large, were selected and collated by Dr. Guido Trasimeni, at the University of Rome (La Sapienza), who also interpreted the observations. This is the natural course of events now that the neurosurgeon is no longer... also a neuro-radiologist. I think this has strengthened considerably the overall value of the text.

The list of contents – introduced by its *specific philosophical message, as all chapters of the book* – has been extensively re-worked by the copy-editor to provide as detailed a flowing sequence of the order of presentation as possible. A careful reading of this presentation before undertaking a systematic study of the treatise or only consulting it in search of an answer to a particular question is recommended. It expresses telegraphically the structural development of the narrative content.

Positioning introduces immediately the reader to all three of the reasons cited in the first paragraph of this Preface for writing a Second Edition: correcting errors or inexactitudes, updating the text, a more complete and graphic expression of my message. It integrates imaging studies into the exercise of conceptualizing the target area and sets them as closely as a two-dimensional image may be placed to a three-dimensional representation: the reader looks at the image as though he were the surgeon. The glaring error of rotating the semi-seated child around the axis of the tabletop is corrected, with illustrations showing how this brings the child into a complete vertical position. The answer to the position problem is presented.

The same illustration techniques are used in **Incisions**, integrating even more completely the images into the half-tone drawings, adding new color drawings for skull-base procedures. **Hemostasis** was integrated with the **Incisions** chapter to eliminate repetitive and confusing descriptions, since with an incision one begins hemostasis. **Flaps**, **Suturotomy**, and **Dural Openings** remain unchanged in content: no perspectives for performing these have been developed, and access to the base of the child's skull is fully achieved without resorting to the osteoclastic techniques recommended by some for surgery on adults. In **Cerebral Retraction**, anatomical drawings of normal structures are added to give depth to the descriptions, a section of the surgical technique for preserving the anatomical integrity of the olfactory nerves is added. **Cerebrotomy** and **Cerebral Resection** are unchanged.

Epilepsy is a new chapter, added because of the great interest in the surgical treatment of this affliction during the past 10 years. It was written with Dr. Francesco Cardinale, who put together all of the material. Since this treatise is on pediatric neurosurgery, this chapter is written informatively: it is not a treatment of epilepsy as a clinical and surgically amenable disease state. This must be dealt with by epileptologists, who write for those who understand the principles and are qualified to apply the surgical techniques. Pediatric neurosurgeons, with rare exceptions, limit themselves to referring children to highly specialized centers. Hence, this chapter is an in-detail, but very elementary, reference source. It provides all the information a generalist needs, and stresses the indications for referring the children to surgical epileptologists.

In **Tumors**, one finds a very extensively re-written and expanded text. Clinical information, such as incidence and percentages of individual tumor types, documentation of incidence of complicating hydrocephalus along with the results of treating it pre-craniotomy. Tabular summaries of tumor resection results and survival rates compose a significant portion of the Introduction. This was done to provide a base upon which the clinical considerations could be built. Then, at the very beginning, skull base and bony tumors are dealt with in the new style of integrating imaging studies into the clinical and operative planning methods... as already commented upon for the earlier chapters and as is continued throughout the subsequent ones.

Weaving epidemiologic and clinical data into surgical considerations and technique set a lively reading pace which I hope will permit the reader to dwell upon each tumor type, confronting one with the idea of evaluations concerning treatment options. It is here that I have tried most to establish a personal contact with the reader, to welcome him to an open and spontaneous "discussion" with this book. The full breath of pediatric brain (eye included) and mesenchymal tumors unfolds from external (chordoma) to internal (choroid plexus papilloma), as the extensive literature review and re-evaluation of my personal cases are brought into the original text, as standard middle and posterior fossa approaches are revisited and developed into the more modern "skull base" terminology and perspectives. The critical review component is present throughout, most affirmatively in medulloblastoma, craniopharyngioma, ependymoma, intramedullary tumor.

In **Vascular Disorders** the continuing value of cerebral angiography, the role of MRI, and dramatic shift to intravascular approaches to such complex problems as arterio-venous malformations of the galenic system are broadly treated. Advances in anatomic and embryologic knowledge form the basis for new treatment methodologies, which are not within the surgical armamentarium of the neurosurgeon. The confrontation of surgical and intravascular approaches seems close at hand, for microsurgery (especially of the "keyhole" variety) brings us to a strong enough position to reconsider freshly the issues at hand. **Infections** opens with a thorough review of all – primary and secondary – surgically related infections, and then proceeds to discuss surgically treatable diseases, and **Trauma** is expanded considerably to bring newer concepts in coma classification and socio-economic causes of the "battered child" syndrome into our areas of knowledge. Post-traumatic vascular pathology is considered here rather than in **Vascular Disorders**.

Congenital Anomalies and **Hydrocephalus** are extensive re-workings, incorporating the most recent theoretic and technical consensuses ranging from craniosynostoses through the lipoma and into a totally new perspective of hydrocephalus (its pathogenesis primarily, but also classification). The Chiari malformations, syringomyelia, arachnoidal cysts are integrated into a readily comprehensible group of anatomo-pathological clinical entities which appear to be inter-related.

As in the First Edition, I made all of the drawings in the conceptive stage. These sketches were then presented to Ruth Daly (for the First Edition) and Marina Longani (for this, the Second Edition). Each of these two extraordinarily gifted young ladies then patiently and skillfully worked the sketches into the finished products this book holds.

It is my hope that the owner of this book will read it, at first as though it were a novel, and then refer to it as he would to a friend, a colleague, a teacher – but always conclude by "trusting his own crooked eye".

Acknowledgements

Five people – four already mentioned – contributed significantly to this book, to the extent that their names need special emphasis. The four already mentioned are Ruth Daly, Marina Longani, Guido Trasimeni, Francesco Cardinale. The unmentioned one, Lucia Duran Raimondi, my wife, scanned the literature, collated the bibliography, typed the manuscript repeatedly, integrated the illustrations into the text, and picked up all the pieces I dropped... re-inserting them quietly and efficiently into their proper places.

Preface to the First Edition

"Better is a dinner with herbs where love is, than a stalled ox and hatred therewith."
SOLOMON, Kings – The Holy Bible

Lest the preface become an essay, lest it stand alone and independent of the text, I shall limit it to presenting the What, the Why, and the How of this work.

The What is the subject of the Introduction. There, the reader will find a narrative integrating selected fundamentals of this book's contents into specific perspectives of what Pediatric Neurosurgery is as a theoretic and a technical discipline.

The Why results from the development of our field as a speciality and, as such, is beyond communicating its many messages only through articles, conferences, chapters, and sections of "Handbooks." Pediatric Neurosurgical centers have been established, providing much clinical experience and varying amounts of experimental opportunities to integrated groups of workers, carrying on their activities, more or less, in a collegial manner. Though not recognized as an independent speciality by international, regional, or national adult neurosurgical organizations, Pediatric Neurosurgery is *de facto* recognized by pediatricians, by *all* other medical and surgical specialities, and by Society. It is taught at the undergraduate level in medical schools, and at the graduate level in such specialities as neurosurgery, neuroradiology, neurology, pediatrics, and anesthesiology. Its principles are being established, its limits extended and defined, and its practitioners indentified. Texts are needed.

By and large, multiauthored books dealing with an entire field are disjointed, eclectic works, allocating limited pages to almost unlimited numbers of subjects, each dealt with by different clinicians. A common thread is wanting; the woof and the warp often fail to strengthen one another or to form a pattern. Bibliographic and clinical reviews abound.

I chose to undertake this work alone, attempting always to remember Donald Matson's words when I asked his advice concerning what I should include in my book Pediatric Neurosurgery: "Tony, write only what you know and express it as you see it." Therefore, the reader will find some subjects and chapters that are strong, some that are weak, some that satisfy him and some that don't. He will encounter throughout the text a direct and consequential relationship between diagnosis and surgical indications, between recommending surgery and knowing what result (quality of life) one may reasonably expect, between surgical technique and attaining the desired therapeutic goal.

The How of a book such as this (how it is conceived, composed, constituted, and completed) determines its effectiveness. The author's contribution to the subject, in turn, is a mosaic of mentality and motivation, experience and readings, analysis and synthesis. All of these are inspired by a composition of oneself, his teachers, his students, and his patients. By the time a physician enters the third phase of life, his learnings and the absorbable messages of his teachers have blended into a single cog-

nition. His emotions and volitions are the stuff that permit him to make the transition from student to teacher.

Perceptive and constructive students contribute by their very presence: by learning quickly and well, they first reward and then stimulate the teacher to move on, to identify new problems and to solve them, to formulate new clinical perspectives, and to give substance to new surgical techniques. I have been blessed with many such students who, by virtue of intelligence, diligence, and total dedication also supplemented one another's education... and training. Their very different national origins and ultimate goals in life formed the basis for a truly humanistic school of Pediatric Neurosurgery: we always were foresquare in front of the reality that sick children are sick children everywhere in the world, that their parents suffer equally irrespective of the gravity of the illness, and that their needs for neurosurgical care *must* transcend the economic or academic needs of the physician. The bittersweet: from time to time I am discomfited by the truth of Saadi's recollections... "Never have I taught a student archery without, in the end, becoming his target" (Saadi, *The Rose Garden,* circa 1280 A.D.).

Still, when everything is said and done, how does a book that hopes to be a humanistic treatment of a scientific discipline, one composed of theoretic and technical elements, come into existence? I have no answer to this question. I do have a need to express, in a very few words, the humanistic part of my being, as a Preface to what the reader will find as he encounters the scientific aspects put forth in words, illustrations, and photographs. In holding myself out to treat diseases or injuries of a developing, and growing, brain, and to teach these perspectives and arts to others, I have never ceased, not for a moment, to be overwhelmed by this awesome responsibility, this unique privilege – both having resulted in polarizing my conscious efforts to my life's work. These latter came from my love for my own children. My vow to dedicate myself to Pediatric Neurosurgery was made when I spent two months with my newborn son Marco on 2-East at Children's Memorial Hospital. Marco, Laura, and Paolo (my children) remain the most human, powerful, forces I have ever felt; they have given me the perspective to see my parents, Vito Orazio and Leona, and the understanding that a child and his parents are one: neither dominates, neither may decide for the other without deciding for himself. Pediatric Neurosurgery, sociologically, is Family Neurosurgery, and I have found that it cannot be practiced with equanimity without becoming a member of the Family.

Acknowledgements

I wish to express my gratitude and recognition to Lucia Duran, Ruth Daly, Elizabeth Sachs, Deborah Crocket, Barbara Stevens, Barbara Ann Quintero, and Jack Leb for their full measure of participation in various aspects of the preparation of this work. Theirs was not just a duty discharged, it was in every way an expression of understanding, support: a sincere application of individual and personal perception and skills to an undertaking which required today's sacrifices for tomorrow's results. I hope they will be satisfied.

Koreaki Mori prepared the section on shunt characteristics and external ventricular drainage, Luis Yarzagaray the ventriculo-gallbladder shunt, and J. Thomas Brown the intrauterine shunt. Yutaka Maki made me aware of Monk Kukai (priest who contemplated sea and sky).

The staffs at Springer-Verlag and Stürtz were truly professional and human, understanding my desires, concepts, and apprehensions.

The last impression I wish to leave the readers, who are careful enough to peruse this section, is that an author cannot complete a book such as this unless he has been motivated continuously throughout its conception, writing, editing. In my case, that motivation is the result of a desire to pass on to the medical world the benefits I received from my patients, their families. I thank them for their trust and their confidence.

List of Contents

All too often we get so lost in our work on Earth
that we forget life is a transition between sea and sky.

1 Positioning

"If you have planted a thistle, do not expect jasmin to sprout –."

SAADI, The Fruit Garden

It is not realistic to specify the single most important aspect of an operative procedure, namely, diagnosis, anatomical localization, blood volume control, flap selection, exposure, or head and body position. It is realistic, however, to assert that, if the surgeon positions the child's head and body properly – taking into consideration the location of the lesion, the planned skin incision, and bone flap – he will, throughout the operation, be oriented anatomically – he will always have the lesion at the center of his operative field.

One of the most significant equipment/instrument advances hitherto made for neurosurgery, neuronavigation, is still of no value in operating on newborns and infants, and of very limited value in operating on toddlers. There are two principal reasons for this conclusion: the thin skull does not lend itself to the use of rigid frames, and the commonly present secondary or primary hydrocephalus predisposes the brain to major shifts within the cranium once the dura is opened.

On the other hand, the wide range of applicability of endoscopic procedures finds many indications in the pediatric ages. Neither of these methodologies, however, simplifies the matter of positioning the child. In fact, as we move forward with minimally invasive surgery, we find it ever more important to obtain precise positioning.

Positioning for pediatric neurosurgery varies considerably with the age of the child (newborn, infant, toddler, juvenile), the number of surgeons (one surgeon alone, surgeon and assistant, etc.), the location of the anesthesiologist and amount of monitoring equipment used, and the target area.

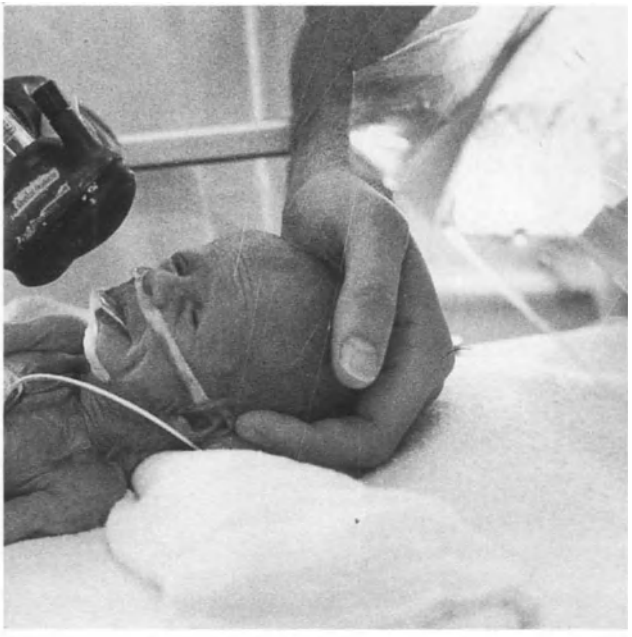
These variables are generally not applicable to neurosurgical operative procedures on adolescents and adults because of their uniform size, and the constant relationship between brain and skull. Also, there is no need for such anatomical considerations as open fontanelles and sutures, relatively larger basal cisterns, continuity of the periosteum with the outer layer of the dura at the sutures, and the presence of ossification centers. Therefore, this chapter is organized to present general and specific considerations concerning each age group, individual body positions, relative position of surgeon vis-à-vis the child, and positioning of the head. The recommended positions for specific operative procedures are then discussed before they are described.

General Discussion

Age

The relative sizes of the surgeon's hands and the head of the newborn, infant, toddler, and adolescent place into relief the remarkable differences in dimension of skull and brain in the different pediatric age groups. This range in overall head size is expressive of a proportionate range in individual anatomical structures (lobes of the brain) or compartments (basal cisterns), since they vary individually, and disproportionately, from the newborn to the toddler.

The head of a premature newborn may be so small as to fit within the palm of the surgeon's hand (Fig. 1.1A), whereas that of a term newborn rests comfortably within the fully cupped adult hand (Fig. 1.1B). The heads of the infant and toddler (Fig. 1.1C,D) are proportionately larger (the same hands are used in all four photographs). This change in volume occurs pari passu with changes in dermal (skin, connective tissue, and aponeurosis of the scalp) thickness, inversion of relative amounts of diploic and lamellar components of the skull, diminution in volume of cisternal cerebrospinal fluid and increase in cerebral volume, and closure of fontanelles and narrowing of the sutures. In all four of these age categories the air sinuses are not yet developed, and the second dentition tooth buds occupy the



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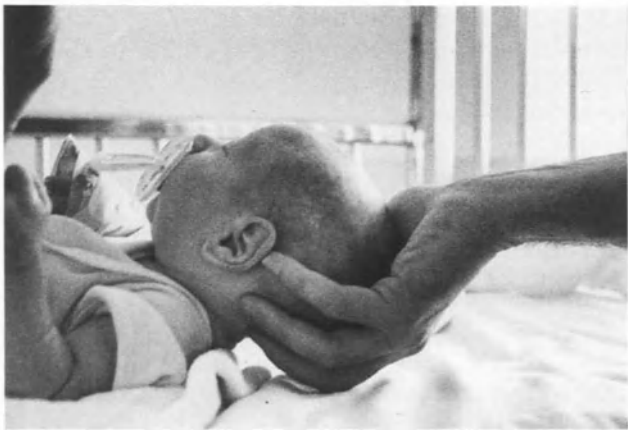

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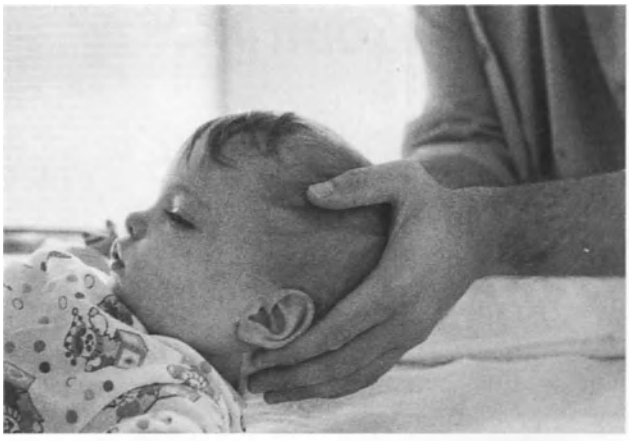

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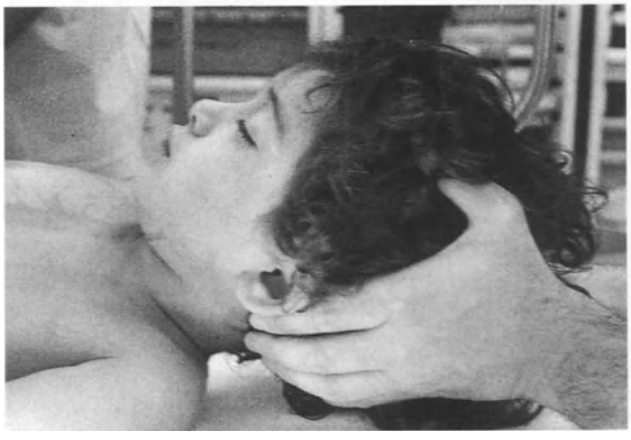

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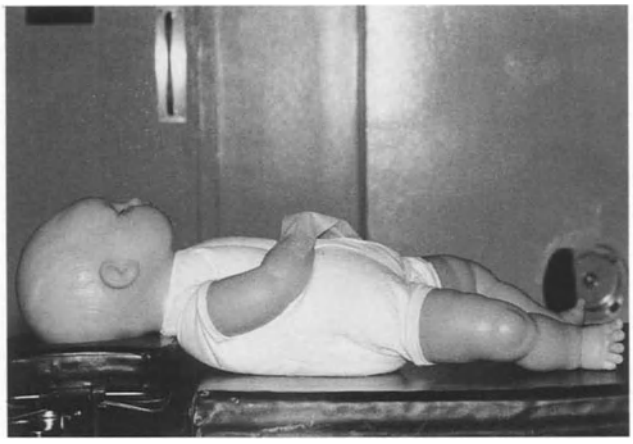
Figure 1.1. (A) Premature newborn. Not only is the skull size so small as to fit completely within the palm of an average adult hand, but the individual membranous bones slide freely over one another at the sutures. The scalp is very loose, and the fontanelles may be full or actually bulge. (B) Term newborn. There is a firmness to the term born child's skull which distinguishes it immediately from that of the premature, the fontanelles are invariably depressed and pulsatile, and the scalp is somewhat less mobile over the underlying membranous bones. The skull volume is significantly larger. (C) Infant. The infant's fontanelles are closed, as are the sutures, and the subgaleal space is much more a potential than a true compartment. An average human hand from fingertip to thenar eminence is its measure. (D) Toddler. The toddler has a rigid skull and developed mastoid eminences; the membranous bones cannot be digitally depressed. Both hands are necessary to hold it firmly.

jaw bones. The pacchionian bodies have not yet appeared, and the cortical bridging veins are both exceptionally fragile and easily converted into rectilinear structures.

Premature Newborn

To all intents and purposes, and with only the rarest exceptions, neurosurgery on the brain of the premature newborn is limited to placing an external ventricular drain or inserting a ventriculoperitoneal shunt. Consequently, the supine position, with the head turned to either side, is all that is used in this age category. The exception is the prone position for posterior fossa hematoma secondary to birth injury.

Figure 1.4. (A) Access to the cervical spine, the craniovertebral junction, and the occipital lobes may be gained with the newborn or infant prone. It is necessary to place pillows or sandbags under the shoulders and to flex the head. The shoulders should be taped in the caudad direction (*arrow*), distracting the neck, and the head should be taped to the headrest (B). This is a lateral view showing the degree of flexion of head on neck. Note that the frontal eminences, not the face, nestle into the headrest. ►



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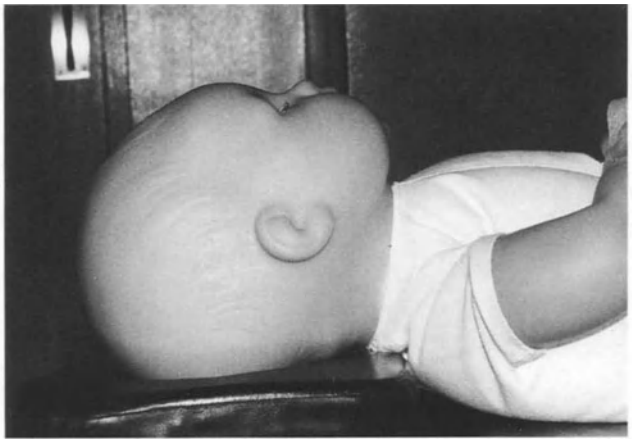

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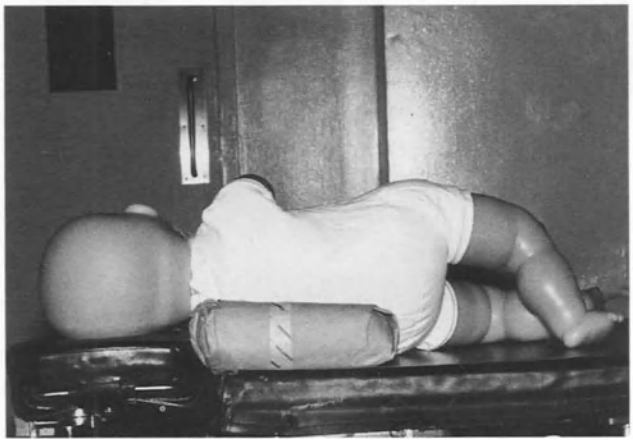
Figure 1.2. (A) The body is positioned supine, with the arms taped loosely across the thoracoabdominal junction. This permits ready access to the chest wall and avoids compression of the abdomen. (B) This is a view of the head's position when the child is supine. The head is neutral. One may either flex or extend it for access to the anterior frontal or posterior parietal areas, respectively. Turning the head slightly to one side or the other (either from the neutral position, flexed or extended) facilitates more direct access to the lateral surfaces of the hemispheres.



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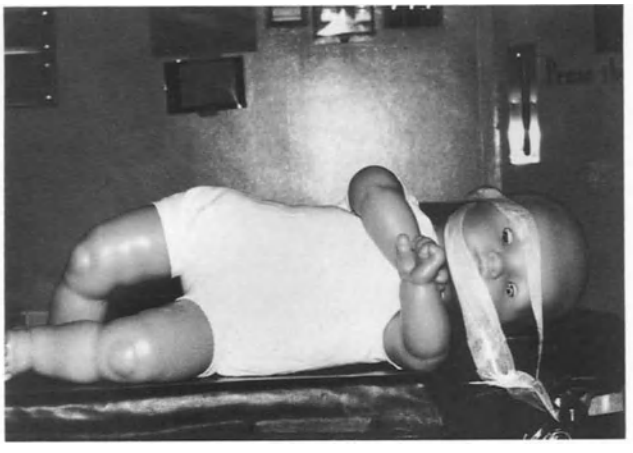

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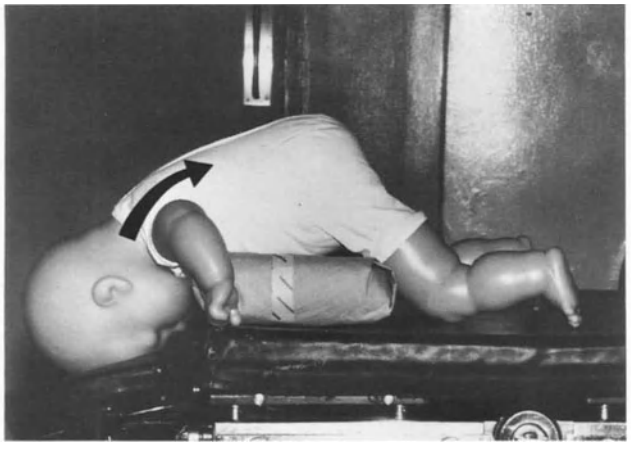
Figure 1.3. (A) The supine newborn may be rotated from its back to its side and held firmly in position with a single sandbag. (B) The arms are folded across the shoulders. Tape suffices to fix the head firmly in the desired position. Placing the superior leg behind or in front of the inferior leg thrusts the body more into the supine or prone positions, respectively. The anesthesiologist may increase his access to the thorax by taping the superior arm to the side and extending the inferior arm. This will not shift the body.



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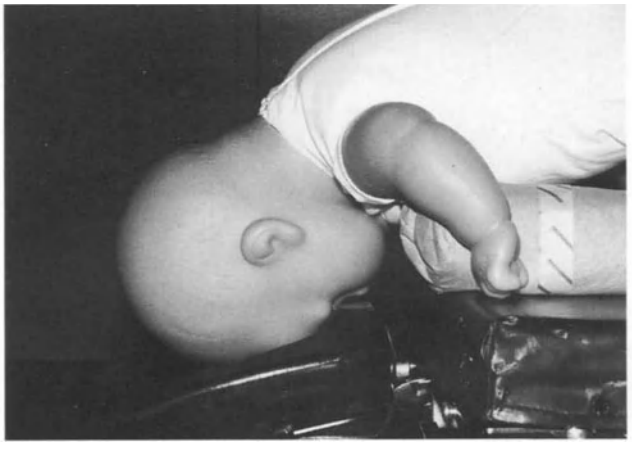

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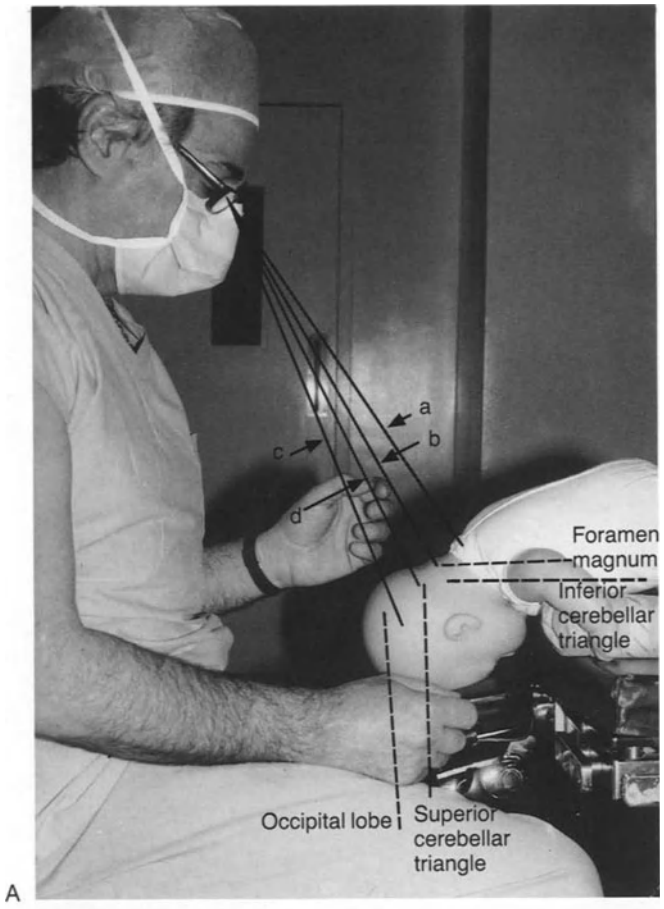

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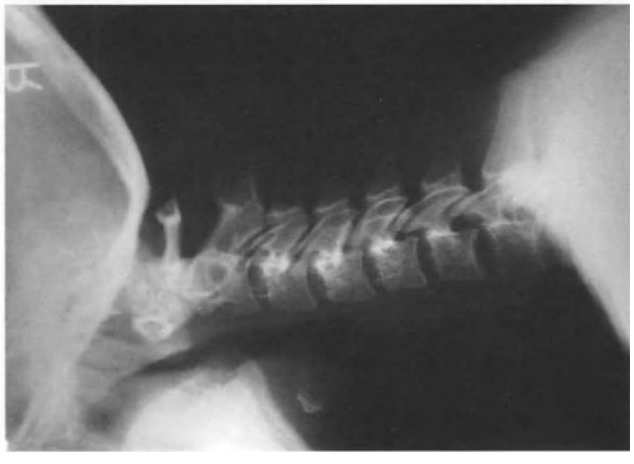

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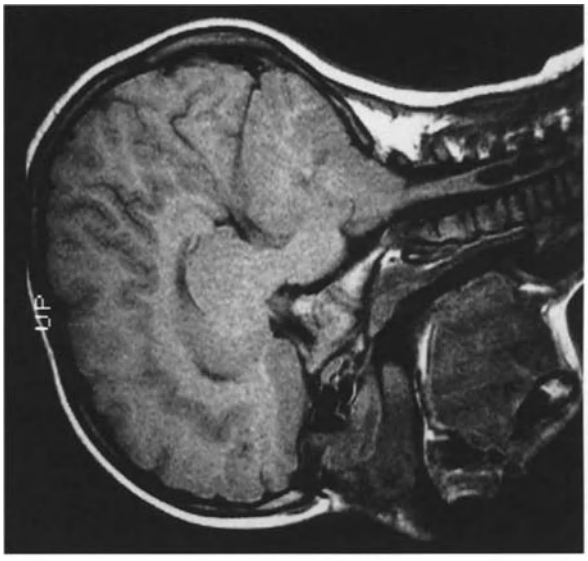

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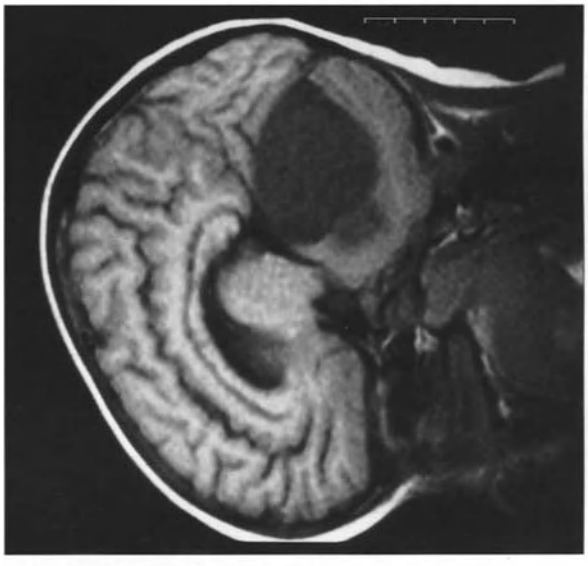

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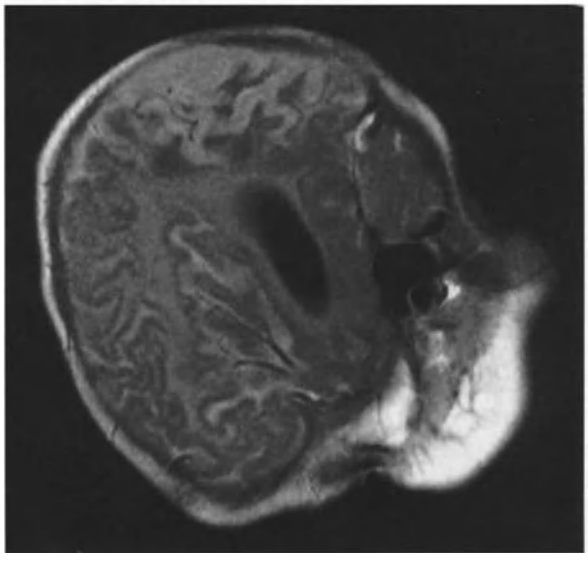


E

Figure 1.5. (A) The relative positions of the surgeon and the child's head, with the newborn prone, illustrate: (a) an *excellent* line of vision to the cervical spine; (b) an *adequate* line of vision to the foramen magnum (except in Chiari II children); and the inferior cerebellar triangle; (c) an *unacceptable* line of vision to the superior cerebellar triangle area because of the ledge of squamous occipital bone and the transverse sinus; (d) a good line of vision to the occipital lobes. Lateral cervical X-ray (B), MRI of chiari I with syringomyelia (C), MRI of superior vermian tumor (D), and MRI of occipital lobe lesion (E) to put into relief the points made in 1.5A.

Term Newborn and Infant

The term newborn and the infant may suffer the full range of neurosurgical diseases, so that it may be necessary to operate on children in these age ranges in the supine, the prone, or the sitting positions. The sitting position in the *newborn* is extremely difficult to maintain (the infant keeps slipping away from the drapes). The *infant* may be more securely positioned sitting. The need to arrive at the region of the culmen monticuli of the cerebellar vermis, fortunately, does not occur often. One may, consequently, perform most intracranial procedures on the term newborn using either the supine or prone positions. Indeed, one is advised to avoid the sitting position in these ages if at all possible. The anterior fossa, orbits, frontal and parietal lobes, and metopic suture may be exposed with the newborn or infant supine and its head in the neutral position (Fig. 1.2A,B). Rotating the head to the opposite side, and placing a pillow or sandbag along the back from the shoulder to the hip, provide immediate access to the entire hemispherical convexity (Fig. 1.3A,B). Elevation of the shoulder by a pillow or sandbag avoids both stretching and compression of the jugular veins, and intervertebral foramen impingement/compression/occlusion of the vertebral arteries.

The prone position (Fig. 1.4A,B) is for occipital, craniovertebral junction, and some posterior fossa lesions. It permits optimal exposure of the occipital lobes and craniovertebral junction, but the anatomical structures within the posterior fossa are so located as to permit one to work effectively only in the inferior cerebellar triangle. The position of the surgeon, vis-à-vis posterior fossa contents, makes this obvious (Fig. 1.5). The disadvantages of this position are most notable when performing a suboccipital cranictomy for decompressing the foramen magnum in children with the Chiari II malformation. *One is not able to work efficiently, either in the superior cerebellar triangle for posterior fossa masses or at the foramen magnum in Chiari II children, with the newborn child in this position.* It is also difficult to gain a direct line of vision to the superior cerebellar triangle because of the short posteroanterior (clivus-squamous occipital) and the long superoinferior (tentorial opening-foramen magnum) distances. These anatomical characteristics impair significantly the surgeon's ability to visualize the superior aspect of the culmen monticuli.

Toddler

The *toddler* may be put, safely and effectively, into either the sitting or lounging position because the trunk is long enough to sit the child up, and the skull, generally speaking, is thick enough to offer purchase to the pins of standard headholders. It is fortunate indeed that this is true, since there is a high incidence of posterior fossa pathology after the 2nd year of life. Such lesions as superior and inferior cerebellar vermis tumors, arteriovenous malformations of the galenic system, pineal tumors, arachnoidal cysts of the quadrigeminal and superior cerebellar cisterns all occur in this age category. Figure 1.6 illustrates relative positions of the surgeon and patient with midline occipital, some pineal, and superior cerebellar triangle lesions in the newborn (Fig. 1.6A), toddler (Fig. 1.6B), and juvenile (Fig. 1.6C).

Specific Positions

Supine Position

The supine position is for frontal, frontopterional, parasellar, and orbital lesions. Placing the head in the neutral position, and extending it slightly, eliminates the need for lowering the head of the table when working at the chiasm or optic foramina. Conversely, flexing the head slightly provides more direct visualization of the cerebral convexity along the posterior frontal and anterior parietal regions of the brain. With the head neutral and slightly flexed, the supine position offers immediate access to the convexities and parasagittal areas of the frontoparietal, parietal, and parieto-occipital lobes. Turning the head to either side (bringing the coronal suture parallel to the sagittal plane of the body) affords access to the convexity of the hemisphere, exposing the frontal, temporal, or occipital poles, and to the floors of the anterior and middle fossae, the tentorium, and the lateral surface of the opticocarotid region. It also puts the child into perfect position for a ventriculoperitoneal shunt, permitting the surgeon to insert the ventricular end either into the occipital horn and trigone or the frontal horn.

Although extension of the head around an axis running through the auditory canals does not interfere with venous drainage, flexion may cause the horizontal rami of the mandible to compress the internal jugular veins. Distraction of the skull prior to flexion minimizes this risk of jugular compression (Fig. 1.7A–C).

Prone Position

By placing the child prone with the head in the neutral position, one may expose the lambdoidal suture (the parieto-occipital region) for immediate access to the occipital lobes. Flexing the head and distracting it at the craniovertebral junction provides access to the squamous occipital, craniovertebral, and cervicothoracic regions. This position is used for occipital, inferior cerebellar triangle, foramen magnum, and superior cervical cord lesions (Fig. 1.8A,B). When the child is prone, as when it is supine, particular care must be taken to

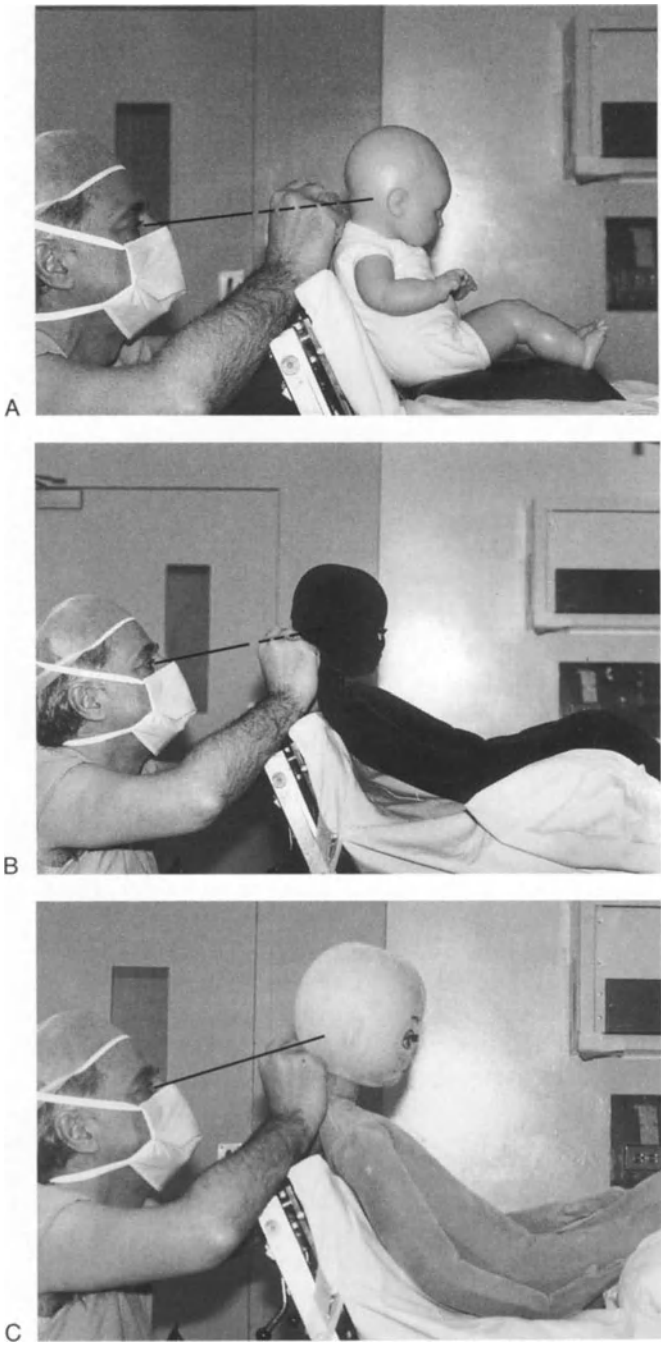


Figure 1.6. Note the relative sizes of the surgeon's hands and the child's head as shown here with (A) the term newborn: the surgeon's hands, in the neutral position, must be held apart to permit visualization of the operative field between the hands, since the newborn head is so small that one is not able to look over them and into the posterior fossa; (B) the toddler; (C) the juvenile: the head size is such as to enable the surgeon to look over his hands, giving him a wider range to pronate and supinate them with wrist or elbow movement. With increasing age and body size, it becomes decreasingly necessary to look between one's hands, increasing the operative field to vision and manipulation. The head size of the toddler and juvenile are relatively the same, but the neck and body sizes are so different as to alter the relative working space.

distract the skull from the cervical spine prior to flexing it around the axis that runs through the auditory canals.

The pressure exerted by the weight of the skull tends to jam the mandible against the jugular veins, greatly diminishing cerebral venous drainage. This is worsened by the horseshoe headrest (which must be used in newborns and infants), but somewhat facilitated by the skull-pin Mayfield or Gardner-Wells (Codman, Inc., Johnson & Johnson Company, Randolph, Massachusetts, USA) headholder (which may be applied to toddlers and older children). Whether using the horseshoe or Gardner-Wells headholders, adequate clearance between the *symphisis mentes* and the body mat must be provided so that the endotracheal tube is not compressed. If this happens, it may either kink or be forced into one of the main stem bronchi. The weight of the drapes, especially as they become soaked during the procedure, may be enough to cause a decubitus of the chin. One must leave enough room for the anesthesiologist to check and manipulate the endotracheal tubing.

Lounging (Sitting) Position

The "lounging" position is ideal for access to the posterior III ventricle, the superior cerebellar triangle, and the falx tentorial junction. *Irrespective of the physical inconveniences to the surgeon and the truly negligible risk of air embolism if appropriate anesthesiologic precautions are taken, it is a safe way to operate on lesions in the superior vermis, brachium conjunctivum, superior cerebellar hemispheres, opening of the aqueduct into the IV ventricle, the pineal region, and the great vein of Galen.*

The same problems concerning mandibular compression of the jugular veins are encountered, to a much greater extent, when operating on the child in the lounging position (as in either the supine or the prone positions). Here, again, the head must be distracted in order to avoid compression of the jugular veins. It must then be flexed around the axis of the auditory canals to provide the surgeon with a direct line of vision to the superior portion of the cerebellar vermis and the tentorial opening. Fixing the head securely holds it suspended against its own gravitational force. Figure 1.9A shows this with the use of a horseshoe headholder in an infant, and Fig. 1.9B with the use of the skull-pin headholder, which have not been designed for the thinner calvarium or relatively more voluminous diploic spaces. The risk of air emboli through the diploë remains very real, so care is given to adequate use of bone wax.

At times one must adapt. It may be necessary to place the headholder very close to the operative field in order to assure solid purchase, and then to use a jugular vein for a central venous pressure line (essential in either the sitting or lounging positions). Some form of

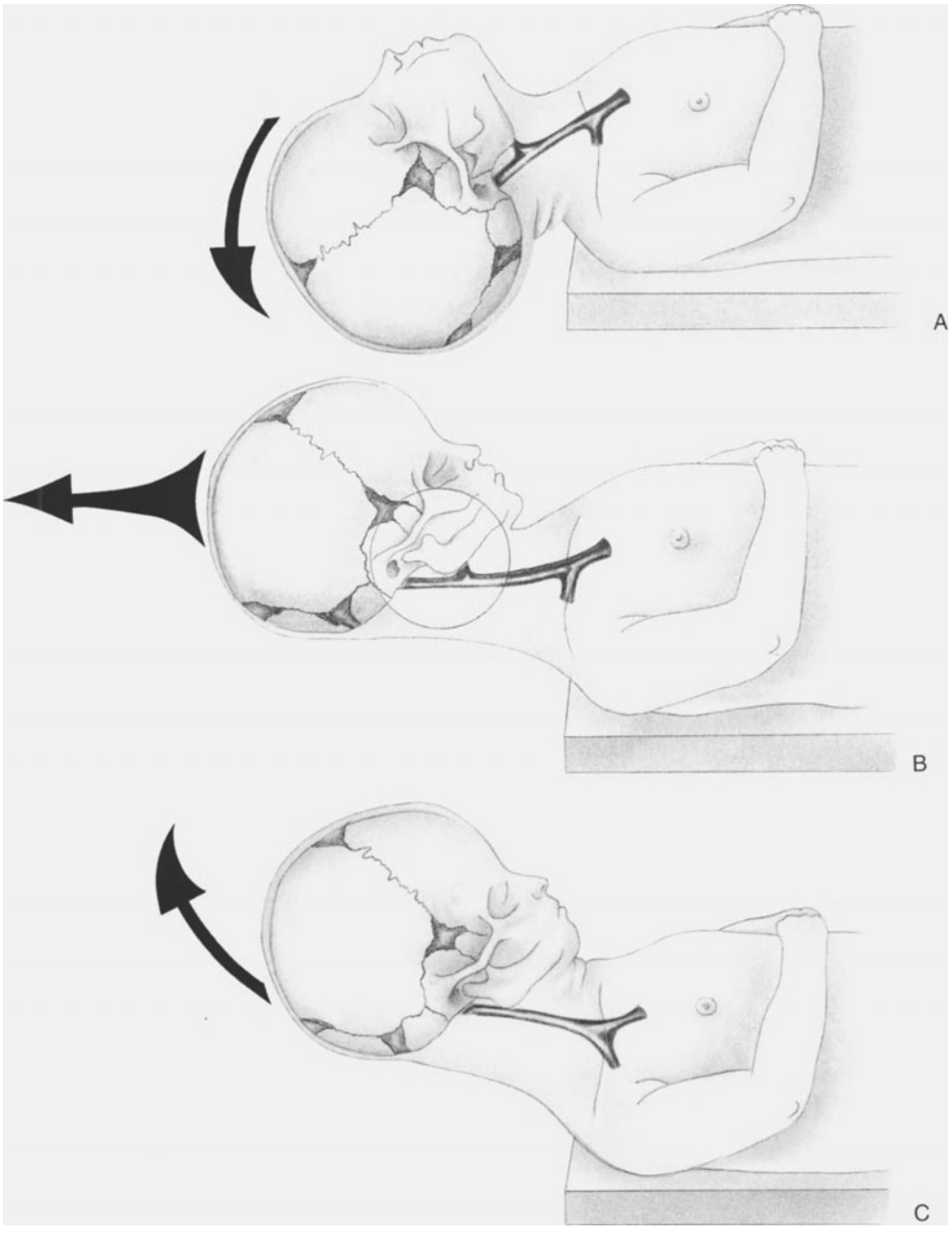


Figure 1.7. The head is drawn schematically, illustrating that, in extension, one need take no particular precautions to avoid compressing the jugular veins by the horizontal rami of the mandible, but that it is necessary to distract the head to avoid this in flexion: (A) The head is extended around an axis running through the external auditory canals; (B) the head is flexed around the same axis and distracted prior to being secured in position (either onto a headrest or in pins), avoiding jugular vein compression; (C) the same as in (B), but the head has not been distracted, resulting in compression of the vertical ramus and angle of the mandible against the internal jugular vein. Diminished venous return and increased intracranial pressure are the consequences of this compression.

plastic draping may be used to cover the tubing. Such a situation is illustrated in Fig. 1.10, which also cones down on the distracted head, allowing one to appreciate how this separates the rim of the foramen magnum from the arch of C-1. All too often, consequently, because of the very wide range in body and head size of the pediatric population and the standard size of operating tables, the surgeon must improvise in positioning and securing the child firmly in place. Pillows, sandbags, sheets, and so on are pressed into service even in the best-equipped pediatric operating rooms. A well-constructed car seat, which may be purchased almost

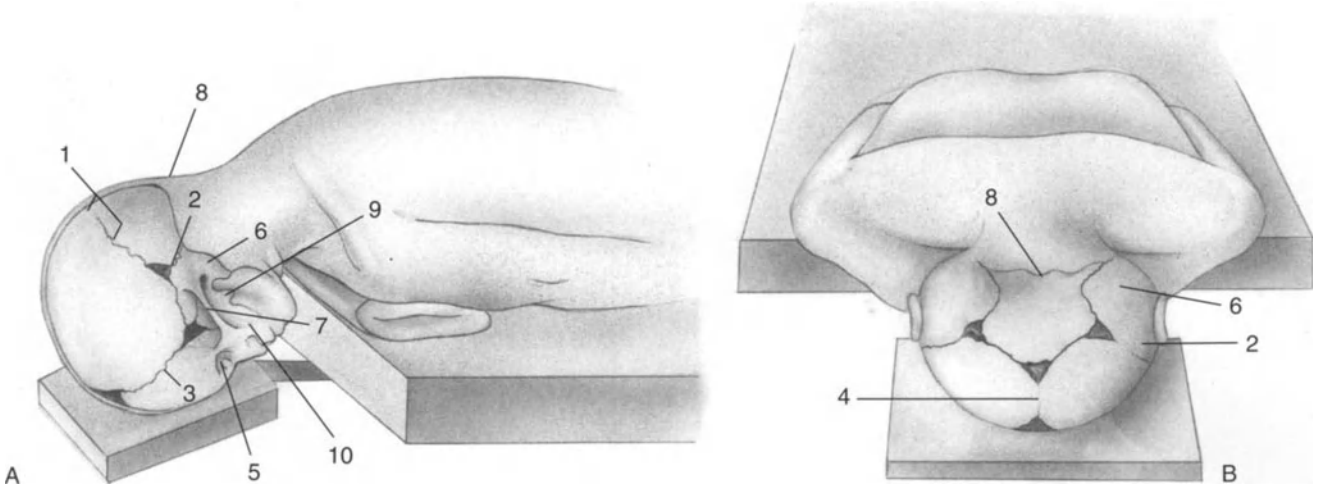


Figure 1.8. (A,B) The prone position as herein illustrated demonstrates the lamboidal (*1*), occipitomastoid (*2*), coronal (*3*), sagittal (*4*), and zygomaticofrontal (*5*) sutures: the mastoid (*6*) and the zygomtic arch (*7*); the rim of the foramen magnum (*8*); and the squamous temporal (*9*) and greater wing of the sphenoid (*10*) bones.

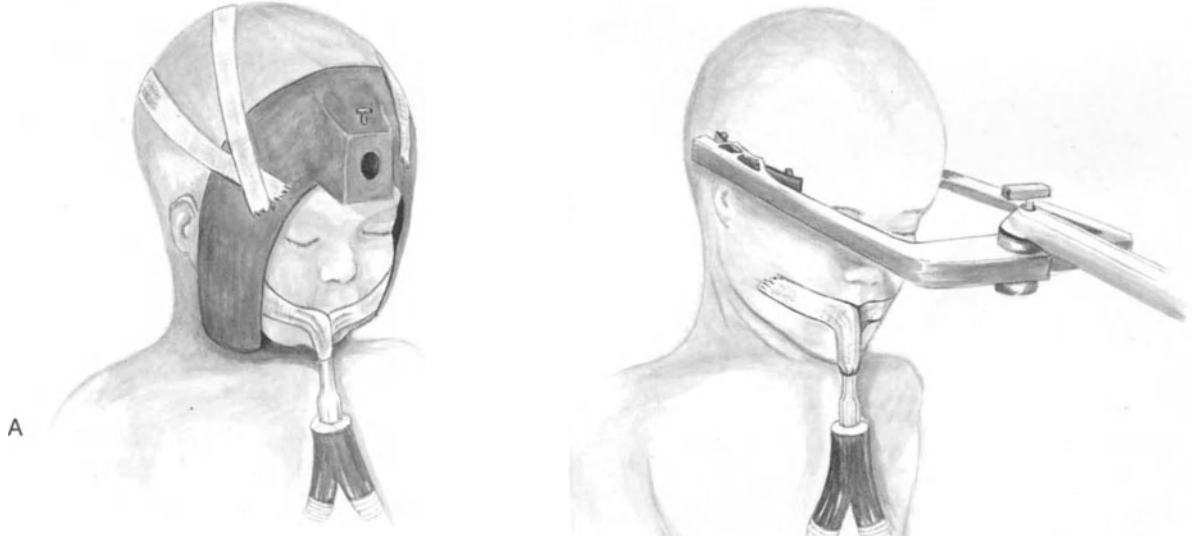


Figure 1.9. Fixation of the head in the lounging position is extremely important. In order to ensure maintenance of craniocervical junction distraction in the infant, one should tape the head to the horseshoe headrest and nestle the chin into the bottom of the headrest (A) or use the Gardner-Wells headholder on a toddler (B).

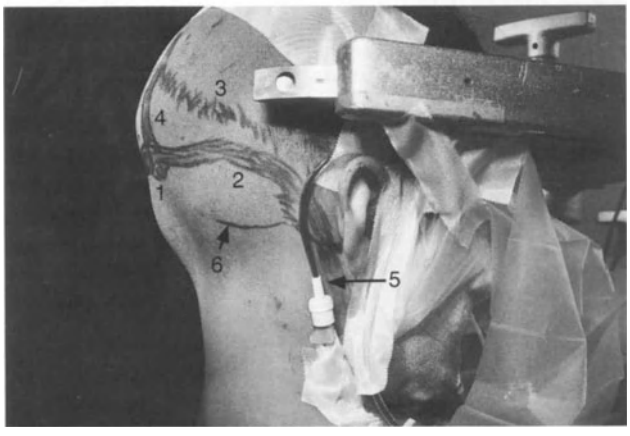


Figure 1.10. After distraction, the headholder is locked, maintaining head and neck position throughout the operation. In this child, one of the pins had to be set close to the operative field, something which at times is unavoidable, and the tubing for the central venous line had to be brought superiorly and curved around the helix of the ear. The torcular Herophili (*1*), transverse sinus (*2*), squamosal suture (*3*), superior sagittal sinus (*4*), internal jugular vein (*5*), and horizontal plane of the squamous occipital bone (*6*) have been drawn on the scalp to provide orientation.

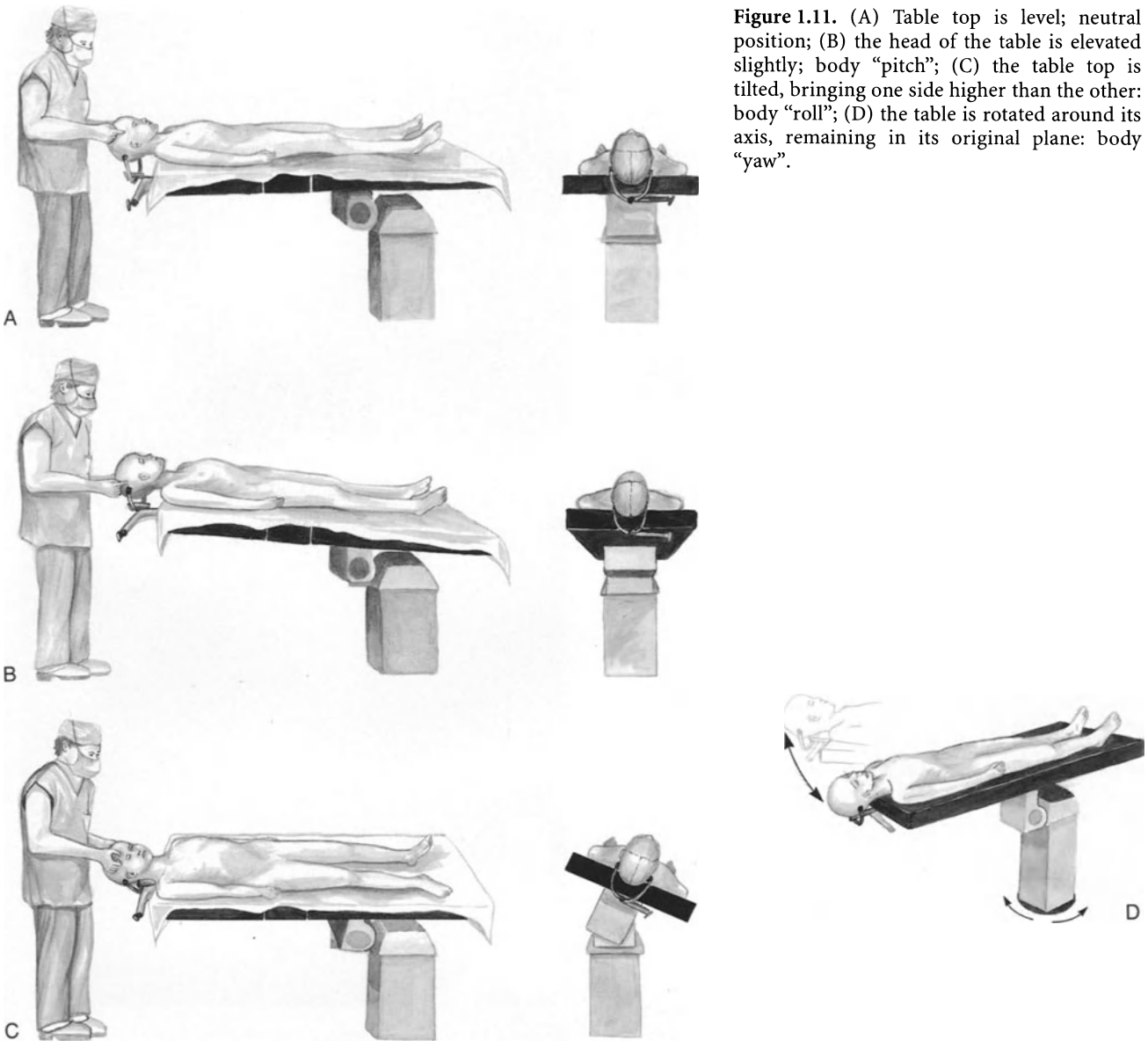


Figure 1.11. (A) Table top is level; neutral position; (B) the head of the table is elevated slightly; body "pitch"; (C) the table top is tilted, bringing one side higher than the other: body "roll"; (D) the table is rotated around its axis, remaining in its original plane: body "yaw".

anywhere, serves this purpose well. It may have to be cut, molded, or padded, but it is far superior to anything else available or to any combination of pillows, towels, and sandbags. It is ideal for moving the child onto the operating table, and from it to the cart at the end of the procedure or in the event of an emergency. Most importantly, it facilitates fixing the infant or tiny toddler in position. The vacuum-regulated beanbag serves the same purpose, equally effectively, for children in these age categories.

All this work of positioning must be done relatively quickly because the anesthetized child, especially the newborn and infant, loses body heat rapidly.

Positioning of the Child Vis-à-Vis the Surgeon's Line of Sight

The most important consideration in positioning the child for surgery is not to complicate the already diseased or injured central nervous system. The second most important consideration is to position the child securely on the operating table so that the surgeon may move the child at will, bringing it into a variety of positions throughout the procedure, so as to realize the primary goal of successful positioning: bringing the target area *for the specific aspect of surgery being performed at that moment* along the surgeon's line of vision. When this is accomplished, the operative exposure is optimal.

If the child is positioned properly, and if the surgeon takes advantage of the full range of motion (body pitch,

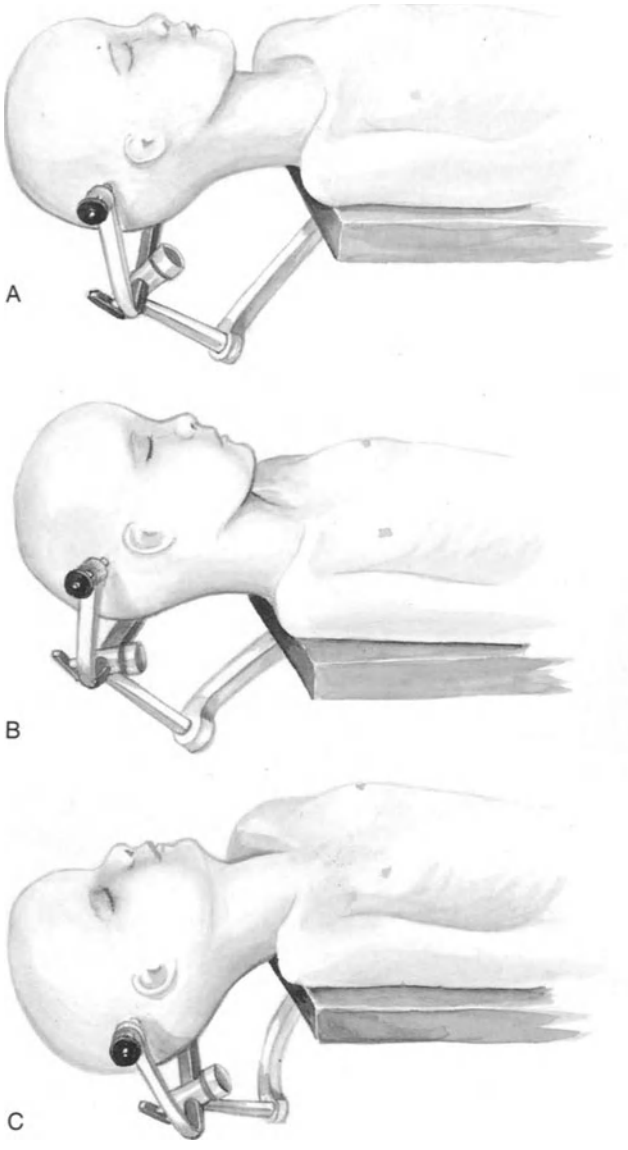


Figure 1.12. The head is shown in the neutral position (A), elevated but not flexed (B), and depressed but not extended (C).

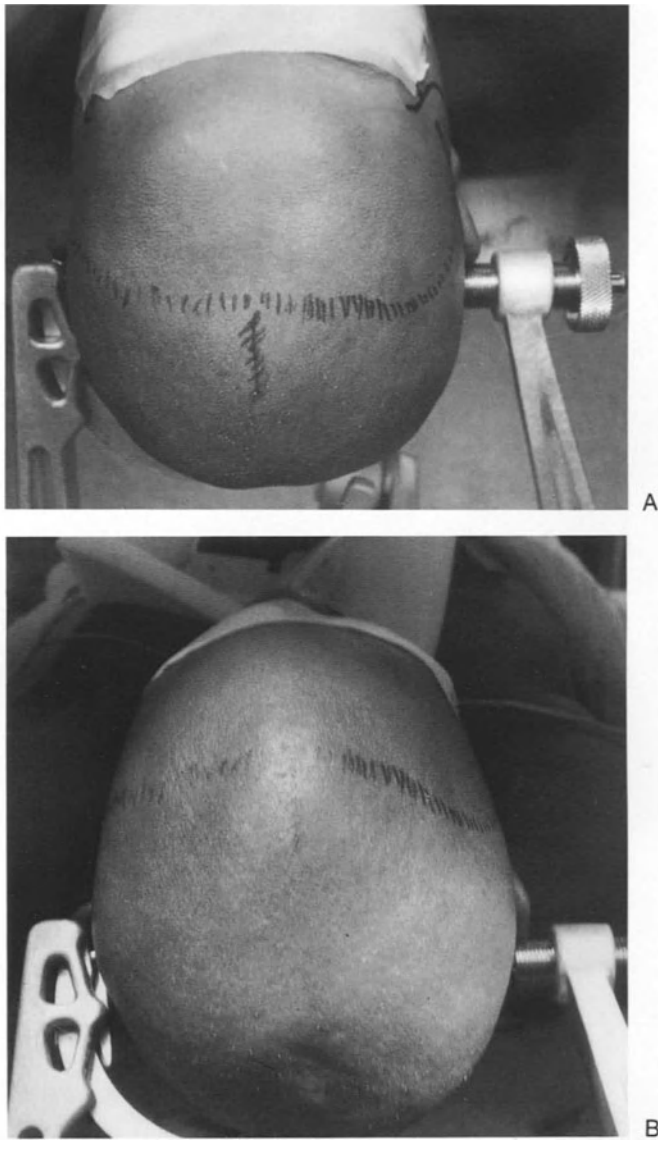


Figure 1.13. (A) The surgeon's view of the frontal bone, which is provided by using body "pitch" or head lowering with the child supine. (B) The surgeon may change his target area (shown in A) without changing his own body position, simply by altering body "pitch", lowering or elevating the head.

roll, yaw, and slight elevation or depression of head and/or body), he may work comfortably with elbows relaxed at his sides, and with his line of vision extending directly to the target area. This diminishes fatigue so that it allows the surgeon to work with his body in its natural posture. There should be little need to move about continuously, to use platforms, and to stretch or stoop during the operative procedure.

In Fig. 1.11A–D, the operating table is moved from neutral so as to provide body "pitch," "roll," and "yaw." In Fig. 1.12A–C, one appreciates the mobility of the headholder from neutral to elevation and depression of the head. All these changes in position may be obtained during the procedure, moving the operative site directly into the surgeon's line of vision or bringing a desired intracranial structure more clearly into view. This is crucial when working in the parasellar area (so as to take advantage of the bifrontopterional exposure), in the pineal region, within the trigone of the laterale ventricle, or within the region of the IV ventricle. It is important for spinal cord lesions but of little value for convexity lesions or shunts. An example of how one may change one's line of vision as the target area changes during the operative procedure is illustrated in Figs. 1.13 and 1.14.

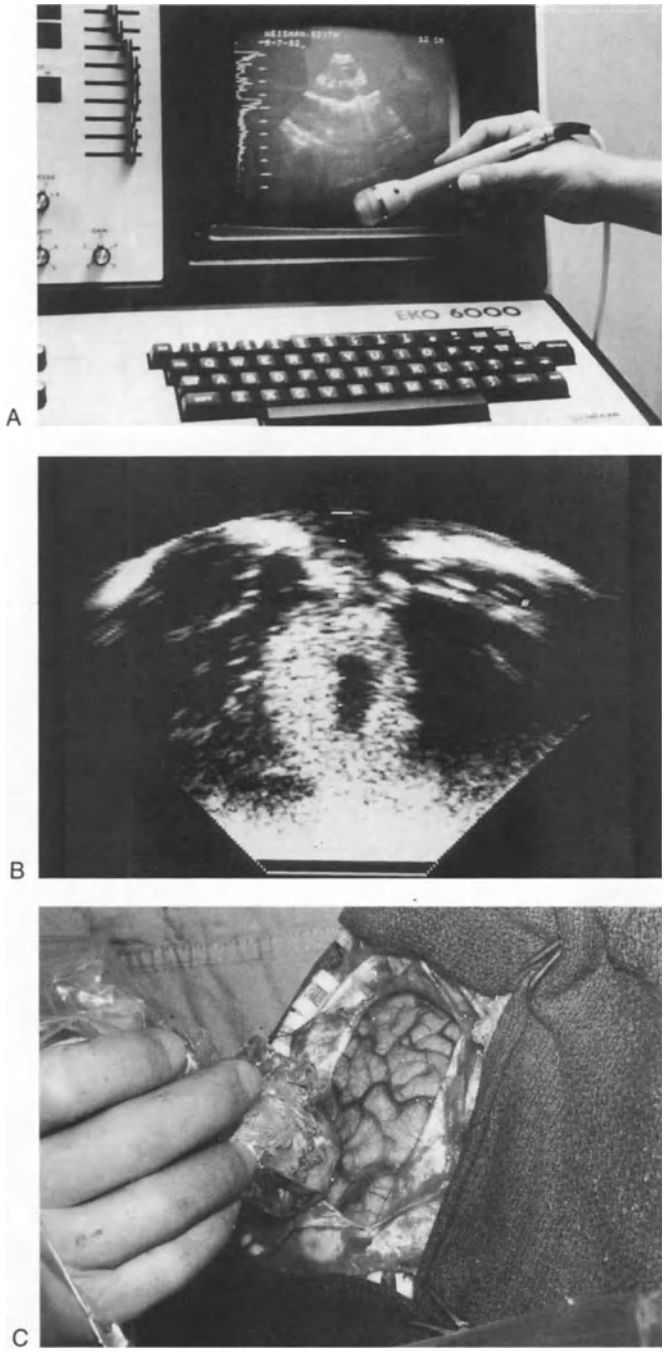


Figure 1.14. (A) Note the relatively large size of the handle and head of the echoencephalography unit and the monitoring screen. (B) This is an echoencephalographic image of a cerebellar astrocytoma that contained a small, centrally located, cystic cavity. (C) The ultrasound head has been applied to the cortical surface.

Positions of Surgeon, Assistants, and Nurse Around the Patient

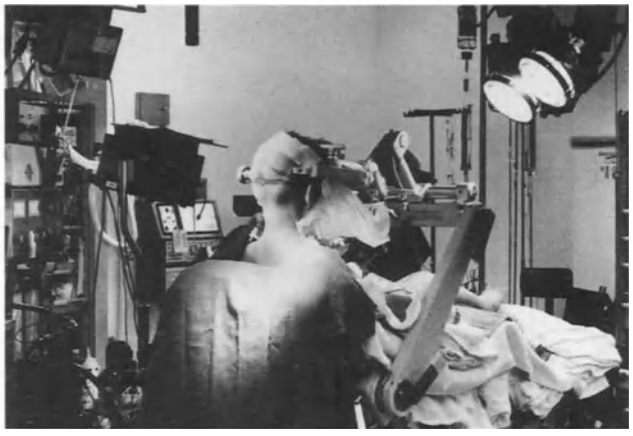
There is no need for two assistants in (pediatric) neurosurgical procedures. In fact, since the operating microscope has become a standard piece of surgical equipment, one does very well in neurosurgery *without* an assistant. When assistants are used, there is little room for them. Consequently, the scrub nurse may serve as the first assistant, passing suction or bipolar cautery to the surgeon along with other instruments. Self-retaining retractors are now standard in all neurosurgical procedures, so that it is not necessary to have someone scrubbed into a procedure to hold them. Similarly, motor-driven (either electrical or hydraulic) chairs, connected to the operating microscope, permit the surgeon to control optical and mechanical magnification, zoom, elevation and lowering of the microscope and mechanical chair, 35-mm camera shutter release, electrosurgical activation, and angulation ("roll") of the optical field by using foot controls. Assistants should scrub in for pediatric neurosurgical procedures only to learn operative technique and surgical instrumentation.

The placement of television monitors within the direct line of vision of the nurse (across the operative field) and the assistant and anesthesiologist (across the length of the patient's body) permits everyone to observe directly the details of the micro- and macrosurgical procedures. The coordination of the operating microscope with the operating table and surgeon's chair greatly facilitates access to such diverse neuroanatomical locations as the parasellar area, trigone, pineal region, and foramen magnum, without encumbering the surgeon or cluttering the operative field.

In setting up the operating table and equipment around it, and then positioning the essential operating-room personnel, attention should be given to keeping the floor as clear as possible of such lines as electrical cords and suction tubing. A central, ceiling-mounted operating microscope or universal power outlet facilitates this, as does placement of the auxiliary electrosurgical unit and suction bottles at the base of (and beneath) the operating table, so that the cords may go directly from the operative field to these outlets, rather than passing from the field and across the floor to wall-mounted outlets.

Accommodating Anesthesia

Although the methods described so far for positioning the child on the operating table permit the surgeon maximum exposure of, and access to, the operative site, great attention is also given to providing the anesthesiologist full access to the child throughout surgery.



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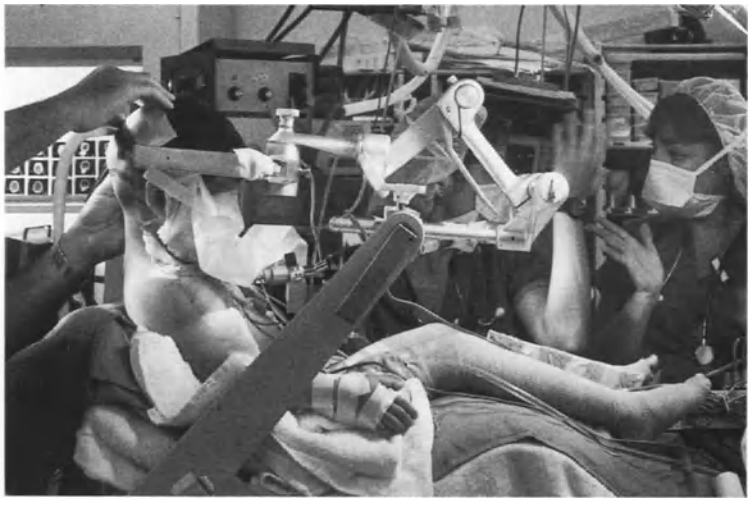


B

Figure 1.15. The heat lamps have been positioned at a safe – to avoid burning – distance, so as to provide warming (A and B). The child's eyes are covered and the endotracheal tube securely taped (B) in place. Note the wrapped legs and lounging position.

Setting heat lamps (Fig. 1.15A,B) at a safe, yet warming, distance from the child during positioning provides protection against potentially dangerous hypothermia, as does placing the plastic drape around the operative field before beginning the prep. This prevents heat loss and insulates the child from the cooling effects of spillage of surgical soap and water over the uncovered skin. It is particularly helpful if extensive body areas, as in prepping for a ventriculoperitoneal shunt, are to be included in the operative field, since a 10-min scrub would allow large amounts of surgical soap and water to accumulate along the recumbent surfaces of the child's trunk and neck.

The drapes should be placed and fashioned around the child, extending from the operative field in such a manner as to allow the anesthesiologist full access to the face, neck, chest, and limbs. This entails the use of an overhead table, either for the instruments or to which the drapes may be fixed. Placement of a Mayo stand, preferably mounted to the operating table, slightly above the most superior aspect of the head or face, depending upon whether the child is sitting, supine, or prone, suffices if an overhead instrument table is not available. Then, intravenous poles must be placed to one side or the other of the operating table. This permits tenting of the drapes from the operative site in such a fashion as to leave the anesthesiologist full access to the child for the purposes of controlling the position of the endotracheal tube, functioning of central venous pressure lines, and monitoring of intravenous fluids and thermistor probes.

Irrespective of the body position for the operative procedure, care must be taken to maintain the head as close to the level of the right atrium as possible, so as to avoid cerebral venous stasis, air emboli, and hypotension.

The provision of a color television monitor permits the anesthesiologist to observe directly the operative procedure (Fig. 1.16).

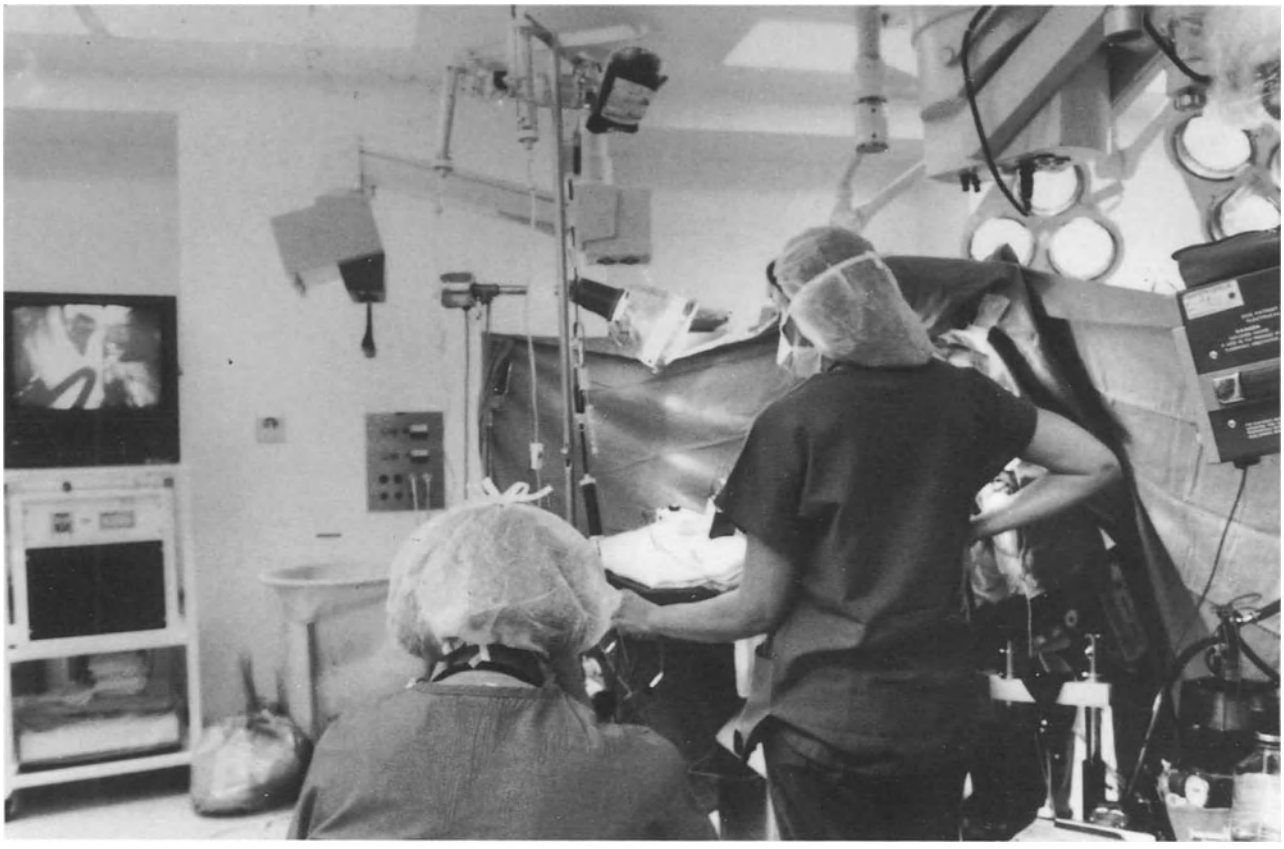


Figure 1.16. Placement of TV monitor so the anesthesiologist may observe the entire (macro and micro stages) operation.

General Positions

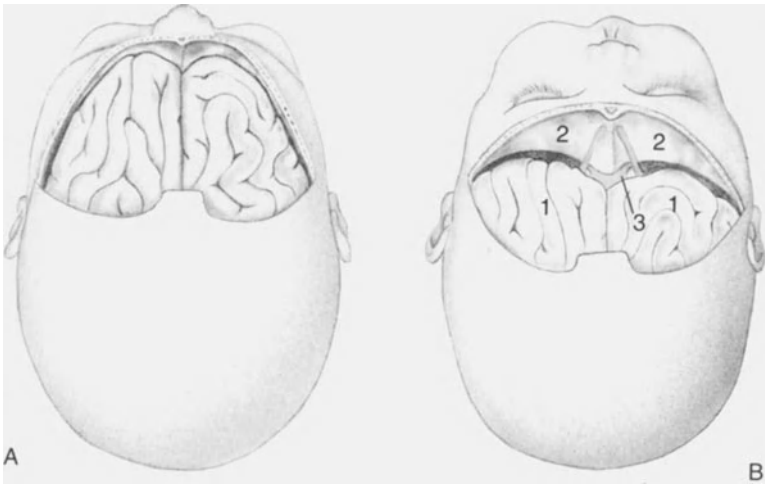
The three basic operating positions – prone, supine, lounging – refer only to the body, not the head. After the body has been positioned, the anesthesiologist finalizes arrangement of his tubes to assure himself easy access to the face. The head is then positioned on the body, by the surgeon, to permit access to intracranial areas and anatomical structures.

In conceptualizing the operative procedure the surgeon must "visualize" the lesion, or desired anatomical area, *within* the head for the operation: head extended on a supine body for performing a bifrontopterional craniotomy to expose the optic chiasm; head distracted and flexed at the craniovertebral junction with the body supine for exposure of the tentorial opening and pineal region; child's body in the lounging position with head distracted on C-1 and slightly flexed, for visualization of lesions in the superior cerebellar triangle; and so on.

This overview guarantees correct anatomical orientation. It is used consistently throughout this volume so as to transmit an holistic concept of positioning, structural anatomy, and surgical technique.

Supine Position

The head may be manipulated on the supine body for performance of frontal, bifrontopterional, craniofacial, parietal, biparietal, and temporal craniotomies as well as procedures demanding access to the craniocervicothoracoabdominal areas (such as for ventriculojugular or ventriculoperitoneal shunts). The transmaxillary ("face splitting"), transmandibular, retropharyngeal approach to the clivus and sphenoid sinus may be performed when the patient is supine and should be considered when dealing with lesions of or about the anterior foramen magnum, the clivus, and the sphenoid body. The pure pterional approach has no place in pediatric neurosurgery, since the extraordinarily rare aneurysm of the circle of Willis is best approached through a frontal, a bifrontal, or a frontotemporal craniotomy: the small size of the frontal lobes and very large basal cisterns allow for immediate and secure access to the circle of Willis.



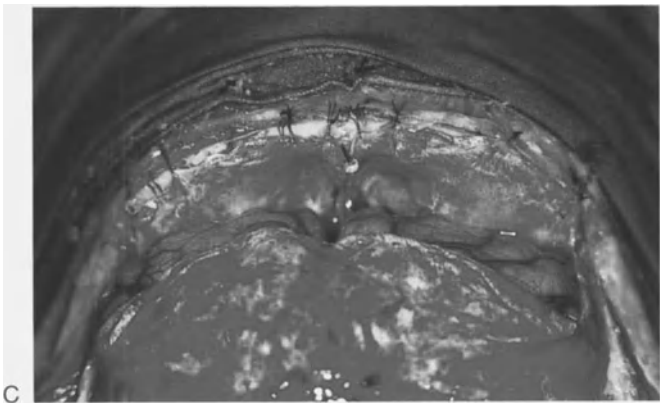


Figure 1.17. The falling away of the frontal lobes results from extending the head around its interauricular axis, *not* by lowering it beneath the level of the right atrium. On occasion, however, to facilitate CSF drainage for optimal exposure, especially if a preoperative shunt has not been inserted, it may become necessary to lower the head by manipulating the headholder or to pitch it inferiorly by manipulating the operating table. (A) The surgeon's view of the floor of the anterior fossa with the head in the neutral position. The *stippled area* indicates anterior fossa floor beneath the frontal lobes. (B) After the head has been rotated, extending it around its interauricular axis, the frontal lobes (1) fall posteroinferiorly, and the entire floor of the anterior fossa (2) with the chiasm (3) come into view. (C) The surgeon's line of vision of the chiasmatic region with the head in the neutral position.

Anterior Fossa and Parasellar Area: Frontal Craniotomies

The supine position is best used for access to the frontal lobes, the orbit, the optic foramina, the intraorbital and intracranial optic nerves, and the optic chiasm. Retrochiasmatic extension of such parasellar tumors as craniopharyngioma may be resected more effectively through the interopticocarotid space, with the child supine, by resorting to the use of "pitch" and "roll" to gain access to tumor lodged between optic nerves and chiasm superiorly, the internal carotid and posterior communicating arteries inferiorly, the basilar artery posteriorly, and the III cranial nerves inferolaterally on either side. The bifrontopterional craniotomy is ideal for this lesion.

When the patient's head and body are placed high and in the neutral position, the surgeon has a good line of sight for the skin opening, but visualization of the parasellar area is blocked by the frontal lobes (Fig. 1.17A). Consequently, slight extension of the head on the neck, as illustrated in Fig. 1.17B, allows for gravitational retraction of the frontal lobes, complete visualization of the roofs of the orbits and the lesser wings of the sphenoid, and as good an exposure of the region of the optic chiasm as one may possibly attain (Fig. 1.17C).

Since slight extension of the head makes for somewhat awkward hand positioning (Fig. 1.18A–E) for the scalp flap, one may simply elevate the head of the table for the skin incision and the posterior bur holes. The head of the table then is lowered to the desired level for placing the "keyhole" and glabellar bur holes, passing the Gigli saw and reflecting the osteoplastic bone flap, whether unilateral or bifrontopterional. The dural opening may be somewhat facilitated by lowering the head of the table approximately 50°. After retraction of the frontal lobe(s) and exposure of the roof(s) of the orbit, anterior clinoid(s), optic nerve(s) and chiasm, one may "roll" the operating table from right (Fig. 1.19A) to neutral (Fig. 1.19B), to left (Fig. 1.19C) so as to facilitate visualization of the lesion and parasellar anatomical structures.

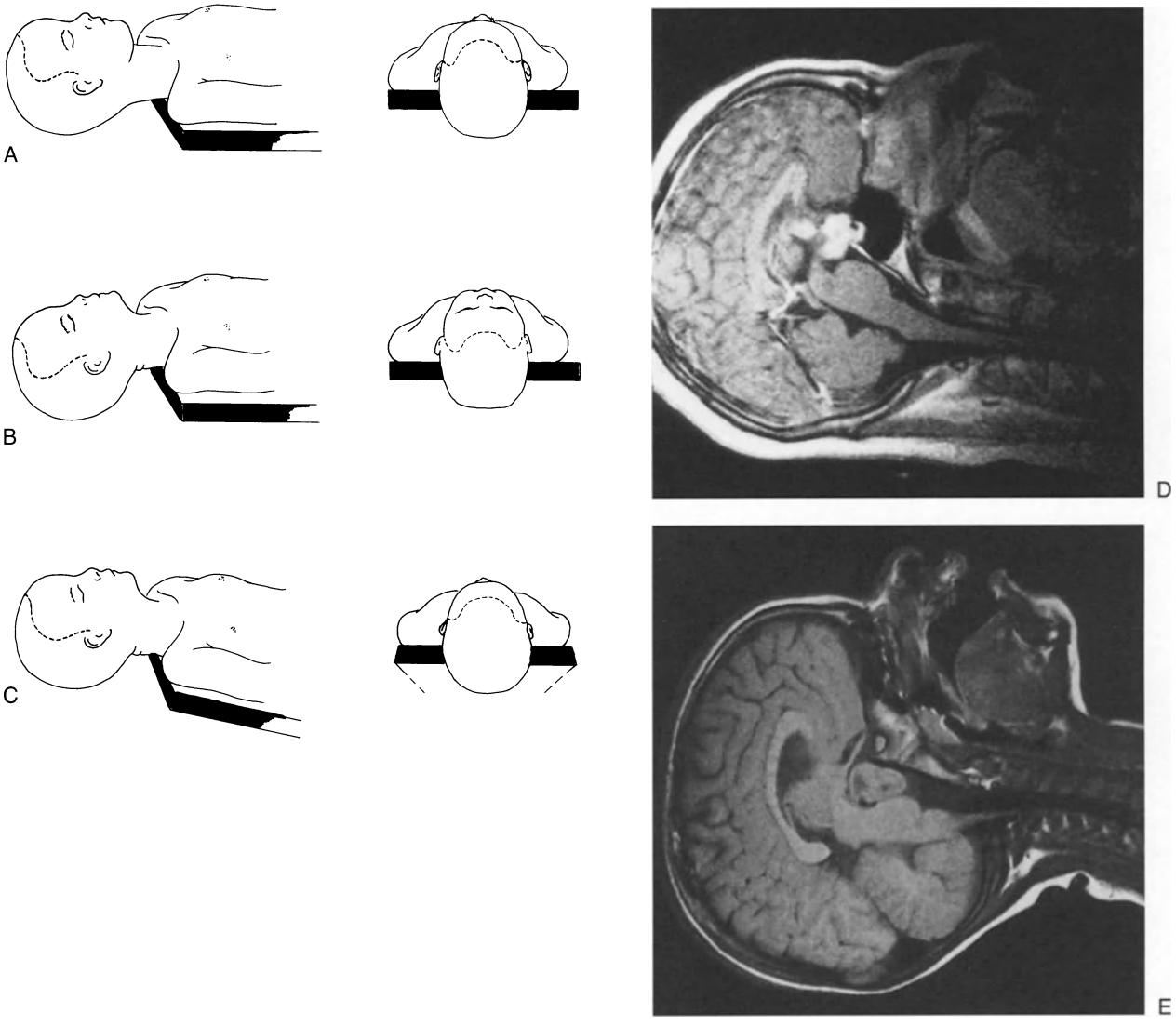
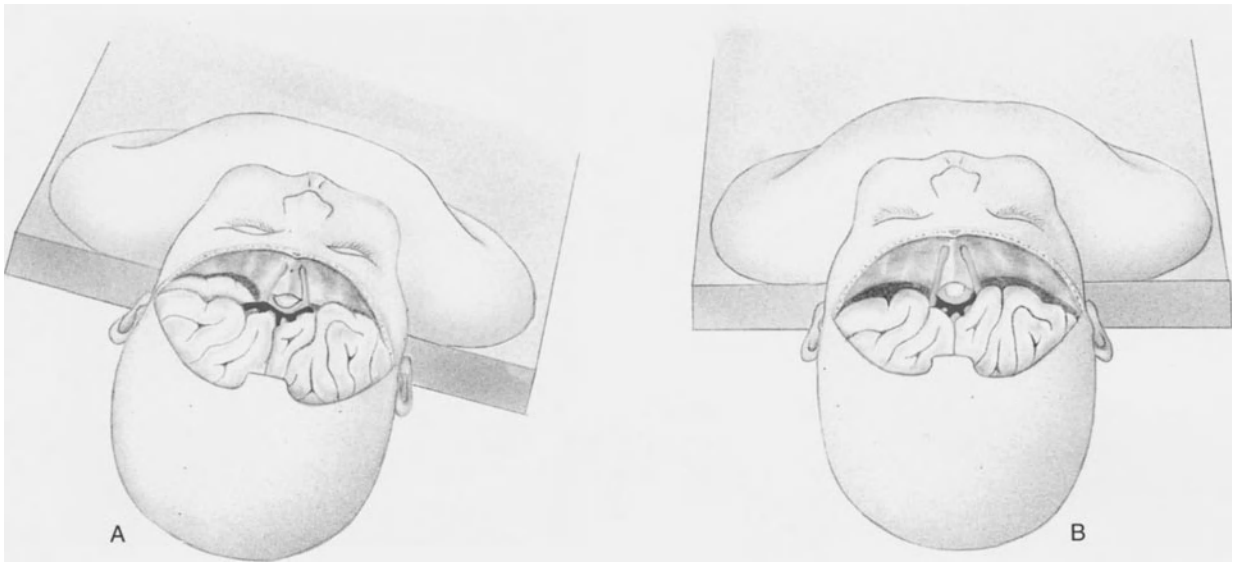
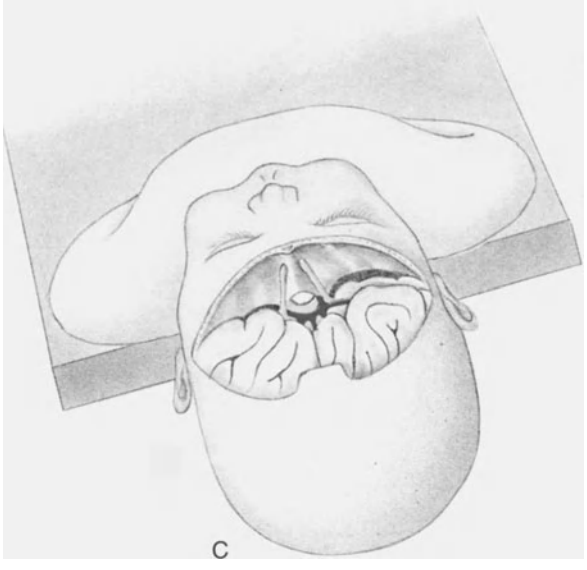


Figure 1.18. (A) Head in neutral position, (B) extended, and (C) extended but elevated using table pitch. The (D) suprasellar *craniopharyngioma* and (E) retrochiasmatic *hypothalamic hamartoma* extending inferiorly into the prepontine cistern are ideally approached through a bifrontal pterional flap with the child supine and the options to hold the head neutral, to extend it, or to elevate it in the extended position by using table pitch. In (D) and (E) intra III ventricular extensions of, respectively, a craniopharyngioma and an *hypothalamic glioma* are shown. Visualizing these two MRI studies as the tumors would be "seen" in (A), (B), and (C) helps considerably in formalizing tumor location/bone flap/skin incision. Though the details will be elaborated upon in the chapter on tumors, for this section it is important to recognize that *parasellar* tumors extending into the III ventricle may be resected very effectively through this approach by using the *1.* prechiasmatic, *2.* left interoptical carotid, *3.* right interoptical carotid, and *4.* translamina terminalis access routes. It is essential to open the lamina terminalis for the intra-III ventricular extension, but the parasellar locations require neurovascular dissections.





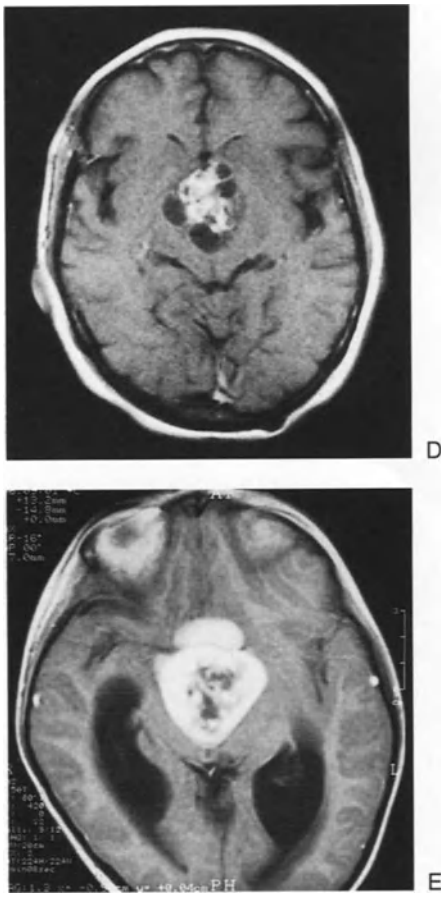


Figure 1.19. (A) Roll to the right permits visualization of the left sphenoid wing from the pterion to the anterior clinoid, internal carotid, and middle cerebral arteries after bifrontopterional craniotomy has been performed. (B) Neutral position is best for planum sphenoidale, optic nerves and chiasm, carotids, and anterior cerebrals. (C) Roll to left permits exposure of right homonyms to those illustrated in (A). (D) and (E) are MRI images of parasellar masses approached with the child in this position.

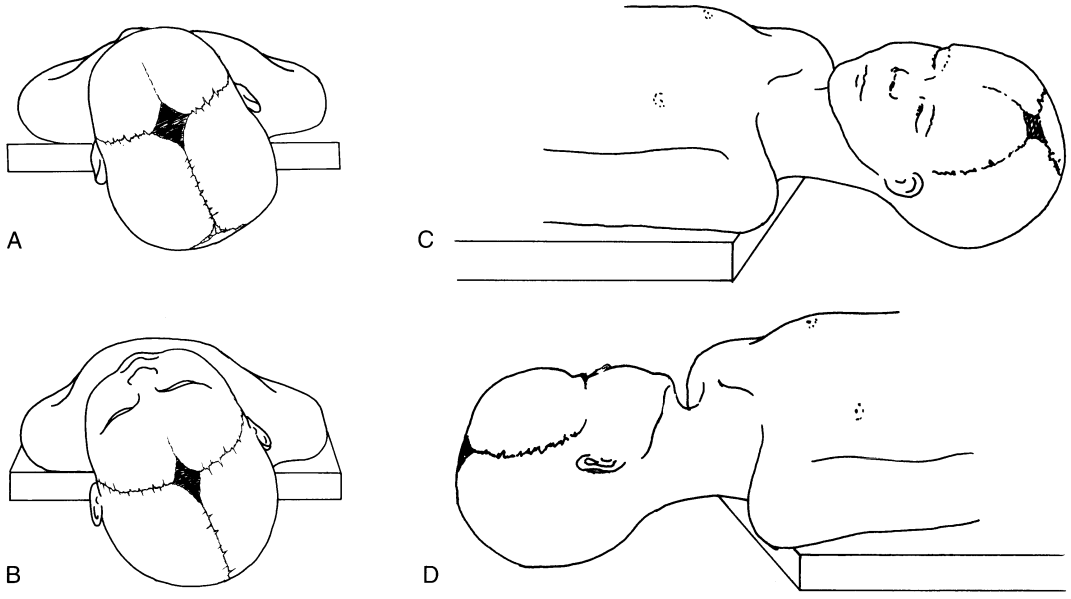
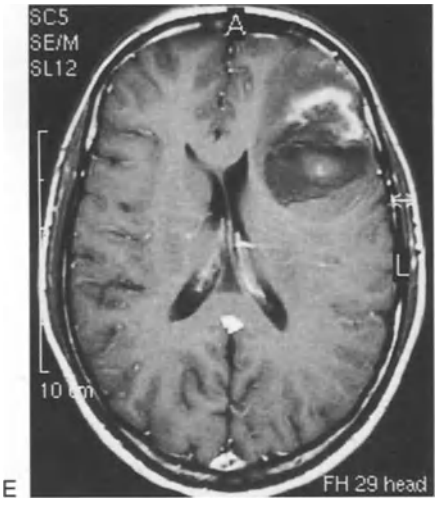


Figure 1.20. Unilateral frontoparietal craniotomy. The child's head is positioned for unilateral craniotomy, showing the frontal eminence, coronal, and sagittal sutures. (A) Coronal view of skull. (B) Surgeon's view of (A). (C) Lateral view of (A) from the left. (D) Lateral view of (A) from the right, showing pterional area. (E) *Prerolandic frontal lobe* tumors, whether limited entirely to the white matter surfacing along the sagittal plane, at the frontal pole, or surfacing at the convexity, are very conveniently resected with the child supine through a unilateral frontopterional craniotomy.



Unilateral Frontopterional Craniotomy

The head is extended on the body and rotated slightly (Fig. 1.20A–D) so as to allow the surgeon to visualize directly the frontal eminence of the side to be operated on. This represents the neutral position for a unilateral frontopterional craniotomy, one from which the surgeon may work comfortably to obtain access to the desired frontal and pterional cranial and intracranial areas simply by manipulating the table position.

Bifrontopterional Craniotomy

The head is extended from the neutral position (Fig. 1.21A) so that the surgeon's direct line of vision is at the metopic suture (Fig. 1.21B). Rotation of the head from side to side, by using body "roll" of the operating table, permits equal access to the posterior and inferior portions of the frontal bone on either side and, subsequently, to the sphenoid wings and anterior clinoids (pterional perspective). Care should be taken not to confuse extension of the head on the neck with lowering the head beneath the level of the shoulders! Extending the head entails movement at the craniocervical junction (Fig. 1.21C) and maintains the cervical spine in the normal anatomical position; lowering it entails extending the head and the neck, en bloc, at the cervicothoracic junction (Fig. 1.21D).

Craniofacial Procedures

The head is positioned on the body in identically the same way as for bifrontopterional craniotomies, since one must expose the entire frontal bone, both orbits, and the cribriform plate; there must also be ready access to the lateral rims of the orbits and the zygomatic arches. More accentuated extension of the head, with minimal lowering, brings the face into the surgeon's view (Fig. 1.22).

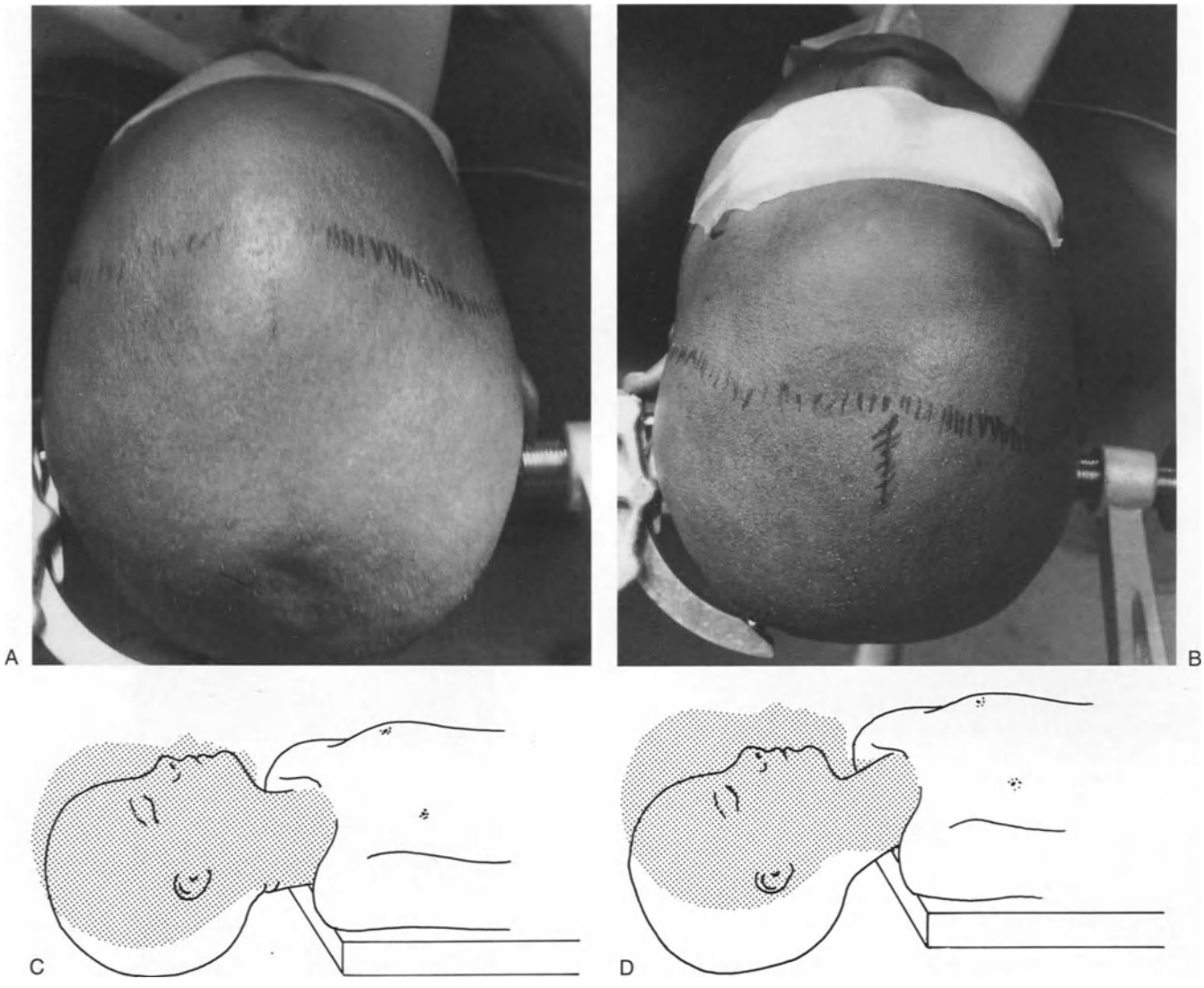


Figure 1.21. Bilateral frontopterional craniotomy, showing surgeon's view with (A) head neutral and (B) extended. (C) *Shaded area* illustrates the head in the neutral position, from which it may be extended to expose the anterior frontal area. (D) Position attained by lowering the head rather than extending it. Here, one may appreciate the differences between extending the head (C) and lowering it (D), with regard to movement at the craniocervical junction.

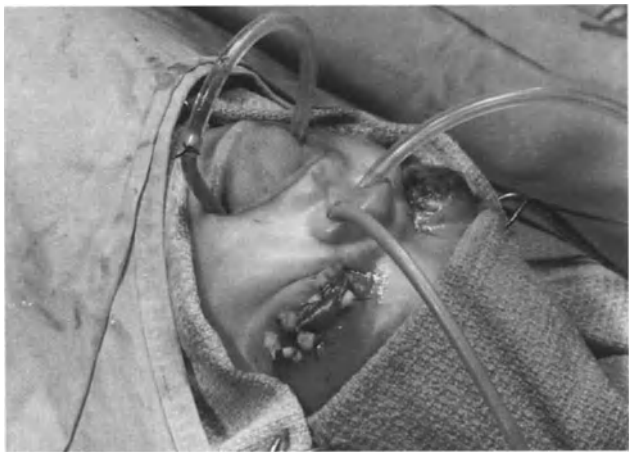


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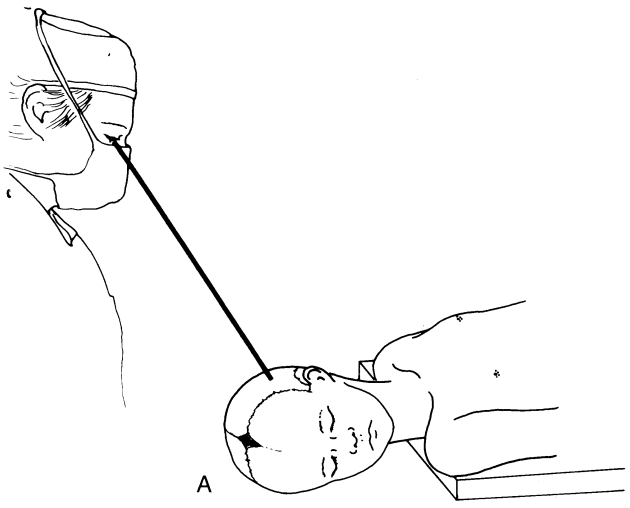
Figure 1.22. Exposure of the orbital and malar areas, which may be attained by extending the head.



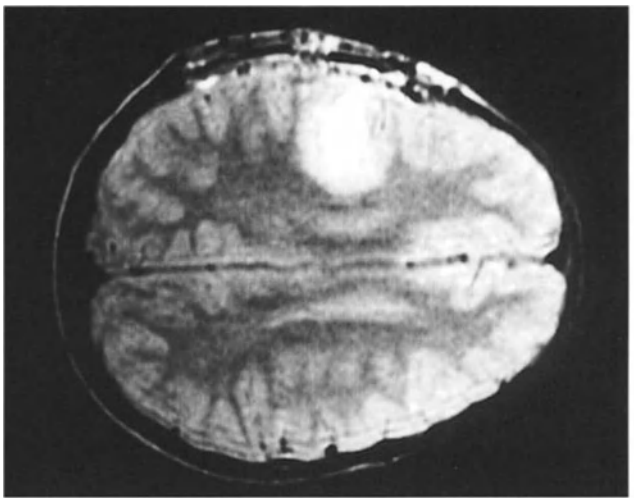


Figure 1.23. (A) The child is supine with the head flexed slightly after having been rotated so as to bring the sagittal suture parallel with the coronal plane of the temporal lobe, but not to the floor of the middle fossa. (B) This position permits a direct line of vision to the superior temporal line convexity of the hemisphere, putting into relief either the superior temporal convolution (by lowering slightly the head of the table) or the region of the frontal and parietal operculae (by raising slightly the head of the table). Hence, direct access to the posterior portion of the sylvian fissure, and after opening it, to the insula is afforded. This magnetic resonance image illustrates a *glioma* surfacing at Broca's area, which was successfully completely resected, with the child in this position without residual aphasia.

Parasagittal and Parietal Areas

The supine position also allows one to work effectively in the parasagittal and parietal areas for access to the superior sagittal sinus (SSS) either for head injuries or lowering the SSS in cases of chronic subdural collections of fluid (Chap. 13, p. 433). Parietal lobe and corpus callosum lesions, as well as those within the III ventricle region of the great vein of Galen, may be effectively approached with the child in this position. This is also a desirable position for placement of temporal, frontal, and parietal bur holes. However, access to the parietal-parasagittal areas obliges one to flex somewhat the head on the neck, at the craniovertebral junction, or to keep the head and neck in the neutral position and flex the cervical spine on the thoracic spine at the C-7–T-1 junction: raising the head.

Parietal Craniotomies

Positioning of the head for parietal craniotomy differs remarkably when one performs a unilateral or a bilateral procedure. In the former the head is rotated completely to one side and slightly extended on C-1, whereas in the latter the head remains in the anatomical plane but is slightly flexed on the neck. The unilateral parietal craniotomy position is also good for parieto-frontal parietotemporal, and parieto-occipital lesions.

Unilateral Parietal Craniotomy

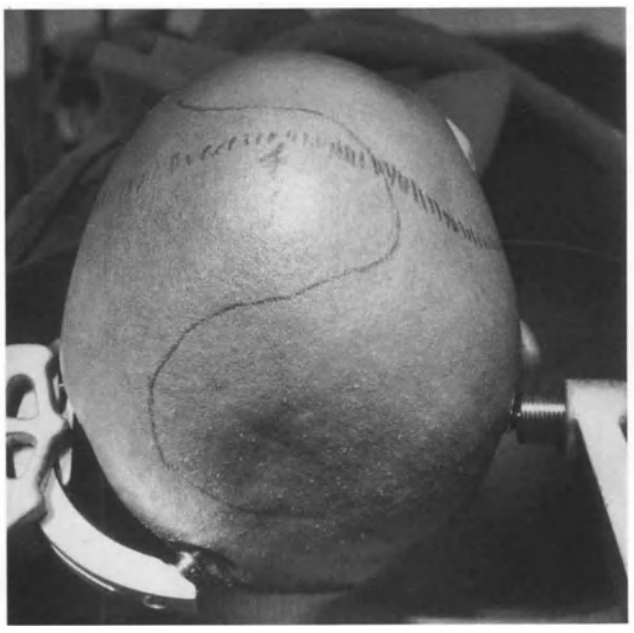
The head is rotated 90° so as to bring the operative side into the mid-sagittal plane of the body: the sagittal suture is positioned parallel to the coronal plane of the body. Flexing slightly the head on C-1 brings the parietal bone and superior temporal line into orthogonal planes, giving a direct line of sight to the superior temporal line (Fig. 1.23).

Biparietal Craniotomy

The head is flexed slightly on the neck and kept in the neutral position, with the surgeon having direct visualization of both the frontal and parietal eminences (Fig. 1.24A–C). This permits equal access to the sagittal sinus, both parietal bones and lobes, and, intracranially, the falx cerebri on either side down to the inferior longitudinal sinus and pericallosal cistern.

Convexity and Middle Fossa

By positioning the child supine and rotating the head a full 90° so as to bring the coronal plane of the head parallel to the sagittal plane of the body, one may expose completely the convexity of the skull and cerebrum. Flexion of the head on the cervical spine to approximately 5°, after it has been turned fully, places the lateral aspect of the calvarium in a plane perpendicular to the surgeon's line of vision. This provides the possibility of exposing the lateral aspects of the frontal, parietal,



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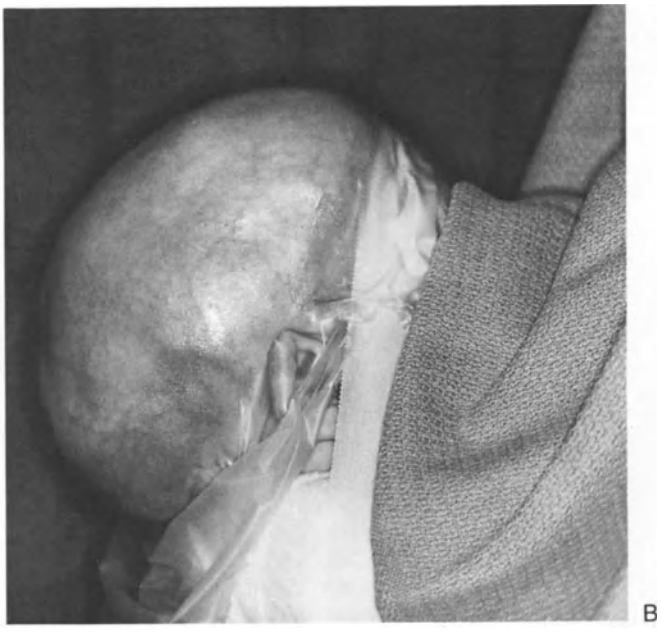

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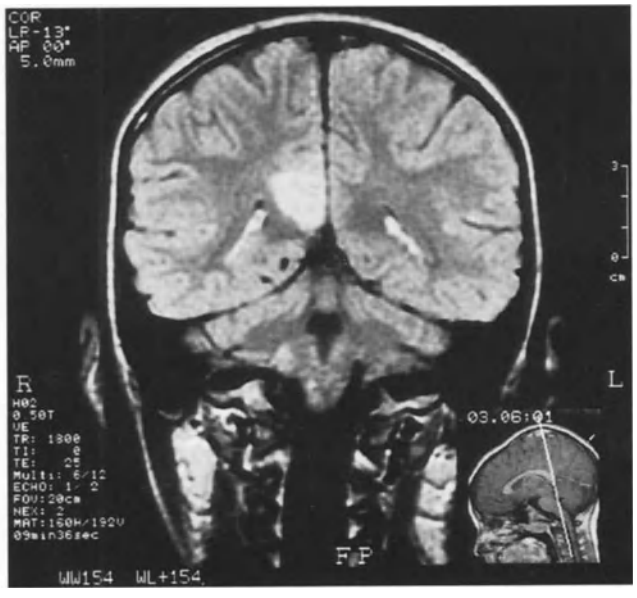

C

Figure 1.24. (A) Biparietal craniotomy. Slight flexion of the head on the neck rotates the superior surface of the skull into an excellent position for the "S"-shaped incision, and access to the frontal and parietal areas bilaterally. (B) A lateral photograph of an infant with the head in this position, illustrating exposure of the inion. (C) *Medial parietal lobe tumors*, especially those expanding within the cingulate gyrus superior to the corpus callosum or, as in this case, immediately posterior to the splenium, are approached with the child supine, sagittally, along the falx cerebri, keeping the sagittal plane of the skull along the sagittal plane of the body. Turning the skull to one side or the other obliges the surgeon either to elevate the parietal lobe or, less desirably, to operate on the recumbent site with the parietal lobe falling away with gravity: this puts undue stretch on the bridging cortical veins. Whether an S-shaped incision, as in this figure, or the horseshoe parietal incision, as illustrated in Fig. 2.15A,B, is used, is of importance only as an expression of tumor volume and vascularization.

temporal, and occipital lobes, as well as the transverse sinus and the tentorium as far medially as the tentorial edge and the ambient cistern. It is useful for approaching intraparenchymal lesions; masses within the frontal horn, body, temporal horn, or trigone of the lateral ventricle, and III ventricular tumors, either through the foramen of Monro or via the interval between the body of the fornix and the thalamus. It is the ideal position for performing either a ventriculojugular or a ventriculoperitoneal shunt (Fig. 1.25A–D).

Temporal Craniotomy

The positioning of the head for a temporal craniotomy is much the same as that for a unilateral parietal craniotomy (Fig. 1.24), with the exception that orthogonal planes of vision to the squamous temporal and greater wing of the sphenoid bones and the underlying temporal lobe necessitate lowering the head slightly (Fig. 1.26). This provides the surgeon a direct line of vision to the superior and inferior temporal lines, the zygomatic arch. Lowering the head facilitates access, respectively, to the sylvian fissure and the entirety of the tentorial surface and ring.

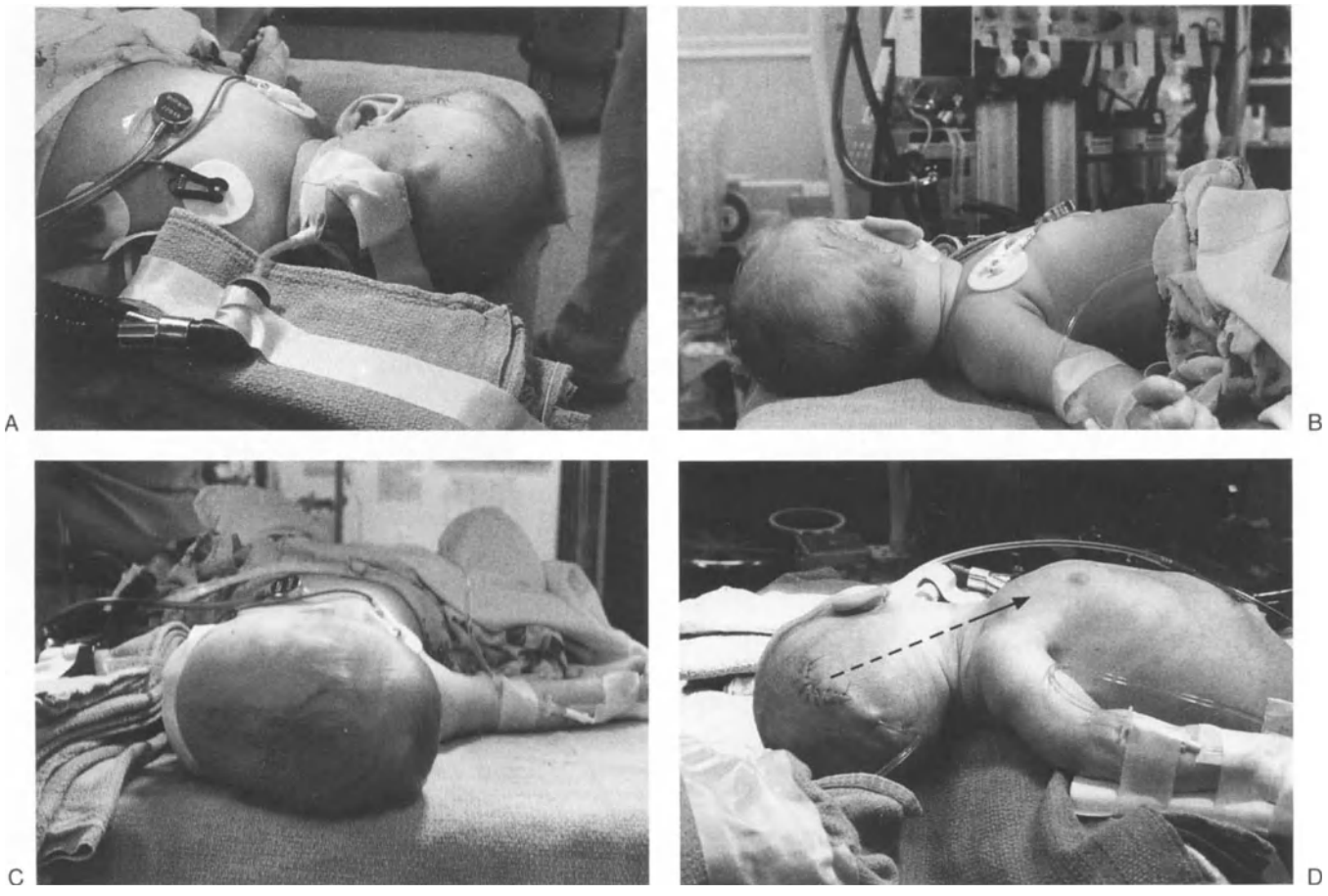

Figure 1.25. (A) Antero-oblique view. Supine child with head rotated 90° to the left, bringing the coronal plane of the skull parallel to the sagittal plane of the body. In this child, this position was used to remove a dermoid tumor, but it is ideal for a ventriculoperitoneal shunt (in which event, however, the head is lowered somewhat to put slight stretch on the neck as illustrated in (D) of this figure. (B) The same child and position as in (A), but viewed posteriorly to allow one to appreciate access to the parietal eminence. (C) The same child as in (A) and (B) viewed here from the vertex of the skull. (D) When a ventriculoperitoneal shunt is to be performed, a roll should be placed under the neck so as to "unfold" it, permitting easy passage of the supraclavicular area (*arrow*). In this child a shunt revision revealed ventriculitis, so it was converted to an external ventricular drain.

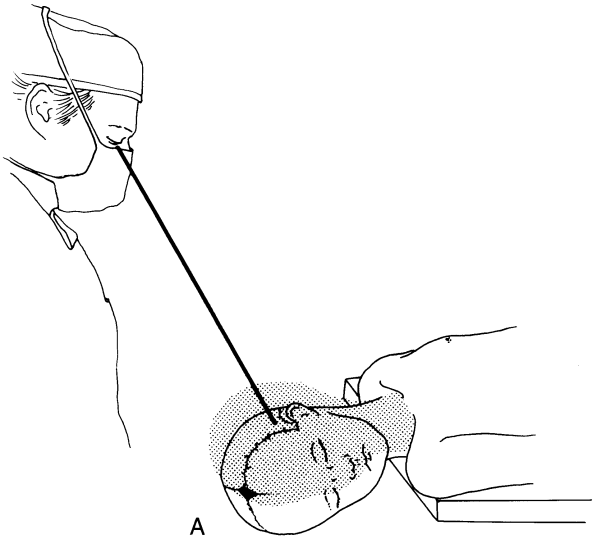

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Figure 1.26. (A) The head has been lowered slightly, approximately 15°–20°, giving the surgeon a direct line of vision to the squamous temporal area. (B) and (C) see p. 22.

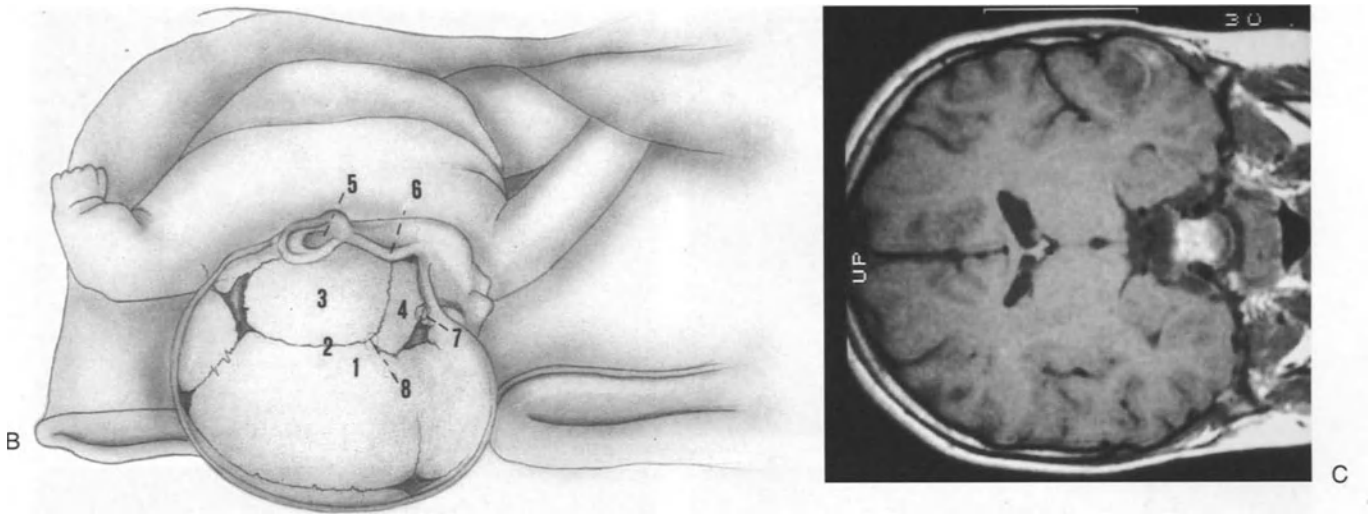


Figure 1.26. (B) Surgeon's view of the operative area. The bony landmarks are the superior (*1*) and inferior (*2*) temporal lines, the squamous temporal (*3*) and greater wing of the sphenoid (*4*) bones, the external auditory canal (*5*) and zygomatic arch (*6*), the "keyhole" area (*7*), and the pterion (*8*). (C) Though *superior temporal convolution gliomas* may be resected as indicated in Fig. 1.26B, middle (as in this case) and inferior temporal lobe masses require the supine child to have the head rotated 90° to one side and slightly lowered so as to bring the entirety of the squamous portion of the temporal bone into as perfectly and horizontal a plane as possible. Looking at this MRI study permits the surgeon to contemplate the implications of the resection, orienting himself from the sylvian fissure to the floor of the middle fossa.

Craniocervical and Thoracoabdominal Positioning for Ventriculojugular or Ventriculoperitoneal Shunts

Good access to the head, neck, thorax, and abdomen entails rotating the head 90° to one side, bringing the coronal plane of the skull parallel to the sagittal plane of the body, without flexing or extending the head (Fig. 1.27A). Placing a roll, or sandbag, under the shoulder blade on the side from which the head is rotated diminishes the degree of "in-axis" cervical rotation (Fig. 1.27B). This position permits access to the parietal eminence as well as visualization of the vertex and inion posteriorly (Fig. 1.27C), and to the glabella anteriorly (Fig. 1.27D). These landmarks are important for orientation purposes when planning to insert a catheter or the proximal portion of the shunting system into the lateral ventricle. It also permits dissection of the facial and jugular veins for placement of the distal end of a ventriculojugular shunt, and access to the abdomen for placement of the distal end of either ventriculogallbladder or ventriculoperitoneal shunts. One may also use this position for placement of a ventriculopleural shunt.

Prone Position

As in the supine position, if the head is kept neutral when the child is prone, the surgeon is obliged to lower it an inordinate (*dangerous*) distance in order to visualize directly those craniocerebral regions best exposed with the child prone: the occipital bone and the medial surfaces of the occipital lobes, the craniovertebral junction, inferior cerebellar triangle lesions, and masses within the inferior portion of the IV ventricle and the upper cervical cord. Cisterna magna lesions are also exposed to advantage with the child prone.

The surgeon's best view of the posterior parietal region is with the head in the neutral position. Exposure of the occipital lobes, inferior cerebellar triangle, and the craniovertebral junction necessitates lowering considerably the child's head and, thus, increasing intracranial venous pressure. In addition to this, the horizontal portion of the squamous occipital bone presents a visual obstacle, a ledge, separating the surgeon's line of sight from the craniovertebral junction. It puts the surgeon in an undesirable position for exposure and removal of upper cervical cord masses, decompression of the foramen magnum, and cisterna magna lesions extending into the region of the vallecula. If the head is distracted from C-1 and then flexed on it, exposure of the medial surfaces of the occipital lobes, the region of the torcular Herophili and transverse sinuses, the foramen magnum and craniovertebral junction, the cisterna magna and inferior cerebellar triangle, and the superior

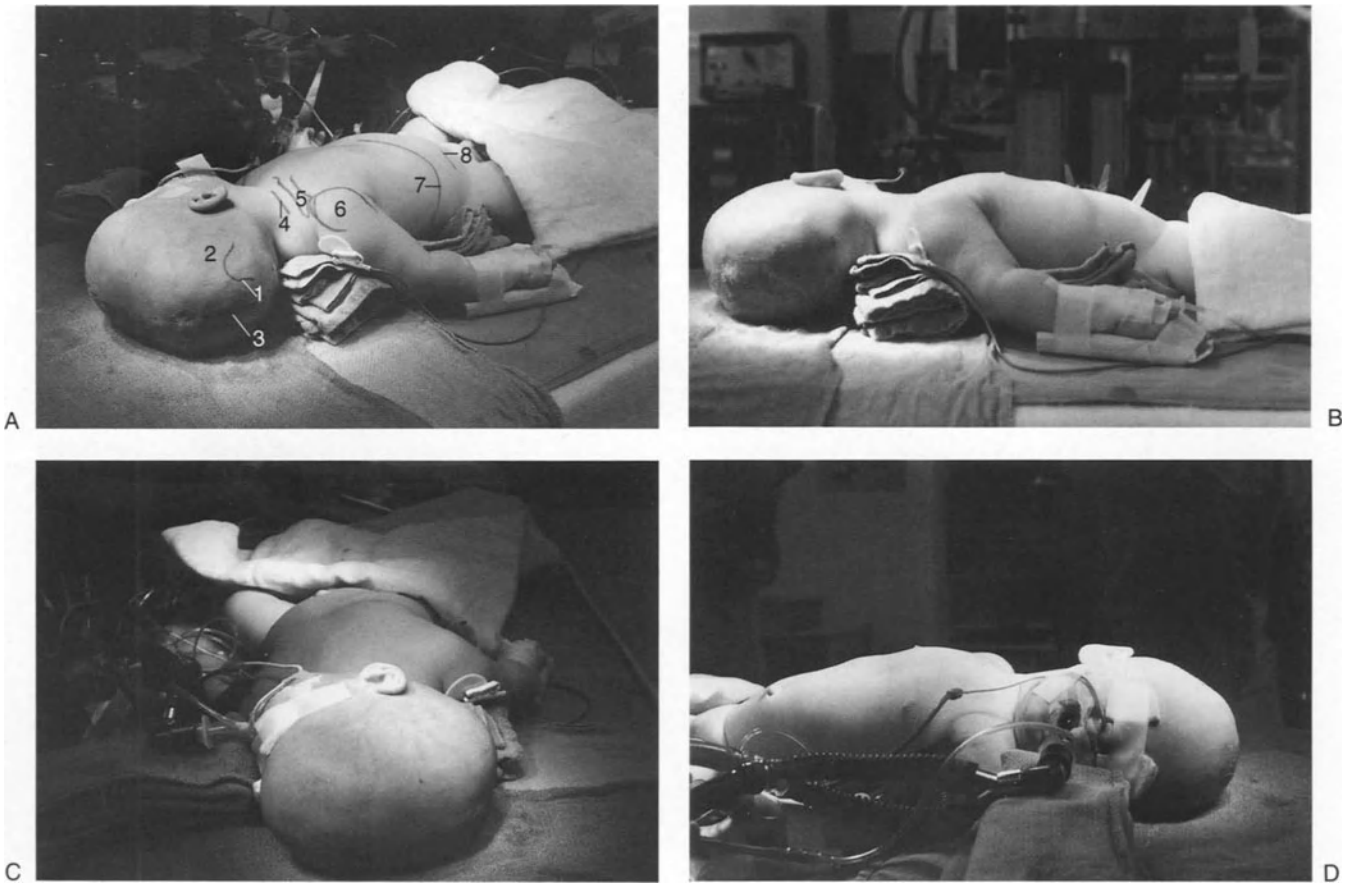


Figure 1.27. Various perspectives of positioning of a child for performance of a ventriculoperitoneal shunt. (A) The horseshoe scalp incision (*1*) is placed midway between the parietal eminence (*2*) and the midsagittal plane (*3*). The supraclavicular incision (*4*) is placed approximately 2.5 cm above the clavicle (*5*). The head of the humerus is drawn in (*6*) for orientation purposes, as is the costochondral arch (*7*). The abdominal incision (*8*) is made at McBurney's point. (B) The child as viewed from the posterior aspect of the skull, the perspective one should have when inserting the proximal end of the shunt into the ventricular system. Note towel under neck. (C) The surgeon's view of the head of the child, illustrating that the midsagittal plane of the skull is parallel to the coronal plane of the body. (D) The same child, viewed *en face*.

cervical cord, all come into a more direct line of vision and, subsequently, may be operated on more effectively.

It is important to consider in detail the anatomy of posterior fossa lesions when deciding whether to operate with the child in the prone or lounging positions.

Factors other than pooling of blood within the posterior fossa warrant consideration if one operates on the child prone. Since the surgeon has no choice but to work standing at the head of the patient, *it is impossible for him to position himself so as to have a direct line of vision to the region of the superior cerebellar triangle, the supracerebellar and quadrigeminal cisterns, the pineal gland, and the posterior portion of the III ventricle, when the child is prone* (Fig. 1.28A–C). Although he may have an adequate line of vision of the inferior portion of the IV ventricle and the vermis (from the fastigium inferiorly to the pyramis and then anterosuperiorly to the nodulus), he is in no position to deal effectively with superior draining veins going to the transverse sinuses and tentorium. Flexing the head upon the neck at the craniovertebral junction, and the neck on the thorax at the cervicothoracic junction, may increase somewhat the surgeon's visualization of the IV ventricle and posterior surface of the transverse sinuses. It offers only partial visualization of the superior cerebellar triangle and, if pushed to the extreme, increases prohibitively intracerebral venous pressure.

In order to visualize the superior cerebellar veins and the structures within the superior cerebellar triangle, one must place oneself so that one's line of vision centers along a plane running 45° from the horizon (Fig. 1.28D). Consequently, the decision to operate on the posterior fossa with the child prone or lounging should not be one of preference of the surgeon, but one predicated entirely upon the location of the lesion which must be dealt with!

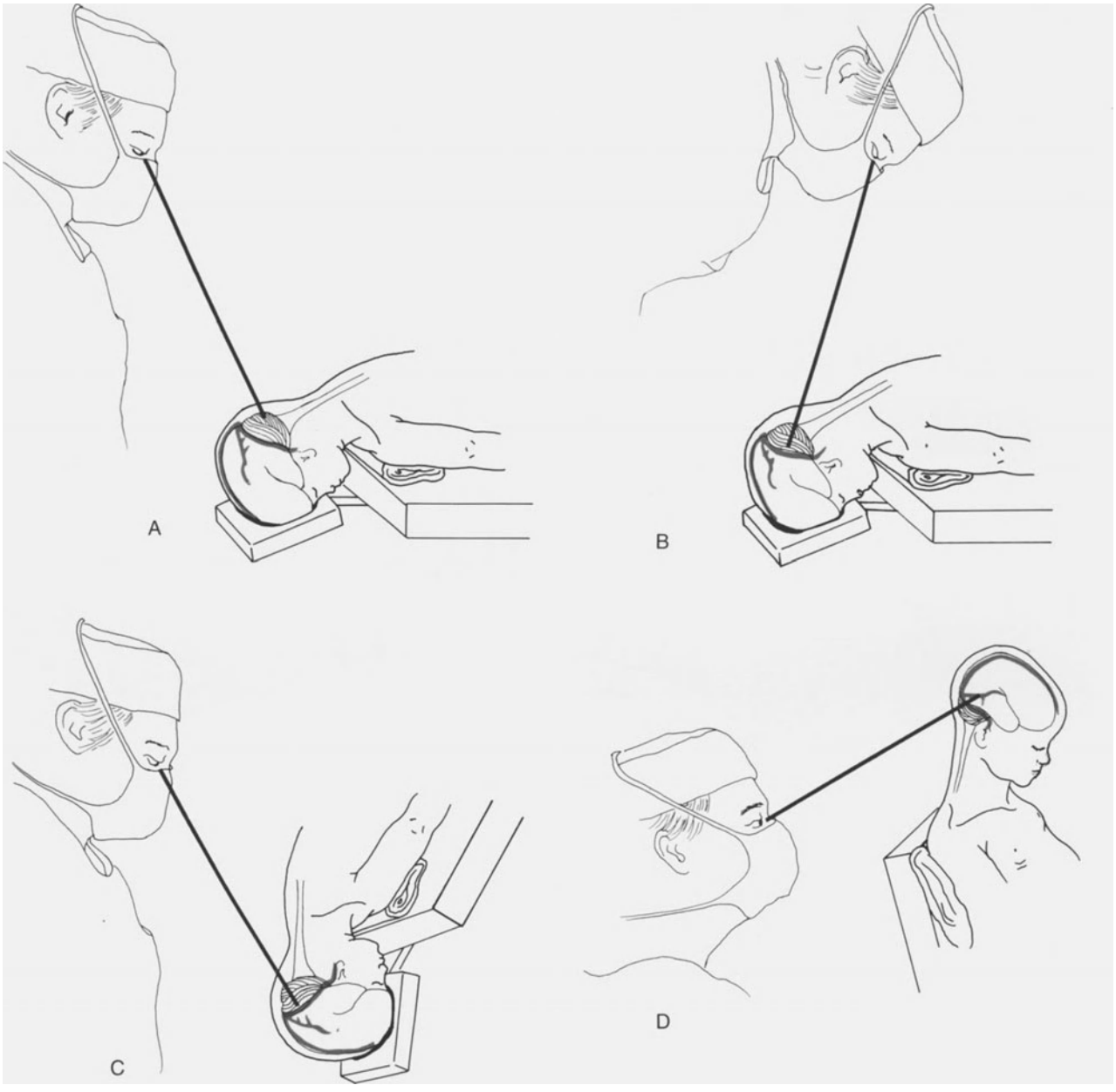


Figure 1.28. With the child prone (A), the surgeon cannot position himself to view directly the superior cerebellar triangle contents, the superior IV ventricle, or the aqueduct because he is obliged to work from the head of the patient. He can, however, and with excellent illumination, see these structures well through the operating microscope. The difficulties are positioning of the surgeon's hands to hold the instruments naturally: extreme wrist flexion and hand supination are required, limiting range of motion and predisposing to fatique. In order to obtain a line of vision to the tentorial opening, without the operating microscope, the surgeon would have to elevate his head and move it caudad (B), or bring the child so low as to have it almost in a "headstand" position (C). (D) In order to view completely the structures within the posterior fossa, the surgeon should place the child in the lounging position and place himself so that his line of vision is 45° from the horizontal.

In 1974 Meridy and coworkers [1] reported complications occurring during neurosurgical procedures with the child in the prone position. They noted 8% incidence of cardiac arrhythmias, 3% incidence of respiratory complications (with one death), 2% incidence of cardiac arrest (with two deaths), and 1.6% incidence of air emboli. They state that "many anesthetists and neurosurgeons advocate the sitting position for posterior fossa exploration. Such positioning provides an excellent view of the posterior cranial fossa with the operative site situated at the surgeon's eye level. In this position gravity effectively drains spinal fluid and blood from the operative wound. It also facilitates venous return to the heart, relieving intracranial venous stagnation, and so controlling venous pressure and ooze and ultimately brain swelling". They also state "many anesthetists and surgeons are fully aware of the disadvantages of venous air embolism, cardiovascular instability leading to systemic hypotension and diminished cerebral blood flow, the possibility of a patient sliding down the table during operation and difficulty with temperature control." Analysis of their results, comparing them to work published by others, especially Michenfelder, *reveals that there is no difference in the incidence of air embolism or hypothermia in the two groups*, with the work reported by Michenfelder et al. [2] including over 2000 patient studies.

The most experienced posterior fossa [3–8] neurosurgeons prefer the lounging position. Bucy [9] stated, "For many years I operated in the posterior fossa with the patient lying prone and with the long axis of the trunk forming an angle of approximately 40° with the floor. I am now convinced that the sitting position is superior to this and less hazardous. Most of the risks of this position, principally those of air embolism and of arterial hypotension, can be avoided with care and are more than adequately compensated for by the advantages."

Occipital Craniotomy

The head is positioned in the same manner for both midline and lateral occipital craniotomies, mainly by flexing it approximately 10° while it is being distracted. Placing small rolls under the shoulders (humeral heads) on either side takes pressure from the chest and elevates the thorax enough so that the head may be slightly flexed without bringing pressure onto the endotracheal tube. Ideal positioning of the head entails flexing it to the point where the surgeon has a direct line of vision to both the vertex and the inion, as well as the parietal eminences bilaterally (Fig. 1.29).

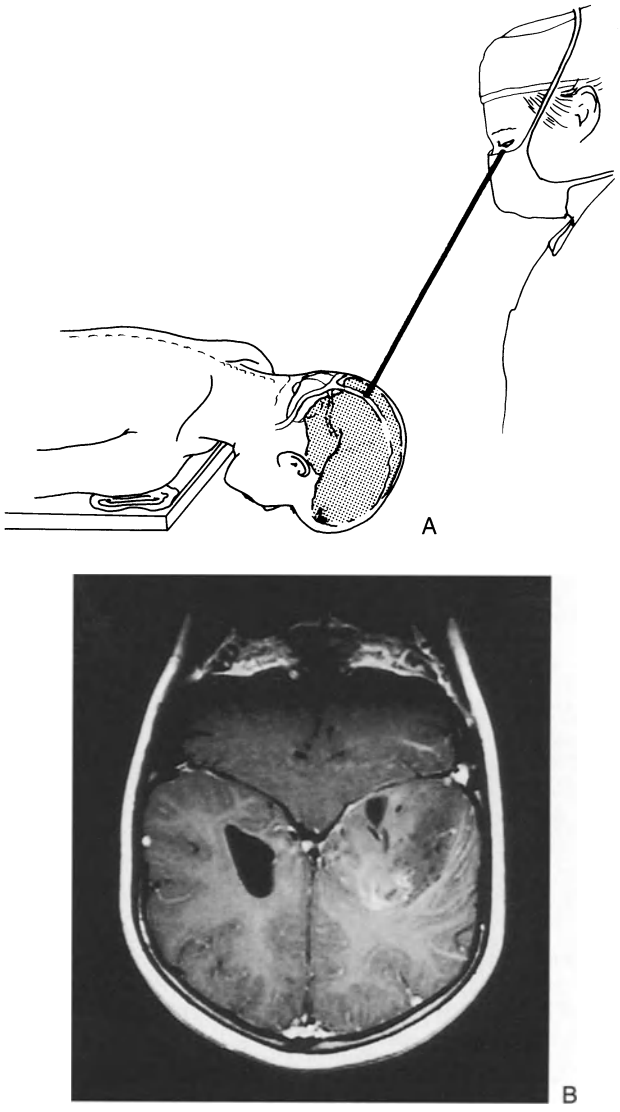


Figure 1.29. (A) Prone position for occipital craniotomy: The head is distracted and then slightly flexed, permitting direct visualization of the inion and both parietal eminences. (B) This child's occipital pole lesion rests upon (and invades) the tentorium. Its resection requires equal access to the medial (superior sagittal/transverse) and lateral (transverse/sigmoid) sinuses.

Suboccipital (Posterior Fossa) Craniotomy

The head is distracted maximally, flexed on the atlas, and lowered so as to verticalize the horizontal portion of the squamous occipital bones surrounding the foramen magnum, and to separate maximally the foramen magnum from the atlas so that this latter structure does not slip into the posterior fossa (Fig. 1.30), to permit the surgeon as cephalad a line of vision as possible (Fig. 1.31). Unfortunately, hanging the head maximally from the trunk does not increase cephalad exposure of the posterior fossa contents. It does increase unacceptably the intracranial venous pressure.

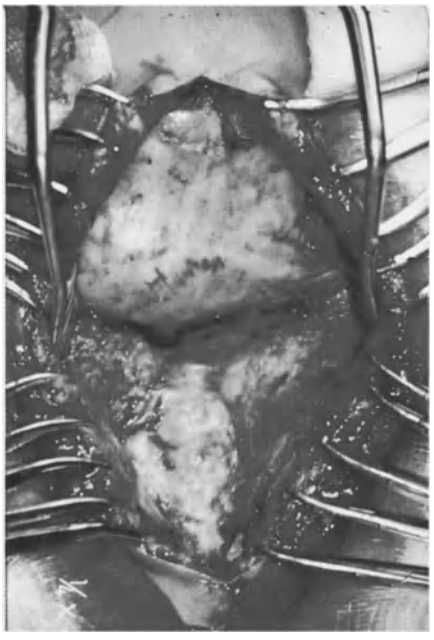


Figure 1.30. The head was not adequately distracted from C-1, so that the latter has come to rest within the foramen magnum. Distracting the head so as to put the atlanto-occipital membrane on the stretch, and to maximize the space between C-1 and the rim of the foramen magnum, provides ideal exposure of the osseous and vascular structures at the foramen magnum.

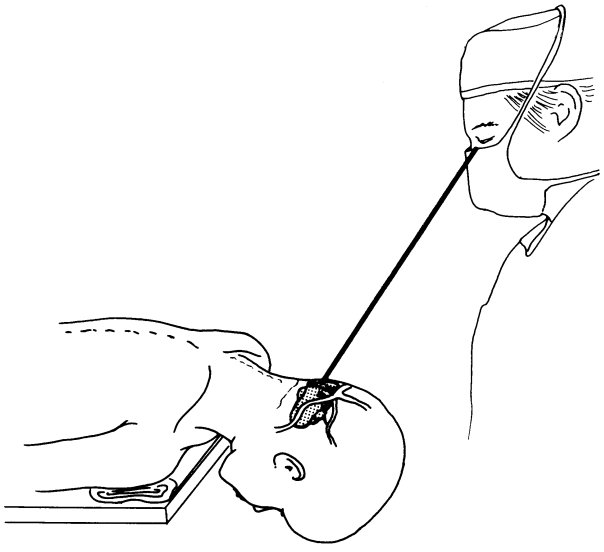


Figure 1.31. The prone position for suboccipital craniotomy, and exposure of the inferior cerebellar triangle, with head distraction, separating widely C-1 from the foramen magnum, provides a maximum view of inferior cerebellar triangle contents.

Laminotomy

The prone position is ideal for cervical, thoracic, and lumbar laminotomies.

Cervical Laminotomy

Maximum exposure of the cervical spine and spinal cord is obtained by positioning the child in an identical manner as for suboccipital craniotomy (Fig. 1.31). This brings the inion into a position such that it does not obstruct the surgeon's line of vision to the atlas, permits entry into the posterior fossa if needed, and gives a very complete and direct view of the entire cervical cord.

Thoracic Laminotomy

For upper thoracic laminotomies it is best to place the head in a neutral position, distracting it only slightly, but not turning it to either side, whereas for mid- and lower-thoracic laminotomies the head may safely be turned to either side, depending exclusively upon the preference of the surgeon and the anesthesiologist. Turning of the head to one side or the other with the child prone rotates the cervical vertebrae on one another so that one can encounter rotation of C-7 on T-1. This is the reason for distracting the head and keeping it in a neutral position for upper thoracic laminotomy.

Lumbar Laminotomy

It is very likely that lumbar laminotomy requires the simplest positioning of any neurosurgical procedure. The head may be turned to either side and the child need only be placed prone with rolls or pillows beneath the shoulders (in children of all ages), and beneath the shoulders and iliac crests (in toddlers, juveniles, and adolescents) (Fig. 1.32).

Lounging Position

The head should be distracted and flexed upon the atlas for both midline and lateral suboccipital craniotomies. The height of the table is then set so as to allow the surgeon a direct horizontal line of vision for making the skin incision (Fig. 1.33A,B) and dissecting the muscles from the skull and atlas, but the setting should allow the table to be elevated when the craniotomy is performed.

To avoid tilting the table forward so as to view adequately the dura mater first and then the superior cere-

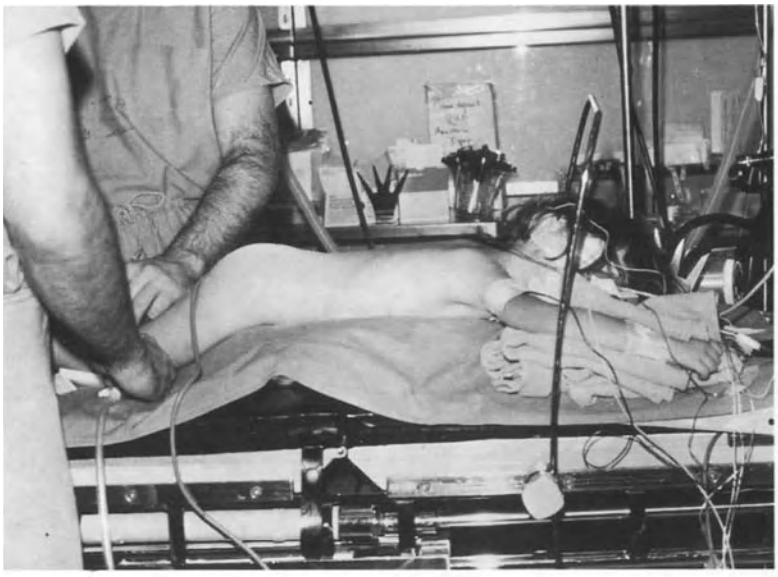
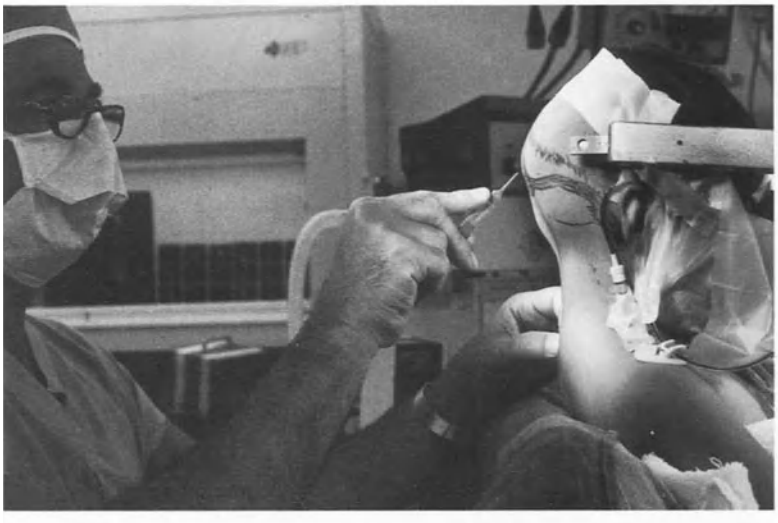


Figure 1.32. Prone position for lumbar laminotomy, thoracic costotransversectomy, or lumbar posterolateral approach for hourglass neuromass extending from the spinal canal into the retropleural or retroperitoneal spaces.



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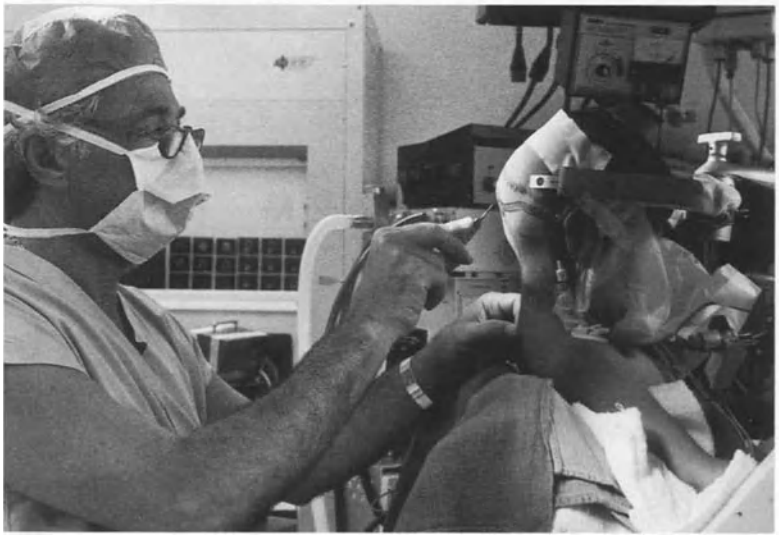


B

Figure 1.33. (A) The table is at a height such that the line of vision is 8°–10° downward to permit viewing the entire skin incision. The arm, forearm, and hand are all in the neutral position, permitting maximum strength and range of movement. (B) This is a cone-down view of the hand in the neutral position, illustrating the potential range of motion from pronation to supination, from flexion to extension.

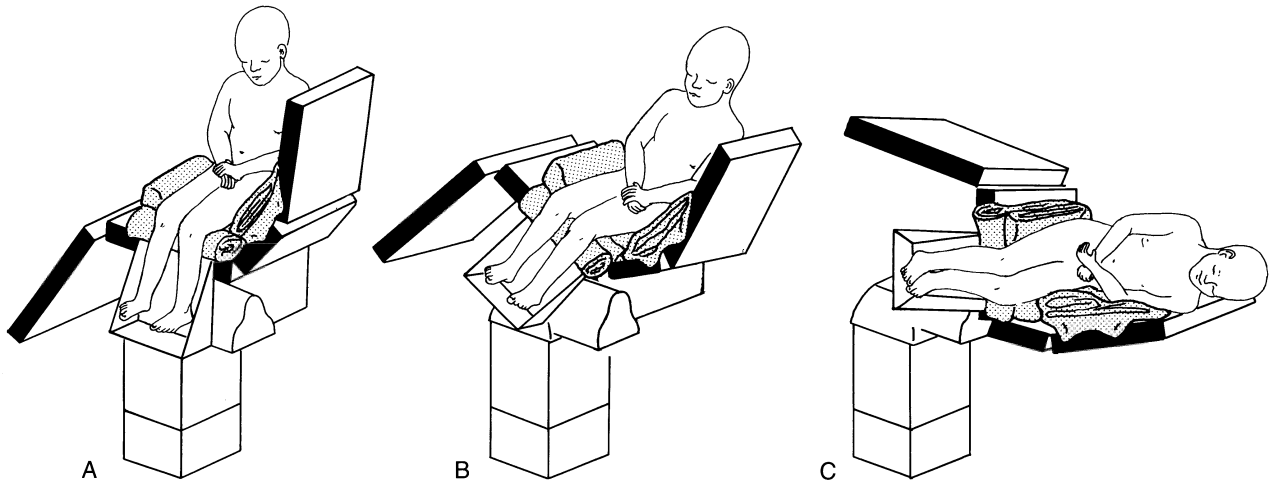


Figure 1.34. (A) The child is positioned sideways, with the vertical axis of its body perpendicular to the long axis of the table, so that the arms are resting on the backrest and the legs are extended perpendicular to the table. Then, pushing the table downward (B) brings the child into the horizontal position (C), without obliging the surgeon to stop the operative procedure. *This illustration is of purely historic value.* This procedure was suggested to permit rapid horizontalization of the patient, who had been positioned "sitting" in the event air emboli developed. Sitting erect, hanging legs are not acceptable at this time in the evolution of neurosurgery.

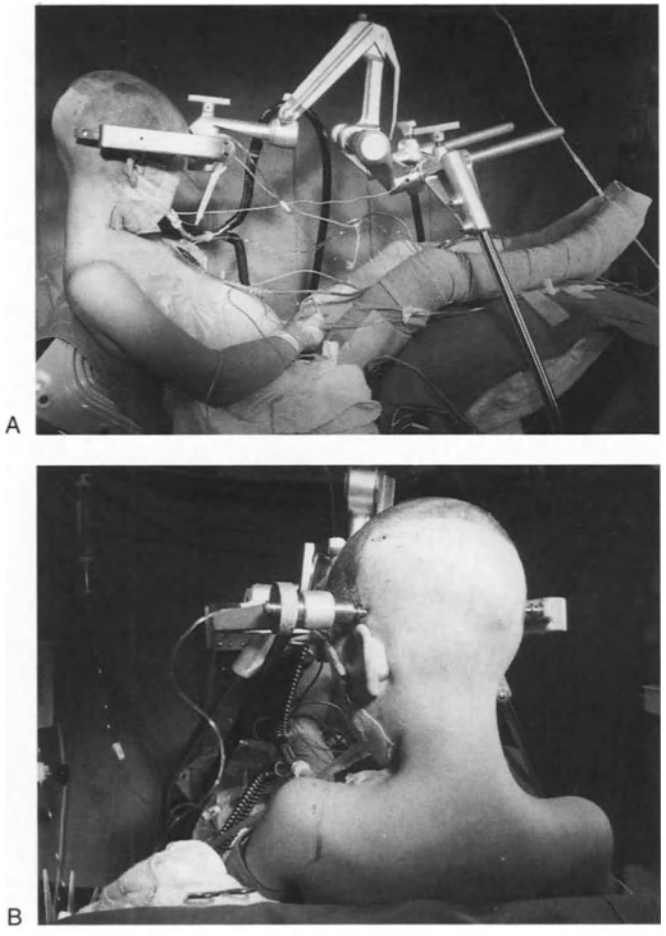
bellar triangle, the surgeon may work sitting. He may lower his chair to augment superior angulation of his line of sight, to bring him comfortably along the tentorial surface of the cerebellum, into the tentorial opening and pineal region. Also, convenient regulation of the surgeon's working height with the operating table, raising or lowering it, adds a significant amount of security to control of the Hudson brace for either perforator or bur use.

Unfortunately, one often speaks of the "*sitting*" position when, in fact, practically no neurosurgeon uses the *sitting* position. Rather, the patient, adult or child, is, in fact, put in the "*lounging*" position. This diminishes greatly the number of complications previously observed with the patient in the sitting position. These include significant diminution of cerebral blood flow, hypotension, and air emboli. Controlled ventilation has resulted in further diminution of the incidence of air emboli and cardiac arrythmias, as have the routine insertion of central venous catheters. In fact, Marshall [10] reported that the incidence of air embolism dropped from 15% to nil when positive/negative ventilation was used. Michenfelder [9] and coworkers [2] reported only a 2% incidence of air emboli in 2002 neurosurgical procedures performed on patients who were positioned "upright." They also noted a significant difference in air emboli in those patients positioned "upright" for cervical laminectomy and temporal craniectomy (less than 0.1%) when compared to those in the same position for suboccipital craniotomy (approximately 2%). When Michenfelder and his associates [11] used the Doppler, they observed that the percentage of "air emboli diagnosed" rose to 6%, although the incidence of clincally significant air emboli did not change. It was Michenfelder's conclusion, consequently, that the Doppler diagnoses incidences of air embolism that would never become clinically significant complications, and that the "threat of air embolism is not sufficient to contraindicate operating on the patient in the sitting position". In his entire series, he observed only 53 patients in whom air embolism was diagnosed. The only death in his series was unrelated to air embolism. Michenfelder's "upright" position is a *semireclining* (lounging) posture.

An historically interesting variant of the "sitting" position was reported by Garcia-Bengochea and coworkers [12]. In brief, it consists of positioning the patient sitting, but seated sideways on the operating table (Fig. 1.34). Lowering of the table in the event of an air embolus, or other intraoperative complication necessitating positioning the patient horizontally, was easily and immediately carried out.

The lounging position may minimize, not eliminate, the theoretical disadvantages of the "sitting" position.

The child is placed horizontal, flexing the elevated calves upon the thighs, and the trunk at the hips, as illustrated in Fig. 1.35. This position is used whenever one wishes to have access to a midline or lateral posterior fossa area. It is easier to place a child, especially an infant, in the lounging position than it is to sit the child up, since one need only distract and flex the head on the neck at C-1, place a pillow or sandbag at the thoracolumbar area, and center a pillow at the popliteal fossae. Though an upper cervical laminotomy may be per-



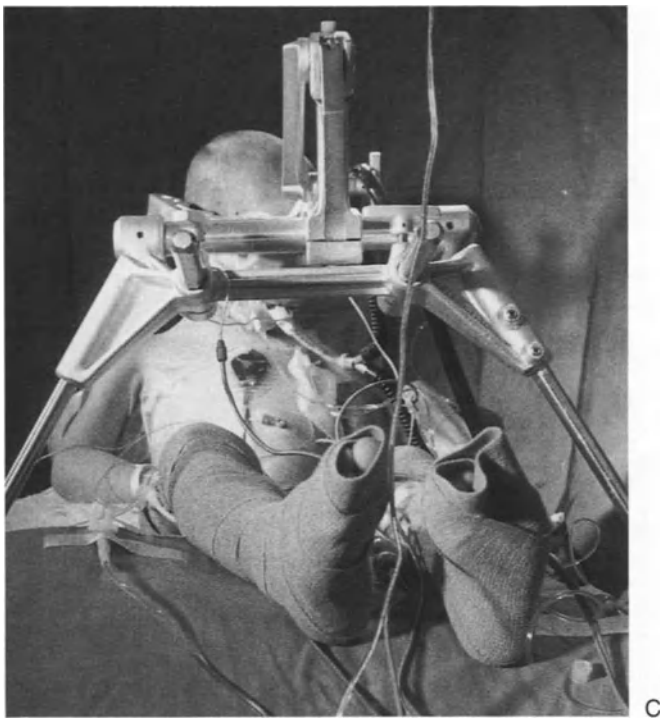


Figure 1.35. (A) The table is pitched backward, bringing the knees in the same plane as the shoulders. (B) Turning the head slightly on the cervical spine exposes the lateral occipital bone for cerebellar hemisphere or pontocerebellar angle lesions. (C) The anesthesiologist's view.

formed with the child in the lounging position, it really is not advisable, since it offers no advantages over the prone position. When the child is in the lounging position and the surgeon seated in a mechanical chair, rotation of the operating table around its axis cocks the head forward. This permits better visualization of the superior cerebellar triangle but does put the child into a sitting position (Fig. 1.36A–D). As the child is rotated forward, the surgeon must both elevate his chair and extend his arms. Two very negative disadvantages result: (1) the child is brought into a sitting position and (2) the surgeon works in a tiring position. It is preferable to elevate the operating table and or lower the surgeon's chair (Fig. 1.37).

This position is ideal for occipital and suboccipital craniotomies (whether midline or lateral), bur holes (unilateral or bilateral, diagnostic or therapeutic), and mid- or lower-cervical shunts (whether shunting from occipital or frontal horns). It is not recommended for upper cervical laminotomy, even if the surgeon suspects that it may be necessary for him to enter the posterior fossa: the prone position is simpler and permits adequate visualization of the craniovertebral junction. The lounging position is considered feasible for mid- and lower-cervical laminotomy only when one expects to encounter either an arteriovenous malformation of the cervical cord or an intramedullary tumor, which may bleed considerably (Figs. 1.38–1.40).

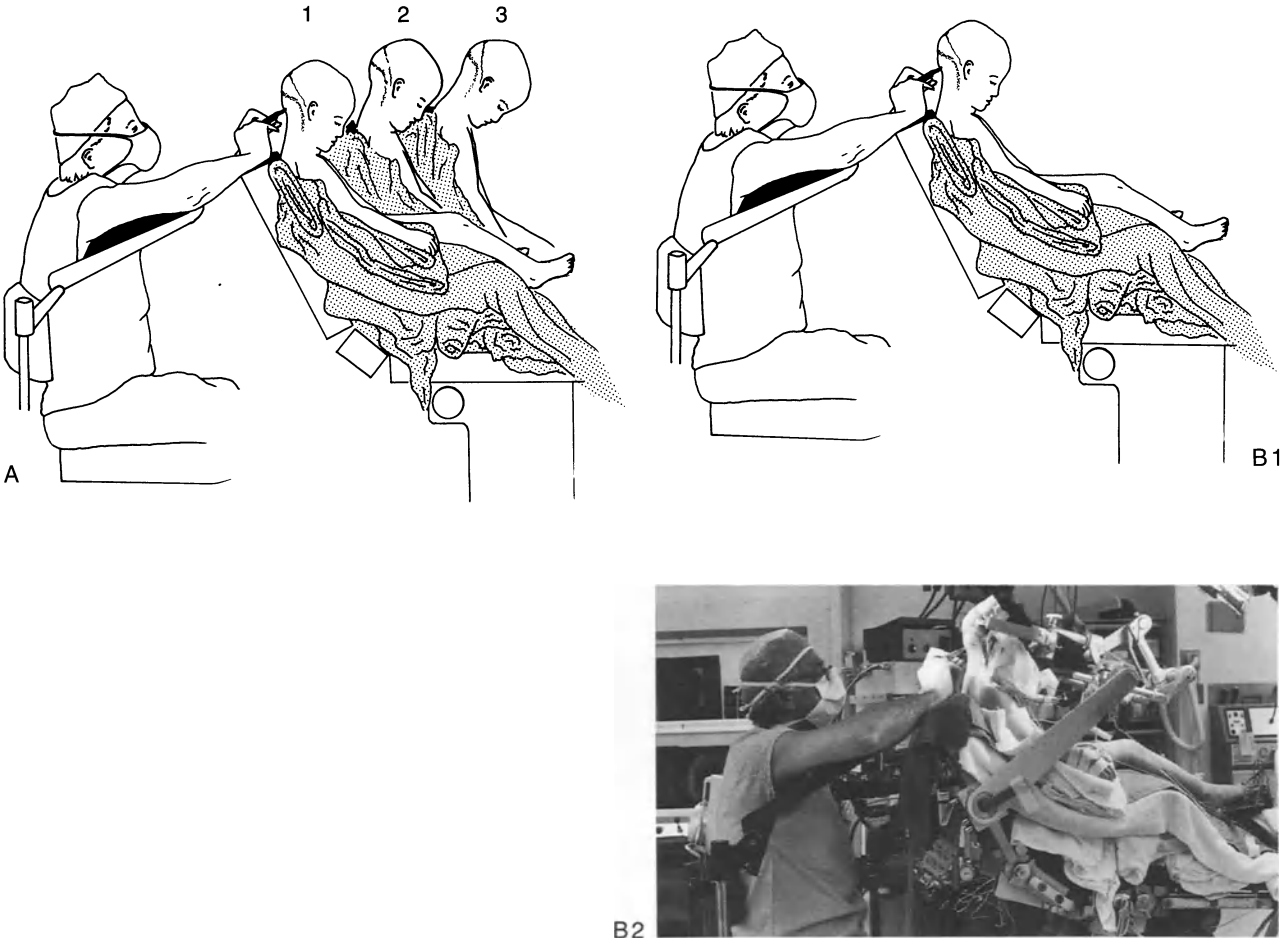


Figure 1.36. The surgeon is seated, his arms on a rest, for skin incision, muscle dissection, craniotomy, and inferior cerebellar triangle work (*1*). Pitching the operating table forward permits access to the IV ventricle and aqueduct (*2*), and superior cerebellar triangle (*3*). Note that, as this is done, the child's head is progressively elevated and its body is verticalized. *This is to be avoided!* The above legend, the drawings in (A), (B1), (C1), (D1), as well as the accompanying photographs (B2), (C2), and (D2) appeared as herein represented in the first edition of this volume. The reader will surely have recognized that as the child is rotated around the central axis of the table, it is brought increasingly into a vertical position. This, indeed, greatly facilitates a straight line of vision to the inferior cerebellar triangle, the IV ventricle, and then the superior cerebellar triangle and pineal region. However, it also verticalizes the child dangerously. This is no longer advised or condoned! In fact, it is to be avoided. ►

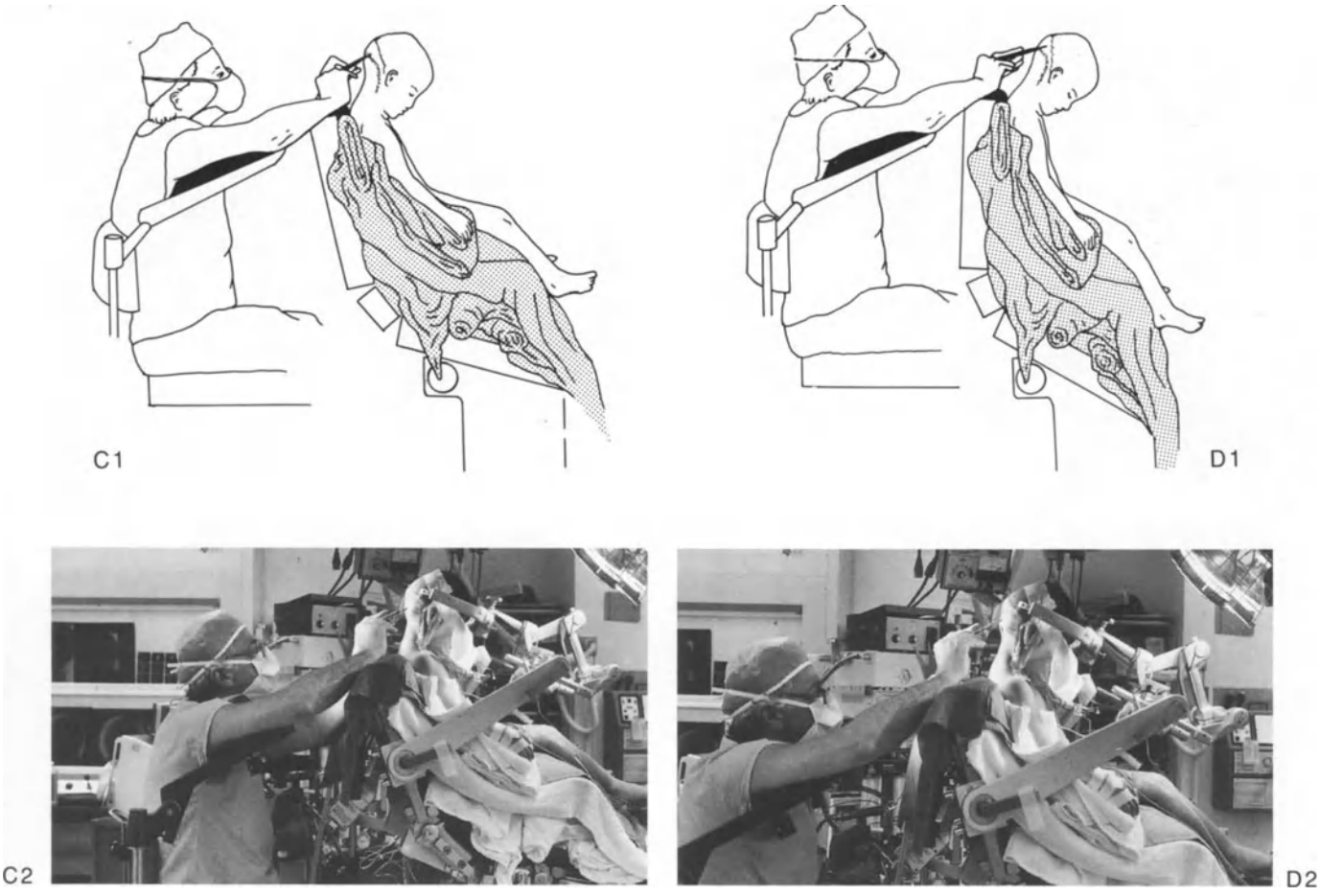


Figure 1.36. Legend see p. 30.

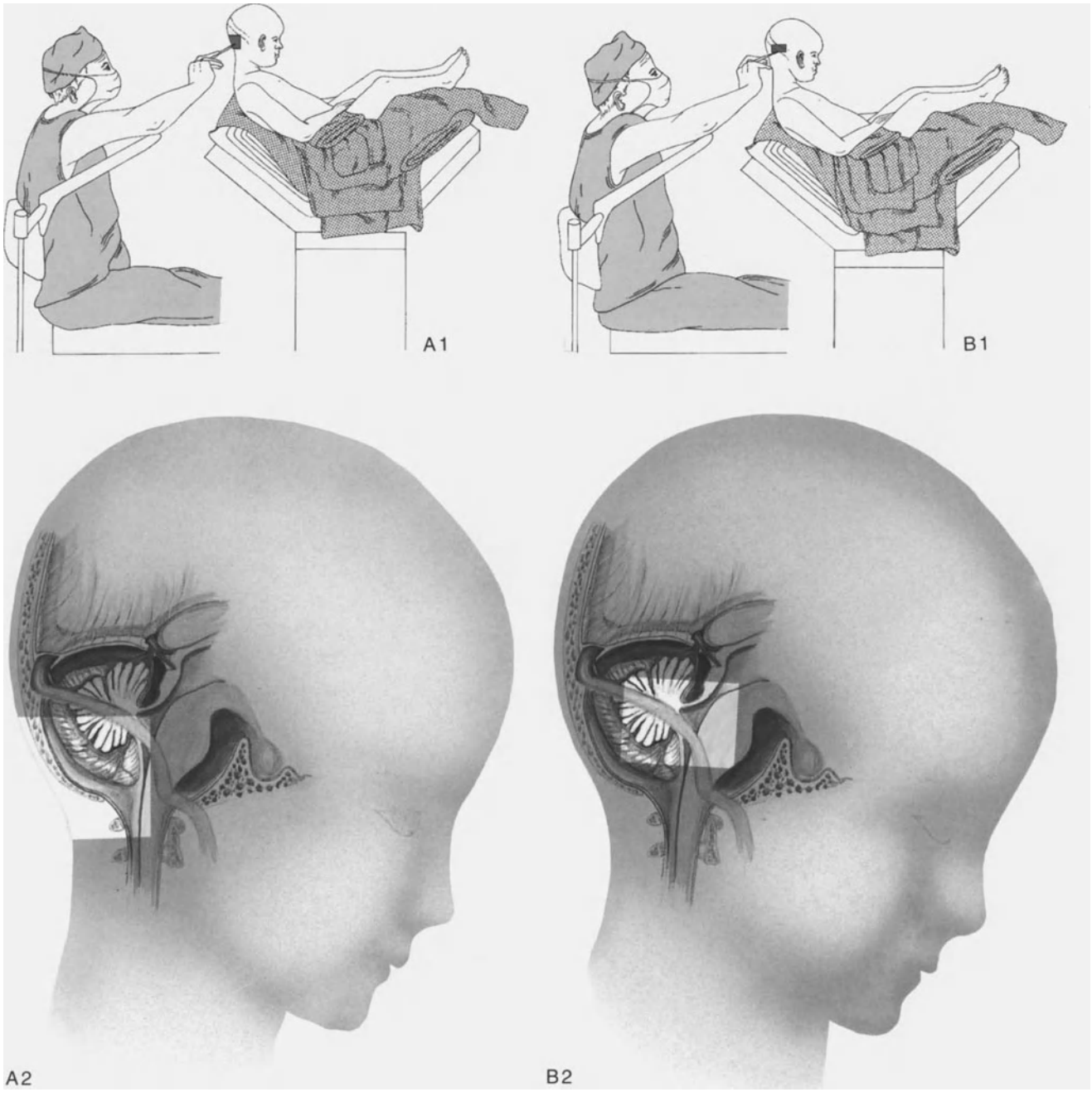


Figure 1.37. In the light of what was shown in Fig. 1.36 and photographically illustrated in Fig. 1.35A–C, the drawings illustrated in this figure place into relief the fact that excellent access to the inferior cerebellar triangle, the fourth ventricle region, and the superior cerebellar triangle including the pineal and quadrigeminal regions may very comfortably be exposed with the child in the "lounging" position. It is sufficient for the surgeon to lower his chair and extend his head slightly as he progressively exposes the inferior cerebellar triangle to the IV ventricle area or over the cerebellar vermis to the pineal/quadrigeminal region if he operates with the naked eye or with loupes. However, using the operating microscope avoids the very real physical inconvenience to the surgeon of extending his head: he need only adjust the eyepiece mount of the microscope. (A1–C1) illustrate diagramatically the changes in surgeon body level (A1) and head extension (B1,C1) to visualize the desired posterior fossa (inferior cerebellar triangle, IV ventricle, superior cerebellar triangle) indicated by the *blue rectangles on the child's head.* The operating microscope, though invariably used by the author for posterior fossa surgery, has not been included in these three drawings for schematic purposes only. (A2), (B2), and (C2) place into relief the structures visualized with ease in the inferior cerebellar triangle, IV ventricle area, and superior cerebellar triangle. ►

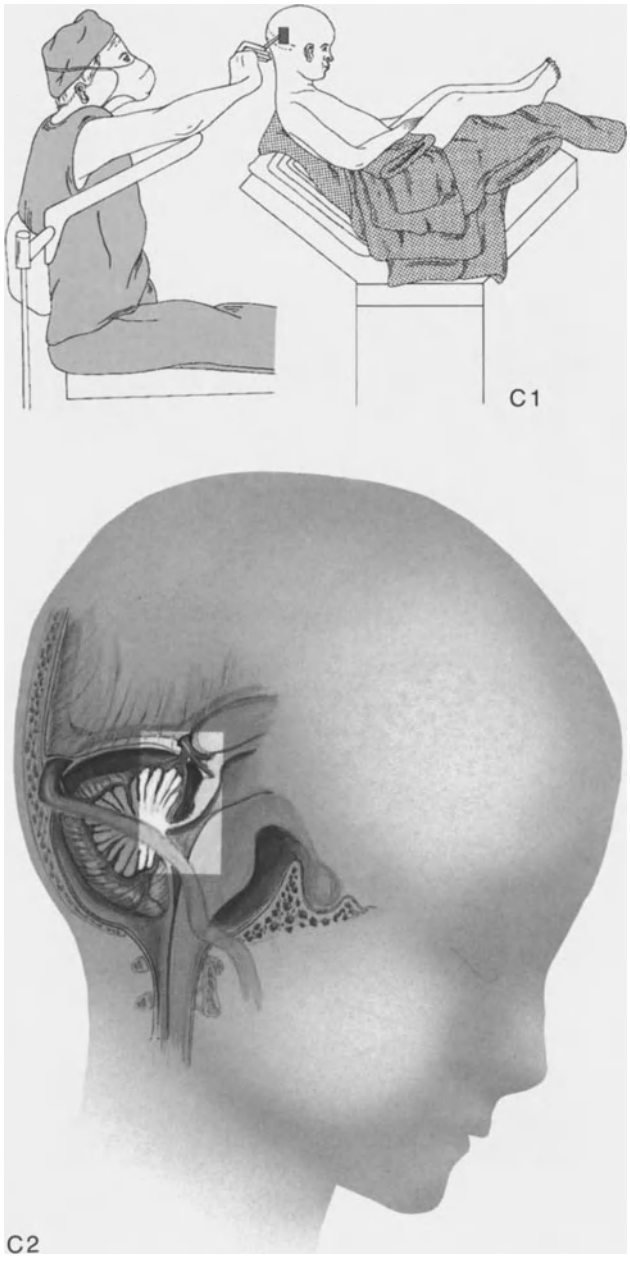


Figure 1.37. Legend see p. 32.

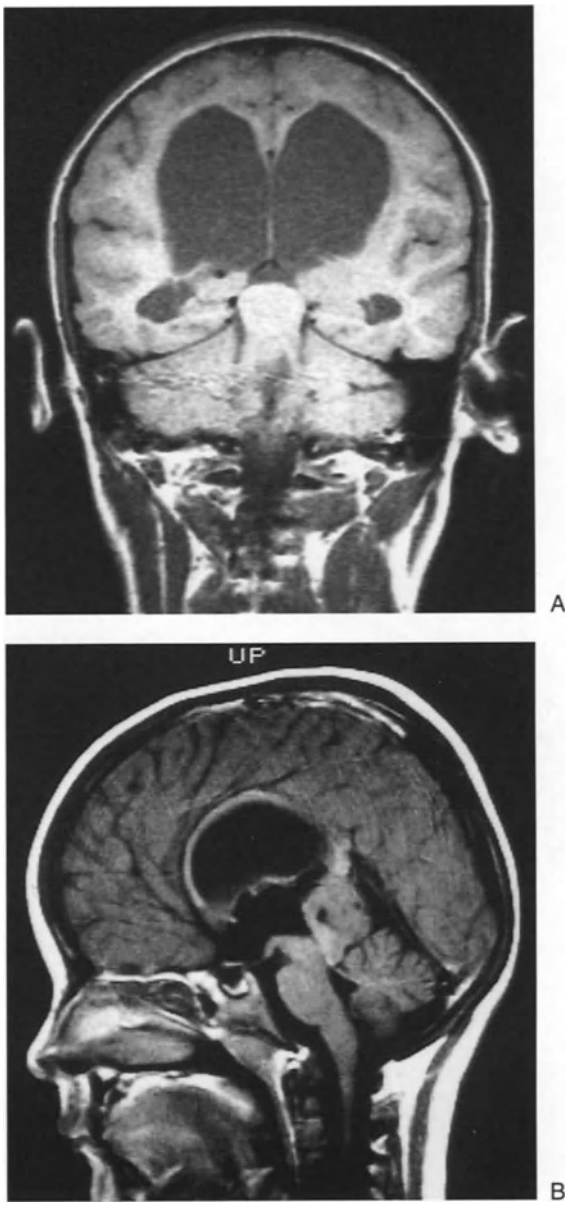


Figure 1.38. *Pineal region tumor*, with the child in the lounging position, may be resected effectively either through a medial occipital flap or using the transtentorial approach. Examples of pineal region tumors which may be resected, depending upon the preference of the surgeon, either through a medial occipital/transtentorial or through a suboccipital/supracerebellar approach, are (A) collicular plate glioma and (B) pineal tumor fungating into the anterior cerebellar cistern and coming to rest upon the collicular plate.

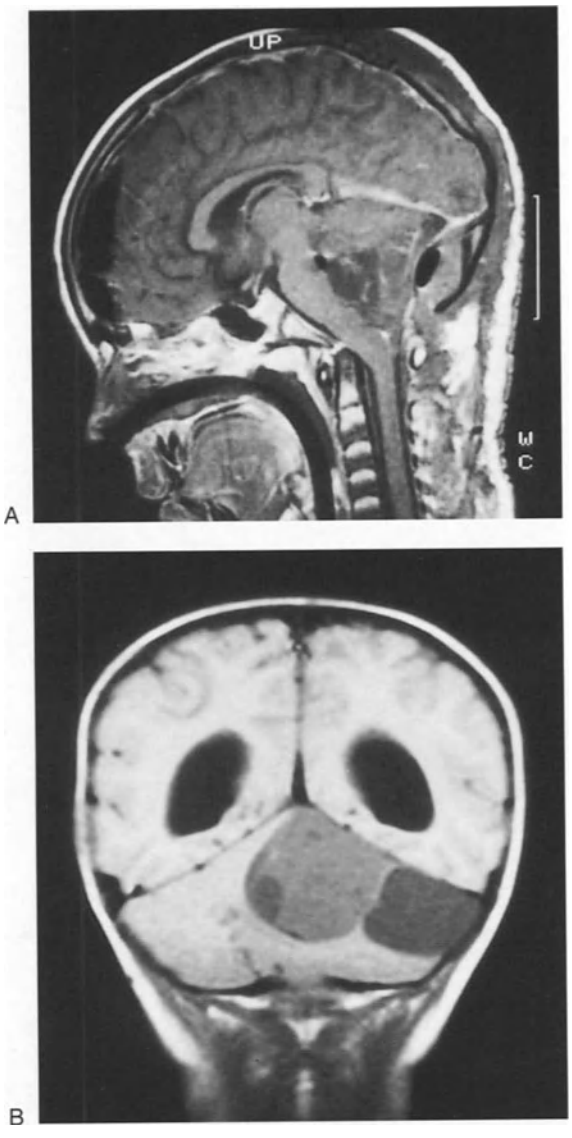


Figure 1.39. Examples of *infratentorial tumors* which occupy (A) the inferior or (B) superior cerebellar triangles, and which are very much more effectively resested with the child in the "lounging" position, are illustrated here.

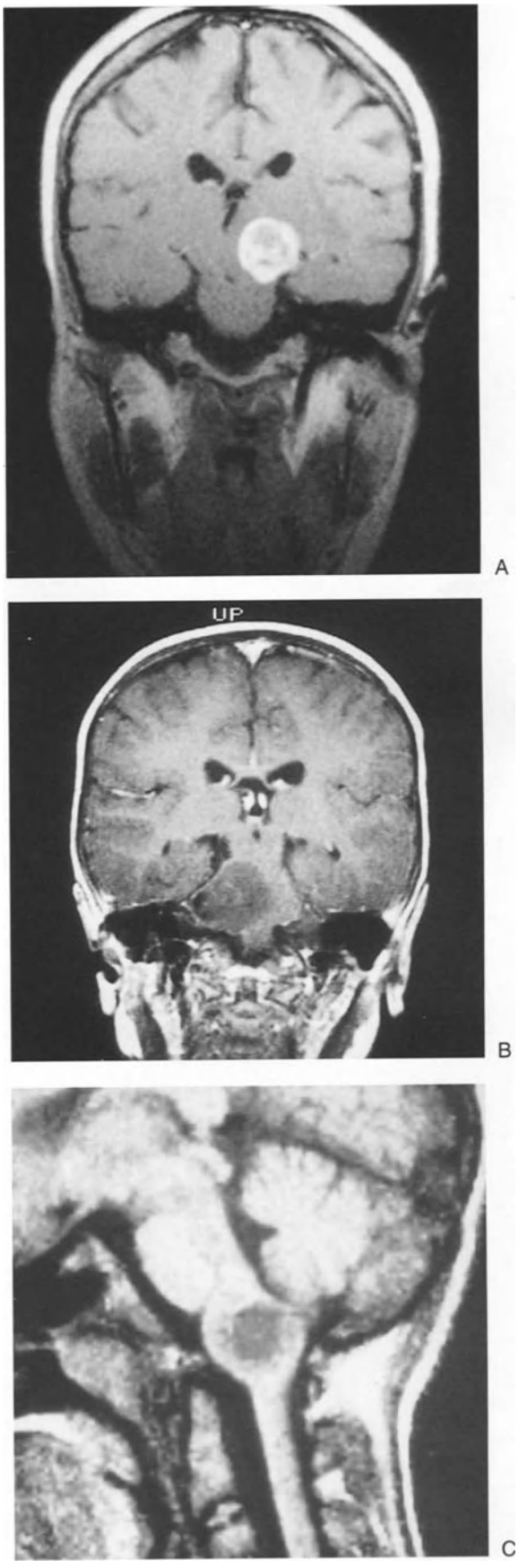


Figure 1.40. The lounging position also provides excellent exposure, in children with tumors, of (A) the midbrain, (B) pons, (C) medulla oblongata. It should be noted, however, that the park bench position, using a subtemporal/transtentorial approach, provides equally good access to *mesencephalic tumors*; that (B) the supine position with the head rotated provides excellent exposure of the superior pons, and with sectioning of the edge of the tentorium of the entirety of the pons; and that (C) *bulbomedullary junction tumors* may very satisfactorily be resected with the child prone.

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2 Incisions

Scalp, Muscle, Tissue, and Tumor Hemostasis

"No matter how sharp it is, the blade of the knife cannot harm its handle."
SOULEYMANE CISSÉ, Thoughts

"Know thou, who'er with heavenly power contends, Short is his date, and soon his glory ends, ..."
HOMER, The Iliad

Specific Incisions for Surgical Approaches

The individual skin incision (bifrontal, suboccipital, temporal, etc.) is planned so as to expose the desired skull area for the craniotomy. Consequently, the nomenclature for skin flaps is generally, but not invariably, identical to that used for bone flaps. Exceptions to this general rule are the frontal skin flap for both medial and lateral frontal craniotomy, the parasagittal skin incision for biparietal bone flaps or sagittal suture resection, the occipital skin incision for medial or lateral occipital craniotomy, and the hemispherical skin incision for frontotemporoparieto-occipital craniotomy.

The frontal and bifrontal skin incisions, and flaps, are used for access to the orbit(s).

These past 5 years have been punctuated, especially in general neurosurgery, by a series of presentations advocating "keyhole" neurosurgery. With the hope that slogans may be avoided, it is recommended that the pediatric neurosurgeon tailor his opening to what needs must be accomplished surgically, avoiding attempts to fit surgical goals into philosophical preconditions.

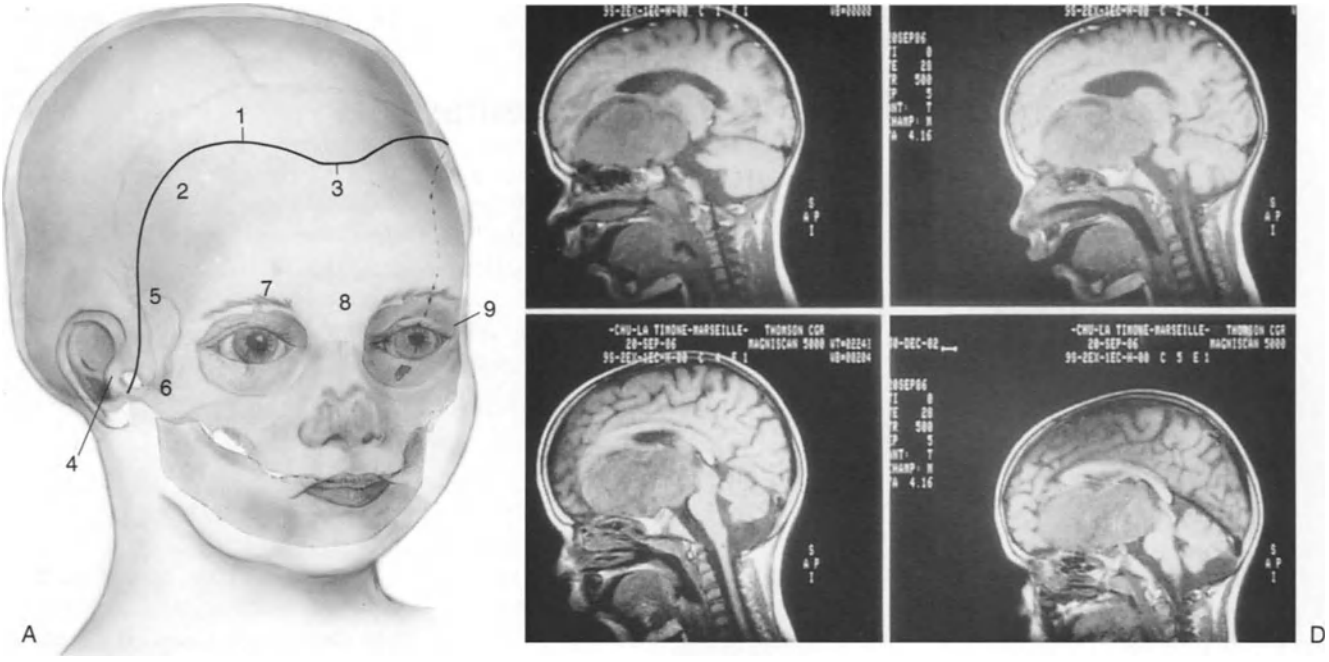
Bifrontal Incision (Figs. 2.1, 2.2)

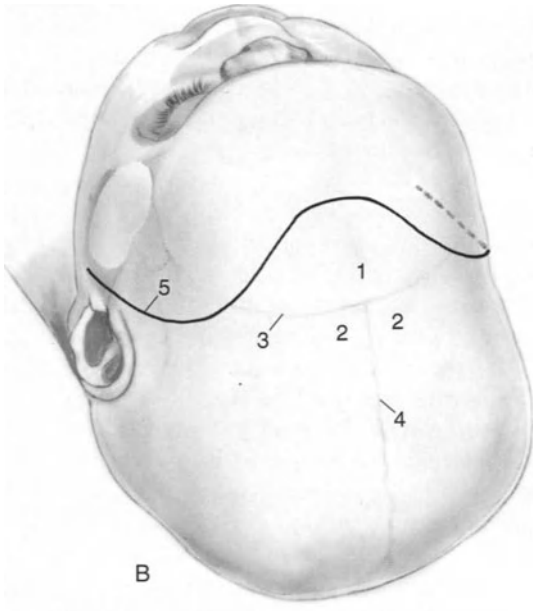
The bifrontal skin incision permits complete exposure of the frontal bone, as well as the squamous portion of the temporal and the greater wing of the sphenoid bone, a *bifrontopterional* bone flap.

Draping. Draping the child for a bifrontal skin incision should be such as to permit covering the supraorbital ridges on either side, over the glabella in the midline, and down the lateral edges of the frontal processes of the zygomas as far inferiorly as the malar bones. One then proceeds posteriorly, along the inferior edges of the malar bones and zygomatic arches, as far as the antitragus of each ear. A single drape may then be brought across the scalp in the coronal plane, extending from the antitragus on one side, around the attachment of the helix of the ear posteriorly, and then across from side to side (in the coronal plane) from the base of one mastoid process to that of the other.

Before planning the skin incision, one should identify the sagittal plane and the significant bony, suture, and muscular landmarks.

Incision. The bifrontal skin incision extends behind the hairline, from one zygomatic arch to the other, beginning approximately 8 mm anterior to the apex of the antitragus, just enough to avoid cutting into the external auditory canal (which courses anteriorly and slightly inferomedially beneath and deep to the antitragus). Following the hairline from the lateral to the superior surfaces of the head gives the skin incision a smooth posterior curvilinear swing, which then turns anteriorly as the sagittal plane is approached. This incision permits preservation of the main trunk of the superficial temporal artery and its anterior branch, as well as the frontal nerve. It allows the surgeon to reflect the scalp anteroinferiorly as far forward as the zygomatic processes and supraorbital ridges of the frontal bone, exposing the glabella; and as far inferiorly, on either side as the zygomatic arches, as to expose both the squamous temporal and greater wing of the sphenoid bones. After dissecting the galea from the periosteum, one may reflect the scalp posterior to the coronal suture. Thus, the entire frontal bone may be reflected, *en bloc,* with both pterional areas coming away with the single bone flap.





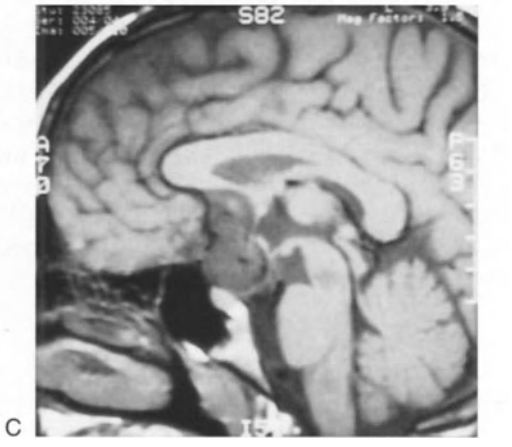


Figure 2.1. (A) If the skin incision (*1*) is made a few millimeters behind the hairline (*2*), it both hides the scar and, because of the horsehoe form it takes in approaching the sagittal plane (*3*), assures complete exposure of the frontal bone and the pterional regions bilaterally. Extending the bifrontal skin incision from one antitragus (*4*) to the other permits exposure of the pterional region (*5*), the zygomatic arches (*6*), the supraorbital rims (*7*) and glabella (*8*), and the lateral orbital rims (*9*). Hence, bifrontopterional exposure is achieved. (B) Bifrontal skin incision viewed obliquely with the child supine. This view permits the reader to appreciate the exposure of the posterior frontal (*1*) and anterior parietal (*2*) bones, and the coronal (*3*), sagittal (*4*), and squamosal (*5*) sutures. Though (C) *perichiasmatic epidermoid tumors* may be easily aspirated, the soft milky-white slightly floccular material coming away quickly, they adhere tenaciously either directly or indirectly to vascular and neural (especially cranial nerve) structures. Also, they extend generously into the entirety of the perisellar cistern (optic, prechiasmatic, olfactory, infrachiasmatic, sylvian, interpeduncular, etc.). Thus a large – bifrontal pterional – flap is indicated. Giant (D) optic chiasm, or anterosuperiorly extending hypothalamic, *gliomas* may fill the entire suprasellar area, displace the lamina terminalis so far posteriorly as to render the III ventricle almost invisible, and fungate out over the dorsum sellae and planum sphenoidale. The occasional softness of these tumors equally occasionally permits one to resect them through a unilateral frontal flap, but the fact that they may be either highly vascular or tenaciously fibrosclerotic makes it advisable to proceed directly with a bifrontal pterional skin incision preparatory to a bifrontal pterional bone flap.

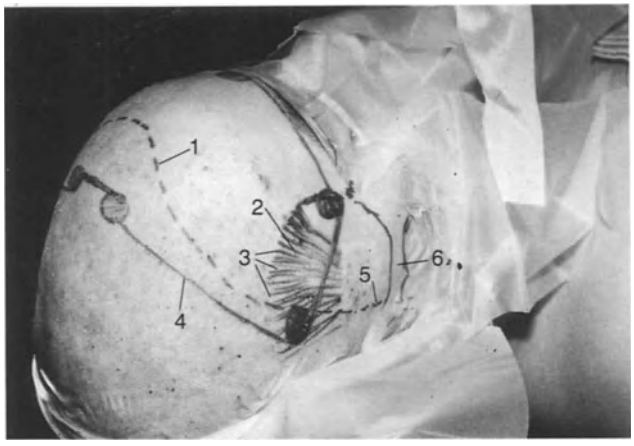


Figure 2.2. The bifrontal skin flap shown from the right. The skin incision (*1*) is represented by a *broken line*. Note the fanning of the temporalis muscle (*2*) and its insertion along the superior temporal line (*3*). The bur holes and craniotomy line (*4*) are drawn onto the scalp to permit the reader to observe that the bifrontal skin incision is placed so as to permit anterior superior reflection of the scalp from the frontal bone and its posterior inferior reflection over the coronal suture. Note that the skin incision is extended to, but not across (*5*), the zygomatic arch (*6*), thus greatly facilitating anterior superior reflection of the scalp over the orbital rims.

Frontal Incision (Fig. 2.3)

The frontal skin flap is used for medial and lateral frontal craniotomies, permitting equally desirable access to the glabella, zygomatic process of the frontal bone, pterion, and coronal suture.

Draping. The draping recommended for a frontal skin incision is the same as for the bifrontal skin incision, since it permits the surgeon immediate access to the opposite side if the need arises and he chooses to proceed with a bifrontal craniotomy.

Incision. The frontal skin incision is a partial bifrontal incision, extending only to the parasagittal plane running through the center of the opposite orbit.

Frontoparietal Skin Incision for Frontoparietal Bur Holes (Fig. 2.4)

If one considers that it is often necessary to reflect a frontotemporoparietal skin flap so as to perform frontoparietal craniotomy after frontoparietal bur holes have been placed, *the desirability of planning skin incisions for frontoparietal bur boles which may be extended into a skin flap becomes clear.* Therefore, curvilinear skin incision for frontoparietal bur holes are preferable because they may serve as either limb of the frontotemporoparietal flap if this becomes necessary.

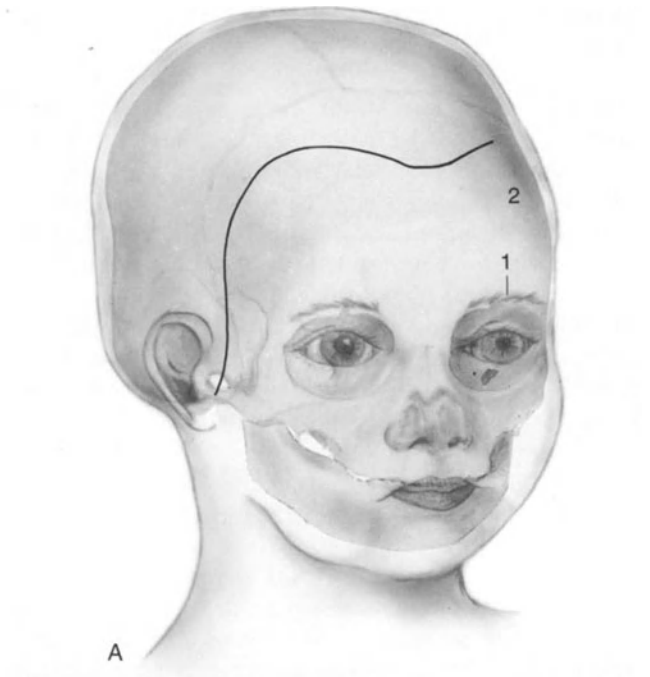
Draping. The draping for frontoparietal bur holes should be such as to permit parietal, temporoparietal, and frontotemporoparietal craniotomies. One drape is placed with its edge along the sagittal plane, from the glabella to the inion. The other is placed across the side of the scalp, from posterior to anterior, extending from the base of the mastoid bone, around the insertion of the helix of the ear, over the zygomatic arch to the malar bone, and then along the zygomatic process of the frontal bone and over the supraorbital ridge to the glabella.

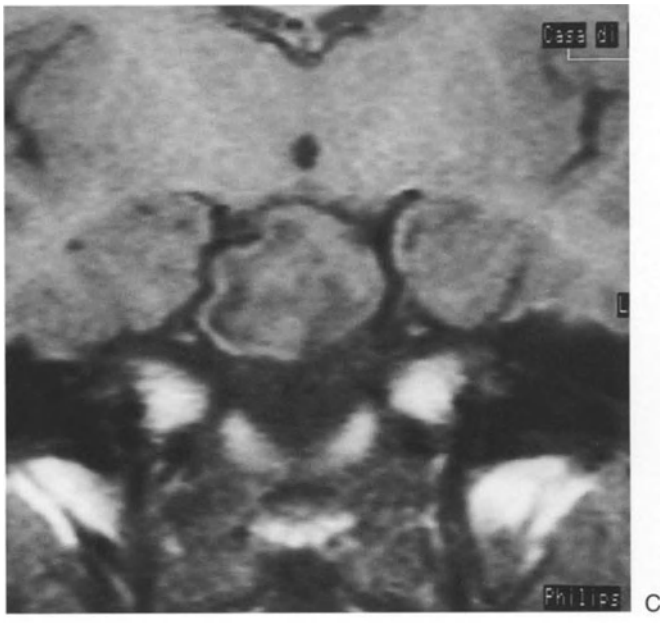
Incision. The anterior skin incision is curvilinear, with convexity facing anteriorly; the posterior incision is also curvilinear, but with convexity facing posteriorly. This curvilinear incision permits placement of the self-retaining retractor so as to expose the underlying skull for the bur holes. In the event the surgeon finds it desirable to reflect a parietal flap, the superior aspects of the anterior and posterior incisions are simply connected. The skin incision is then extended inferiorly along either limb to the proper level, depending upon how low one finds it necessary to proceed. Since the incisions may readily be brought posterior to the occipital artery and anterior to the superficial temporal artery, one need not be concerned about scalp necrosis, even if one chooses to go as far inferiorly as the zygomatic arch in order to perform a temporal craniotomy/craniectomy.

Frontoparietal Incision for Posterofrontal or Anteroparietal Lesions (Fig. 2.5)

Lesions within the posterior frontal or anterior parietal areas are approached through a frontoparietal flap.

Incision. The skin incision extends from approximately 1 cm anterior to the antitragus superiorly and medially behind the hairline, to just across the midsagittal plane. It is then run posteriorly to the midcoronal plane of the head: the plane running through the external auditory canals, not the coronal suture. (The awake patient may be tested for the location of this plane simply by using a pinwheel and asking him to tell you when he feels the pin anteriorly and when he feels it posteriorly.) It is then curved broadly back to the original side and extended as far inferiorly as the superior temporal line.





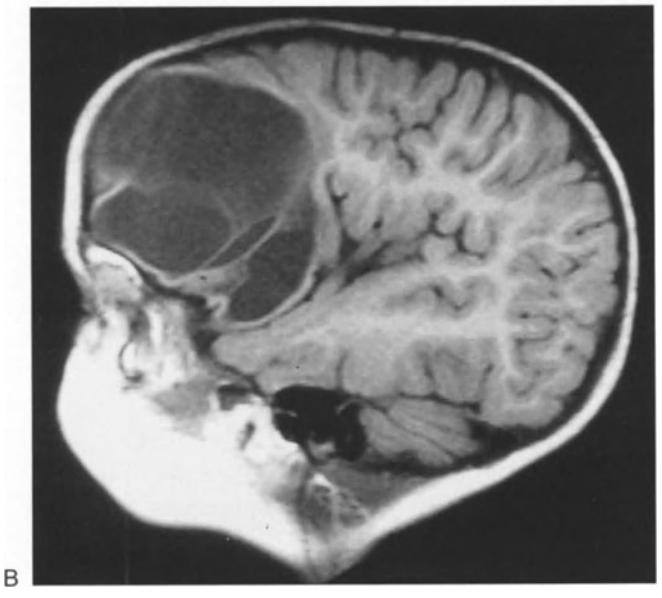


Figure 2.3. (A) Here the skin incision for a unilateral frontal craniotomy is extended across the midline to the parasagittal plane running through the center of the opposite orbit (*1*), just medial to the frontal eminence (*2*). (B) Some neurosurgeons use the (unilateral) frontal incision even for such parasellar masses as the *craniopharyngioma* or *epidermoid tumor*, which I discourage because of the diffuse adherences and/or surface-structure invasiveness of which both of these lesions are capable. However, (B) frontal lobe masses, even such large ones as herein illustrated, or (C, *right above*) pedunculated *hypothalamic hamartomas* are most effectively resected through the flaps this skin incision permits.

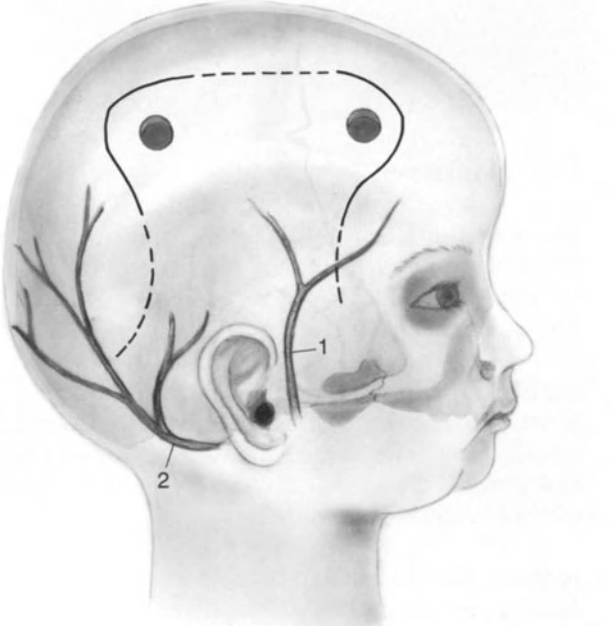


Figure 2.4. The incisions for the frontal and parietal bur holes are marked with the *solid lines*. The *interrupted lines* indicate extension of the skin incision to permit reflection of a fronto-temporoparietal flap if the operative findings suggest this to the surgeon. The superficial temporal (*1*) and small branches of the occipital (*2*) arteries are included in the flap to assure healing without necrosis along the flap's edges. Diagnostic bur holes are a procedure of the past in the industrialized world, but not in the developing countries.

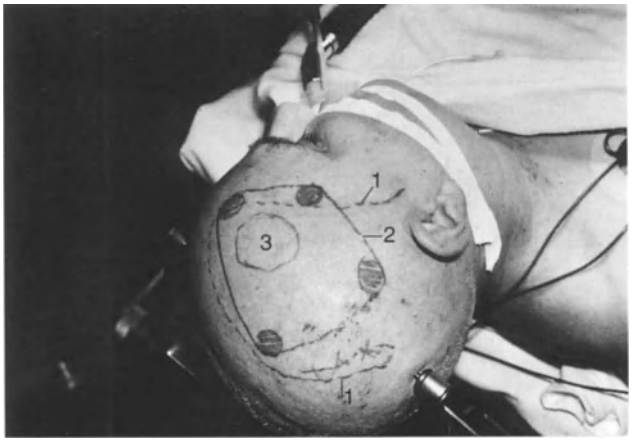


Figure 2.5. For frontoparietal flaps the anterior limb of the skin incision is identical to that for frontoparietal flaps, but the medial limb curves posteriorly (contralateral to the midsagittal plane) to the midcoronal plane of the head, where it is redirected inferiorly and then back over the original side *(1)*. This incision is extended posteroinferiorly to the superior temporal line. A trapezoidal bone flap *(2)* is marked off and a posterior frontal lesion *(3)* is drawn in. This incision permits bone flaps for large frontal lobe tumors, access to the foramen of Monro through the lateral ventricle, or trans-sylvian fissure access to pedunculated hypothalamic hamartomas ◄

Parietal Incision (Fig. 2.6)

The parietal skin incision permits access to the entire parietal bone, from coronal to lambdoid and from sagittal to parietotemporosphenoidal sutures. Thus, one may reflect a medial (superior) parietal bone flap for access to the superior sagittal sinus and the falx, or a lateral (inferior) parietal flap for access to the convexity of the parietal lobe and posterior portion of the sylvian fissure.

Draping. The draping should extend in a sagittal plane from the contralateral zygomatic process of the frontal bone, first superiorly, then posteriorly, and finally inferiorly to the base of the mastoid. It should be brought across the operative side, extending along the highest nuchal line to the base of the mastoid. From here it is run horizontally, over the ear, to the zygomatic process of the frontal bone, before proceeding over the frontal eminence on the operative side to the frontal eminence and zygomatic process of the frontal bone on the contralateral side.

Incision. The skin incision preserves both the superficial temporal and occipital arteries. It extends behind the hairline from just above the pterion to 1 cm across the midsagittal plane, where it turns posteriorly, always running parallel to the midsagittal plane, before being swung back to the operative side behind the parietal eminence and then extended inferiorly and posteriorly. The incision provides a wide pedicle, access to the entire parietal bone, and exposure of the sagittal suture.

Parasagittal Incision (Fig. 2.7)

The parasagittal incision offers excellent exposure of the superior surface of the posterior portion of the frontal bone, and the medial third of the coronal and lambdoid sutures.

Draping. Draping for parasagittal incision should consist of laying towels across a line drawn from one frontal eminence to the other anteriorly, from the frontal to the parietal eminences on either side, and from the base of one mastoid process to the other posteriorly.

Incision. The parasagittal incision is convenient for biparietal craniotomies and sagittal suture resection. (Lowering of the superior sagittal sinus, however, necessitates an extension from the convex portion of the anterior limb of the S-shaped parasagittal incision.) The surgeon should take care to assure the gentle curvilinear course of the parasagittal incision, rather than cutting sharp angular routes: the former permits adequate blood supply to both aspects of the flap; the latter puts the extremities of the flap at risk to necrosis. The flap itself is begun behind the hairline at a point posterior to the frontal eminence on one side, and then extended gently toward the contralateral side before being turned back on itself and extended across the midline. Finally, it is brought onto the contralateral side.

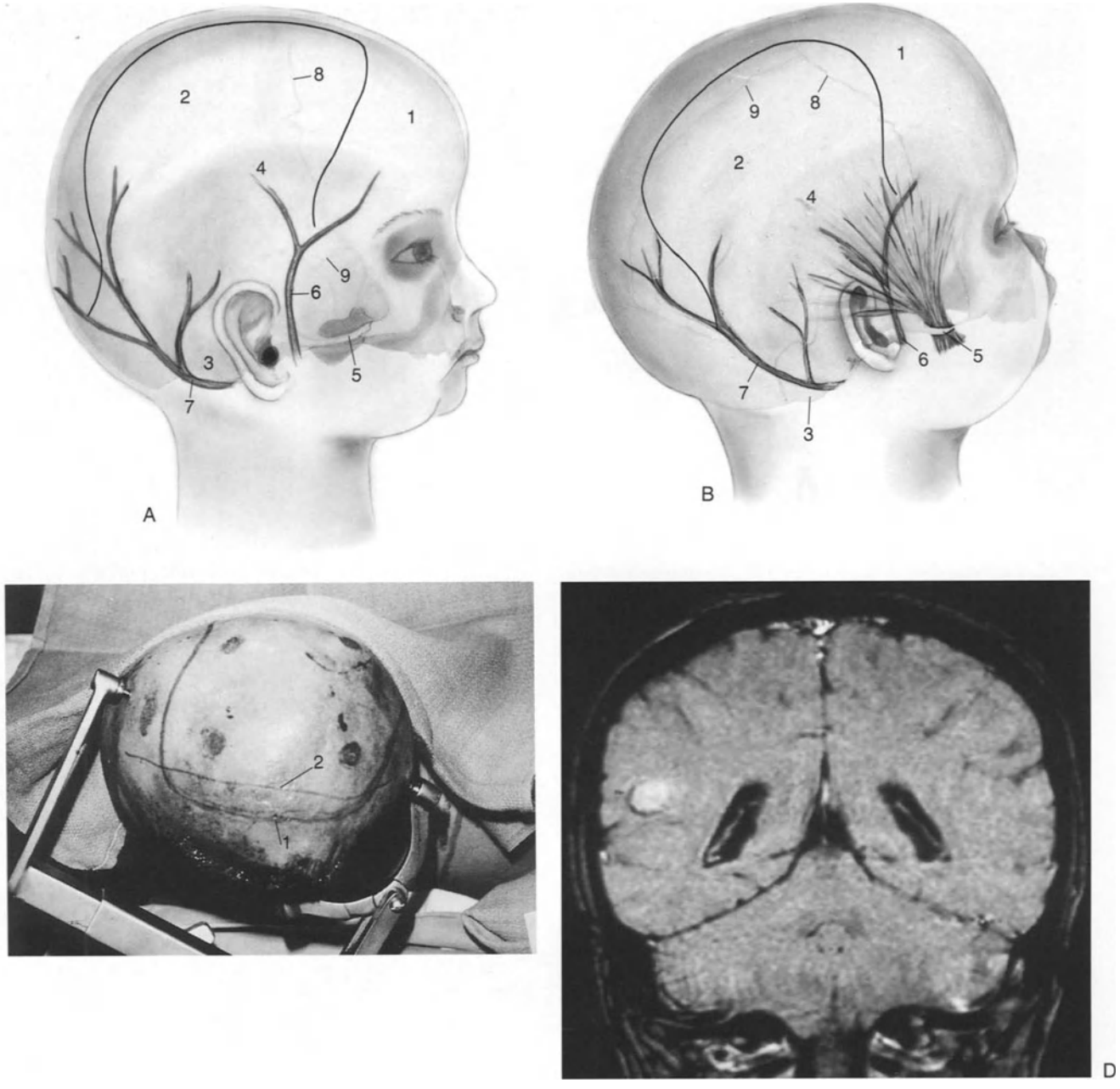
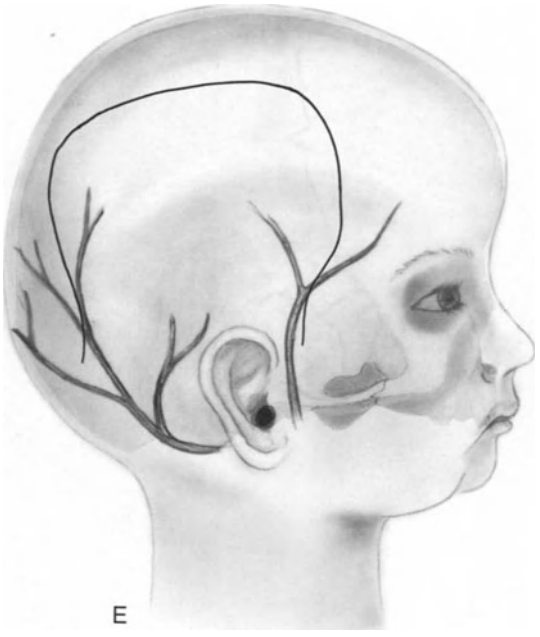
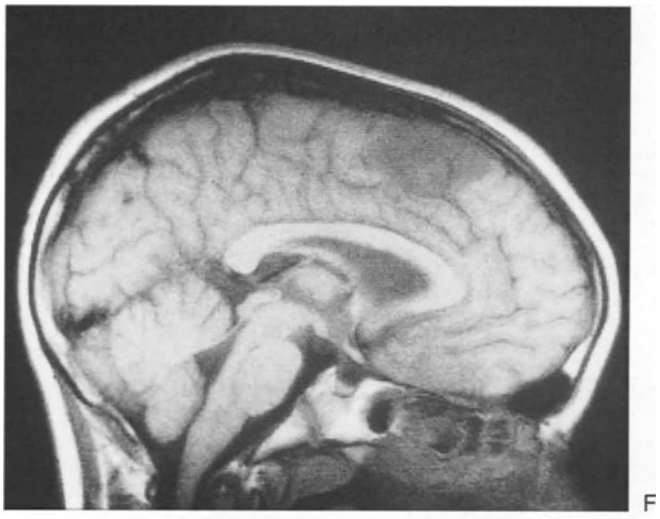


Figure 2.6. The parietal skin incision, as viewed from the lateral (A) and posterior oblique (B) perspectives, shows the frontal (*1*), parietal (*2*), and mastoid (*3*) eminences, the superior temporal line (*4*) and zygomatic arch (*5*), the superficial temporal (*6*) and occipital arteries (*7*), and the coronal (*8*) and sagittal (*9*) sutures. (C) This perspective, from the vertex of the skull, illustrates extension of the medial limb of the skin incision (*1*), which has been extended onto the contralateral side of the sagittal plane (*2*). (D) For an inferior parietotemporal flap, the incision begins about 2 cm above and 1.5 cm anterior to the antitragus. It then remains behind the hairline as it is extended superiorly to a level 2 cm above the superior temporal line and is run posteriorly behind the parietal eminence and then inferiorly to the base of the mastoid. This incision may be lateral, extending superiorly midway between the sagittal plane and the superior temporal line to provide access to the mid and inferior surfaces of the parietal lobe for lesions such as the *cavernoma* herein illustrated. One notes that this cavernoma rests approximately within the angular/supramarginal areas. Therefore it may be best resected using the inferior parietal temporal flap illustrated in (E) of this figure. However, if it were located superiorly, the flaps indicated in (A) or (B) would be preferable. (*Continued* on p. 43).





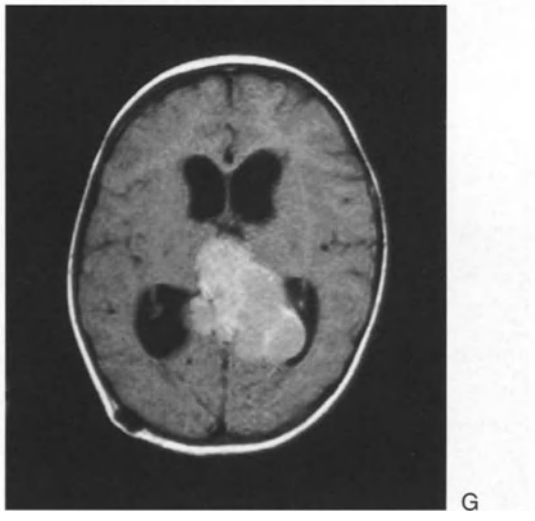


Figure 2.6. (E) For an inferior parietotemporal flap, the incision begins about 2 cm above and 1.5 cm anterior to the antitragus. It then remains behind the hairline as it is extended superiorly to a level 2 cm above the superior temporal line and is run posteriorly behind the parietal eminence and then inferiorly to the base of the mastoid.
Bringing the medial plane of the parietal skin incision to or across the imaginary line projected along the sagittal suture converts this into a medial parietal skin incision, one which permits bone flaps for access to (F) medial frontal parietal tumors or (G) those *pineal region tumors* which extend above the roof of the III ventricle and either rest upon or surround the internal cerebral veins.

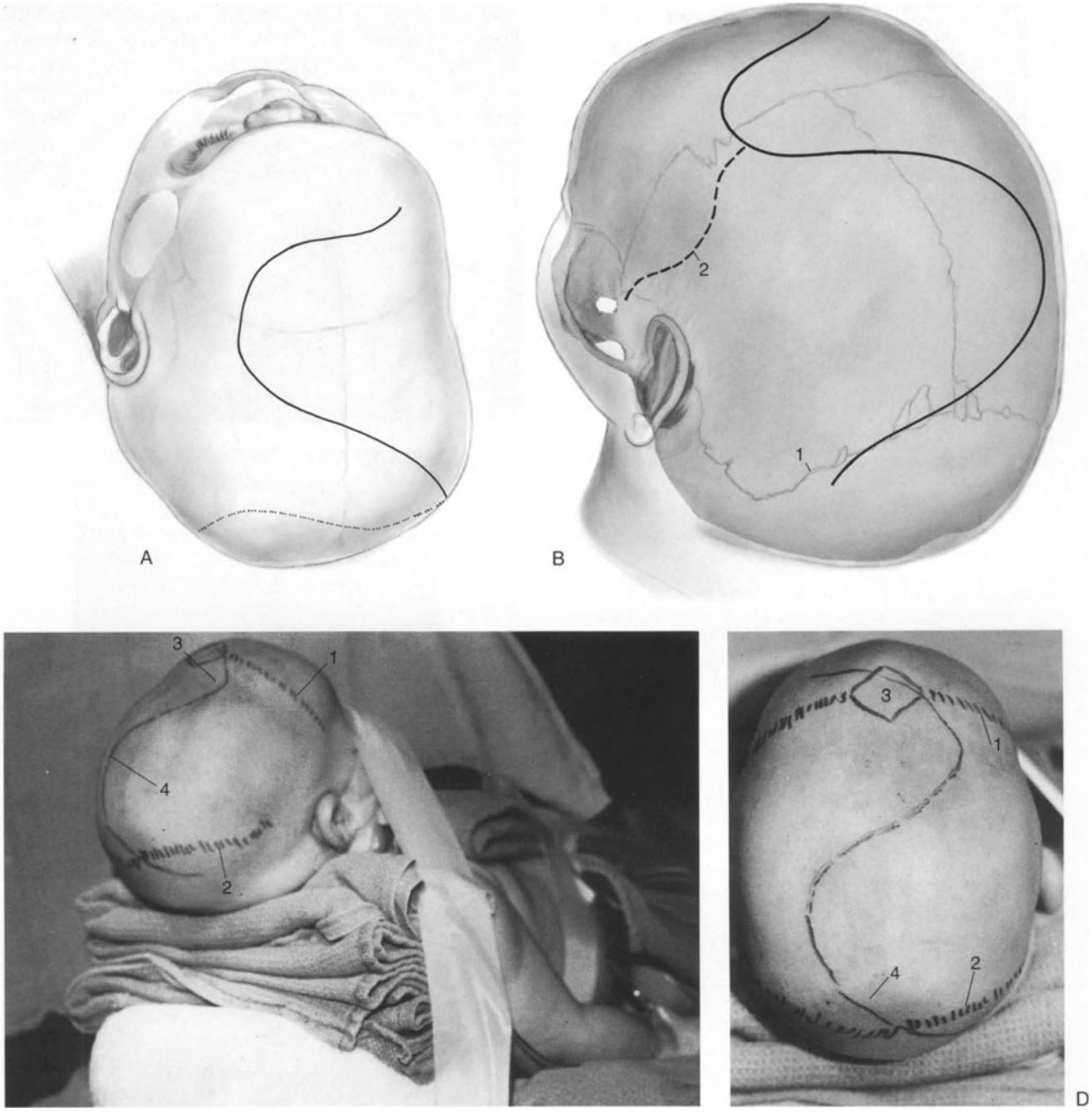


Figure 2.7. (A) This shows the line of the sinusoidal parasagittal skin incision with the child in the supine position, as the child would be during the operative procedure. The *broken line* indicates extension of the incision along the squamous occipital area, *from right to left.* (B) This drawing illustrates the importance of bringing the posteroinferior extremity of the incision beneath the lambdoidal suture (*1*), a matter of particular importance when performing a resection of the sagittal suture in children with sagittal synostosis. It also illustrates the extension (*2*) that must be made if one wishes adequate exposure to lower the superior sagittal sinus and perform a reduction cranioplasty. (C) The placement of towels beneath the child's shoulders and neck (so as to bring the inion into working distance), the coronal (*1*) and lambdoidal (*2*) sutures, and the open anterior fontanelle (*3*). The S-shaped incision (*4*) begins just in front of the coronal suture *on the left*, crosses *to the right* where it is gently curved, bringing it back to the pareital eminence *on the left*, from whence another curve is begun. This latter brings the incision across the inion and beneath the lambdoidal suture *on the right*. This perspective (D) permits one to identify the coronal suture and the anterior fontanelle, so as to appreciate the course of the anterior limb of the skin incision and the swinging of the posterior limb beneath the inion and lambdoidal suture. The labeling is the same as in (C). (E) see p. 45.

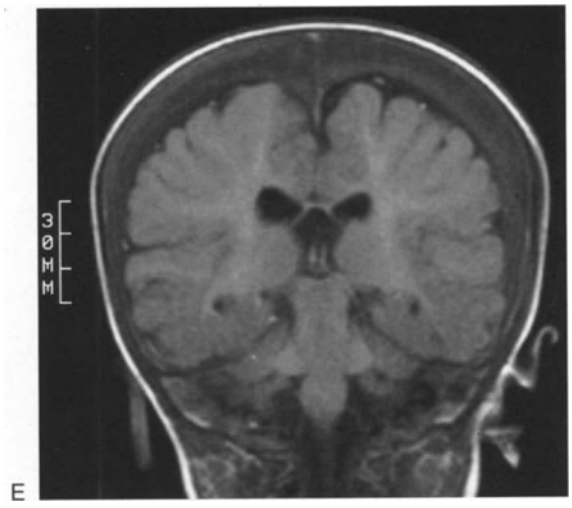


Figure 2.7. This magnetic resonance study of (E) a child with a bilateral *chronic subdural hematoma* offers one of the clearest indications for an "S"-shaped parasagittal incision preparatory to bilateral frontal or bilateral frontoparietal bone flaps which permit the extensive exposure necessary to resect, in one sitting, the entirety of the convexity and parasagittal subdural membranes and/or perform a reduction cranioplasty.

Temporal Incision (Fig. 2.8)

Temporal incisions are used to expose the temporal lobe, either in its entirety or (separately) its anterior, medial, or posterior portions. The temporal skin flap may be placed anteriorly if one wishes to expose only the anterior portion of the temporal lobe, or posteriorly for its posterior portion. A greater space between the anterior and posterior limbs of the incision permits full exposure of the temporal lobe. The important consideration is that the temporal skin incision must bring the surgeon over the full fan of the temporalis muscle, permitting him to reflect an osteoplastic instead of a free temporal bone flap. It is injudicious to reflect a free flap when given the option of reflecting an osteoplastic flap, since the latter affords greater protection against physical and bacterial noxae. Exposure of the greater wing of the sphenoid, the squamous portion of the temporal bone, and the inferior portion of both the frontal and parietal bones is possible through temporal skin incisions.

Draping. Draping for a temporal flap should include placement of a towel in the sagittal plane, from the frontal to the parietal eminences. The inferior drape extends from the frontal process of the zygoma, along the zygomatic arch to the antitragus. Then it runs around the insertion of the helix of the ear onto the scalp, down to the mastoid process and across the base of the skull to the superiorly placed drape.

Incision. Integrity of flap vascularization is assured by respecting the superficial temporal and anterior branch of the occipital arteries. The illustration shows the incision line, reflected flap, temporalis muscle, and superficial temporal and occipital arteries, permitting one to visualize the difference between free and osteoplastic bone flaps: portions of the frontal, parietal, squamous temporal, and greater wing of the sphenoid bones may be seen beneath the temporalis muscle as they would be reflected with an intact muscular insertion.

The incision runs from the zygomatic arch, 8 mm anterior to the posterior spur of the antitragus, behind the hairline superiorly and anteriorly with a gentle posterosuperior curvature, along the superior temporal line, to just beneath the parietal eminence, where the incision turns inferiorly once more, extending to the base of the mastoid bone. In this manner, both the superficial temporal and occipital arteries may be spared. Small and large temporal skin incisions may be made with the former by cutting anteroinferiorly to the occipital artery, and with the latter by cutting posteroinferiorly behind the occipital artery.

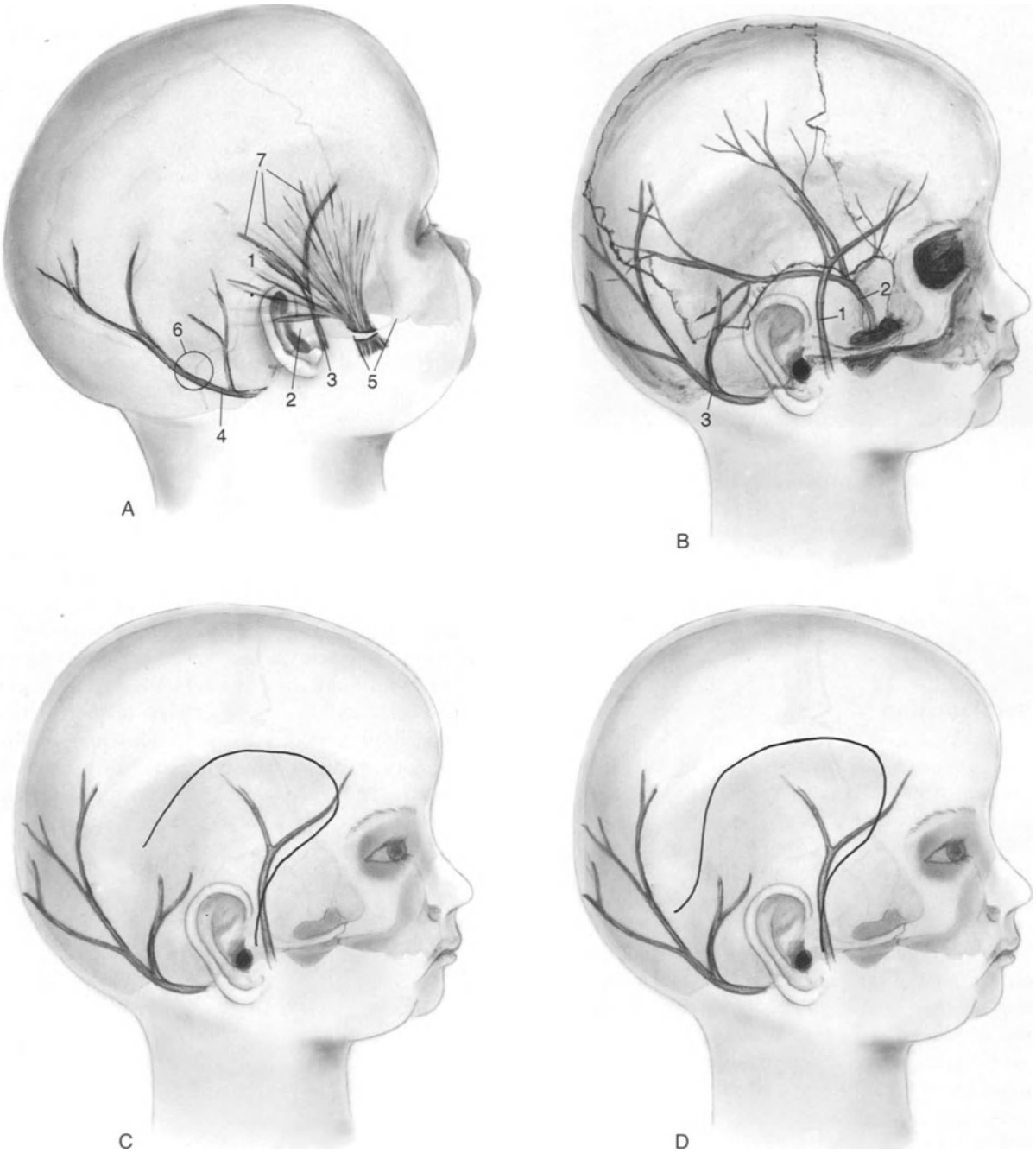
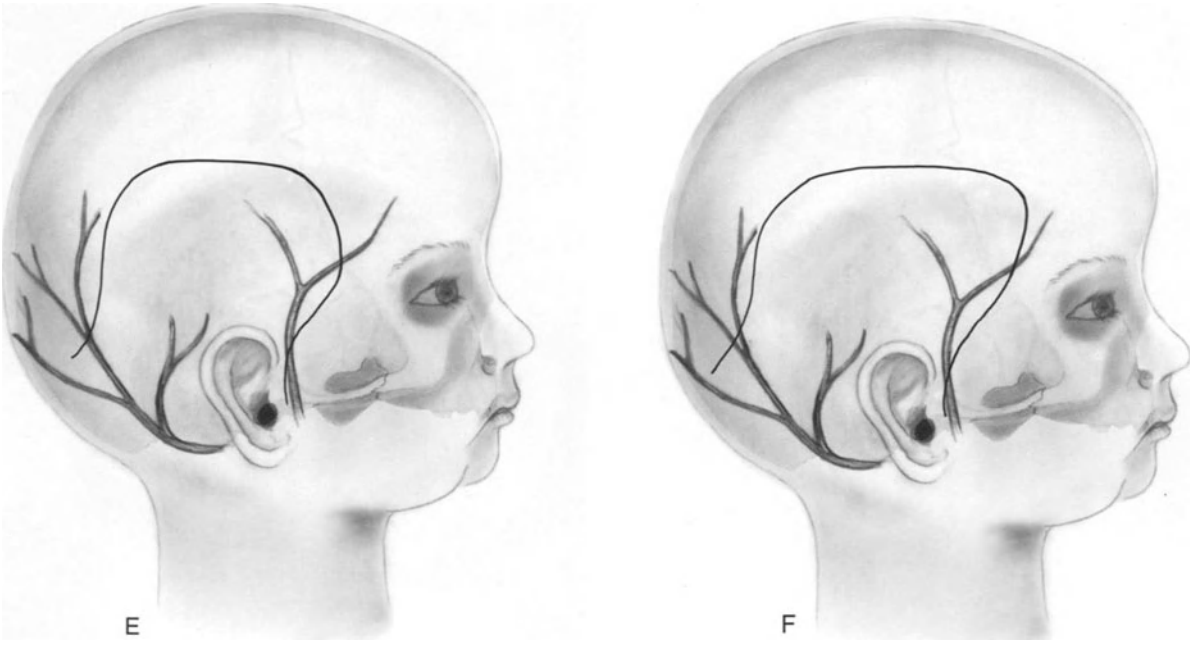


Figure 2.8. Relative location of extra- and intracranial branches of the external carotid vessels. (A) The posterior oblique view illustrates relative locations of the temporalis muscle (*1*), antitragus (*2*), superficial temporal (*3*) and occipital (*4*) arteries, zygomatic arch (*5*), and occipitotemporoparietal sutures (*6*). Note the insertion of the fan of the temporalis muscle along the superior temporal line (*7*). (B) This straight lateral view, with temporalis muscle removed, shows the relative location of superficial temporal (*1*), middle meningeal (*2*), and occipital (*3*) arteries. (C) Incision line for exposure of the anterior temporal lobe. (D) This illustrates a wider, more posterior sweep of the incision, extending it downward behind small branches of the occipital artery, for exposure of the middle portion of the temporal lobe. This incision preserves both the superficial temporal and occipital arteries within the flap pedicle. (E–G) see p. 47.



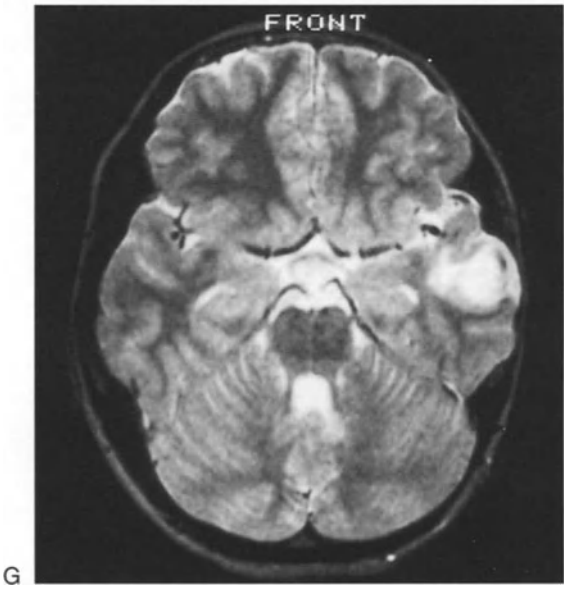


Figure 2.8. (E) This incision line descends more posteriorly behind small branches of the occipital artery for exposure of the middle and posterior portions of the temporal lobe. (F) This flap permits one to perform an enlarged temporal craniotomy. (G) The many alternatives of *temporal incisions* permit selective exposure of (G) gyral, or lobar, anterior or posterior, medial or lateral temporal lobe lesions.

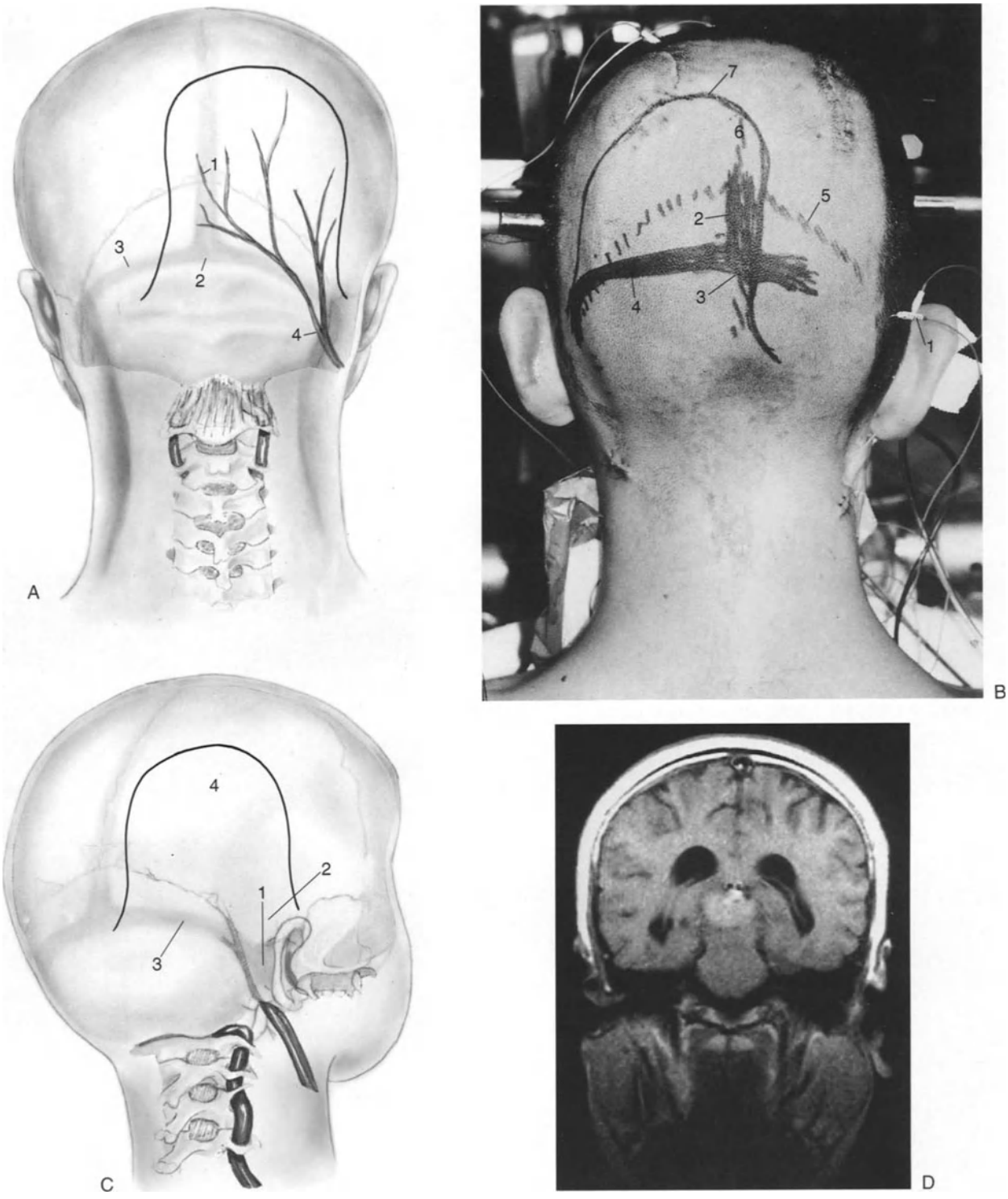


Figure 2.9. (A) The medial occipital skin incision, viewed posteriorly. The inion (*1*), torcular Herophili (*2*), transverse sinus (*3*), and occipital artery (*4*) are shown beneath the skin incision. (B) A child in whom the incision depicted in (A) was made. The electrodes for MEPs have been placed (*1*). The superior sagittal sinus (*2*), torcular Herophili (*3*), and transverse sinus (*4*) are drawn in as are the lambdoidal (*5*) and most inferior portion of the sagittal (*6*) sutures. The incision line is marked off (*7*). (C) The lateral occipital skin incision, viewed obliquely to appreciate extension of the lateral limb of incision anterior to the sigmoid sinus (*1*) at the pneumatized portion of the mastoid process (*2*), has a base across the transverse sinus (*3*) and crosses over the parietal eminence (*4*). The medial occipital skin incision is ideal for (D) pineal region tumors if one wishes to perform a transtentorial approach to resect extensions posteroinferiorly into the supracollicular cistern and the precentral cerebellar area. (E, F) see p. 49.

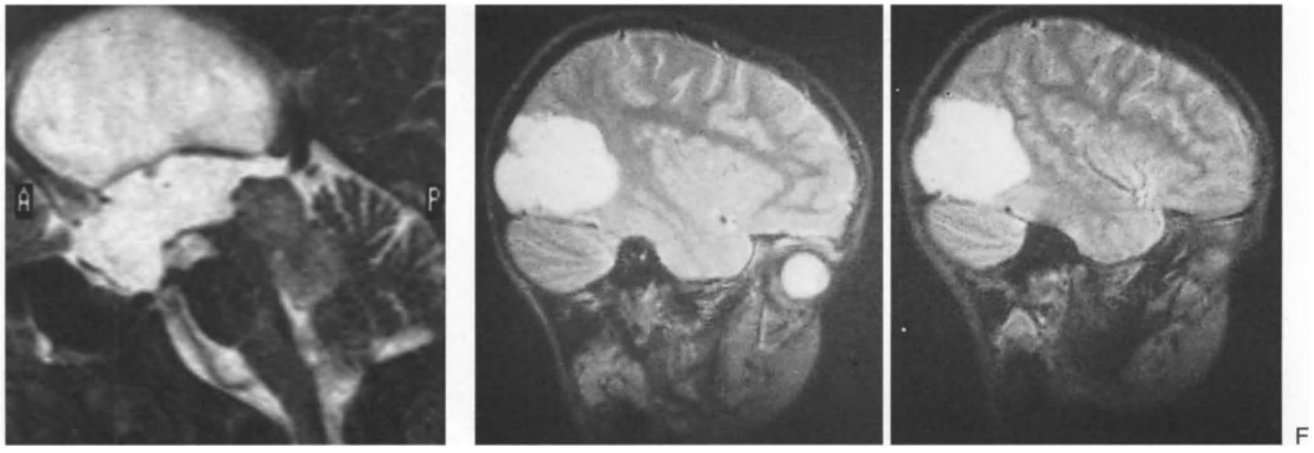


Figure 2.9. The same incision, consequently, permits access to the collicular plate for resection of (E) *collicular plate gliomas.* If the medial limb of the flap is not brought across the line of the sagittal suture, a lateral occipital bone flap is adequate for resecting (F) occipital pole lesions.

Occipital Incision (Fig. 2.9)

The occipital skin incision is placed for exposure of the posterior portion of the parietal bone and the most superior portion of the squamous occipital bone, as well as the inion, the posteroinferior portion of the sagittal suture, and the entirety of the homolateral lambdoidal suture. There are medial and lateral occipital incisions, depending upon whether one wishes access to the falx and pineal region, or to the convexity of the occipitotemporal lobes.

Draping. Draping for either incision should parallel the incision line and extend across the base of the skin flap, allowing approximately 3 cm on all sides of the skin flap.

Incision. The *medial* incision extends from the inion: first, superiorly across the contralateral side of the sagittal suture, and then, horseshoe fashion, to the parietal eminence, before proceeding inferiorly to the base of the mastoid bone. This assures integrity of the occipital artery. The *lateral* incision extends superiorly, and parallel to the superior sagittal sinus, from just lateral to the torcular Herophili to over the parietal eminence. It is then extended inferiorly and anteriorly to just above the helix.

Suboccipital Incision (Figs. 2.10, 2.11)

Suboccipital skin flaps may be either medial (midline) or lateral, depending upon whether one must reflect the squamous portion of the occipital bone for a vermis tumor (medial), a cerebellar hemisphere or pontocerebellar angle tumor (lateral). The medial incision permits exposure of either the inferior cerebellar triangle (beneath the great horizontal fissure of the cerebellum) or the superior cerebellar triangle (above the great horizontal fissure of the cerebellum). The lateral incision permits a craniotomy, exposing the most lateral portion of the cerebellar hemisphere and the pontocerebellar angle.

Draping. Draping for both the midline and lateral incisions should allow for exposure of the skin to approximately 3 cm to either side of the incision.

Incision. The *midline* skin incision extends from approximately 1 cm above the inion to C-6. The *lateral suboccipital* incision extends from just above the lambdoidal suture down to the level of C-5, in a parasagittal plane, midway between the midline and the mastoid process.

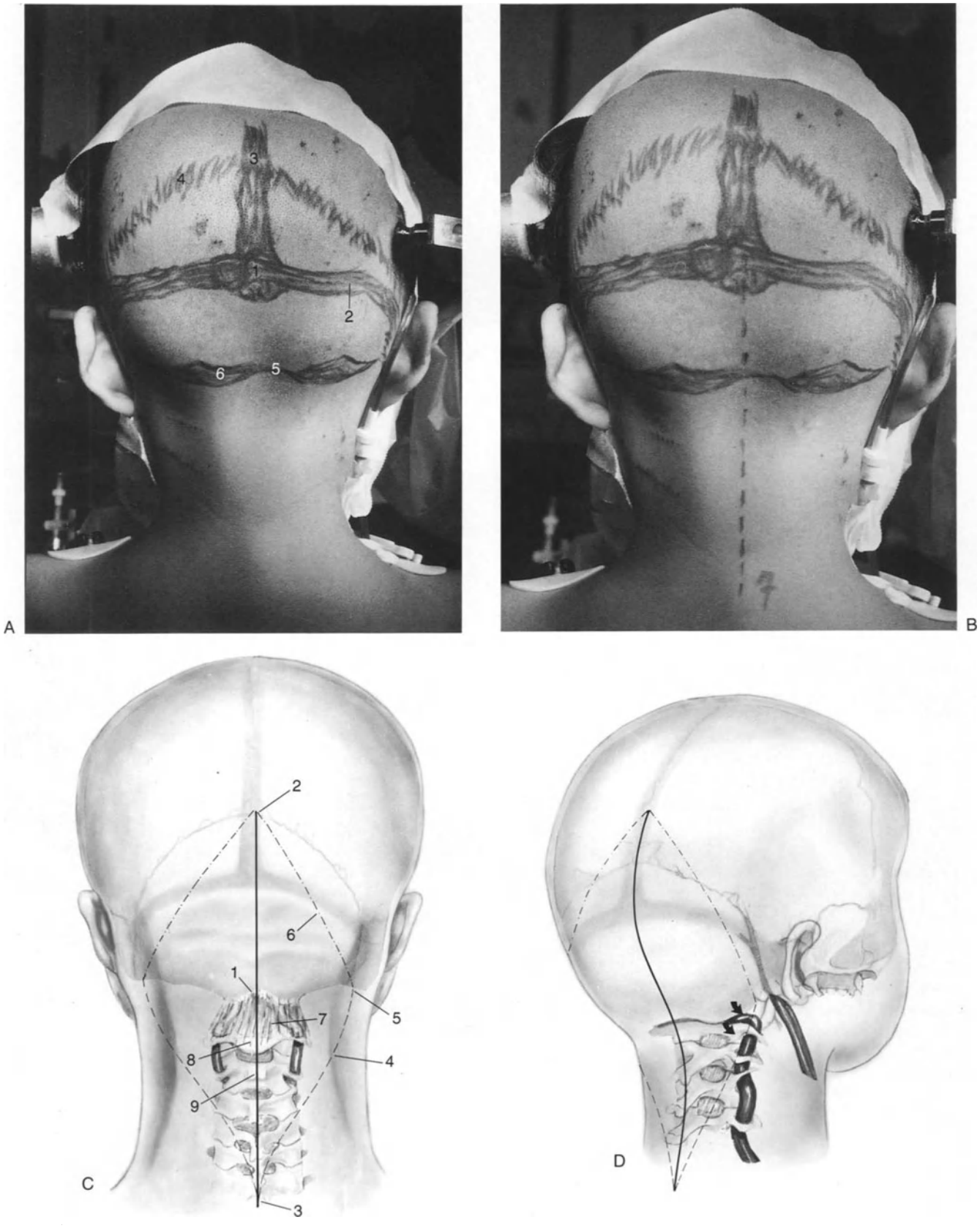
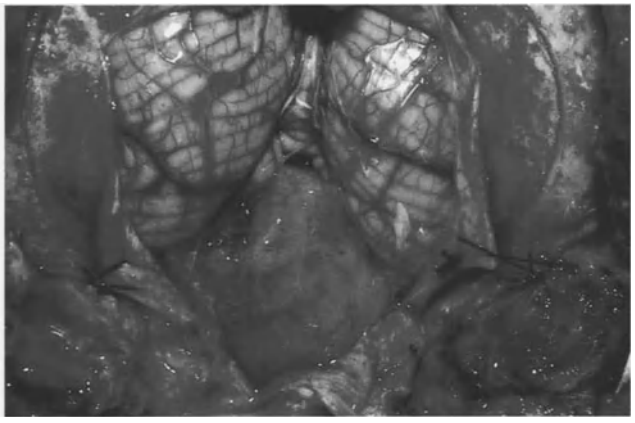


Figure 2.10. Legend see p. 51.



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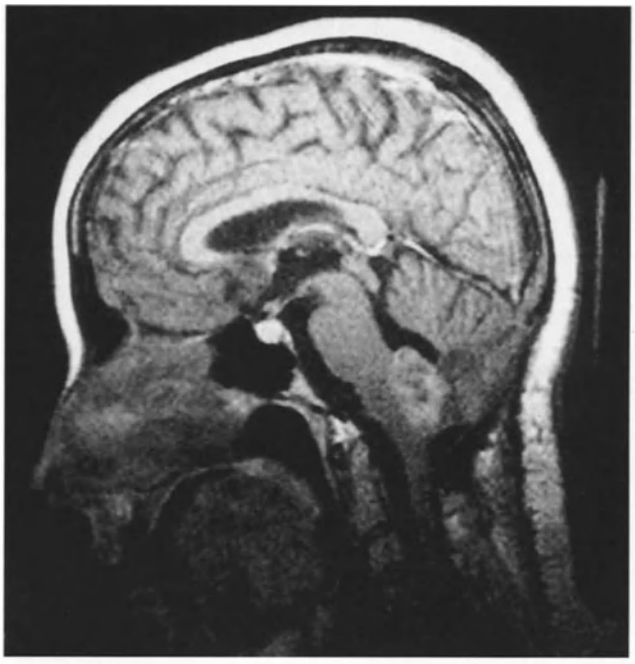

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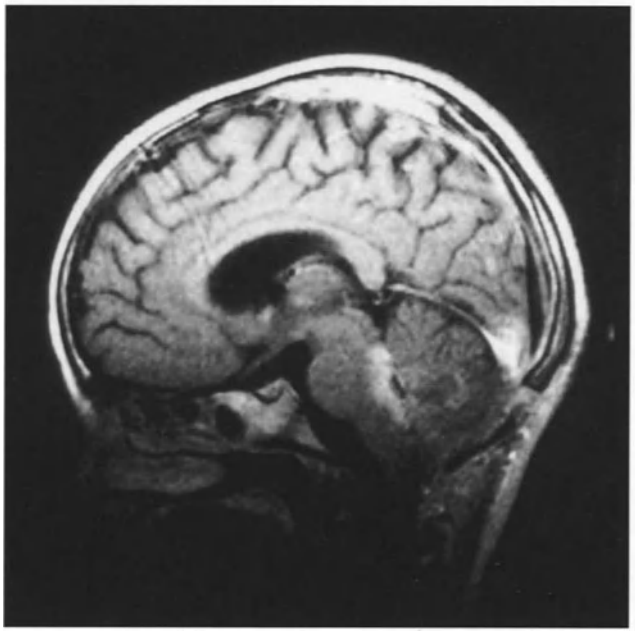


G

◀ **Figure 2.10.** (A) The significant landmarks for a suboccipital craniotomy have been drawn in. They are the torcular Herophili (*1*), the transverse sinus (*2*), the superior sagittal sinus (*3*), the lambdoidal suture (*4*), the rim of the foramen magnum (*5*), and the projection of the occipital condyles (*6*). Visual conceptualization of these landmarks permits one to plan for exposure of foramen magnum and inferior and superior cerebellar triangle lesions. (B) The skin incision (*broken line*) has been drawn in. It extends from the inion to the level of C-7. (C) The skin incision (*black line*) is shown in this transparency drawing of the skull and cervical vertebral column, as seen from the surgeon's point of view. The incision's center is at the rim of the foramen magnum (*1*), its upper extremity at the inion (*2*), its lower extremity at about C-7 (3). One may envision that retracting it (*4*), and the underlying erector capitis and cervicis muscles, as far laterally as the digastric grooves (*5*), exposes the entire squamous occipital bone (*6*), the atlanto-occipital membrane (*7*), the arch of C-1 (*8*), and the bifid spinous process of C-2 (*9*). The retracted skin and erector capitis muscles are indicated (-o-), as is the retracted skin (–) inferior to the level of the foramen magnum. It is not necessary to dissect the erector cervicis muscles from C-2, C-3, C-4, etc. (D) This oblique transparency drawing permits one to envision the curvilinear course of the skin incision from over the squamous occipital bone, onto the craniovertebral junction, and then along the spinous processes of the upper cervical vertebrae. The retracted tissue is indicated as in Fig. 2.13. The vertebral artery, and the entrance of Batson's plexus into the dural sinuses, is at the most lateral exposure of the field (*arrows*). Looking at (A) and (B) in the planning of the skin incision to remove (E) a *medulloblastoma*, (F) an *ependymoma* growing from the posteromost portion of the floor of the IV ventricle, or (G) an *astrocytoma* growing along the brachium pontis and expanding within both the ventricular surface of the vermis and that of the floor of the IV ventricle, an overall mental view of the muscular, extracranial vascular (venous and arterial) posteroinferior cerebellar artery and hind cranial nerves, brain stem, dural sinuses and foramen magnum is essential. (*Continued* on p. 52).

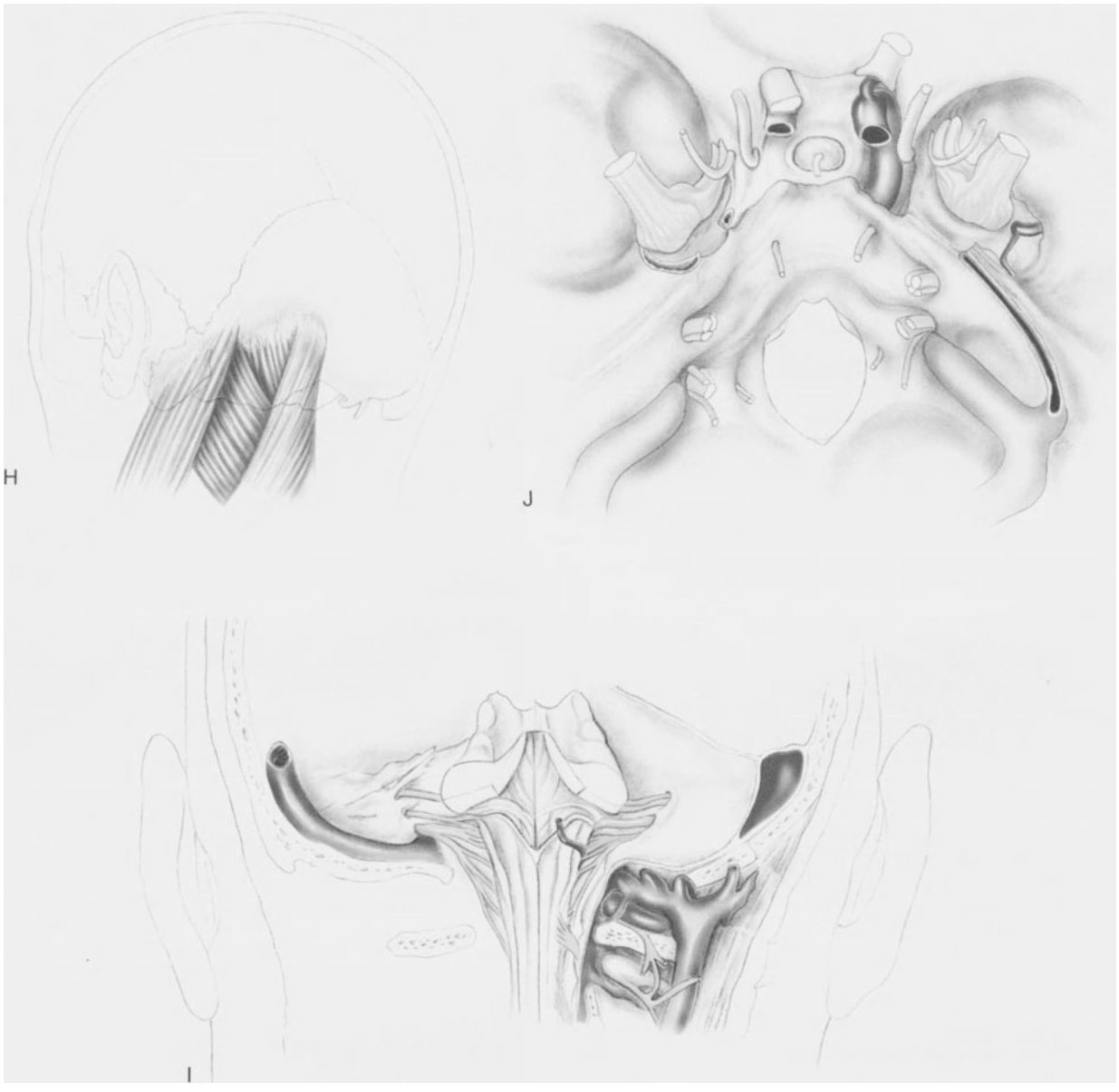
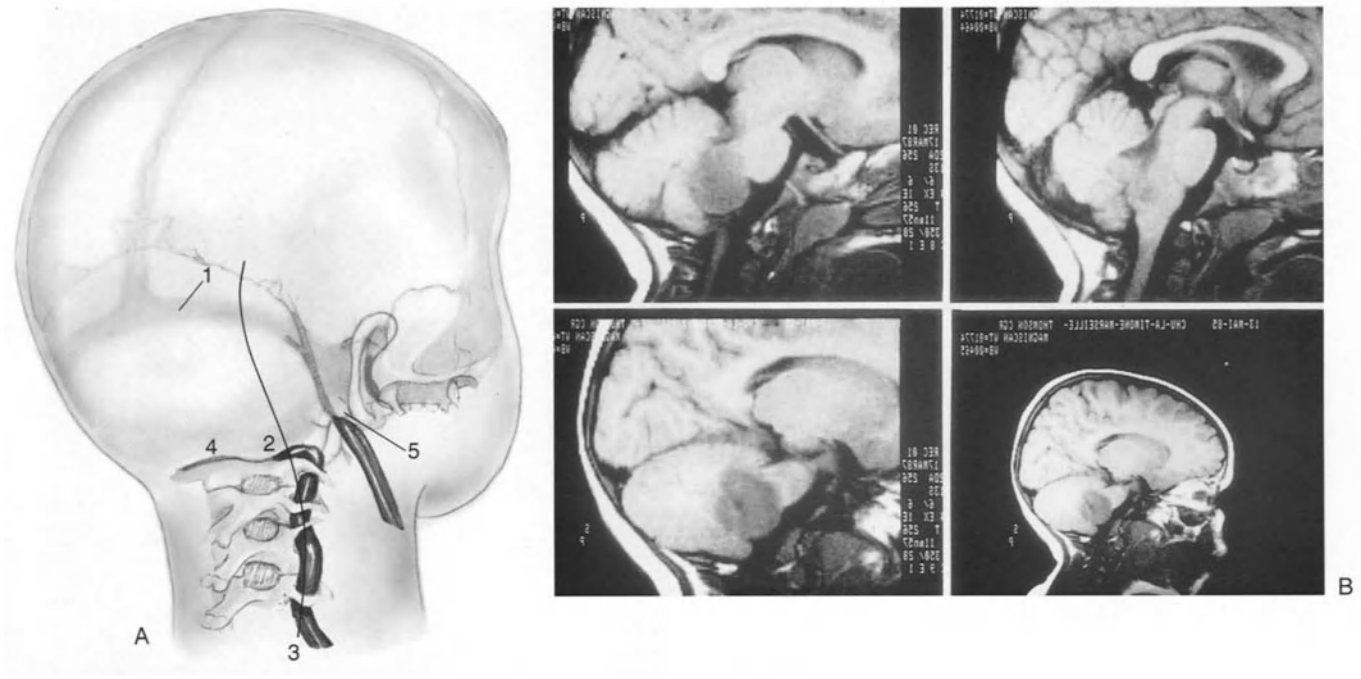
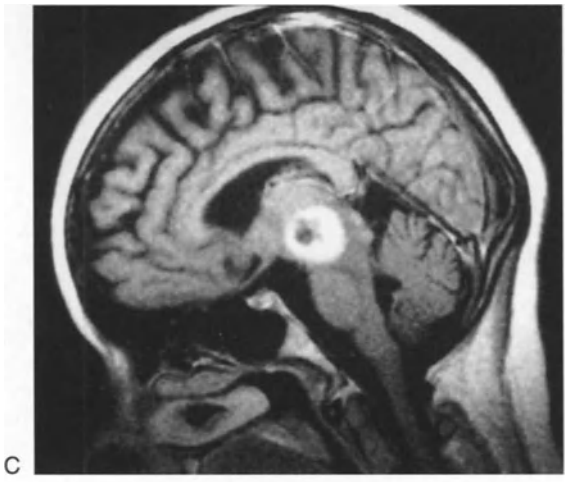


Figure 2.10. The muscular structures which must be taken down from the squamous portion of the occipital bone and the base of the mastoid process, separating their insertions from the bone and dissecting them laterally are, from medial to lateral, illustrated in (H). In preparing the squamous occipital bone, atlanto-occipital membrane, and the posterior arch of C1 for the suboccipital craniotomy and the dural opening, one considers continuously (I) the epidural venous plexus at the base of the skull laterally and the vertebral artery wedged between it and C1. Though the dural opening is no longer ever brought below the level of C1 for such tumors, this drawing (based upon the illustrations in Pernkoff's Atlas) permits an isolated conceptualization of IX, X, XI, posterior inferior cerebellar artery (PICA), the bulbospinal junction, and that portion of the floor of the IV ventricle which is either the origin, wrapped around, or invaded by the tumors illustrated in (E), (F), and (G). It is very unusual for the PICA not to be adjacent to or engulfed by these tumors. Lastly, this drawing permits the reader to identify the restiform body, the brachium pontis, and the brachium conjuctivum, coming thereby to the immediate realization that tumors which take origin in or invade the lateral surface of the IV ventricle invariably involve to a greater or lesser extent these three cerebellar peduncles. (J) is a drawing also made from the anatomical dissections illustrated in Pernkoff's Atlas; it is possible to envision how lateral extensions, through the foramina of Luschka, of *IV ventricle or brain stem tumors* displace or compress XII, XI X, and IX often; and VIII and VII occasionally. Bilateral extension of a IV ventricle tumor or inferior extension of a *cerebellar hemisphere tumor* involves anatomically the sigmoid sinus and/or the jugular bulb. It may be of help, or give very real satisfaction, to stop now and look back at (A)–(D) of this figure, interposing the tumors illustrated in (E)–(G) to appreciate fully the anatomy and pathology of these lesions, and the physiopathology of the pre- or postoperative symptoms and signs.





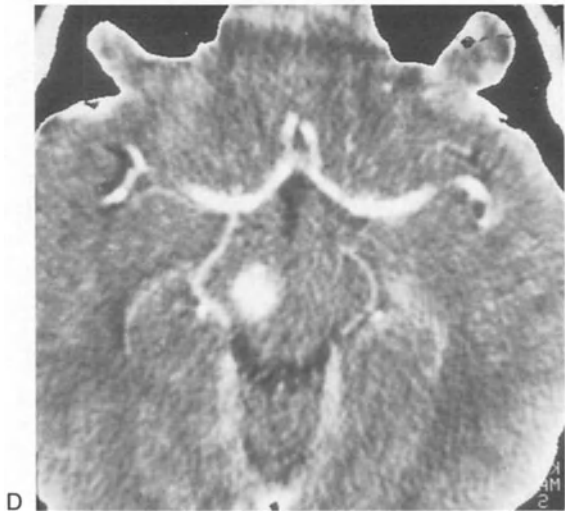


Figure 2.11. Children have as yet incompletely developed mastoid eminences, so the lateral suboccipital skin incision is more than adequate for inferolateral occipital craniotomy. In fact, one may, through this incision, work as effectively as through a far lateral approach in adults. (A) The lateral suboccipital skin incision is illustrated diagrammatically, here from an oblique view, in a transparency drawing. The incision extends from above the transverse sinus (*1*), across the junction of horizontal and vertical segments of the squamous occipital bone (*2*), and down to the level of the base of the neck (*3*). The incision is placed midway between the posterior rim of the foramen magnum (*4*) and the mastoid process (*5*). The (B) *bulbar glioma* shown here was "completely" resected through this opening and the (C) *pontine tumor* subtotally resected. The (D) midbrain *cavernoma* shown in this imaging study, however, must be approached through a skin incision which permits supra- and infratentorial exposure of the peduncles and the inferior mid brain, as illustrated in Fig. 2.22, for the lateral suboccipital skin incision illustrated in (A) only permits access to the inferior half of the pons and the reverse questionmark incision in Fig. 2.21 is much too extensive for such a discreet and small lesion as this. (E) see p. 54.

Figure 2.11. This (E) drawing of the view one has of the medulla oblongata, pons, and lower half of the mid brain with cranial nerves V, VII, VIII, IX, X, XI, and XII shows these structures projected within the infratentorial compartment as seen through the lateral suboccipital skin incision and craniotomy. The ease within which one may access the medulla oblongata and inferior half of the pons is as readily understood as the difficulties involved in working within the superior half of the pons and the mid brain.

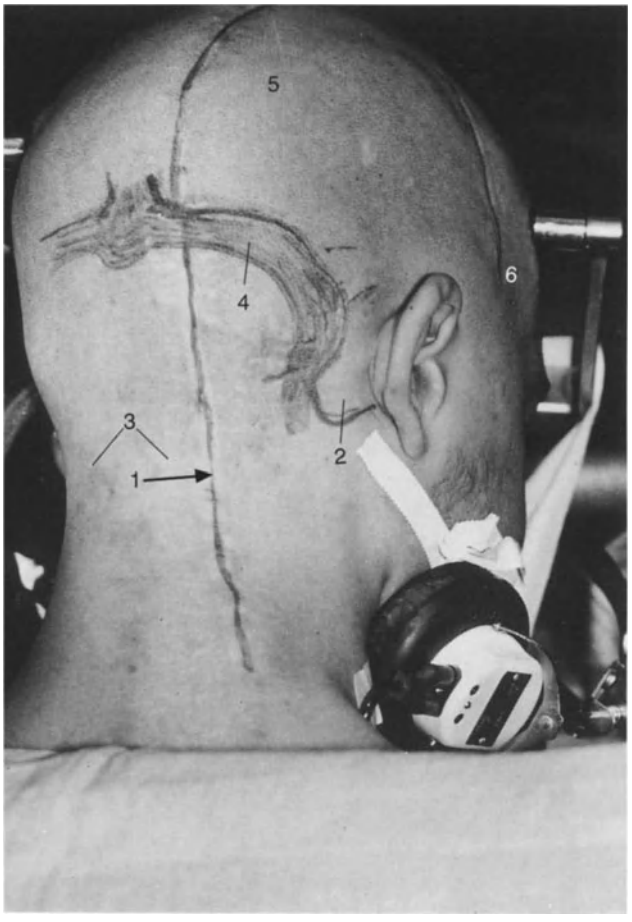


Figure 2.12. The questionmark skin incision for combined supra- and infratentorial approach to the tentorial ring at the anterior clinoid and the pontocerebellar angle is illustrated here, photographed from the anterior lateral perspective. Note that the vertical limb of the skin incision (*1*) runs midway between mastoid eminence (*2*) and the posterior rim of the foramen magnum (*3*). It crosses the transverse sinus (*4*), and then curves anterior to the parietal eminence (*5*) extending to the pterion (*6*).

Combined Supra- and Infratentorial Incision
(Figs. 2.12, 2.13)

Draping. The draping is for a lateral suboccipital incision beneath the horizontal line of the base of the mastoid, and for an occipital incision above this line.

For such tumors as meningioma, acoustic neuroma, and glomus jugulare, which may grow within the supra- and infratentorial spaces as independent tumors, dumbbell tumors growing on either side of the tentorium, or particularly large extraparenchymal tumors extending into the supratentorial compartment from the pontocerebellar angle or into the posterior fossa from the rim of the tentorium, *one of two combined supra- and infratentorial incisions* may be used:

1. For lesions involving the tentorium, a questionmark incision may be used, whose vertical limb extends superiorly from approximately the level of C-4 to the inion, and whose curvilinear limb extends anterior to the parietal eminence and then inferiorly to over the temporalis muscle. It is not necessary for the vertical limb to be located in the midline. In fact, since tumors which extend into both the supra- and infratentorial compartments either grow from the tentorium or from the pontocerebellar angle, much is in favor of the vertical limb being located midway between the midsagittal plane and the apex of the mastoid bone.
2. *Access to glomus jugulare tumors* also requires consideration of a supra- and infratentorial flap. However, since the glomus jugulare tumor begins within the temporal bone, it is essential to place the skin incision so as to have access to the mastoid, petrous, squamosal, and styloid portions of the temporal bone. A "sine-wave"incision is used.

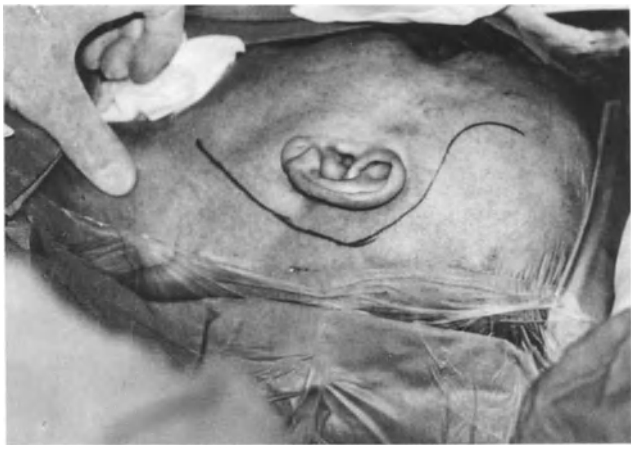


Figure 2.13. The "sine-wave" incision has been marked off. Note that it extends from the angle of the mandible, posteriorly to the mastoid apex, and then superiorly to the base of the mastoid, before turning anteriorly across the occipital and parietal bones to curve over the superior temporal line. This allows one to retract scalp, cervical skin, and ear anteriorly; and scalp posteriorly, thus exposing the mastoid bone and permitting entrance into the petrous bone. One has direct access to VII, VIII, IX, X, XI, and XII; to the jugular vein and bulb; to the carotid and ascending pharyngeal arteries; to the external auditory canal and the entire mastoid bone. Once these bones are removed, one has visualization of the sigmoid sinus, transverse sinus, the supratentorial and infratentorial compartments.

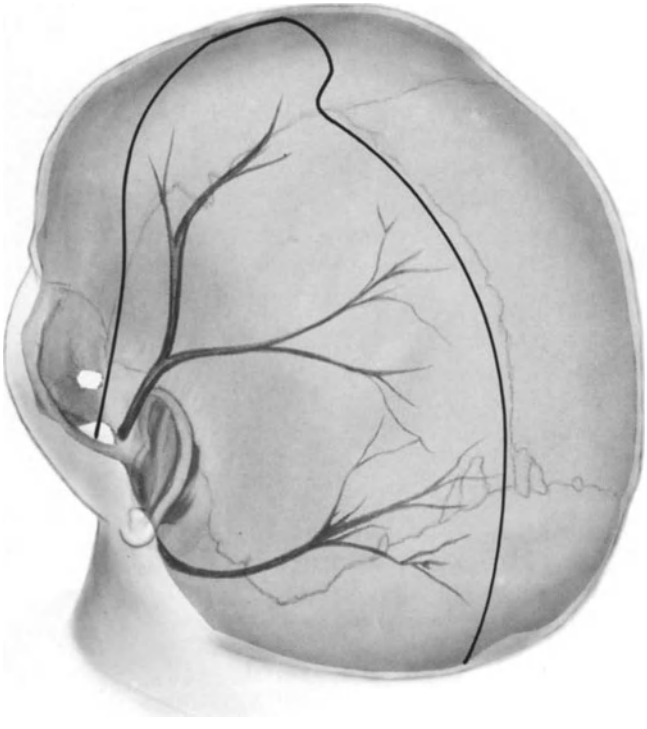


Figure 2.14. Placement of the skin incision for a hemispherical craniotomy.

Hemispherical Incision (Fig. 2.14)

The hemispherical skin incision permits the surgeon to expose half of the frontal and occipital bones and the entirety of the parietal bone, as well as portions of the greater wing of the sphenoid and the squamous temporal bones. This incision is used for hemispherectomies or hemicranial decompression.

Draping. The draping runs in a sagittal plane from the center of the contralateral supraorbital rim posteriorly to the highest nuchal line, then along this line to the base of the mastoid process on the operative side. From here, it is run around the insertion of the helix of the ear to the zygomatic arch, then along the lateral and superior rims of the orbit, medially, over to the opposite side.

Incision. The incision extends from the zygomatic arch approximately 8 mm anterior to the posterior spur of the antitragus, behind the hairline, and across the midline to the contralateral side. It is then brought back to the homolateral side and run approximately 1 mm lateral to the sagittal suture, across the inion and down to the external occipital protuberance. This assures adequate exposure and preservation of the integrity of the superficial temporal and occipital arteries.

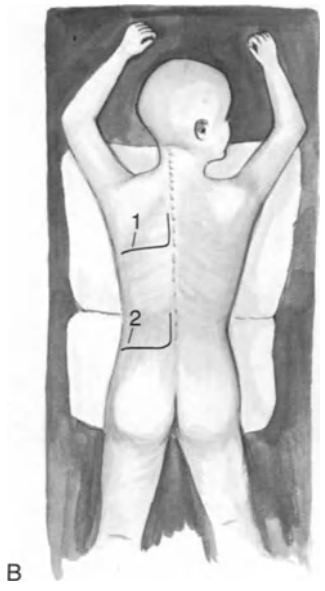
Laminotomy (Fig. 2.15)

Multiple-level laminectomies are an acceptable surgical approach to spinal cord lesions in adults, *not* young children. In most adult patients the procedure is not followed by instability of the spine. In children, however, multiple-level laminectomies may cause kyphosis, scoliosis, anterior subluxation, and instability of the cervical, thoracic, or lumbar spines. The development and physiological anatomy of laminectomy and laminotomy are discussed later, under those headings.

Draping. The draping is simple: paramedian, exposing the sagittal plane, and 2 cm laterally on either side.

Incision. The skin incision is midline, extending the full length of the planned laminotomy plus 4 cm cephalad and 4 cm caudad. If one is operating on a neuroma that extends into both the spinal canal and either the retropleural or retroperitoneal spaces, a "hockey-stick" incision is ideal. Its short limb is placed over the spinous processes, the long limb extended, with a curvilinear arch, over the rib cage or abdominal wall.

Several structures provide for the stability of the spinal column: intervertebral joints, laminae, lingamentum flavae, spinous processes, interspinous and supraspinous ligaments, and paraspinal muscles. In the adult,



◀ **Figure 2.15.** (A) The periosteal elevator has been used to strip the paraspinous muscles from the spinous arch of an infant (note the very small spinous processes). There is almost no resistance to the use of the periosteal elevator, since the paraspinous muscles attach along the median plane, not to the laminae. (B) The thoracic (*1*) and lumbar (*2*) "hockey stick" incisions permit access to the costotransverse and transverse processes, respectively, as well as the spinal canal, retropleural, or retroperitoneal spaces. Single-stage resection and dumbbell neurofibromas are possible with these hockey-stick incisions.

stability depends mostly on the anterior and posterior longitudinal ligaments, and the intervertebral joints, while the role of the other structures is relatively less important.

The vertebrae of the child are developing structures for which balanced mechanical stimulations are necessary to ensure normal growth. Spinal deformity and/or instability result from conditions in which bone and ligamentous deficiencies or neuromuscular imbalance occur. Such conditions may be caused by multiple laminectomies that destroy growing bony structures (laminae and spinous processes), that separate interlaminar and interspinous ligaments from adjoining vertebral arches, and that *substitute scar tissue for insertion of paraspinal muscle masses onto the laminae and spinous processes.*

After the skin incision has been made and clips applied to the subcutaneous connective tissue, the very thin paraspinous muscles are cut from their insertion along the midline of the vertebral arch and then stripped free.

Muscle and ligamentous attachments are separated from the spinal arches, leaving the periosteum and interspinous ligaments intact. The dissection is carried laterally to just beyond the articular facets, with care taken not to open into the joint or strip the capsular ligaments. The closure is facilitated if one leaves a ruffle of muscle and ligament on the spinal apophyses.

In newborn and infants, there is no, or very little, spinous process, and the laminae are both narrow and thin. The paraspinous muscle masses are minuscule. Hence, one should use a small periosteal elevator to separate the paravertebral muscles from the vertebral arches, which are encountered immediately the incision is made.

Techniques for Scalp Hemostasis in Various Ages: Newborn, Infant, Toddler

Skin (Figs. 2.16, 2.17)

The scalp is composed of *s*kin, (dense) *c*onnective tissue, *a*poneurosis (galea), *l*oose connective tissue, and *p*eriosteum. Hence the acronym *scalp*. In this text, "scalp" will be used to designate all of these anatomical layers as a single group, and each will be given its specific anatomical name (e.g., skin, dense connective

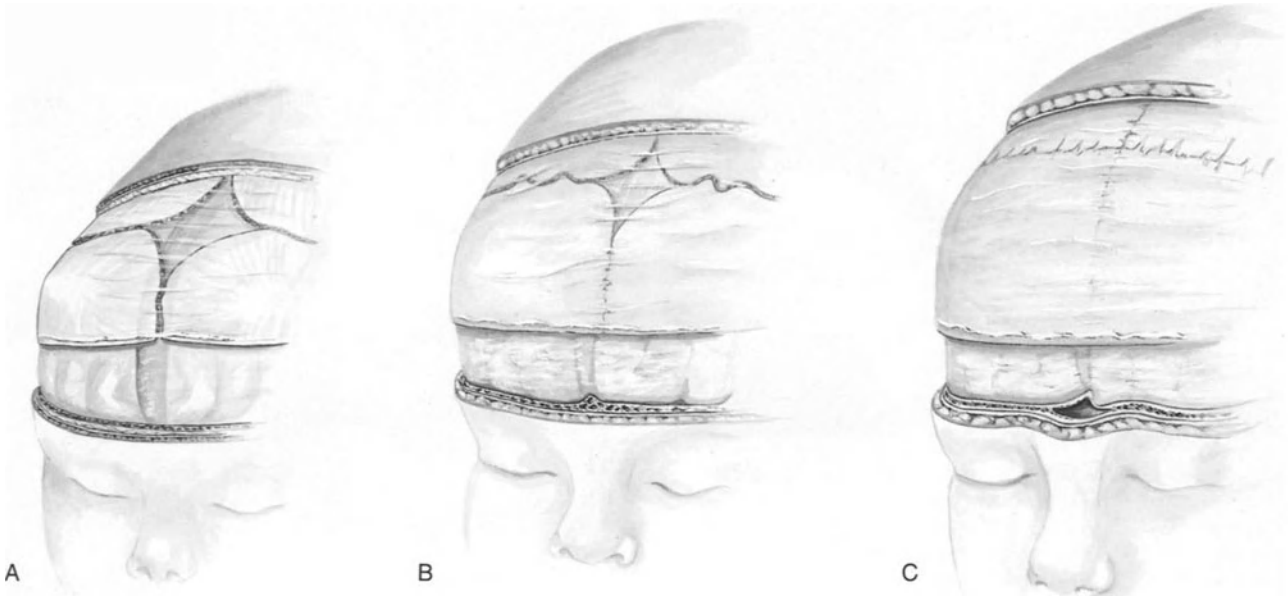


Figure 2.16. The relative thickness of scalp, presence of fontanelle, and state of the sutures in the newborn, infant and toddler. Note that in the newborn the skin, connective tissue, and aponeurosis (galea) are quite thin, whereas the fontanelles and sutures are open. With increasing age, the skull thickens, the sutures close, and the periosteum and the outer layer of the dura become readily separable from the skull. (A) Newborn: the skin and connective tissue are very thin, and there is an abundance of loose connective tissue, explaning the mobility of the outer layers of the scalp, the periosteum, and bone. (B) Infant: the connective tissue thickens and becomes more vascular, the fontanelles diminish in area first and then ossify, and the sutures remain open though they are firmly adherent to one another. (C) Toddler: both skin and connective tissue thicken, as does the aponeurosis (galea). The sutures close, and the skull develops readily identifiable tables (outer and inner) and diploë.

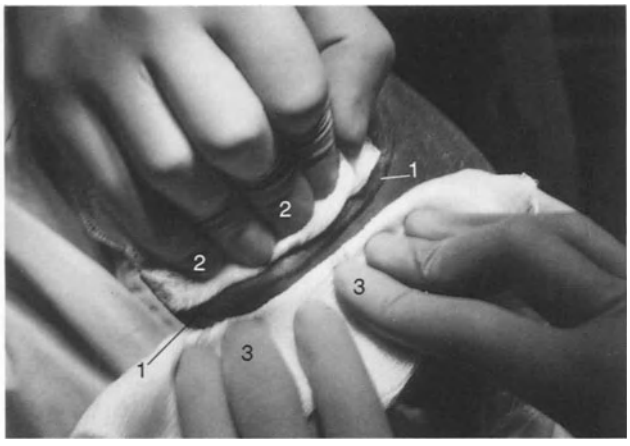
tissue, galea) when referred to as individual components of the *scalp*.

The anatomical differences of the scalp, the suture lines, and the state (open or closed) of the fontanelles in the newborn, infant, and toddler determine the differences in technique for skin incisions, hemostasis, and closures. In the newborn the scalp is palpable as two distinctly different functional entities: (1) a very thin and highly mobile entity composed of skin, dense connective tissue, and relatively avascular galea; and (2) an equally thin anatomical continuum of periosteum and skull, which is eggshell-like in compliance and highly vascularized. The direct continuity of periosteum and outer layer of the dura with one another at the suture lines, the very real mobility of one squamous (occipital, parietal, temporal, etc.) skull bone upon the other, and the presence of fontanelles of varying size offer little or no safe resistance to a blade cutting through the mobile portion of the scalp (skin, connective tissue, galea).

In Fig. 2.16, showing scalp characteristics, the presence or absence of a fontanelle, and the state of sutures, are illustrated in a comparative manner so as to put into relief the existence of thin scalp, the presence or absence of fontanelle, and opened or closed sutures. The very thin scalp, open fontanelles, and sutures are illustrated in Fig. 2.16A, thickening of the scalp and disappearance of the fontanelles are illustrated in Fig. 2.16B, and further thickening of the scalp with closure of the sutures is illustrated in Fig. 2.16C.

Although use of the #10 blade may be acceptable in the adolescent, it is heavy, and consequently the cut may be too deep for use in either the newborn or infant. Depending upon the size of the toddler, either a #10 or a #15 blade may suffice, but, by and large, the #15 blade provides a greater degree of safety and precision.

When using either the #15 or #10 blade, apply simultaneous compression and retraction, compressing the scalp with the pulp of the fingertips, and pulling the scalp away from the line of incision. This allows the surgeon to cut through the skin, dense connective tissue, and the galea to the level of the loose connective tissue without incising periosteum. It is more important to do this in the newborn and infant than in the toddler because of the great vascularity of the periosteum in the former two age categories. Also, in the newborn deep cuts may not only penetrate the periosteum but, at suture lines, may perforate the cranial and dural barriers, cutting through to the cerebrum. Because of the pathological thinness of the skull and scalp in hydrocephalic newborn and infants, particular caution must be taken when incising the scalp in these children, lest one cut through periosteum and suture. Suffice it to remember



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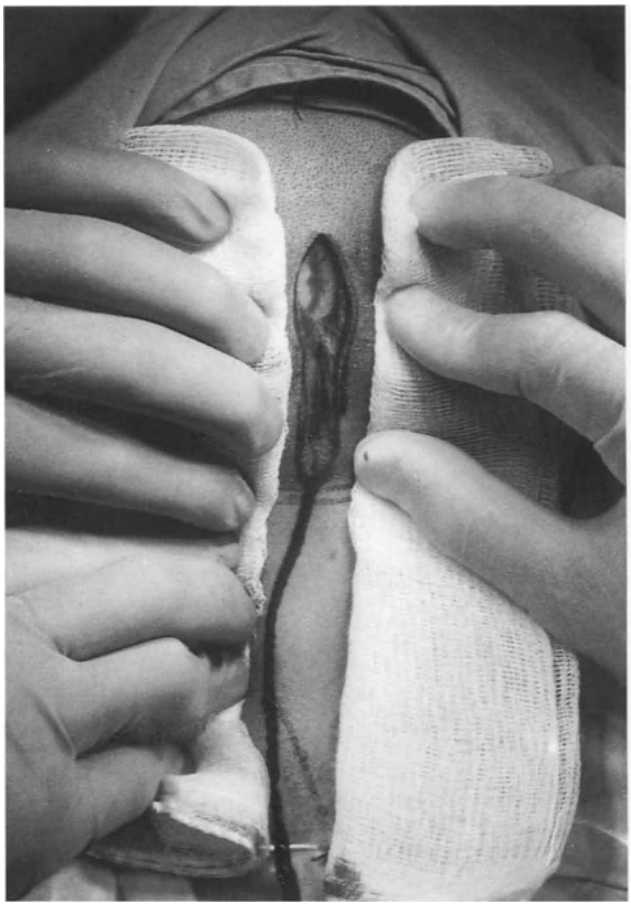


B

Figure 2.17. Digital compression along incision line. (A) The incision line (*1*) has been drawn with a marking pen. The force of the compressing distal phalange's pulp is exerted at the top (*2*): desirable; the pressure and digital spread at the bottom (*3*) are not such as to assure maximal hemostasis: undesirable. (B) Simultaneous compression and retraction with the fingertips. Note that the fingers compressing the scalp *on the reader's left* are "cutting" into the skin, with the compression being exerted by the fingernail and fingertip. This is not as effective in occluding intradermal vessels as when the pulp of the surgeon's distal phalanx is used, as illustrated *on the reader's right*. The distance between fingers (*on the right*) is undesirable, but that illustrated *on the left* is most effective.

that the sutures may be open from a few millimeters to centimeters, so that the suture line – namely, periosteum and the outer layer of the dura – a purely membranous structure, is the only anatomical barrier between galea and the arachnoid over the surface of the brain.

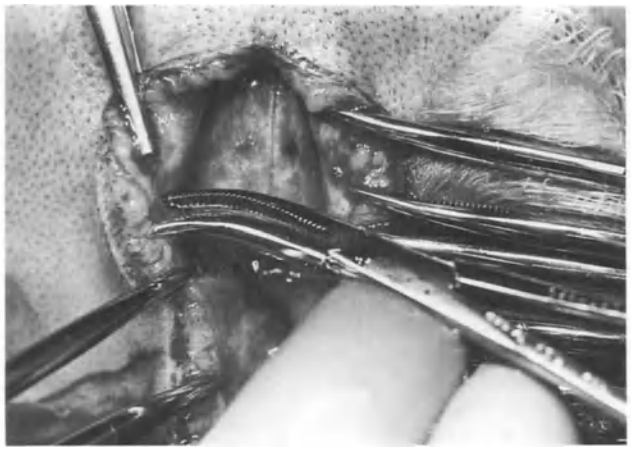
When compressing the scalp (during the time of skin incision), one must take care to apply *gentle* pressure, since the underlying skull is thin and fragile, susceptible to fracture. Consequently, the pressure put upon the scalp should be just enough to allow the surgeon, or his assistant, to feel the underlying skull, so that, in this manner, the underlying squamous bones will neither be fractured nor pushed into the surface of the brain.

In the toddler, irrespective of the degree of intracranial pressure and the presence or absence of split sutures, fully formed bone (with periosteum on its external surface and outer layer of the dura on its parenchymal surface) is interposed between the galea and the brain. Therefore, one may bring the cutting blade to the bony surface. In the toddler, juvenile, and adolescent, scalp compression must be more forceful, so as to wedge smartly the vessels within the loose connective tissue of the scalp between the surgeon's fingers and the skull.

Galea (Fig. 2.18)

The application of Kolodny and Dandy clips (to the galea) for scalp hemostasis in the newborn and infant is of no value because of the relative enormity of these instruments, the thinness of the vessels within the dense connective tissue between the skin and the galea, and the frailty of the galea. Consequently, toothed galea clamps permit one to grasp the cut galeal edge without forcing the jaw of the clip into the dense connective tissue. These clips are also advantageous in that they are half the size of the Dandy or Kolodny clamps, and are made of light aluminum alloy. They neither take up a great amount of space over the small cranium nor weigh heavily on it.

After the clips have been applied to the galea, at approximately 4-mm intervals, and fastened to one another by a rubber band, they may be used to retract the scalp flap. Caution must be taken not to pull on them, since the galea in the newborn and infant is almost membranous and, consequently, may easily be torn, allowing bleeding to continue. The clips are applied serially, at 10-mm intervals, and then fashioned into a retractor, as they splay the scalp flap over an underlying roll of gauze which both facilitates hemostasis and pre-



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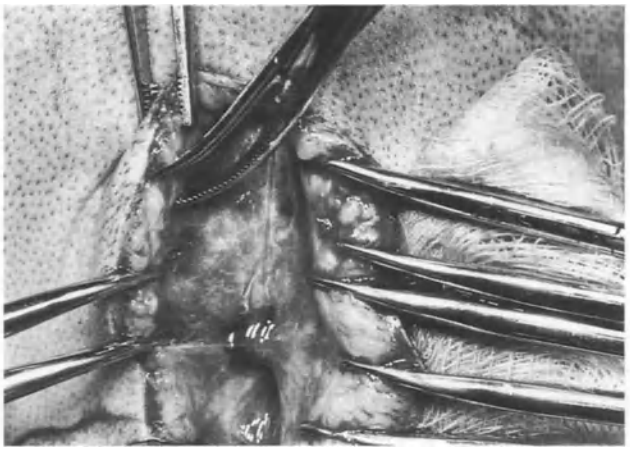

B

Figure 2.18. (A) Technique for applying the galea clips, taking care to grasp only the galea with the jaws of the clip so as to avoid digging into the dense connective tissue or penetrating the skin. (B) Same as A, but a cone down view.

vents kinking of the scalp flap, something that could result in devascularization of the flap.

Loose Connective Tissue and Periosteum
(Figs. 2.19, 2.20)

After the skin incision has been made, the separation of the galea from the periosteum is quite easy and may very often be completed by blunt dissection with a gauze sponge. This is rough and damages the tissue. Moreover, it causes bleeding from the galea and the periosteum, tearing vessels within and between the tiny vessels over a large surface area, which escape damage when a cutting blade is used. In bifrontal flaps, for example, tearing the periosteum and stripping it from the underlying bone causes added bleeding from the bone surface, frays intact periosteum which may be needed at the end of the procedure for dural reconstruction, and contuses or disrupts the superior frontal branches of the facial nerves. This occurs because these branches pass into the scalp at the superior orbital rim, just as they exit the orbit through the supraorbital foramen or groove.

One may save the frontal artery, nerve, and vein by using sharp dissection of the loose connective tissue which bridges the potential space between the periosteum and galea. If this is done, the preparation of a periosteal flap may be effected, since it is fully preserved.

The periosteum should be cut with the sharp, not the flat, edge of the monopolar thermocautery unit. Specifically, the periosteum may be incised 4 or 5 mm distal to the supraorbital ridge and then stripped from the frontal bone down to the supraorbital ridge, taking care not to extend the dissection into the supraorbital groove. In children old enough to have a developed frontal air sinus, the periosteal flap is extended from the glabellar region so as to preserve enough periosteum to sew over the air sinus and onto the dura in the event the air sinus is opened. This periosteal flap, if not used to cover an opened air sinus, is then reapproximated over the supraorbital ridge and sutured to the periosteum of the free bone flap at the time of closure, approximating the periosteum to the craniotomy line and bringing it over the bur holes. This assures complete union of the free bone flap to the surrounding skull.

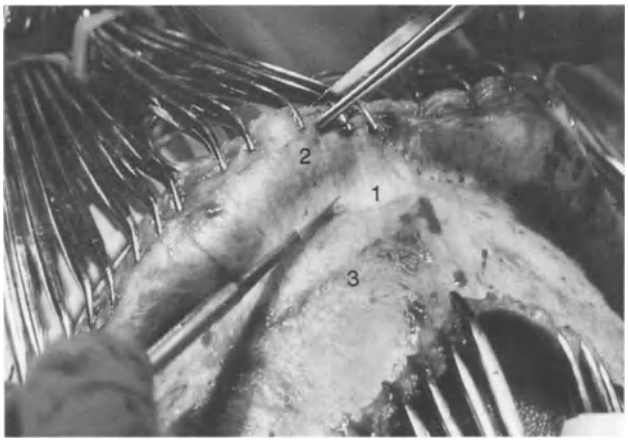

Figure 2.19. The loose connective tissue (*1*) is cut with a #15 blade, preserving galea (*2*) and periosteum (*3*). The frontal portion of a bifrontal flap is being separated.

As will be described subsequently in the section on craniotomy, the craniotome is not to be used to cut bone because it produces a gutter that impairs healing and leaves the cranial vault weak. Frontal, bifrontopterional, temporal, and suboccipital flaps all necessitate incising fascia and separating it, at one point (along one line) or another from the periosteum, with which it is continuous. This provides an adequate amount of tissue purchase for anchoring the free flap during the closure.

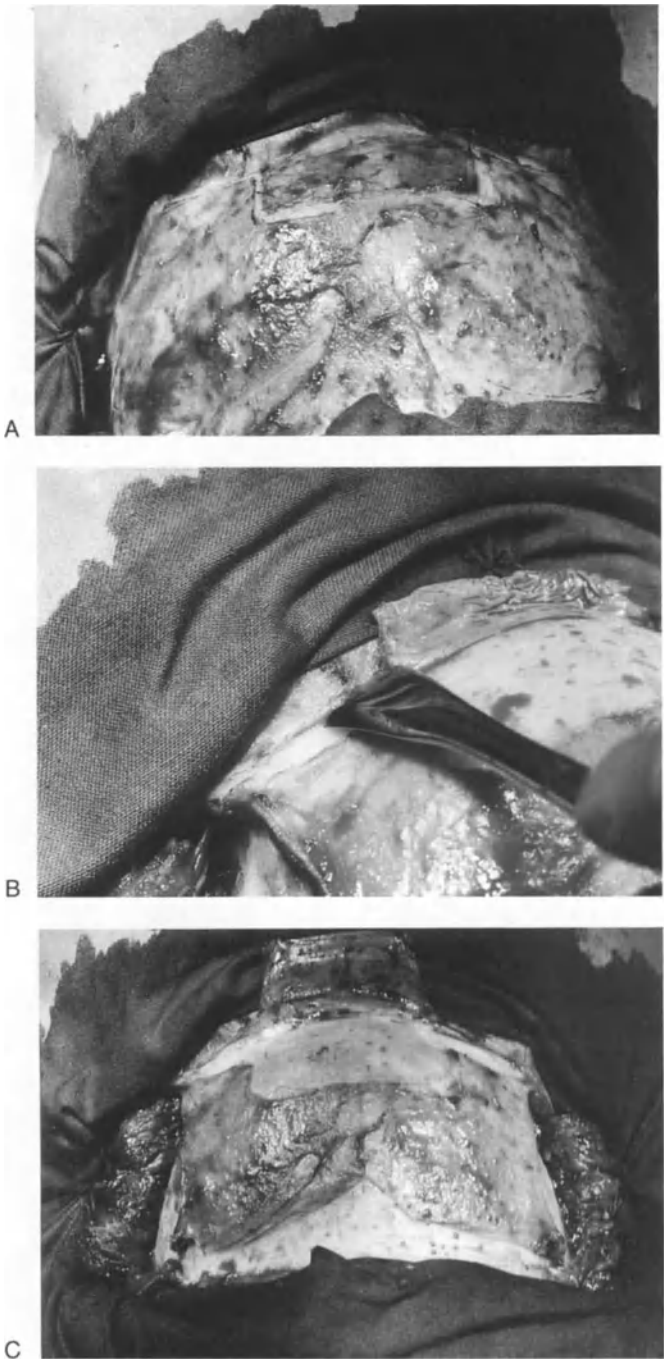

Figure 2.20. A periosteal flap, large enough to be sewn over the frontal air sinus and onto the dura, so as to protect the epidural space from empyema, is being fashioned. (A) The periosteum has been incised. (B) The periosteal elevator is separating periosteum from the superior orbital ridge without damaging the superior orbital artery, nerve, or vein. (C) The periosteum has been incised and stripped from the skull along the planned craniotomy line.

Of great importance to healing and protection from infection is leaving the periosteum on the bone flap, since stripping it away with the galea, dense connective tissue, and skin devitalizes the bone completely by separating it from all macro- and microvascular channels. Preserving the periosteum on the bone flap, and incising the fascia just before it passes into periosteum, assure purchase for future closure without resorting to drilling holes. It also affords maximum protection against bone (periosteal) bleeding and postoperative infection.

Temporalis Muscle (Figs. 2.21, 2.22)

When soft tissue preparation for reflection of the bone flap entails incision of the temporalis muscle, the latter may be performed in a relatively bloodless manner by using a unipolar cutting blade to incise the fascia and muscle immediately posterior to the zymaticofrontal suture. This cut should run superiorly for about 5 or 6 mm, and then extend posteriorly and parallel to the superior temporal line. If the incision is made at a distance of approximately 3 mm from the superior temporal line, the bleeding will remain minimal. This provides the surgeon an adequate amount of fascial tissue on the free frontal bone flap for purchase for the sutures, which will bring the temporalis muscle back into anatomical position at the time of closure. The temporalis muscle should not be cut through its belly nor along its origin from the zygomatic arch. This devascularizes it.

Erector Capiti Muscle (Figs. 2.23, 2.24)

Dissection of the erector capiti muscles and the trapezius from the lowest and highest nuchal lines results in stripping the periosteum from the squamous portion of the occipital bone. If these are stripped with chopping or sawing movements, it becomes impossible to reapproximate them at the time of closure, adding to the dead space and increasing the risk of fluid collection. Reapproximation of the two muscle groups to one another at the midline and of both to the skull at the time of closure is greatly facilitated if the surgeon incises the fascial insertion parallel to the highest nuchal line and approximately 1 cm inferior to the inion. He may then extend this incision lateralward on either side for a distance of approximately 2 cm from the midline. It allows one to strip completely the squamous occipital bone of the erector capitis muscular attachments to the inion, leaving four flaps of musculotendinous tissue for closure. The stripping is then extended well lateral and inferior to the lambdoidal sutures, as far as the digastric grooves on either side, preserving musculotendinous integrity for complete anatomical osteomuscular reconstruction at the time of closure.

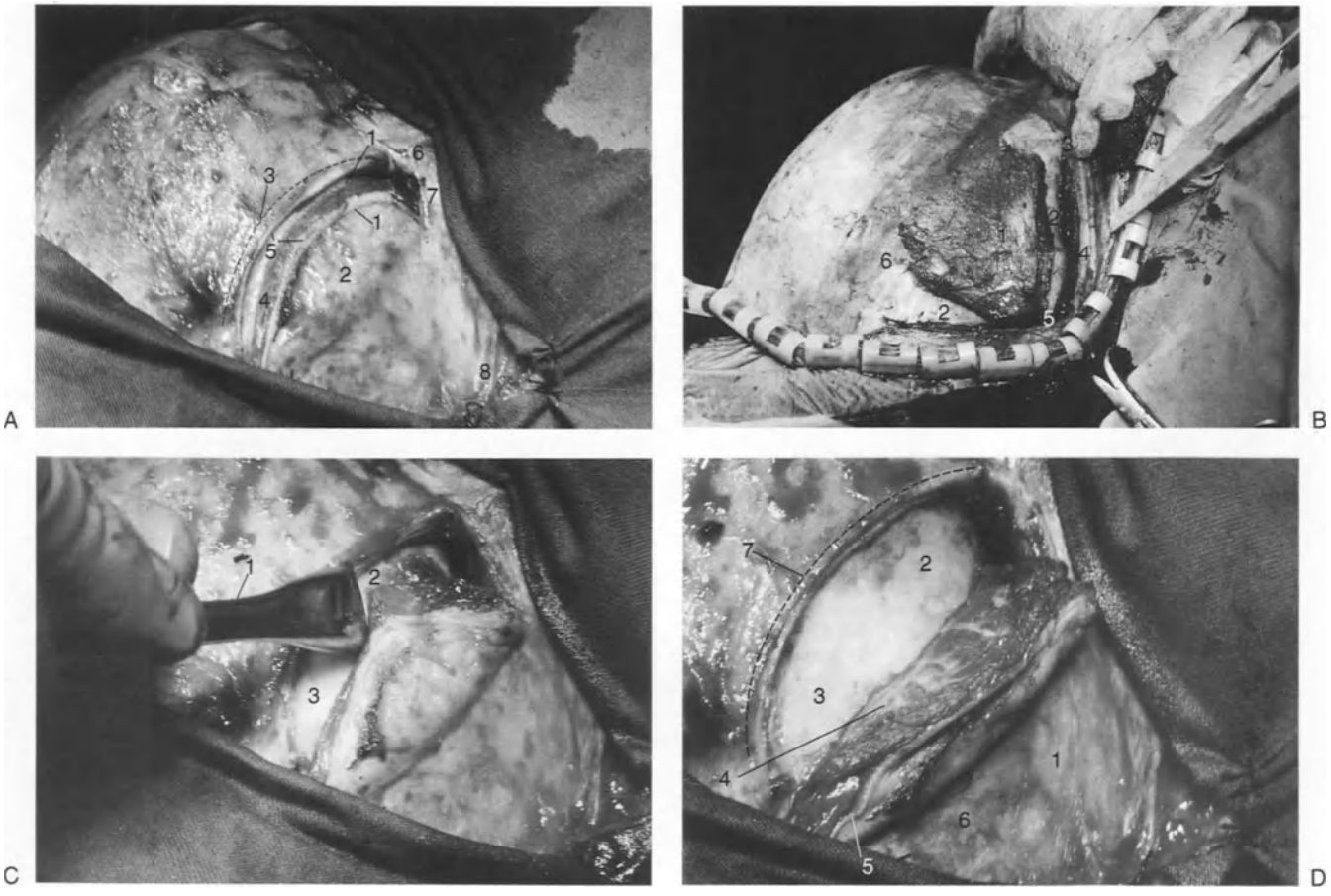

Figure 2.21. (A) Line of incision (*1*) in temporalis muscle (*2*), so as to minimize bleeding and facilitate anchoring bone flap into position at the time of closure. The fascia of the temporalis muscle has been cut about 3 mm from the superior temporal line (*3*) and then reflected to show full thickness of the bipenniform temporalis muscle (*4*). The cut fascial edges (*5*) will be sewn together at the time of closure, avoiding the need to perforate the bone flap edges. For orientation purposes, the zygomatic process of the frontal bone (*6*), frontal process of the zygoma (*7*), and zygomatic arch (*8*) are labeled. (B) The temporalis muscle (*1*) has been cut improperly (*2*), since the incision line extends from the frontal process of the zygoma (*3*) posteriorly across the zygomatic arch (*4*) to the pterion (*5*), and then superiorly to the superior temporal line (*6*). This brings the incision through the most vascular portions of the temporalis muscle, and, of still more negative and dangerous value, effectively devascularizes the mass of temporalis muscle adherent to the skull. (C) Suggested technique for use of periosteal elevator (*1*) to strip the periosteum from the greater wing of the sphenoid (*2*) and squamous temporal (*3*) bones, so as to avoid fraying the periosteum: cutting edge held firmly and run parallel to muscle insertion. (D) The bipenniform temporalis muscle (*1*) has been dissected from the greater wing of the sphenoid (*2*) and the squamous temporal (*3*) bones, preserving the deep (*4*), intermediate (*5*), and superficial (*6*) fascial layers. One may now appreciate the ease with which a suture may be brought through the full thickness of the temporalis muscle, to anchor it to the lip of periosteal and muscular tissue 3 mm from the superior temporal line (*7*) at the time closure.

Periosteum-Suture Lines

Within the limits imposed by the desired location and size of the bone flap in a toddler (and especially in an infant), it is preferable to dissect the periosteum across the sutures when the line of craniotomy runs perpendicular to them, but to leave the suture intact when the line of craniotomy runs parallel. If the suture is left intact, the craniotomy edges of adjacent bones (frontal-parietal, parietotemporal, etc.) may simply be lifted from their attachment to the suture line. Since the outer layer of the dura is continuous with the periosteum at the suture line, it is not possible to separate adjacent squamous bones as a single unit across a suture line as one does in an adult.

If the dissection is extended across the suture line, it should be blunt and performed with the use of a periosteal elevator. Sharp dissection should only be per-

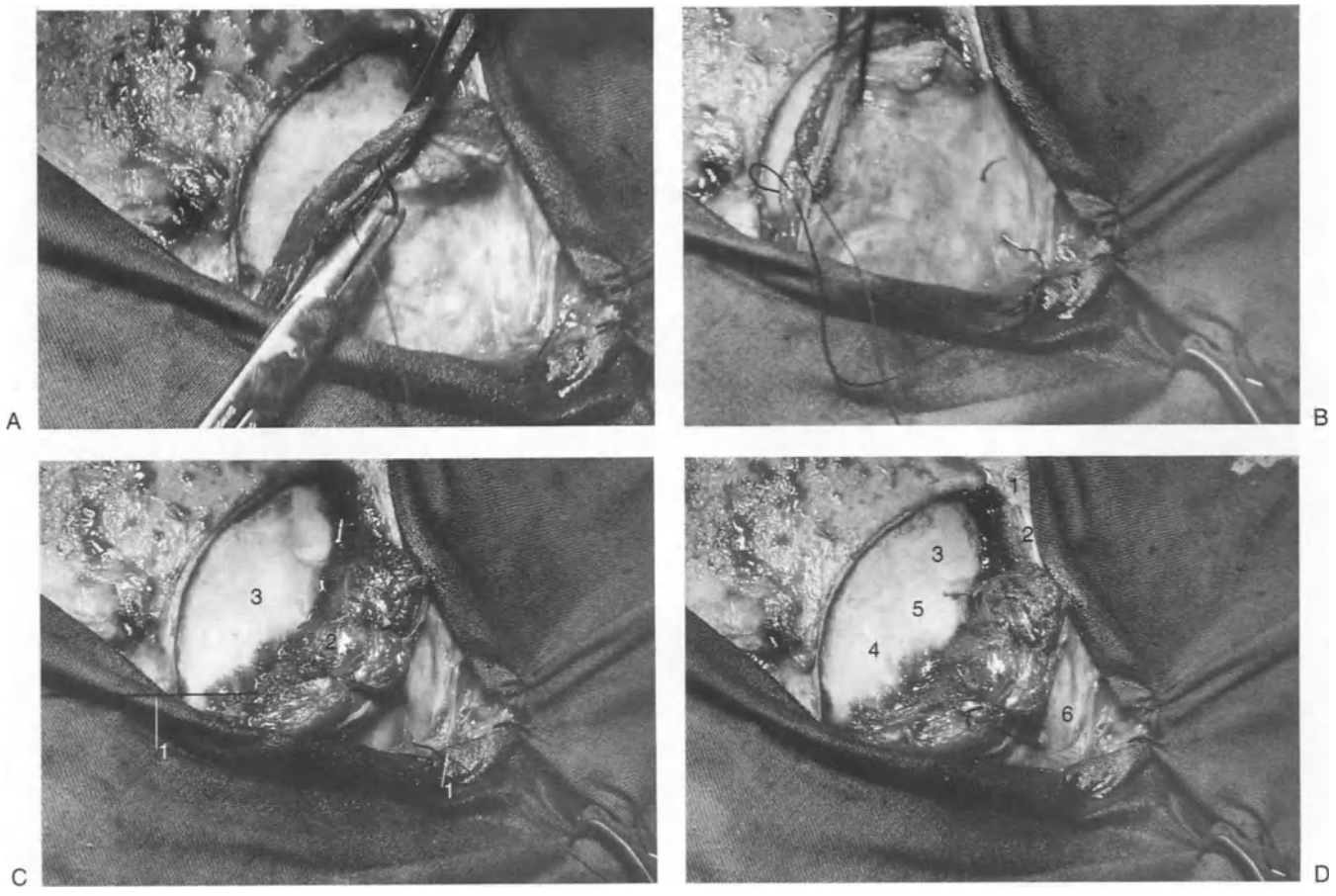

Figure 2.22. Technique for retraction of the temporalis muscle from the pterional region. (A) A suture is sewn through the full thickness of the temporalis muscle, which is held with a tooth forceps. (B) The suture is then brought over the surface of the temporalis and sewn onto the same muscle immediately over the zygomatic arch. (C) After the suture has been sewn through the full thickness of the temporalis (*1*) and over the zygomatic arch, it is drawn tautly to bring the flap of temporalis (2) away from bones bordering the pterional region (3). (D) Tying down the suture retracts the temporalis flap and provides hemostasis, as it exposes the posterior surfaces of the zygomatic process of the frontal bone (*1*), the frontal process of the zygoma (*2*), the greater wing of the sphenoid (*3*), the squamous temporal (*4*), and the pterion (*5*). For orientation purposes, the zygomatic arch (*6*) is labeled.

formed with a sharp-edged periosteal elevator (such as an Oldberg) or, at most, with a #15 blade, using it in a sweeping manner. Dissection of the periosteum in this manner minimizes bleeding from the highly vascular suture line. Electrosurgical units should never be used to cut across suture lines in newborn or infants: the cutting effect may extend deeply into the intracranial compartment, possibly damaging vascular or parenchymal tissue. In the newborn, as will be described in Chap. 4, it is much simpler to use heavy scissors, such as Mayo scissors, to cut suture or bone.

Techniques for Stopping Bleeding

The technique for stopping bleeding varies not only from subgaleal space to muscle to bone to artery to vein and sinus, but also with the location of the artery or vein (cortical, sulcal, cisternal, etc.) and the nature of the sinus (sagittal, cavernous). It is a sound principle *in surgery never to proceed to the subsequent level* (skin to bone, born to dura, dura to cortex, etc.) *until complete hemostasis has been attained.* The modalities for doing this are many.

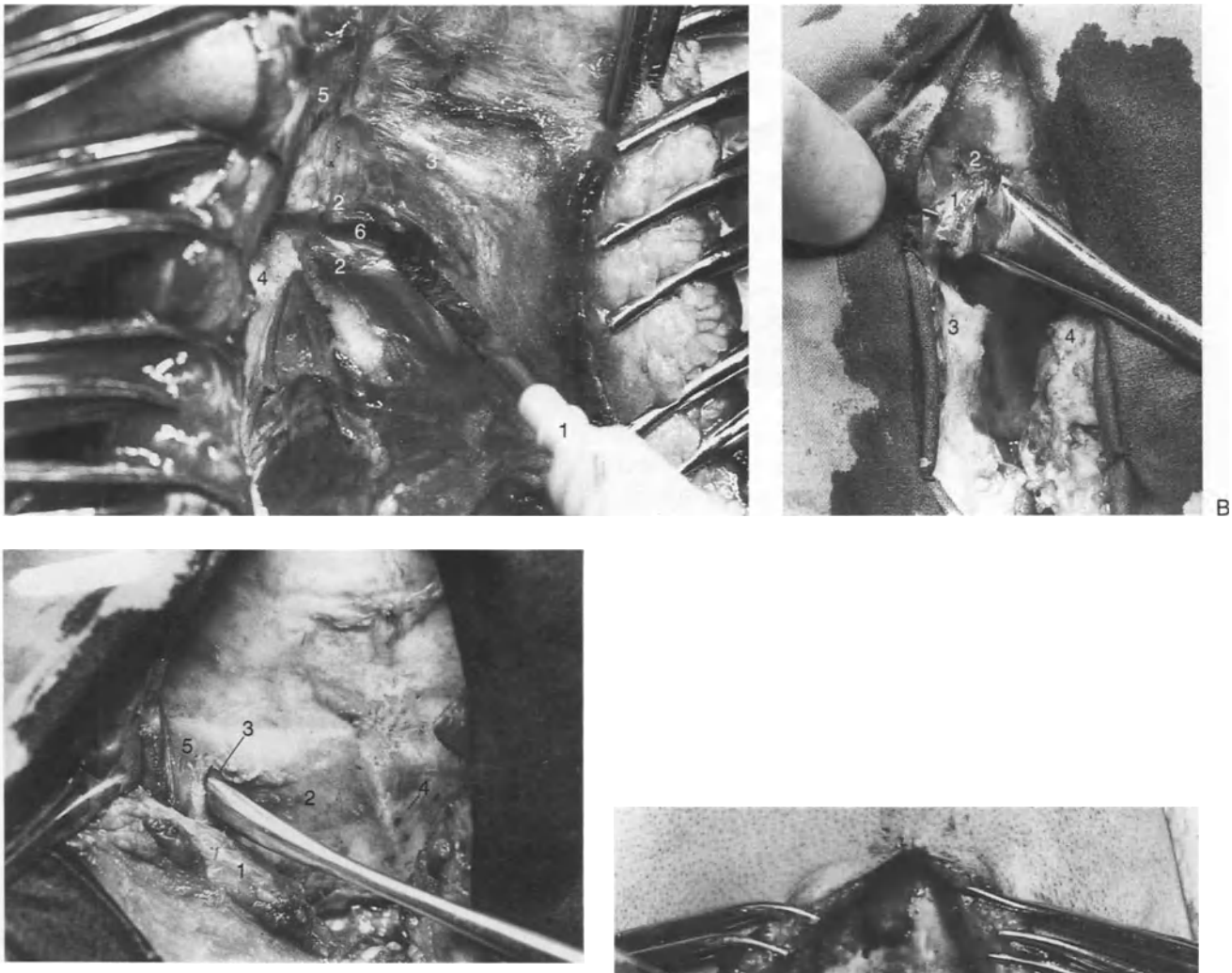

Figure 2.23. (A) Dissection of erector capiti and trapezius muscles from the occipital bone: a monopolar thermocautery unit (*1*) is used to cut the musculotendinous tissue (*2*) just proximal to its bony attachment at the highest nuchal line (*3*); this is done vertically first, and extended superiorly in the ligamentum nuchae (*4*) to the inion (*5*) before small incisions are made perpendicular to that cut (*6*). (B) The periosteal elevator is being used to free the left superolateral musculotendinous flap (*1*) as high as the inferior portion of the inion (*2*). The left (*3*) and right (*4*) inferolateral musculotendinous flaps have already been freed. (C) The periosteal elevator is being used to strip erector capiti muscles (*1*) from squamous occipital bone (*2*) as far laterally as the digastric groove (*3*), exposing the rim of the foramen magnum (*4*) and the mastoid process (*5*).

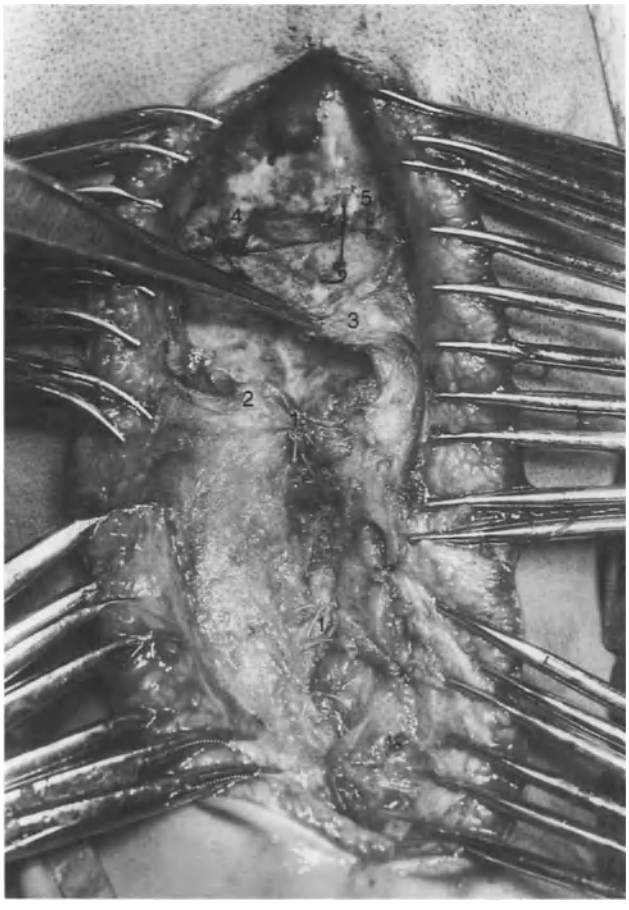

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Figure 2.24. The closure that may be attained by preparing four musculotendinous flaps. The line of closure is brought superiorly along the ligamentum nuchae (*1*) to the left (*2*) and right (*3*) inferolateral flaps, which are, in turn, closed. These latter are then anchored to the left (*4*) and right (*5*) superolateral flaps, thereby bringing the erector capiti muscles back into anatomical position.

Use of Cotton Fluffies (Fig. 2.25)

Irrespective of whether one has minimal bleeding from an arterial or venous structure, or whether this structure is located in the parenchyma, a sulcus, a cistern, or a pathological lesion, the application of soaked cotton "fluffies" to the bleeding area is extremely helpful in controlling or considerably limiting the bleeding. It almost invariably controls bleeding long enough for the surgeon to gather himself, his assistants, and his instrumentation together to deal effectively with the bleeding. The cotton "fluffies" should be fashioned according to the needs of the moment into larger or smaller, wider or narrower, longer or shorter pieces, and then dipped into warm saline immediately before application to the bleeding site. The application to the bleeding site should be performed by the surgeon holding the "fluffy" in forceps before bringing it down onto the bleeding surface. The suction should be brought into the area, but not applied to the "fluffy" until the "fluffy" has been pressed firmly against the bleeding area.

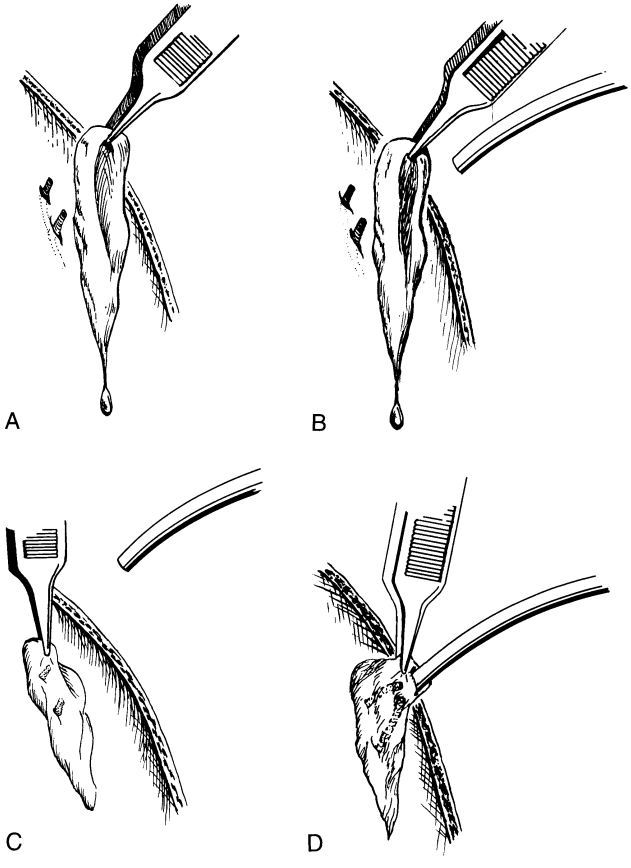


Figure 2.25. Use of fluffy cotton. (A) The soaked fluffy is brought to the bleeding surface. (B) The suction is brought into the field, but not applied to the fluffy. (C) The fluffy is pressed firmly against the bleeders. (D) The fluffy is sucked dry until it becomes glistening white, and then the forceps is released but the suction is held in place.

When this has been done the sucker is applied to it and suction continued until it becomes glistening white. The sucker is held firmly against the "fluffy" so that the "fluffy" gently compresses the underlying bleeding surface. One may, from time to time, remove the suction, leaving the dried "fluffy" in place, in order to see whether a spot of blood appears or whether the entire "fluffy" becomes red immediately. This gives information concerning degree of bleeding and specific bleeding site. Two, three, four, or more "fluffy" applications may be necessary to stop the bleeding. After this has occurred, it is advisable to leave the "fluffy" in place and to proceed with another part of the operation, returning within 3 or 5 min to remove the dried "fluffy" with Cushing forceps, using a Penfield dissector to elevate it from the surface that had been bleeding. At times the "fluffy" may be irrigated away and then lifted out with the forceps.

Use of Gelfoam, Surgicel, and Avitene

If fluffy pressure does not suffice to stop the bleeding, one uses the same technique, but applies Gelfoam, Surgical, or Avitene (microfibrillar collagen hemostat), applying one of these to the bleeding surface (from which the blood is being aspirated) and then immediately applying the fluffy over the hemostatic substance. The chronological order of availability of these substances was Gelfoam, Surgicel, and Avitene, which is in the inverse order of effectiveness. In fact, Gelfoam may be forgotten, Surgicel is of value in preparing the "sandwiches," which will be discussed in detail in the section on dural sinus bleeding, where one can appreciate its effectiveness. Avitene is absolutely essential! The proper techniques for its usage should be mastered. At the present time Avitene may be obtained either as a loosely packed mass or in densely compressed strips. The loosely packed Avitene must be lifted from the bottle and then applied dry to the bleeding surface. This is quite a "sticky" undertaking, since when the Avitene is placed against the brain or touches an instrument, it tends to adhere to them and become unmanageable. Consequently, one should apply the mass to the bleeding area and then cover it immediately with a "fluffy" cotton, which is used to pack the Avitene into the bleeding area. An alternative to this is to lay the Avitene over a wet gauze sponge and then to press it into a flattened form, which may then be more easily applied to a bleeding surface. The compressed strips of Avitene do not stick to instruments; they adhere snugly to the bleeding area. These are very easy to use. Both the loosely packed and solid strips of Avitene are helpful in sealing cerebrospinal fluid leaks at bony and dural levels.

Specific Types of Bleeding

Bone Bleeding

Bone bleeding differs considerably, whether the blood comes from the diploë, or the surface of the skull after the periosteum (internal periosteum) has pulled away. The child's skull is highly vascularized because of its continuous growth. Consequently, there is a great disproportion in bone bleeding from adult to adolescent, adolescent to juvenile, juvenile to toddler or infant. The infant and neonate bleed a great deal from periosteal and epidural (diploic or inner table of skull) hematoma.

Bone Surface Bleeding

Calvarial, inner or outer layer, bleeding is seldom of any real significance. However, clinical entities such as highly vascular tumors, transcranial arteriovenous malformations, and trauma may complicate the surgery by periosteal bleeding from the skull surface. When the bleeding is from the outer table, it is readily identified and may be stopped with relative ease. When, however, it comes from the inner table, it may be cryptic and most difficult to stop. This is the unpleasantly surprising cause and location of postcraniotomy and postshunt epidural hematoma. Bone edge bleeding is readily stopped with bone wax. It is immediate and permanent. Bone surface is less quickly stopped. However, the wax must be warm, so that it may be applied readily, without the need to exert force, so that it may penetrate the vascular channels of the smooth surfaces of the outer or inner tables. These tables are richly supplied by the periosteum; hence the genesis of hematomas when the bone and its periosteal covering, extracranial or intracranial, have been separated. The wax should be applied uniformly to the denuded surface of the skull, attaining a minuscule layer that coats the bleeding surface and penetrates the vascular channels. This is relatively easy for the outer table of the skull, but is almost impossible for bleeding into the epidural space, which comes from the inner table. Hence, for inner table bleeding, one must insert wet compressed strips of Avitene and then immediately sew the outer layer of the dura to the periosteum of the skull, thereby using the dura to tamponade Avitene against the bleeding surface of the inner table. If this is not done, the bleeding continues and pools in the epidural space, allowing the dura to become further dissected from the edges of the bone, producing persistent and irritating oozing throughout the operative procedure. It may even produce clinically significant epidural hematoma.

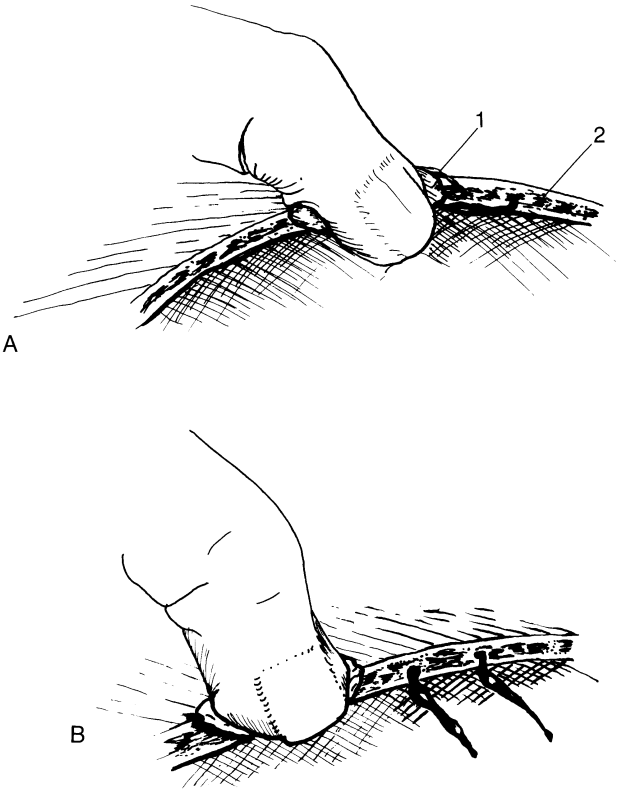


Figure 2.26. Recommended technique for applying bone wax to bleeding diploë. (A) The compressing fingertip is run along the bleeding surface at an angle of about 10°, sliding the wax (*1*) into the diploic (*2*) channels. (B) The pulp of the fingertip is pressed perpendicularly against the diploë as it passes beyond the dural surface and inner table of the skull. This prevents wax from being squeezed between dura and skull.

Diploic Bleeding (Figs. 2.26, 2.27)

The younger the child the more significant the diploic bleeding and the more care which must be taken to stop it. The diploë are relatively larger, and the outer layer of the dura is only loosely adherent to the inner table of the skull. Consequently, one may attempt to jam the wax into the diploë, because of the degree of bleeding, using wax which has not been adequately warmed. This results in stripping the outer layer of the dura from the inner table of the skull.

The correct procedure for applying wax to the diploë, consequently, must be adhered to fastidiously. This avoids packing wax between dura and bone. The compressing finger is angled approximately 10° from it; the tip will press most of the wax between dura and bone. Similarly, if the finger is run along the edge of the diploë, much of the wax will slip under the bone edges.

When wax slips between bone and dura and oozing occurs, the surgeon should stop immediately, take the wax from under the bone edges and tack the dura up to the surrounding periosteum (as already described),

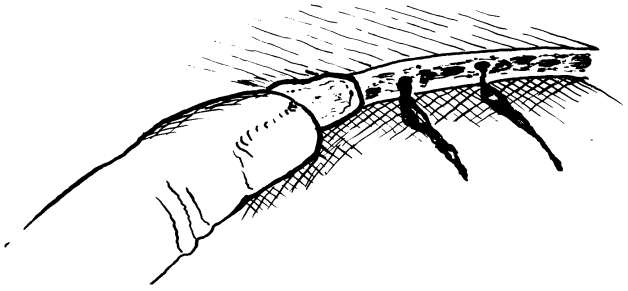


Figure 2.27. Incorrect application of bone wax. When the compressing finger is guided along the surface of the diploë, wax is jammed between dura and inner table, separating one from the other and adding to the degree of skull bleeding.

drawing the sutures tightly so as to stop the ooze. Because of the great vascularization of skull and periosteum in newborn and infants, delay in doing this may result in the formation of a truly significant epidural hematoma.

Dural Bleeding

Arterial Dural Bleeding (Figs. 2.28, 2.29)

The smaller arterial dural bleeders may be coagulated (under water)with bipolar forceps after they have been transected during the dural opening, but larger bleeders must either be clipped or ligated. Do not trust cautery to stop large dural bleeders.

Dural Sinus Bleeding (Figs. 2.30–2.32)

Dural sinuses must be ligated if they are to be transected! Small, almost microscopic, openings in the dural sinus may be occluded by using Avitene, but the use of such topical hemostatic agents is to be avoided if the opening is large enough so that a portion of the agent may bulge into the lumen of the sinus, since this may result in occlusion of the sinus.

If one encounters a large sinus opening in a location that precludes transection and ligature, the bleeding may be controlled by fashioning "sandwiches" made of Surgicel with either Gelfoam or Avitene at the center. The "sandwiches" are placed over the rent in the sinus. They are then pressed onto the surface of the sinus, using soaked cotton fluffies, which are immediately sucked dry with a large suction tip. This draws the walls of the sinus against the Surgicel "sandwich." It generally suffices to stop the bleeding. In the event this technique does not succeed, one may prepare a larger "sandwich," large enough to extend at least 1 cm to either side of the rent in the sinus, and anchor this into place with 4–0 sutures running from the dura on one side of the sinus to the dura on the other side. If the bleeding persists, a

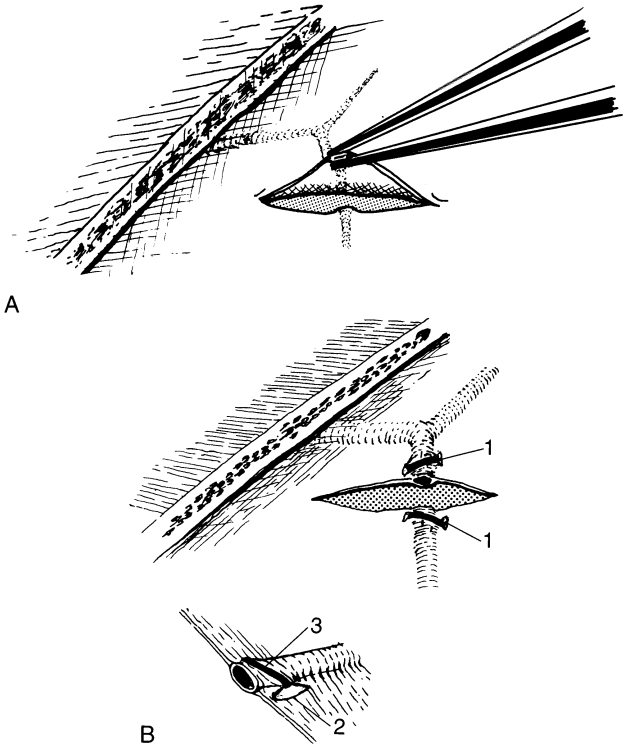


Figure 2.28. Dural arterial bleeders. (A) The smaller ones are coagulated between bipolar forceps, with the surgeon taking care to pick up the full circumference of the transected artery in both blades of the forceps. (B) Larger dural bleeders require clipping or ligature. Here, a hemoclip has been applied to the edge of the vessel (*1*). The clip may be applied by setting it obliquely, occluding the transected intradural artery on the bias. If this proves difficult, nicking (*2*) the dura parallel to the edge of the artery prior to application of the clip perpendicular to the long axis of the artery and parallel to the transected edge (*3*) offers effective and safe access to both sides of the vessel. Clips which are compatible with MRI must be used. The author prefers ligatures.

"sandwich" may be anchored over the dural rent with a pedicle (dural) patch graft swinging from one side of the dura, over the "sandwich" and rent in the sinus, to the other side of the dura, where it is anchored. In extreme instances, it may be necessary to occlude temporarily the sinus on either side of the rent (allowing the surgeon the time and conditions to fashion a dural pedicle graft), and then to sew the graft directly to the edges of the torn sinus in a circumferential manner. Use continuous or interrupted 4–0 or 5–0 suture material to seal the dural opening. When the temporary clips are removed, Avitene should be packed to the surface of the anchored pedicle graft. A cotton "fluffy" is used to hold it in place for a period of 5–10 min. This technique stops the more severe sinus bleeders and does not subject the child to the risk of sinus occlusion by topical hemostatic agents. It is the technique used to repair traumatic lacerations of the sinus.

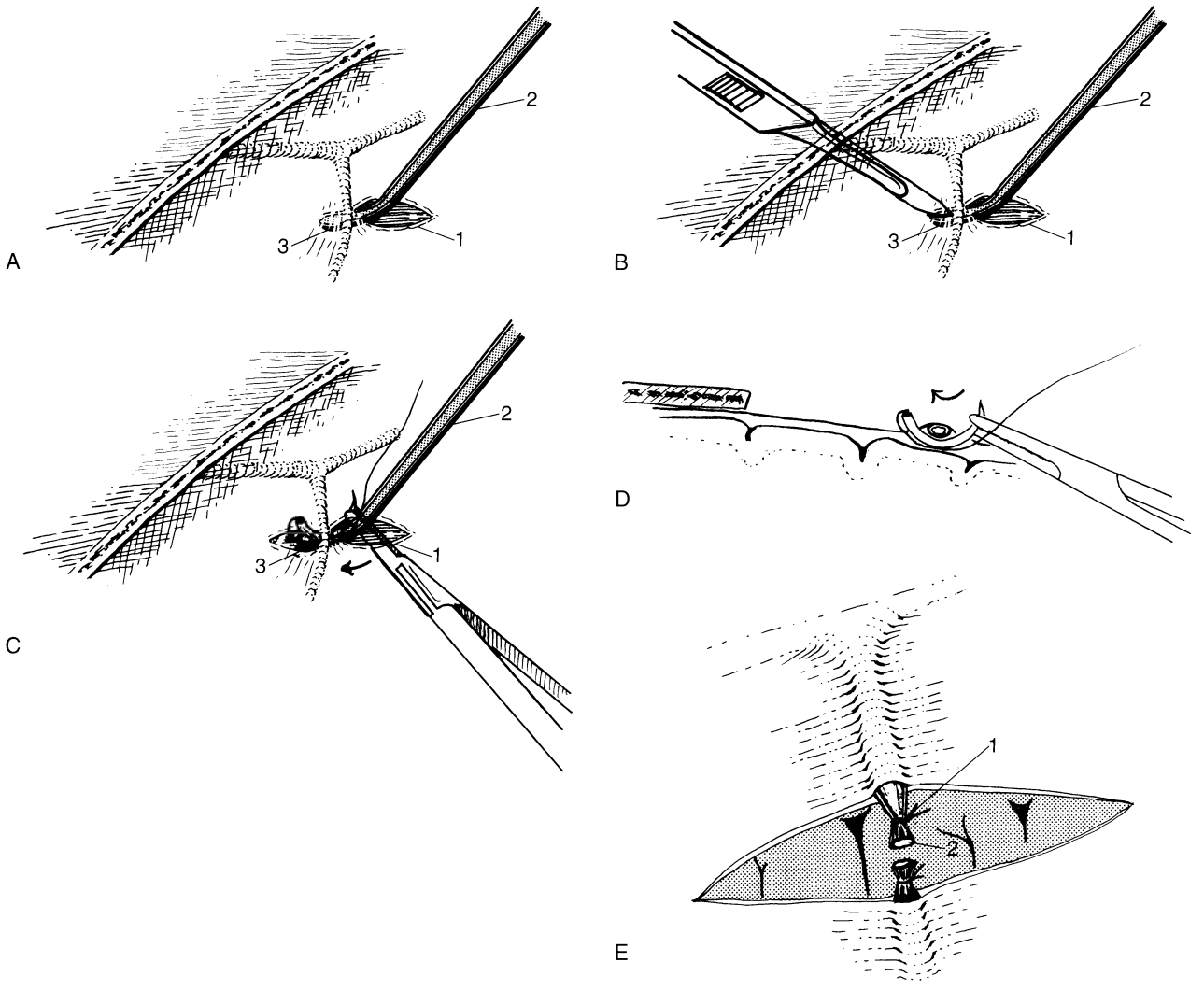


Figure 2.29. Ligature of meningeal arteries. (A) Larger meningeal arteries that require ligature should be approached by opening the dura distal to the artery (*1*) with the scleral hook and a #15 blade, so as to insert a dural guide (*2*), which may be passed beneath the dural artery to the opposite side of the vessel (*3*). (B) After the dural guide has been positioned, a #15 blade is brought down upon the grooved dural guide, cutting through the dura. (C) This allows one to open the dura to the very edge of the intradural artery on either side. The surgeon may now pass the ligature along the grooved dural guide, beneath the artery, from one side to the other. In this drawing the ligature is being passed backward, with the trailing edge of the needle being led along the groove director beneath the dural artery. (D) Cross section of what is illustrated in C. (E) After two ligatures have been passed, separated from one another by approximately 3 mm, they are tied down (*1*) and the artery is transected (*2*).

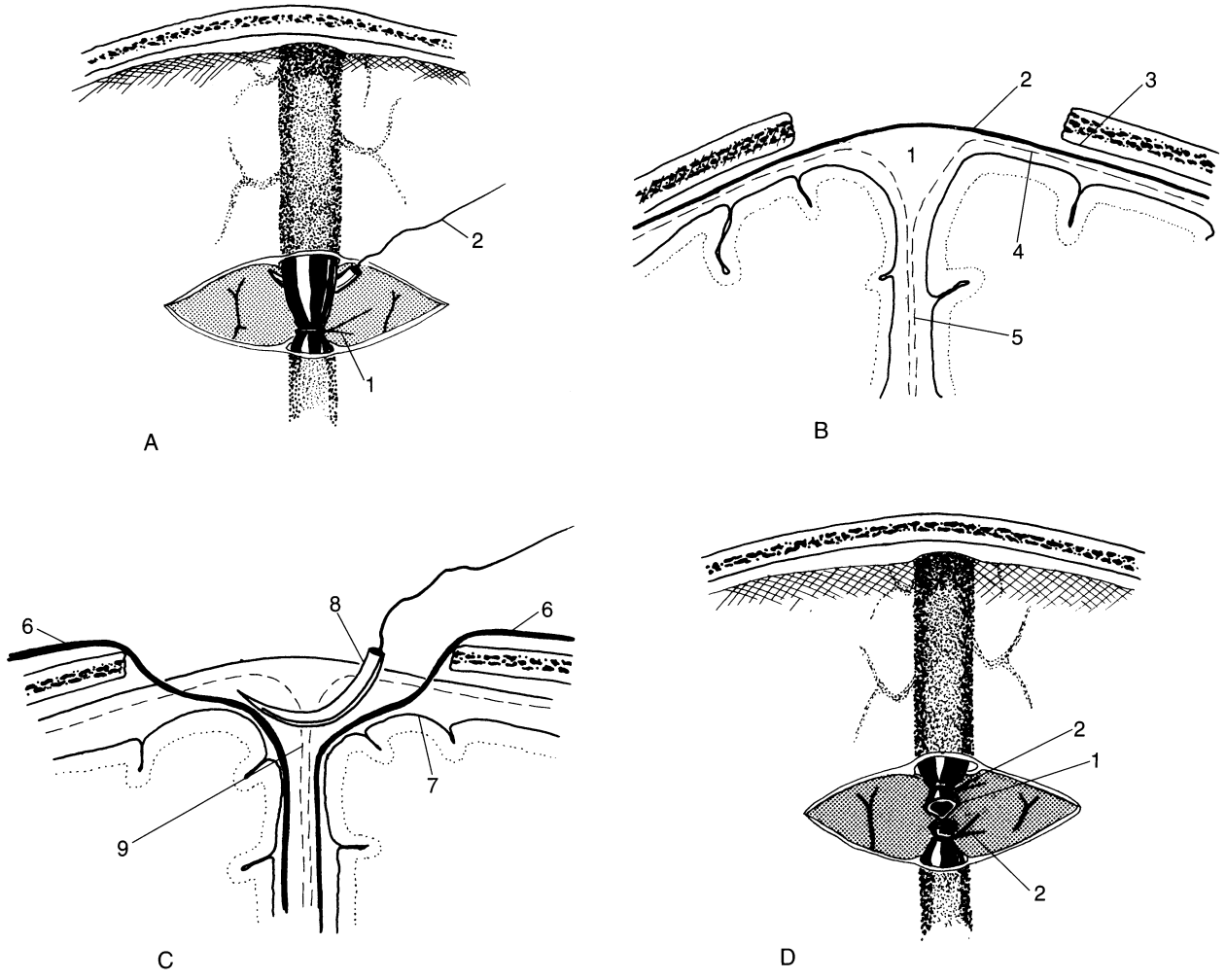


Figure 2.30. Ligature of dural sinus. (A) After the dura has been opened to the very edge of the dural sinus and either the falx or tentorium cerebri identified and exposed bilaterally, a 3–0 or 2–0 suture is passed beneath the apex of the dural sinus. The ligature is tied down (*1*). A second ligature is passed about 3–4 mm distal to the previous one (*2*), and it in turn is tied down. The sinus is cut between the ligatures. Passage of the round needle is illustrated from the surface of the sinus. (B) Anatomical basis for passage of the round needle represented in a cross-sectional drawing. The SSS (*1*) is triangular in shape, a result of the extension of the outer layer of the dura (*2*) along the inner table of the skull (*3*) and of the inner layer of the dura (*4*) into the falx cerebri (*5*). (C) After the dura has been opened to the lateral surface of the SSS, Telfa strips (*6*) are lain over the cortical surface and the convexities (*7*) are allowed to fall away from the SSS. The needle (*8*) is now (visually) inserted through the falx (*9*), at the most inferior point of the SSS. (D) The sinus (*1*) is transected after the ligatures (*2*) have been tied.

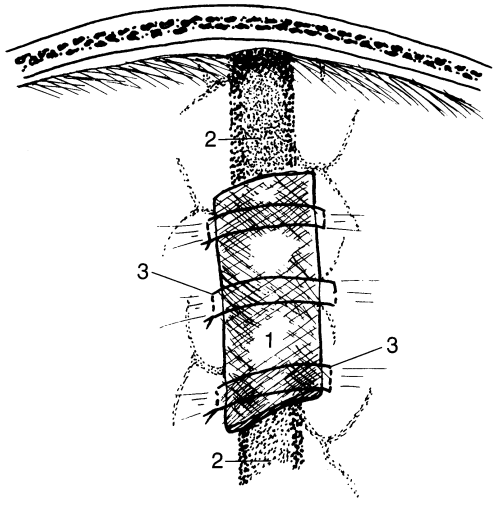


Figure 2.31. A Surgicel-Avitene "sandwich"(*1*) has been lain over the oozing or bleeding sinus (2) and then anchored into place with sutures passing over it from the dura on one side to that on the other (*3*).

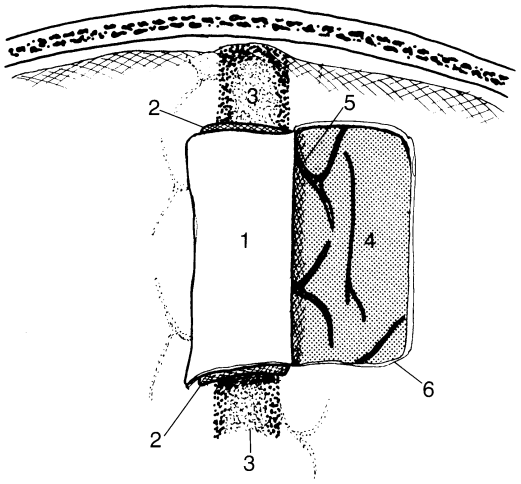


Figure 2.32. In this illustration, a dural flap (*1*) has been sewn over an underlying Surgicel-Avitene "sandwich"(*2*), compressing it against the sinus (*3*). The underlying cortex (*4*), cortical bridging veins (*5*), and edge of dural donor site (*6*) are illustrated. The dural defect is then closed with a periosteal patch graft.

Cortical Bleeding (Fig. 2.33)

Cortical Arterial Bleeding

Cortical arterial bleeding presents quite different problems from sulcal or cisternal arterial bleeding in that the arteries are *within* the cortex, beneath or nestled within the pia mater. They are also thin when compared to sulcal or cisternal arteries. The best technique for coagulating these vessels is to apply the bipolar forceps to either side of the vessel, in a plane longitudinal to it, and then to coagulate as a drop of saline is allowed to fall over the interval between the blades of the bipolar forceps. This gradually constricts and occludes the artery. However, there is commonly an open, though minuscule, channel remaining at the end of the coagulation, so the vessel should be transected and then each stump taken perpendicularly with the forceps and coagulated until the edges have been sealed completely.

Sulcal or Cisternal Arteries

Sulcal or cisternal arteries, on the other hand, bathe freely within the cerebrospinal fluid, have no parenchymal tissue adherent to them, and are thick-walled. Therefore, the cautery forceps should be placed perpendicular to the long axis of the artery, and the coagulation effected with the cauterization beginning just as the forceps touch the arterial wall, before they compress it. This technique is extended over an area of approximately 3 mm before compressing slightly the artery over the same distance. Lastly, the cauterization is carried out with the artery occluded between the blades of

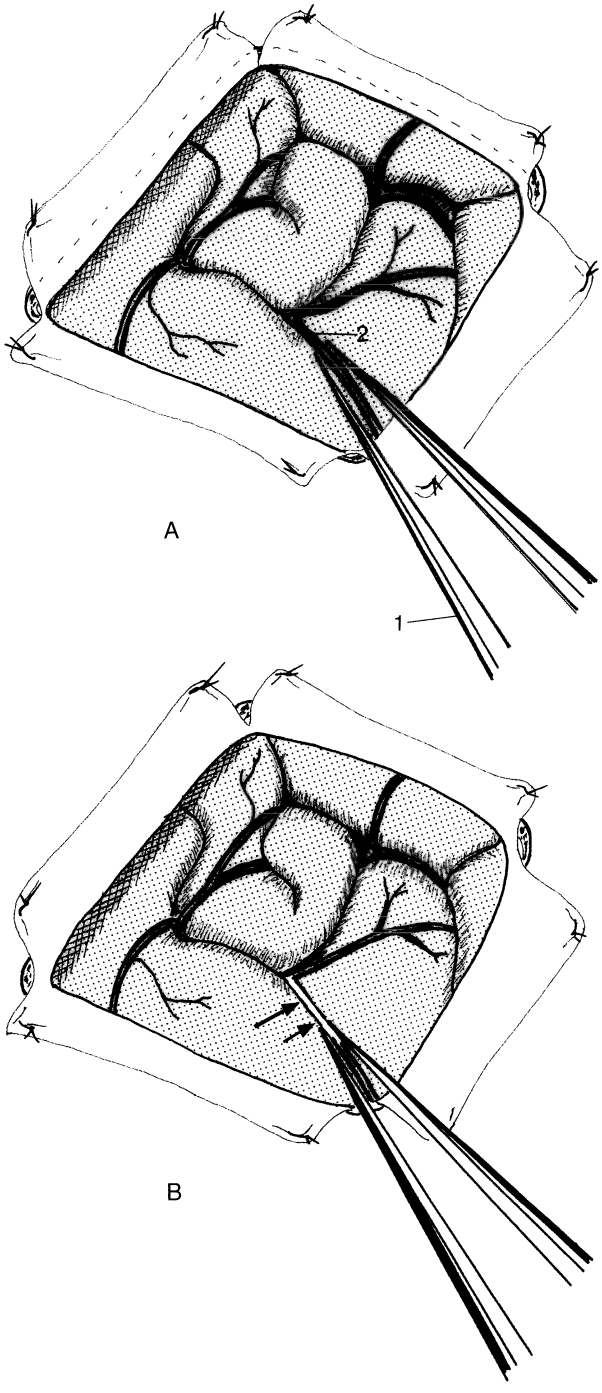


Figure 2.33. (A) The forceps (*1*) are placed parallel to the axis of the vessel (*2*), without being closed completely. (B) The cortical vessel is coagulated over a 3- to 5-mm length (*arrows*) until it is converted into a yellowish band. This coagulated strip should then be transected and each stump coagulated until the edges have sealed.

the forceps. This allows for gentle shrinkage of the artery before it is completely occluded and coagulated. If this coagulation is carried out with the arterial surface and the cautery forceps bathed in either cerebrospinal fluid or a drop of saline, there will be no sticking of the artery to the forceps. However, this latter point is moot, since there are forceps presently available that adhere only minimally to the vessel wall. Transection after coagulation allows for retraction of the artery and assures permanent hemostasis, but one must take care to coagulate the stumps until the vascular circumference has sealed the lumen!

Larger Cisternal Arteries

Larger cisternal arteries demand clipping! Take time to instruct the nurse to lubricate the jaws of the hemoclip applicator before loading the clip, to minimize sticking. The clips should be applied at a distance of approximately 4 mm from one another. If a large artery is clipped in the middle of an operative procedure (such as removal of a tumor or resection of an arteriovenous malformation), one may expect the artery to be in the field for a prolonged period of time. It is advisable, in these instances, to apply two clips at either end of the planned transection site, and to separate these clips from one another by approximately 1 mm. This affords the protection of an additional clip on a cut vessel that may be manipulated repeatedly throughout the procedure.

Application of the hemoclips should be done with care, bringing the artery between the jaws of the hemoclip so as to see both sides of the clip, then gradually closing the clip until the artery begins to be compressed before locking it securely with a steady, not snapping, motion. This avoids slippage of one blade by adhesion of the clip to the applicator: an event that very commonly occurs. Therefore, before withdrawing the applicator from the field, one should grasp the artery with a pair of forceps and hold it snugly, providing countertraction. If the clip is stuck to the applicator, a fine-pointed instrument suffices to slide it out of the jaw to which it is adherent. Silver clips are not reliable.

Large Sulcal Arteries

Very large sulcal arteries, those the size of the middle or posterior cerebral arteries, are occluded by applying a temporary clip first (such as the Yasargil), and then applying the hemoclips. If one chooses, they may be occluded, by ligature. I discourage the use of ligature because the difficulty of tying the knot snugly, when working in the depths of a very small infant's intracranial compartment, increases the risk of tearing the floating artery from its parent vessel.

Venous Bleeding

Cortical veins, sulcal veins, and cisternal or bridging veins require different techniques for rapid and precise coagulation and transection.

Cortical Veins

The cortical veins are coagulated simply by applying the bipolar forceps to the surface of the vein and sliding it along the longitudinal axis, applying a drop or two of saline. The vein coagulates quickly, and converts into a yellowish fibrous band, which may either be transected or left intact, depending upon the direction of the planned dissection. It is seldom necessary to transect these vessels after they have been coagulated.

Sulcal Veins

Sulcal veins, on the other hand, fish freely within a small amount of cerebrospinal fluid in the subarachnoid spaces and, consequently, should be coagulated by applying the forceps perpendicular to the long axis of the vein, but just so much as to touch the vein rather than to attempt to occlude it completely (which very often results in adhesion of the vein to the cautery forceps). Consequently, one risks tearing the veins as the forceps are pulled away. Simply touching the wall assures shrinkage and coagulation of the vein without the risk of having it adhere to the forceps. It is preferable to expose the sulcal vein over a distance of 3 or 4 mm before coagulating it.

Cisternal Veins

Cisternal veins (the deep middle cerebral vein, the sphenoparietal vein at its entrance into the cavernous sinus, the superior petrosal vein) must be exposed over at least 1.5 cm and should be coagulated with the regular bipolar forceps, not the microforceps. The blades of the forceps are applied to the surface of the vein, not compressing it, and then slid along the desired length of coagulation, preferably 3–6 mm, as drops of saline are applied. The vein will gradually convert to a yellowish band, which may then be compressed slowly, continuing the coagulation. It is best to taper the coagulated area from complete occlusion at the center to minimal occlusion at either end, giving the coagulated vein an hourglass appearance. Sharp demarcation lines between normal vein and coagulated vein are to be avoided, for this may result in rupture of the venous wall. The coagulated cisternal vein must be cut, since the fibrous band into which it is converted is dense and may be torn from the vein to which it is tributary during surgical manipulation.

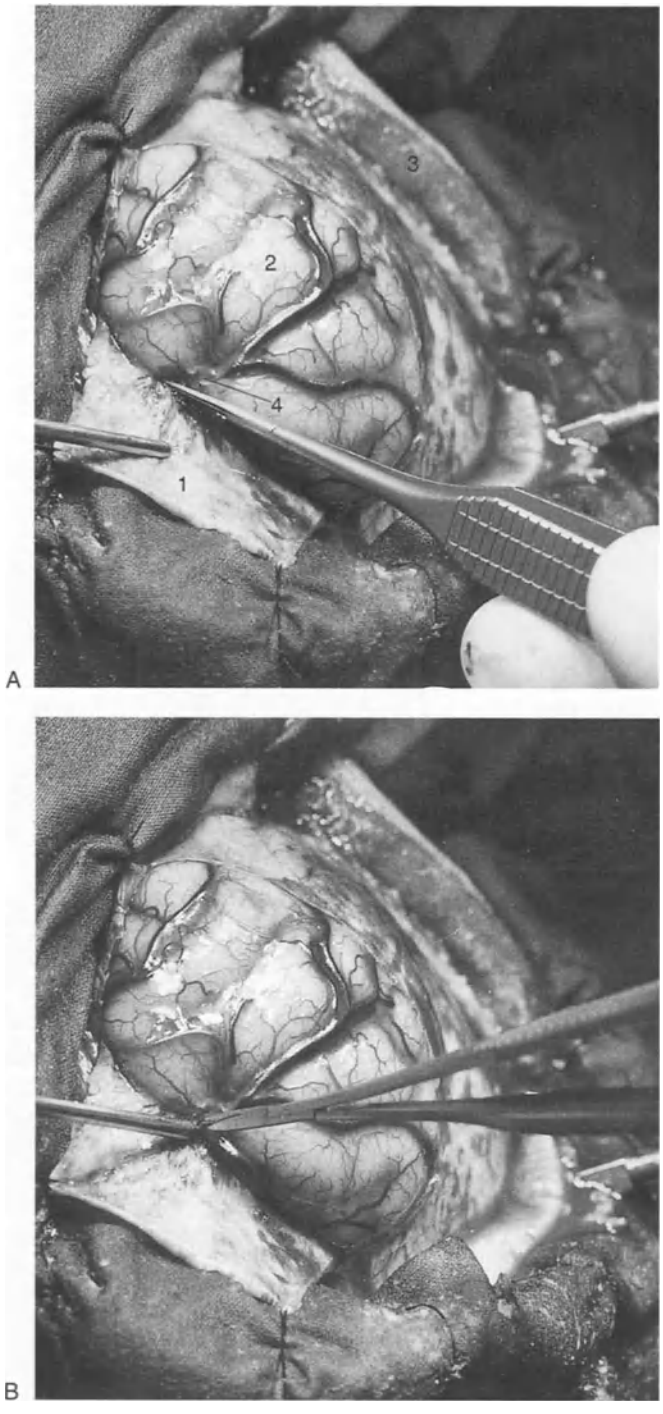


Figure 2.34. Coagulation and transection of a convexity (frontal) bridging cortical vein entering the superior sagittal sinus. (A) The medial dural trapdoor (*1*) has been reflected medially, exposing the frontal lobe (*2*). The temporalis muscle (*3*) offers a point of reference. The microbipolar forceps are placed along the cortical surface of the bridging cortical vein (*4*). (B) The suction is brought into the field at the time of the cut (in the event there is bleeding). This controls the bleeding immediately and helps to minimize the risk of air embolism.

Cortical Bridging Veins (Figs. 2.34, 2.35)

Cortical bridging veins present great challenges to the surgeon. They pass for varying distances through the cortex before entering the subarachnoid space, at which point they may receive tributaries from other cortical veins before they penetrate the dura, coming to lodge between its two, inner and outer, layers. Their entrance into the dural sinus is from anterior to posterior. The surgeon has the impression that the vein is entering perpendicular to the transverse sinus because the hemisphere is retracted from the sinus, thus changing the anatomical relationship between cortical bridging vein, cortex, and sinus. Once the cortical bridging vein has been dissected free and the point of its exit from the cerebral hemisphere, as well as its entry into the sinus, is identified, the coagulation is begun at the cortical surface. *It is preferable, actually safer, to coagulate at the cortical vein or, still less desirable, at the entrance of the cortical vein into the sinus. This does not damage the cortex any more than occlusion of venous drainage.* Coagulation at the cortical surface is begun by applying saline to the surface, and then the bipolar forceps to the borders of the vein, without attempting to occlude the vein. As the coagulation progresses, one notes that the bridging vein shrinks by being turned into a yellowish, fibrous structure. One must avoid continuing coagulation at exactly the same point! Rather, the coagulation should be extended over a surface area of approximately 3–6 mm, the constriction of the vein should be

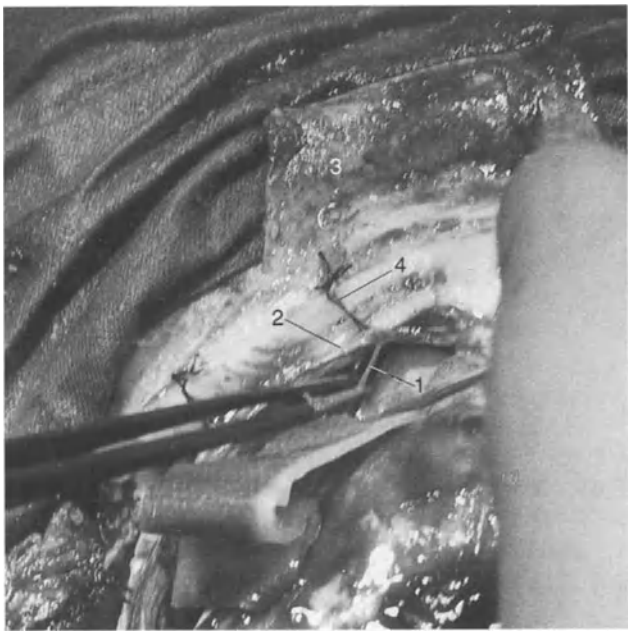


Figure 2.35. Coagulation and transection of a frontopolar bridging cortical vein. The vein (*1*) has the forceps blades touching its surfaces. Note the frontal air sinus (*2*) and the dural flap (*3*), as well as the tack-up sutures (*4*).

observed as the coagulation progresses, and the uniform diameter of the shrinking cortical vein should be maintained. Lastly, when the vein is still identifiable but clearly patent, one may occlude it completely, always coagulating under saline, and proceed to occlude the vein over an area of 3–5 mm. Once the vein is completely occluded it may be transected by using microscissors.

Choroid Plexus (Fig. 2.36)

Choroidal tissue may be safely dissected from some portions of the ventricular surface, as in the area of adhesion of the tela choroidea of the III ventricle, along the surface of the thalamus at the junction between the lateral and III ventricle.

This is not true for choroid plexus because this tissue is highly vascularized. Its vascular supply comes from branches of the carotid and vertebrobasilar systems. Also, the choroid plexus receives small perforating arteries through ependymal substance. It is therefore necessary to identify and occlude the major choroidal arteries (anterior choroidal, medial posterior choroidal, choroidal branches of the posterior inferior cerebellar artery) when planning to resect the entire choroid plexus. One then effects the resection by coagulating along the microvascular pedicle of the plexus, whether it is located within the lateral ventricle, the roof of the III ventricle, or the roof of the IV ventricle.

Bleeding from the surface of the choroid plexus is readily stopped by applying soaked (cotton) "fluffies" and sucking them dry once they have been firmly pressed against the bleeding site, or by coagulating the plexus. Coagulation of the plexus offers no unusual problems. It is performed by applying the forceps to the plexus, again taking great care to avoid compressing the blades snugly against one another. Rather, they should be progressively approximated to one another as, gradually and slowly, the plexus shrinks from the bipolar cautery. Every effort should be made to avoid bleeding from the microvascular pedicle of the plexus. This is difficult to stop because of retraction of the vessel within either the cerebral substance or, worse still, through the choroidal fissure and into the ambient cistern.

Tissue Bleeding

Stopping bleeding from tissue calls for a different technique from that used in coagulating, clipping, or the usage of the ligature to occlude identifiable bleeding vessels. These techniques vary from tissue to tissue because of the differences in anatomical composition.

Galeal Bleeding

The Dandy and Kolodny clamps are too coarse and large for use on a child's scalp, especially in a newborn or infant. The Rainey clips are made for the scalp of a full-grown adult male. Also, the present plastic model is unreliable. Therefore, arresting scalp bleeding coming from the interstices of the dense connective tissue necessitates the use of lightweight, delicate-toothed scalp clips, which may be applied directly to the very thin galea of an infant, grasping the galea between the teeth. Reflecting these galeal clips over the scalp edge and onto a gauze sponge compresses gently the vessels within the loose connective tissue. This is the only safe and reliable form of scalp (skin, dense connective tissue) hemostasis recommended. The scalp flaps, pedunculated and anchored, are then retracted by reflecting them along their bases, using retention sutures strung tautly by rubber bands, and sewn to the skin, respectively. Retraction by the retention sutures, superiorly on the pedunculated flap, and inferiorly only on the anchored portion, of the flap, exposes completely the desired area of periosteum and skull.

Bleeders along the *undersurface of the galea* necessitate coagulation, because (1) they cannot be trapped between galea and skin and (2) they bleed profusely. They are identified by rolling the scalp over, so as to expose the galea and the vessels along its internal surface. Avoid overcoagulation of these vessels, especially in the newborn, since one may penetrate or necrose the scalp.

Parenchymal Bleeding

Parenchymal bleeding is stopped, as already described, by applying "fluffy" cotton, Gelfoam, Surgicel, or Avitene to the bleeding surface.

Tumor Bleeding

It is best to make every effort to stop tumor bleeding before it occurs, rather than attempt to do so on a raw, bleeding, tumor surface. Consequently, identification of bridging vessels from the surrounding tissue to the neoplasm should be undertaken by skirting the circumference of the neoplasm, using fluffy and Telfa to dissect parenchyma from neoplasm, and coagulating the bridging vessels as one proceeds. One should also coagulate the visible vessels on the surface of the tumor as they are seen. Once the parenchyma has been separated from the neoplastic tissue, a wide-bladed forceps may be used to coagulate the surface of the tumor, shrinking it gradually. This may be painstaking and lengthy as a procedure, but it definitely diminishes tumor bulk and reduces considerably tumor vascularity, particularly of ependymoma and medulloblastoma. Choroid plexus papilloma, primitive neuroectodermal tumor, and sub-

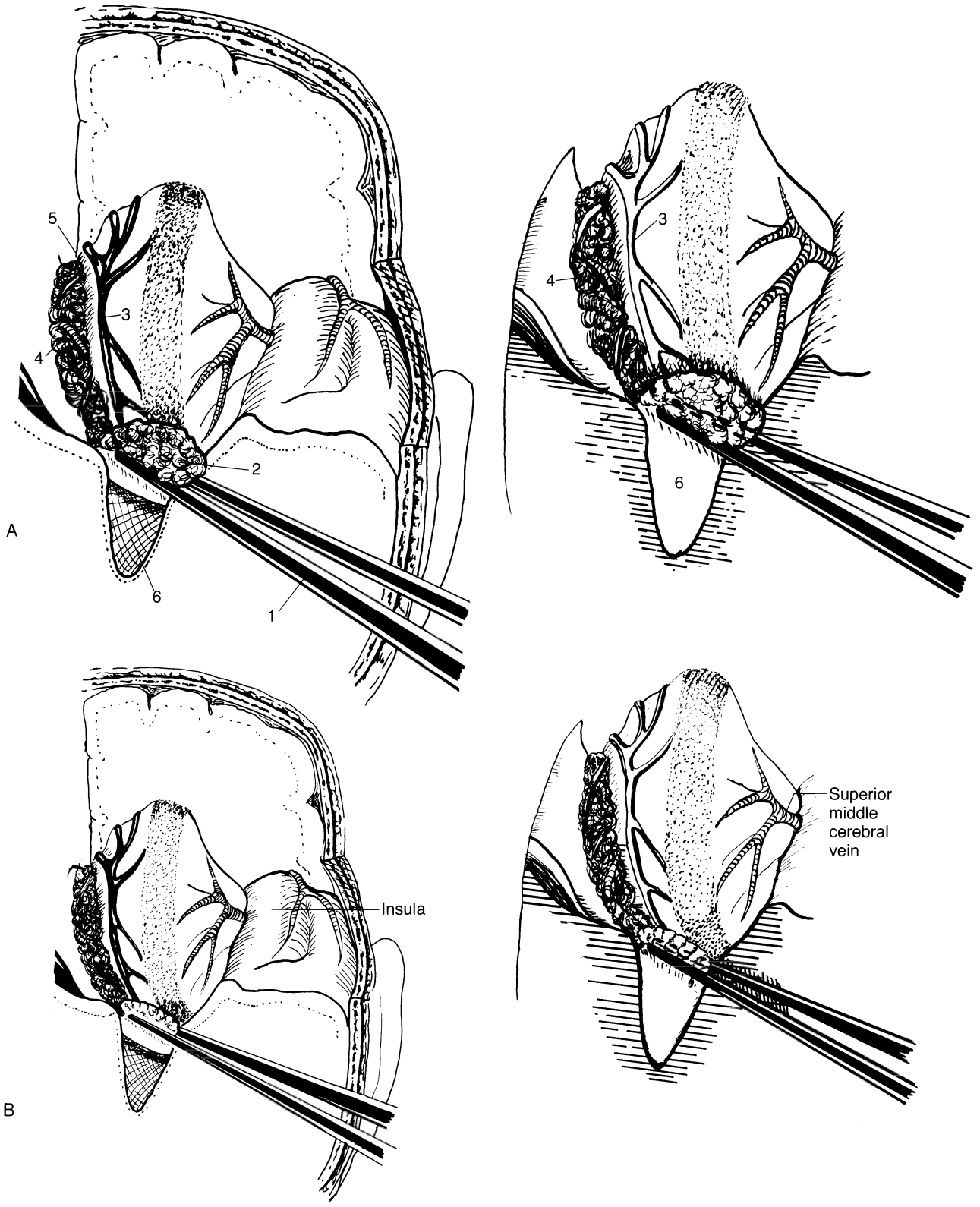


Figure 2.36. Coagulation of choroid plexus. (A) The forceps (*1*) are applied to the surface of the glomus (*2*) and the coagulation begun. One avoids coagulating the terminal vein (*3*), running in the terminal sulcus. If the surgeon chooses, he may extend the coagulation along the entire choroid plexus within the lateral ventricle (*4*), up to the foramen of Monro (*5*). There is no choroid plexus in the occipital horn (*6*). (B) As the choroid plexus shrinks, the bipolar forceps are closed with the blades grasping it snugly between them. The coagulation is continued until the bleeding stops, and a plane of coagulated tissue may be identified for sectioning with microscissors.

ependymal astrocytoma may also be dealt with effectively in this manner.

The CO_2 laser is ideal for vaporizing these tumors almost bloodlessly when they are only minimally vascular, after the surface and bridging vessels have been identified and coagulated. In this manner one reduces considerably the bulk of the tumor, shrinking the intratumoral vasculature as one proceeds. If the tumors are moderately or highly vascular, CO_2 laser is an undesirable instrument. The Cavitron is unsurpassed for tumor removal, but the claim made by many that it respects the "integrity" of vascular structures is incorrect!

Closure

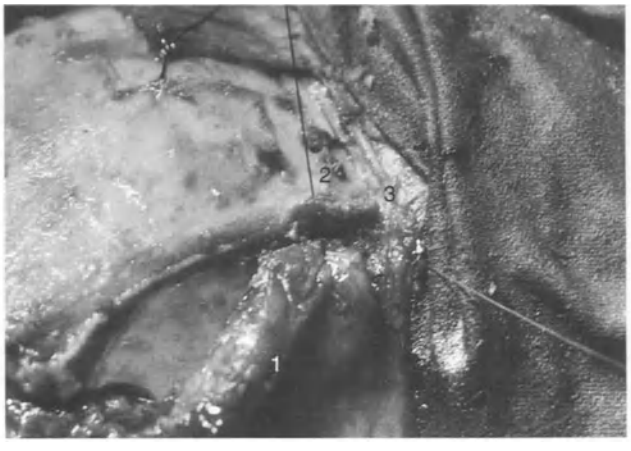
Cranial Closure

Fascia and Muscle Closure (Figs. 2.37, 2.38)

Temporalis Muscle

The temporalis muscle is a bipenniform structure, consisting of two muscular bundles and three fascial planes. Specifically, there are (1) an outer musculofascial layer, (2) an intermediate fascial layer, and (3) an inner musculofascial layer. Bleeding from muscular tissue and transected vessels located within either the inner or outer muscular layers is a common occurrence. One may assure adequate hemostasis only by being certain to include all three fascial layers of the temporalis muscle in the suture.

A



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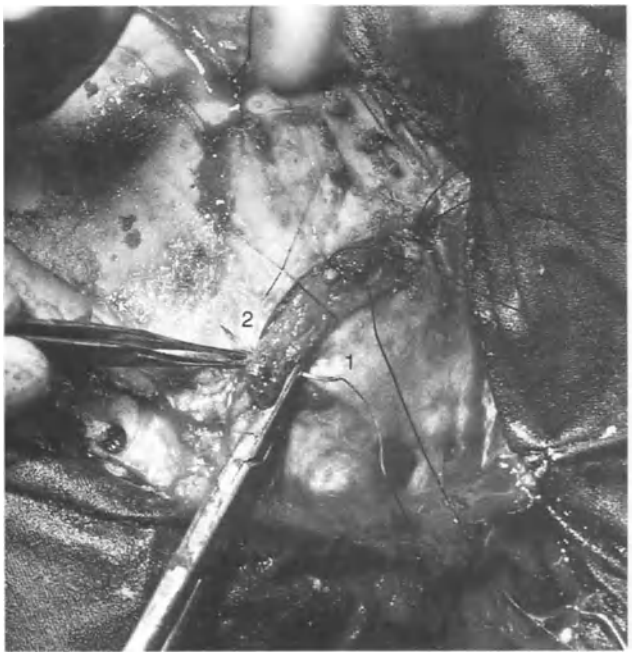


C

Figure 2.37. (A) At the time of closure, the frontal (bifrontal) flap is put back into position, and the temporalis muscle retention sutures are removed. The muscular (*1*) and bony (*2*) edges of the cut temporalis insertion along the superior temporal line will be used to anchor the frontal bone flap back into place. (B) The muscular (*1*) and bony (*2*) edges of the temporalis muscle are being sewn to one another at the posterior rim of the zygomatic process of the frontal bone (*3*), over the "keyhole" bur opening. (C) The last suture is being placed through full thickness (all three layers) of the temporalis muscle (*1*) and the periosteum adherent to the superior temporal line (*2*).

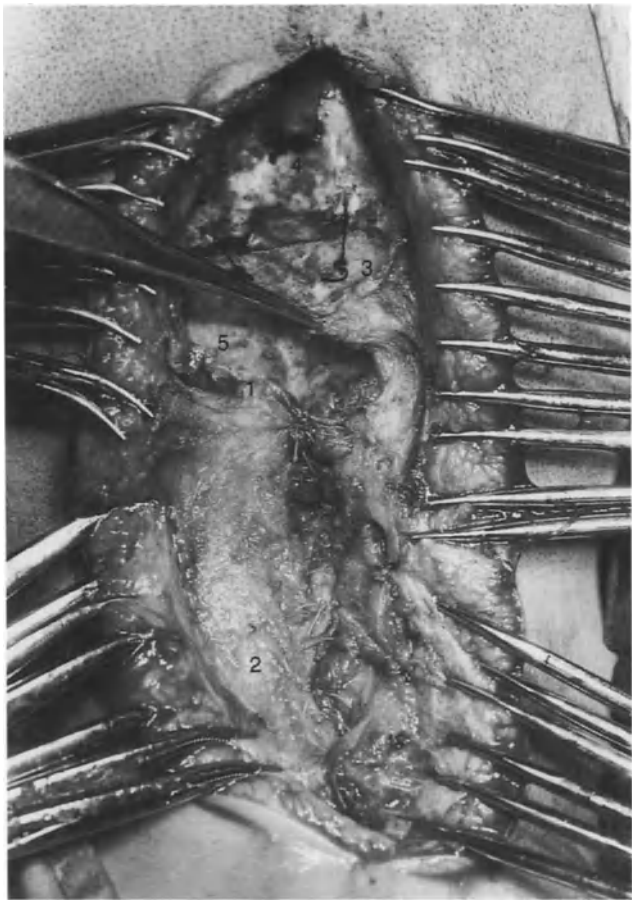


Figure 2.38. The erector capitis muscle masses have been reapproximated and sewn together in the area extending from over the squamous occipital bone (*1*) and C-2 (*2*). Also, the fascia from these masses (*3*) has been sewn to the periosteum, which was left intact, and attached to the squamous occipital bone at the external occipital protuberance (*4*). The tissue forceps holds the fascia of the right erector capitis muscle, which is being brought over the replaced osteoplastic bone flap (*5*) to be sewn to the fascia on the left.

Erector Capitis Muscles

After the suboccipital bone flap has been secured in position, one should extend slightly the head of the child prior to closing the muscular layers in the suboccipital area. This facilitates reapproximation of the erector capitis fascia and muscular tissues that had been preserved at the time of opening (Figs. 2.23, 2.24). Since it is not necessary to remove the erector cervicis muscle from the spinous processes of C-2, C-3, and so on in order to attain excellent exposure of the posterior fossa, one has only to reapproximate the two cut edges or the ligamentum nuchae, starting inferiorly and working superiorly, bringing the trapezius and erector capitis muscles from both sides together at the midline, and continuing this closure superiorly. The horizontally transected musculofascial tissue, about 1 cm beneath the external occipital protuberance, may be closed at this time.

Skin Closure (Fig. 2.39)

The skin closure should proceed one section at a time, *taking care not to remove all the galeal clips at one time because this results in excessive bleeding, which, for the younger child, may cause an unacceptably large volume of blood loss in a brief period.* A section of approximately 4 cm at a time is quite safe. After the galeal clips have been removed, a search should be made for uncoagulated vessels lying along the undersurface of the galea. When these have been individually coagulated, the skin may be closed. Avoid coagulating bleeding vessels within the dense connective tissue, since this may result in necrosis of the skin edges.

Burying of sutures by using an independent galeal closure results in their extrusion, especially in the newborn and infant. Consequently, galeal closure may be assured by the use of the Cloward stitch, which allows the surgeon to close the galea and skin in a single, removable stitch. In order to place the Cloward stitch, run the suture through the full thickness of the skin, through the galea, on the free edge of the flap, and then proceed to sew only the galea on the opposite side. Return now to the previous side to pick up only the galea, and then bring the needle through the full thickness of the skin on the opposite side. The skin edges are now brought together by drawing the two ends of the stitch tightly. This completes the Cloward stitch.

Though this stitch effectively closes the galea and skin, it does not assure approximation of the two edges of the epidermis. Consequently, we recommend completing the Cloward stitch and then continuing, bringing the needle through epidermis from the anchored to the free side before tying the knot.

The skin flap closure, as the dural closure, is performed by tying sutures on either side of the flap alternately, and by inserting the sutures at approximately 4-mm intervals on the free side and 5-mm intervals on the anchored side. This technique provides proper approximation of skin edges when the flap is curvilinear. For closure of straight incisions, one should maintain the same interval on either side.

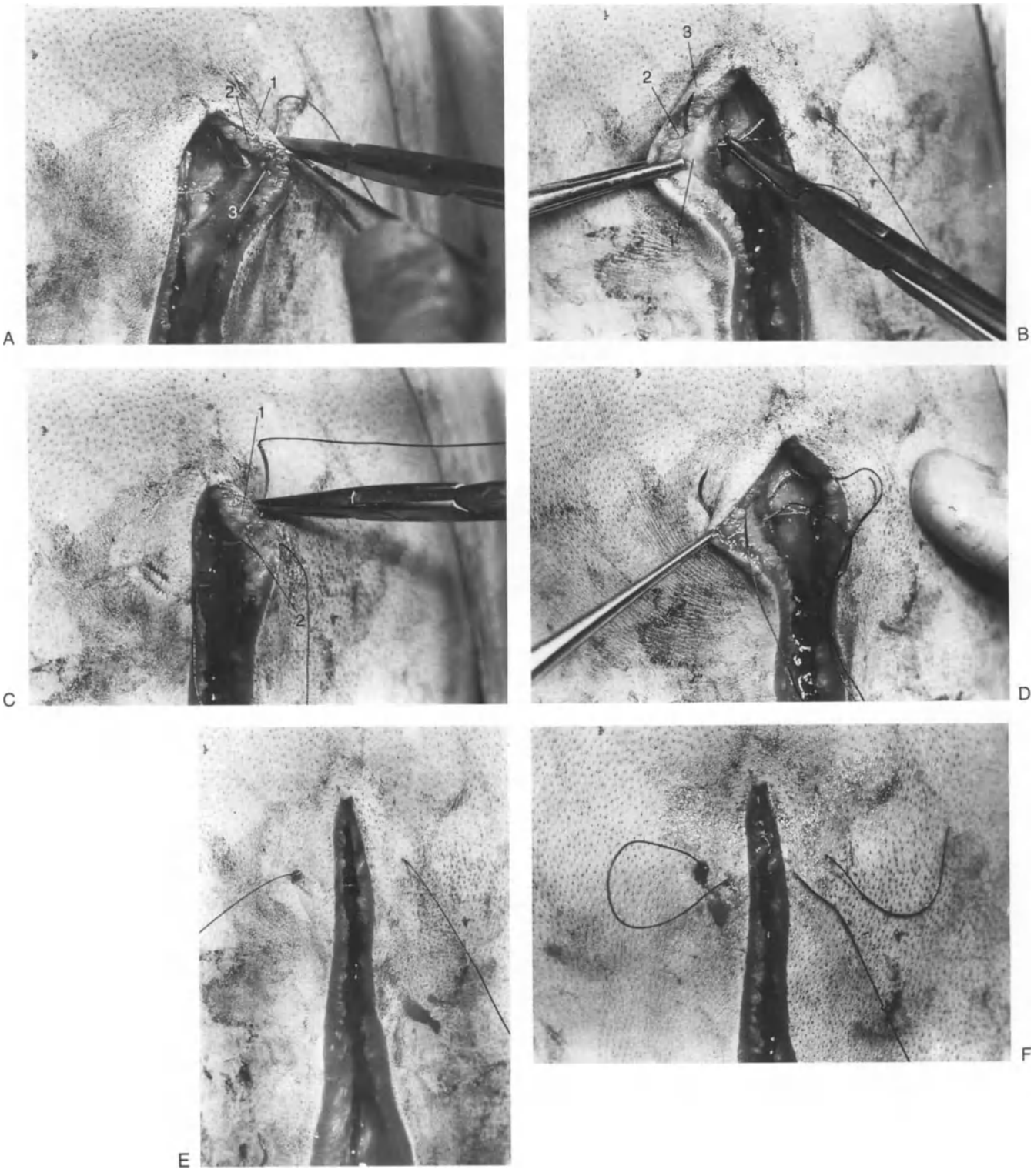


Figure 2.39. Legend see p. 77.

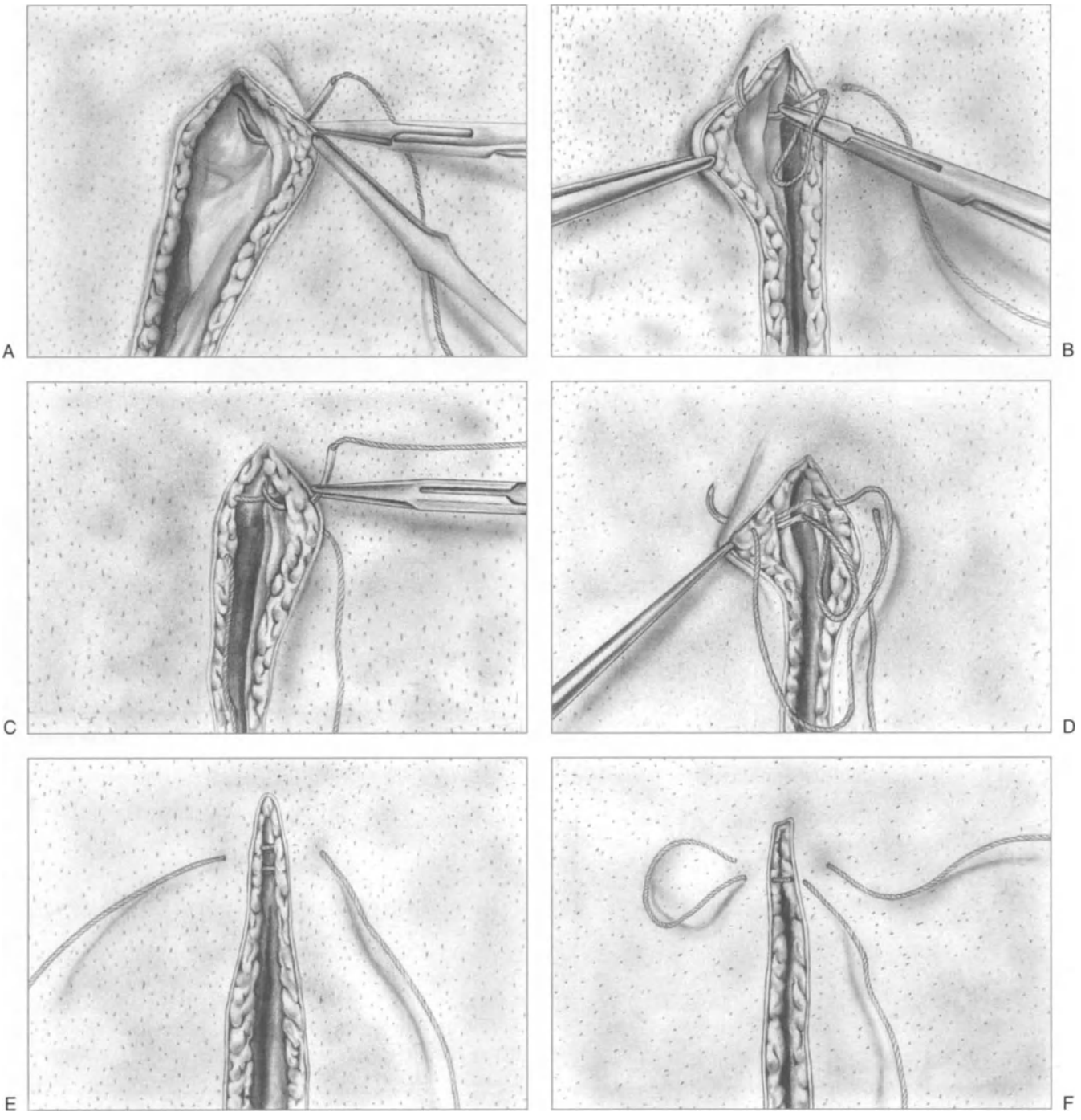
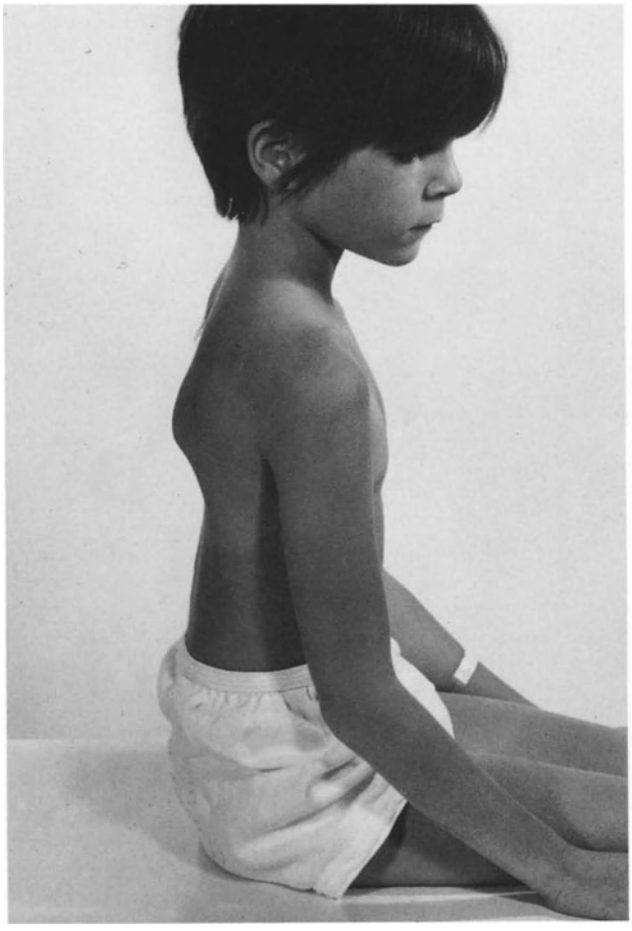


Figure 2.39. Technique for the Cloward suture, which closes the galea and the skin without necessitating burial of suture material. (A) The needle is brought through the full thckiness of skin (*1*), dense connective tissue (*2*), and galea (*3*). (B) The needle is brought only through the galea (*1*) of the opposite side, penetrating the dense connective tissue (*2*), but not the skin (*3*). (C) The needle is brought back to the original side and inserted through the dense connective tissue (*1*) and full thickness of the galea (*2*). (D) It is then returned to the opposite side to perforate the galea, dense connective tissue, and skin. (E) One then has suture entering skin on the left, drawn through dense connective tissue and the galea on both sides and the exiting skin on the right. (F) Passage of the suture through the epidermis of the two sides, thus completing the single suture technique for closure of the galea, dense connective tissue, and skin, an addition to the Cloward suture we recommend for children. This series of drawings, taken directly from the photographs illustrated herein, is presented step by step only to make the technique used for inserting the Cloward suture more readily understandable. Therefore, the reader is advised to follow the legend of this figure comparing the photographs with the drawings.



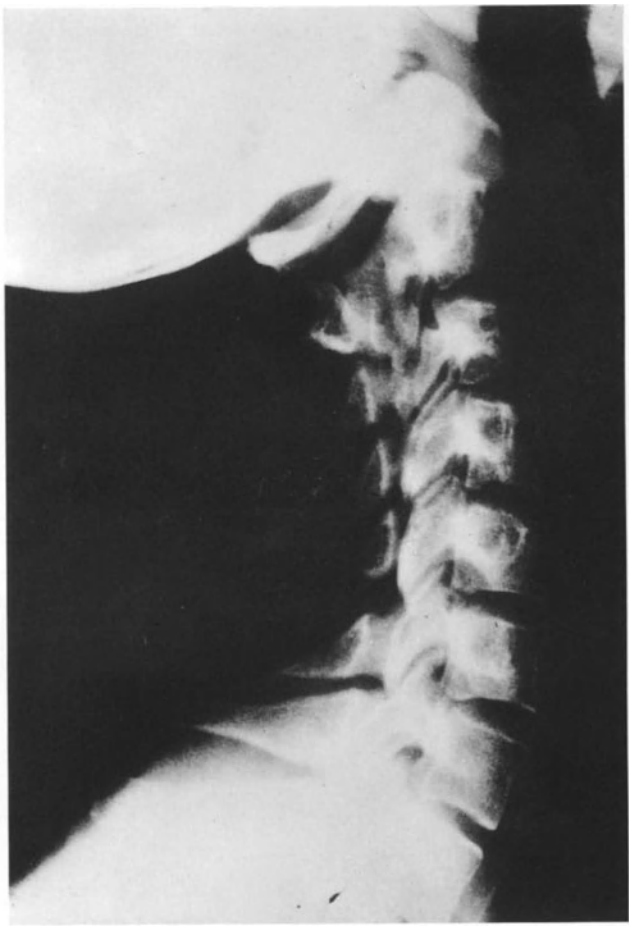


Figure 2.40 (*left*). Postoperative kyphoscoliosis in a child who had a laminectomy.

Figure 2.41 (*right*). Lateral cervical spine film of a child on whom an extensive cervical laminotomy had been performed. No malalignment is present.

Laminotomy (Figs. 2.40, 2.41)

Muscles and Fascia Closure

The paraspinous muscles are allowed to fall into place by removing the self-retaining retractor. Then the muscles are sewn down to the interspinous ligaments of the anchored laminar flap as well as to those above and below the laminotomy. This reapproximates the paraspinous muscles, interspinous ligaments, spinous processes, and laminae, restoring anatomical continuity between them. Very likely, this reapproximation is responsible for avoiding the postoperative scoliosis so commonly observed in children with laminectomy for spinal cord surgery, since it assures uniform muscular pull on the spinal column after healing has been completed. Postoperative scoliosis is uncommon following laminotomy.

Muscle Bleeding

Muscle bleeding is generally stopped by gentle compression, no more. However, especially in the erector capiti group, large arterial or venous bleeders retract deeply into the muscle belly, rendering identification and coagulation tedious and difficult. Those bleeders that have retracted into the erector capiti and erector cervicis muscle groups must be searched out and coagulated. This is not true for temporalis muscle bleeders, since the temporalis, a bipenniform muscle, has three fascial planes entrapping the two bellies of the muscle between them. Therefore, one may stop most of the bleeders at the time of closure by taking care to pass the suture material from the external fascial plane, through the intermediate fascial plane, to the internal fascial plane, before tying down the knot. This constricts the bleeders. See section on temporalis muscle.

Skin Closure

The skin over the spinal column is closed with mattress sutures.

3 Bur Holes and Flaps

"Who makes the Past, a patterne for next yeare,
Turnes no new leafe, but still the same things reads,
Seene things, he sees againe, heard things doth heare,
And makes his life, but like a paire of beads."

JOHN DONNE, Verse Letters: to Sir Henry Goodyere

Unfortunately, the terminology of anterior fossa or posterior fossa craniotomy is too vague for descriptive purposes in a text on operative technique. For example, an anterior fossa craniotomy includes bifrontopterional, medial and lateral frontal, and frontotemporal craniotomies; the middle fossa craniotomy includes anterior and posterior temporal craniotomies and temporoparietal craniotomy; the posterior fossa craniotomy includes access to the superior and inferior cerebellar triangles for medial lesions, and the pontocerebellar and clival approaches for lateral lesions. The parietal, occipital, and parasagittal nomenclature for craniotomies finds no place in the anterior, middle, and posterior fossa classification.

In planning the bone flap, one must correlate the "target area" with the skin incision, so as to attain ideal placement. For example, a bifrontopterional craniotomy for access to a *retrochiasmatic craniopharyngioma* entails exposure to the clinoid processes, optic nerves, and internal carotid arteries, bilaterally; of the optic chiasm and lamina terminalis in the midline; and of both internal carotid artery bifurcations and posterior communicating arteries. Therefore, a bifrontal skin incision permits reflection of a single bone flap, one which allows placement of the operating microscope for visualization along the floor of either fossa, down either sphenoid wing, and along the midsagittal (ethmoidal) plane to the parasellar area.

Another example is the attack on a pineal tumor *expanding over the roof of the III ventricle and beneath the splenium of the corpus callosum.* This surgery is most assured of success and carries a minimum risk of damaging the internal cerebral veins, if it is carried out through a medial parietal craniotomy. This entails a medial parietal skin incision with a horseshoe flap extending slightly to the contralateral side, a quadri- or pentalateral free bone flap, and medial reflection of the dura over the superior sagittal sinus (SSS). Dissection along the SSS and falx cerebri may then be accomplished with ease.

Such correlation of skin incision with bone flap and target area is presented throughout this text. Overlapping drawings (transparent cells), to indicate the various steps in the operative procedure as the surgeon proceeds from skin incision to craniotomy to target area, are used herein to illustrate this manner of conceptualizing the lesion and approaching it.

Bur holes and either craniotome or Gigli saw are not necessary in the newborn or very young infant, since the fontanelles and sutures are open and the skull thickness seldom exceeds 3 mm. The individual bones of the cranial vault are thin and not anchored securely to one another, thereby rendering it easy to cut them with scissors and dangerous to use heavy instruments that require force for penetration or sawing. Consequently, in very young children suturotomy and cutting of the bone are effected with heavy scissors (see Chap. 4). This, of course, does not invariably apply when an infant has craniosynostosis. The techniques for these specific procedures are described subsequently.

The use of power instruments for making bur holes and craniotomy is also to be *avoided* in toddlers, juveniles, and adolescents, since the size of the clutch-controlled perforator is for an adult! The tool is not sensitive enough to guarantee release of the clutch in children with thin skulls. Because it is possible to refill the bur hole with bone dust from the drilling shavings and to fashion a plug from the inner table of the skull, as described later in this chapter in the section "Bone Closure" (Figs. 3.49, 3.50), the surface area of the perforating instrument is only relatively important as a negative factor. The rapid regrowth of bone in the infant makes this size factor even more relative. However, the risk of plunging and of failure of the clutch release mechanism in younger children (as well as the very real compression of the underlying brain, which results from pres-

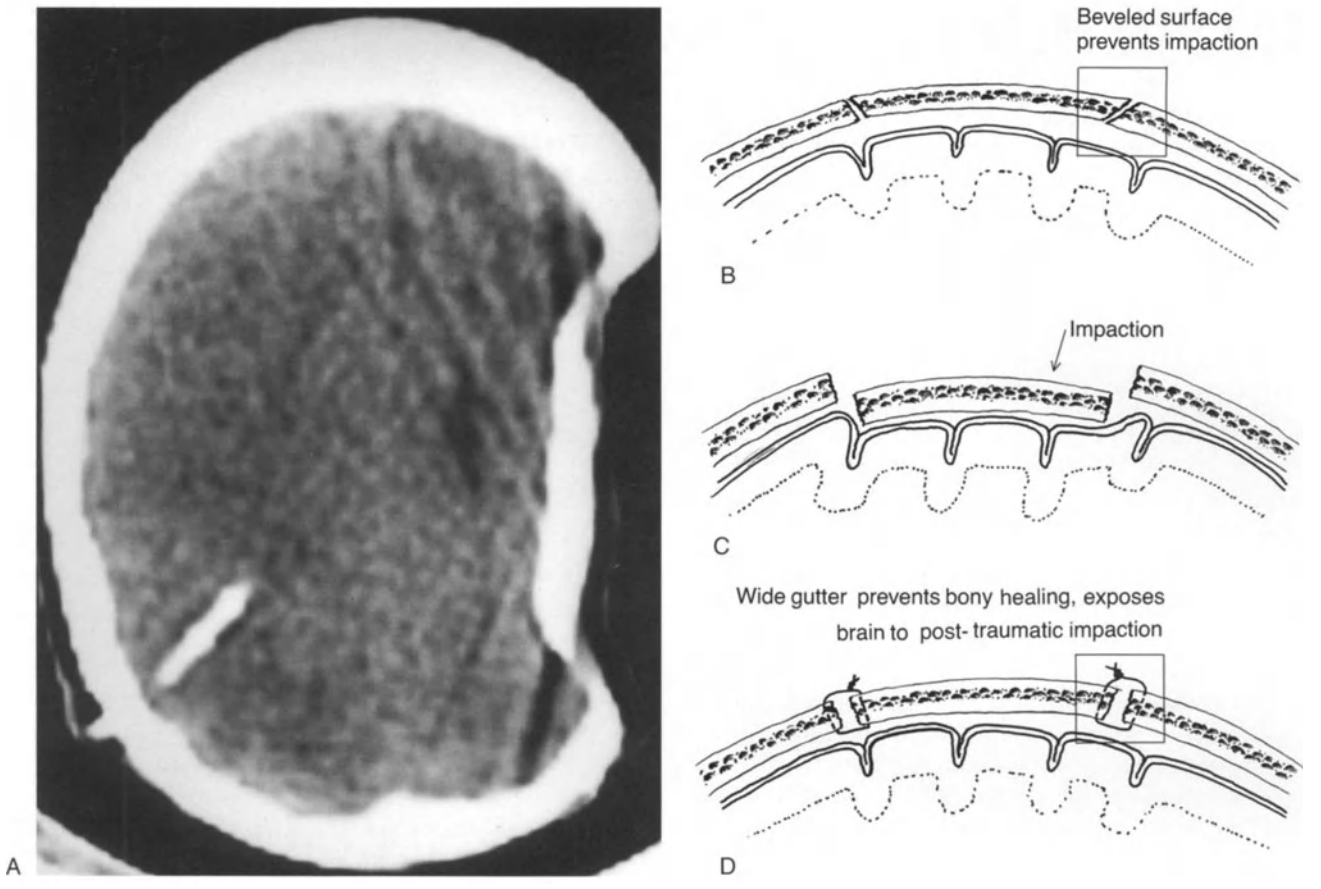


Figure 3.1. (A) This computed tomography (CT) scan shows the unacceptable degree of flap depression that occurs when the power craniotome is used. The Gigli saw permits adequate beveling, so that the flap nestles into place irrespective of placement of sutures through the skull or a functioning shunt system. (B) Using the Gigli saw permits one to bevel the cut, so that the free flap may be solidly seated at the time of closure, assuring perfect flap replacement and bone edge continuity. (C) However, when a power craniotome is used, the gutter is so wide that the flap sinks below the skull surface. (D) Wire suture (*to be avoided*) placed through drill holes may suspend the free flap, but *still (except in infants)* bony growth across the gutter only rarely occurs. The defect is permanent, and thereby susceptible to dislocation, impacting the underlying cerebrum, and injury.

sure applied to the surface of the skull in order to disengage the clutch) are very significant contraindications. It is unfortunate that adequate instrumentation, in regard to size and sensitivity for cutting bone, is not available for pediatric work.

All power craniotomes cut a large (2–4 mm) gutter in the skull (Fig. 3.1), rendering it impossible to reflect bone flaps which have a beveled surface. Flaps cut with power instruments may not be nestled snugly back into place. They rest on the surface of the dura, floating higher or lower, back and forth from one edge of the skull to another. This is a grave problem in hydrocephalic children, whose underlying brain surface may vary with the functional status of the shunt. Fixing the bone flap into place through perforations in the bone is to no avail, the gutter being too wide to permit bony bridging and the movement being such as to stimulate only the formation of scar tissue (except in the newborn and very young infant). Complete healing of the flap, on the other hand, ensues when the Gigli saw is used. One must make every effort to reconstruct the skull of children so that it will heal completely, providing the underlying brain protection throughout life, whether the craniotomy is supra- or infratentorial.

Bur Holes: Frontoparietal (So-Called "Diagnostic") (Fig. 3.2)

The theoretical considerations for placing the skin incisions for frontoparietal bur holes have already been described and illustrated. These skin incisions provide ready, safe, and ample conversion into a full temporoparietal flap in the event the surgeon encounters an epidural or subdural hematoma that should be removed through a craniotomy, or the bur hole openings reveal no visible epidural or subdural hematoma (because the clot may be located between the bur holes). Also, inferior extension of both the anterior and the posterior limbs of the skin incision permit reflection of a temporal flap for exploration and/or removal of an intracranial clot, subtemporal decompression, or partial or total temporal lobectomy.

Flaps

The placement of bur holes and outlining of the Gigli saw osteoctomy lines are discussed in the following sections.

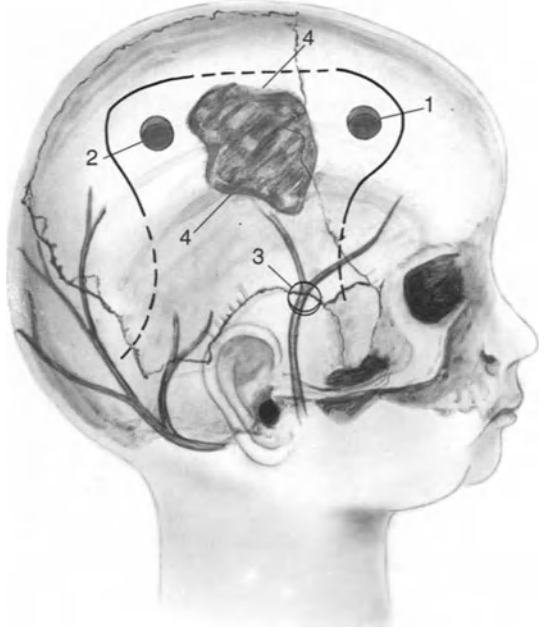


Figure 3.2. This drawing illustrates how frontotemporoparietal bur holes (*1, 2, 3*) may be placed around an epidural clot (*4*), so that the surgeon may not see the clot at the time of surgery if he limits his procedure to bur holes. If the clot is not seen when the bur holes are placed, one is obliged to proceed to reflect a flap so as to inspect completely the epidural, and/or subdural, space.

Bifrontal Flap (Figs. 3.3, 3.4)

The bifrontal (bifrontopterional) bone flap permits access to the entire anterior fossa (ethmoid and orbital), the parasellar area, both orbits, the circle of Willis (with the exception of the basilar fundus and mesencephalic portion of the posterior cerebral artery), and both middle cerebral arteries from the origin to their ramification over the insula. Consequently, it is an ideal flap for surgery for parasellar tumors, access to both orbits, and anterior circle aneurysms. The seven bur holes of the bifrontal flap are located at the glabella, immediately lateral to the SSS on either side, just posterior to the zygomatic processes of the frontal bone (the "keyhole") bilaterally.

For early childhood, the frontal air sinuses are not developed. The bur hole at the glabella runs no risk of penetrating one of them. The two parasagittal bur holes should be placed so that their medial aspects border upon the lateral aspects of the SSS.

The two pterional bur holes need no comment, but we will discuss the two "keyhole" perforations (those located immediately behind the zygomatic processes of the frontal bone). Because of the anatomical fact that the anterior fossa, the orbit, and the middle fossa are in immediate contact with one another at this point, it is essential to make the keyhole opening by directing first the perforator and then the bur in a superior, a slightly posterior, and a medial plane so as to assure entering the anterior fossa and to avoid entering either the orbit (specifically the region of the lacrymal gland) or the middle fossa.

In children with craniosynostosis of the coronal suture (plagiocephaly), the middle fossa is displaced so far anteriorly that one almost cannot avoid entering it if the bur hole is placed immediately behind the zygomatic process of the frontal bone. In these children, consequently, one must place the keyhole opening slightly superior to the zygomatic process of the frontal bone in order to enter the anterior fossa. However, since the lesser wing of the sphenoid is resected as far medially as the superior orbital fissure, there is no problem if one enters the middle fossa, other than that it renders reflecting the frontal flap difficult.

After the Gigli saw guide has been passed from bur hole to bur hole and the Gigli saw is brought into position between the inner table of the skull and the guide, the saw itself should be set snugly at the extreme periphery of each hole so as to connect their *outer* arcs and thus assure maximum size of the bone flap. The exception to this technique is the osteotomy line between the two parasagittal bur holes, which is placed at the most *central* arc. The reason for this is to allow the presence of a spur of bone over the SSS, a spur that may be removed in order to gain access to the proximal portion of the sinus in the event its distal portion is damaged

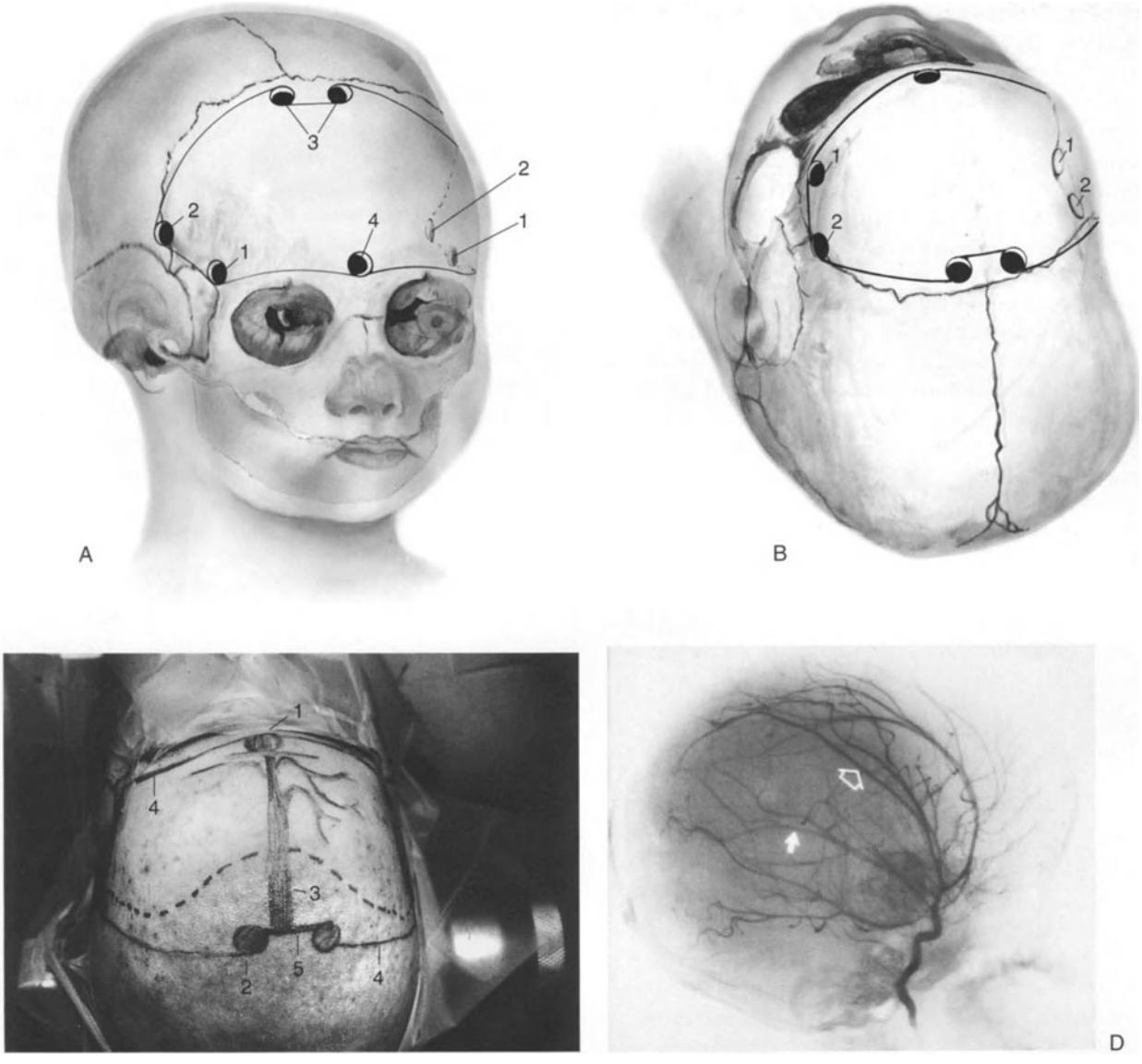


Figure 3.3. (A) Bur holes and osteotomy lines for bifrontal bone flap with child in the anatomical position. The severe bur holes are located at the "keyhole" (*1*), the pterion (*2*), the parasagittal area (*3*), bilaterally, and at the glabella (*4*) in the midline. (B) Illustration of bur holes and osteotomy lines for bifrontal flap with child in the supine position, and projected obliquely to put the "keyhole" (*1*) and pterional (*2*) holes into relief. (C) The skin incision (*broken line*) for the bifrontal flap follows the hairline. The glabella (*1*) and parasagittal (*2*) bur holes are placed, respectively, over the most anterior inferior portion of the superior sagittal sinus (*3*) and to either side of this structure approximately at the coronal suture. Note the craniotomy lines (*4*). They connect the bur holes along the most peripheral arc of the bur holes with the exception of the two parasagittal ones (*5*), where they connect the most medial arcs. In (D) this cerebral angiogram of an infant with a calcified craniopharyngioma, the intimate anatomical relationships between the parasellar tumor and the anterior, middle, and posterior cerebral arteries are shown. In addition to these one has an opportunity to appreciate the remarkable alterations in these vessels which result from the hypertensive biventricular *hydrocephalus* caused by the occlusion of the III ventricle by the calcified *craniopharyngioma*. Such a lesion golfball consistency, which is embraced by the entirety of the anterior circle vasculature, must be resected through a bifrontal pterional craniotomy.

during reflection of the free bone flap. If this does happen, the surgeon need only use a rongeur (to bite away the bony spur) to gain immediate access to the SSS. This precaution is best taken because of the proximity of the coronal suture to these parasagittal bur holes. Since the periosteum, coronal suture, and dura may all be adherent to one another at the suture, rongeuring bone from over the SSS at the point of the coronal suture may be both tedious and dangerous.

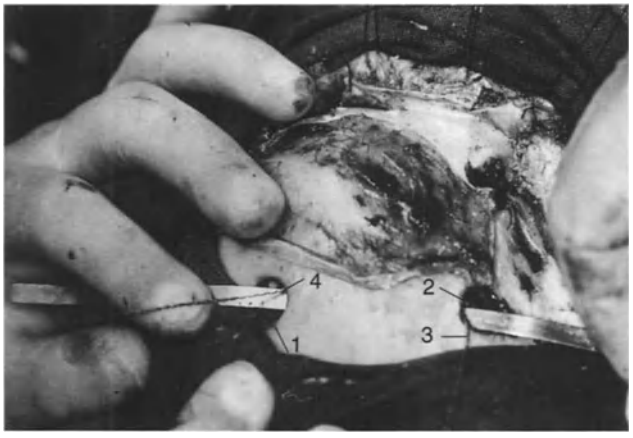


Figure 3.4. The Gigli saw guide has been passed from the parasagittal (*1*) to the pterional (*2*) bur holes and the saw subsequently brought into place. *On the right* the saw is positioned properly, at the peripheral arc of the bur hole (*3*), whereas *on the left* it is positioned improperly, at the central arc of the bur hole (*4*).

Frontal Flap (Figs. 3.5, 3.6)

Unilateral frontal flaps may be placed medially or laterally, depending upon whether the surgeon wishes to approach the parasellar area along the falx from only one side (in which case the medial frontal flap is preferable), or whether he wishes access to the frontal lobe (in which case the lateral frontal flap is preferable).

One notes immediately that the medial frontal flap is almost exactly half of the bifrontal flap. The sole exception is that the medial osteotomy incision extends to the parasagittal bur hole on the homolateral side of the flap, so that the SSS is exposed only at its most anteroinferior point (where it originates within the falx cerebri, extending from the crista galli of the ethmoid). The lateral frontal flap differs from the bifrontal craniotomy in that the anteromedial bur hole is not located over the glabella, but over the medial aspect of the supraorbital ridge. In this instance, no portion of the SSS is exposed.

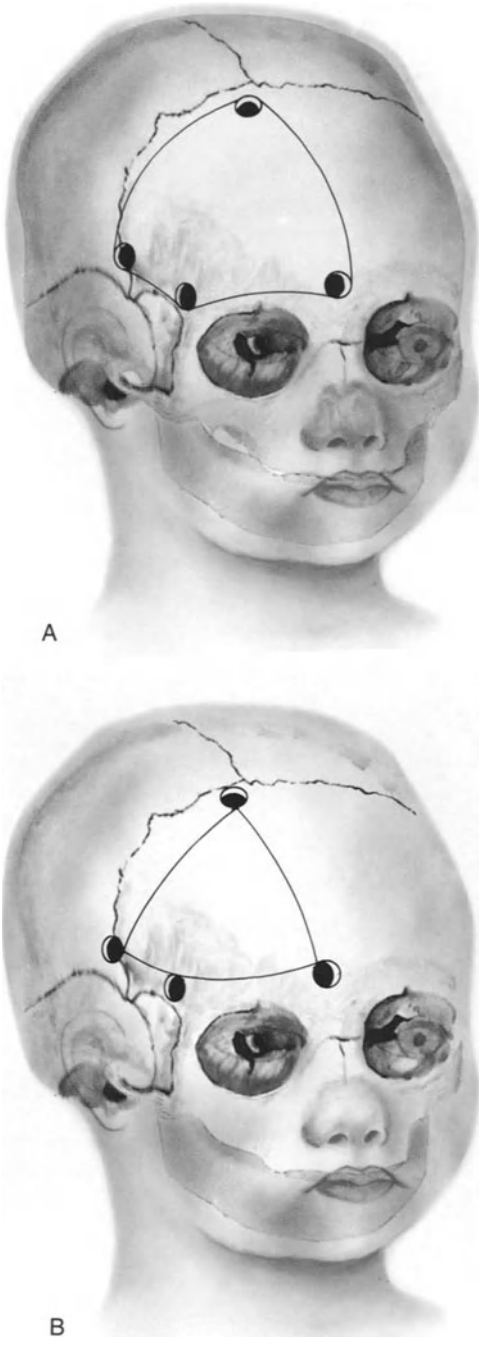


Figure 3.5. Medial frontal flap, illustrating placement of inferomedial bur hole at glabella. Note that the craniotomy lines extending from one bur hole to the other pass from the most distal arc of the bur hole's circumference. This gives maximum surface area exposure. Compare A, the recommended opening, with B, undesirable placement of craniotomy lines, to appreciate how much difference in exposure is obtained if the Gigli saw is properly placed.

Approaches to the Orbit

As general information, without any intention to present specific surgical technique for the performance of approaches to the orbit other than the transcranial, we describe the transethmoidal, superior lateral, lateral orbitotomy (of Krönlein), extended lateral orbitotomy of Jones, and the supraorbital approach of Jane.

The indications for the transfrontal approach are given in the section dedicated to the description of that operative technique. It is generally assumed that tumors

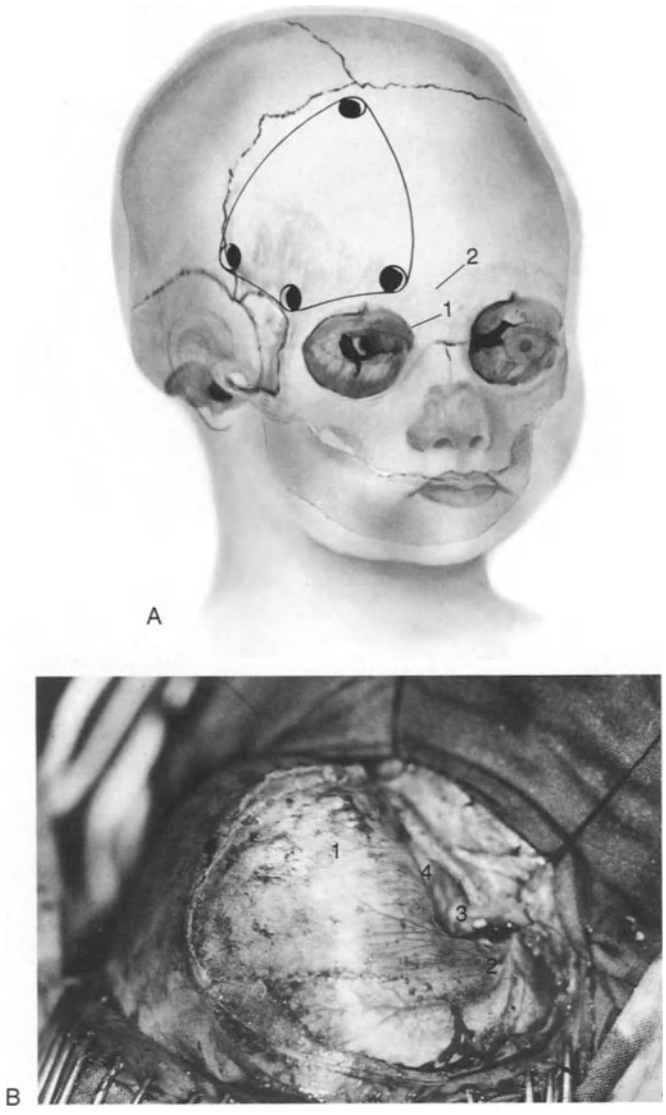


Figure 3.6. (A) Lateral frontal flap, illustrating placement of inferomedial bur hole over medial aspect of supraorbital ridge (*1*), lateral to the glabella (*2*). (B) Exposure of the frontal (*1*) and temporal (*2*) lobes as well as the pterion (*3*) and lesser wing of the sphenoid (*4*), which is attained when one reflects a lateral frontal flap (as illustrated in A).

of the orbital apex and in the superior orbit are best approached through a transfrontal craniotomy. Ophthalmic, plastic, and otorhinolaryngologic surgeons, however, all have occasion to operate on tumors either primarily within or extending into other orbital borders. Consequently, it has been suggested that tumors located along the inner wall of the orbit are best approached through the transethmoidal route, those of the lacrymal region through the superior lateral route, and tumors of the posterior lateral and inferior portions of the orbit through either the lateral orbitotomy of Krönlein or the extended lateral orbitotomy of Jones.

Transethmoidal Approach

The skin incision extends from just distal to the inner canthus, along the side of the nose and over the frontal process of the maxilla, inferiorly to the superior surface of the alar cartilage. The lacrymal sac is displaced laterally after the periorbita has been opened, before resecting the internal walls of the ethmoids. Tumors, primarily osteomas, located within this region are immediately visualized.

Superior Lateral Approach

The incision is made along the lateral two-thirds of the brow, curving around the zygomatic process of the frontal bone down to the lateral canthus, and the periosteum separated superiorly and inferiorly along the entire line of the incision, taking care to incise the periosteum well superior to the exit of the frontal nerve from the supraorbital groove (or foramen). Two bur holes are placed, one at the "keyhole" and the other above the supraorbital rim at the most medial skin incision opening, with attention being given to avoid the frontal sinus. They are connected to one another with the use of a Gigli saw, before the use of a high-speed drill to perform an osteotomy along the orbital roof so as to free a single orbital rim flap. One takes the same precautions to protect the periorbita as described later for the transfrontal approach.

Lateral Orbitotomy (Krönlein Approach) [1]

For the Krönlein approach, a horizontal skin incision is made from the external canthus posterolaterally for a distance of approximately 5 cm, prior to performing a canthotomy to expose the lateral wall of the orbit. The temporalis muscle and fascia are stripped from the lateral wall of the orbit and then incised in a plane parallel to, and immediately beneath, the skin incision, exposing the bony structures of the lateral wall, which, in turn, is opened with a high-speed drill in order to gain access to the periorbita. After the periorbita is opened, the region of the lacrymal gland and the anterolateral portion of the orbit are exposed. Further exposure may be gained by use of a rongeur to nibble away the necessary amounts of the lateral orbital wall. The use of the Stryker drill is discouraged because it may damage underlying periorbita, whereas a high-speed bur (though more time consuming) permits a relatively precise opening with minimal risk of damaging underlying soft tissue.

Extended Lateral Orbitotomy of Jones [2]

For Jones's extended lateral orbitotomy, a skin incision is made from the center of the supraorbital rim (in the brow) along the zygomatic process of the frontal bone and the frontal process of the zygoma, to the malar bone. Then another incision is made, perpendicular to this, extending from the outer canthus to a point approximately 1.5 cm anterior to the base of the antitragus, along the zygomatic arc. The underlying periosteum is dissected free, as are the temporalis fascia and muscle, exposing the entire lateral surface of the orbit. The bony opening is the same as for the lateral orbitotomy of Krönlein, except that it is extended across the malar bone. The inferior cut may come from the orbit lateral to the inferior orbital nerve. Once the periorbita has been dissected free, the bony opening is extended with a cut that dissects the zygomatic arch. This permits reflection of an osteoplastic flap attached to the temporalis muscle. Further opening of the floor and the lateral wall of the orbit may be obtained with the use of a Leksell rongeur.

Supraorbital Approach of Jane [3]

For Jane's supraorbital approach, a bifrontal skin incision is reflected and the underlying periosteum and frontal bone exposed, carrying the dissection inferiorly as far as the glabella and the entirety of the superior half of the orbital rim (medial, superior, lateral) on the involved (exposed) side. The dissection is continued inferiorly as far as the zygomaticofrontal suture and zygomatic arch. Bur holes are placed at the glabella and the "keyhole," and then a craniotomy is used to perform an osteotomy extending posteriorly from the glabella, in a parasagittal plane, for approximately 6 cm. It is then curved laterally coming across the superior temporal line and then inferiorly to the keyhole opening. The Gigli saw guide is passed from the glabella to the keyhole opening, and then the Gigli saw is used to connect these to one another, *bringing the cut through the orbit, not the intracranial portion of the anterior fossa* along the superior orbital rim. This frees a unilateral frontal flap with the supraorbital rim, in a single piece, exposing the frontal lobe and orbit.

Transfrontal Approach to Orbit(s) or Cribriform Plate (Figs. 3.7–3.12)

If a bifrontal bone flap is reflected and then an extradural dissection is performed, one has excellent exposure of the orbital roofs, the planum sphenoidale, and the cribriform plate. The extradural dissection may extend directly posteriorly or from lateral to medial, along the lesser wing of the sphenoid, in order to expose completely the anterior fossa on one side. This approach, along the lesser wing of the sphenoid, assures complete exposure of the roof of the orbit and brings the surgeon safely to the anterior clinoid and planum sphenoidale. The bilateral exposure, with coagulation of the rootlets of the olfactory nerve at their exit from the olfactory bulb, is used in craniofacial procedures and in repair of cerebrospinal fluid leaks through the cribriform plate (resulting from basilar skull fractures). A medial frontal (unilateral) flap may be reflected if one wishes access only to the cribriform plate, the intraorbital contents, or the orbital roof on one side. It is adequate for access to the *planum sphenoidale*, ideal for exposure of one anterior clinoid or optic nerve. Indeed, for intraorbital surgery a medial frontal craniotomy is recommended.

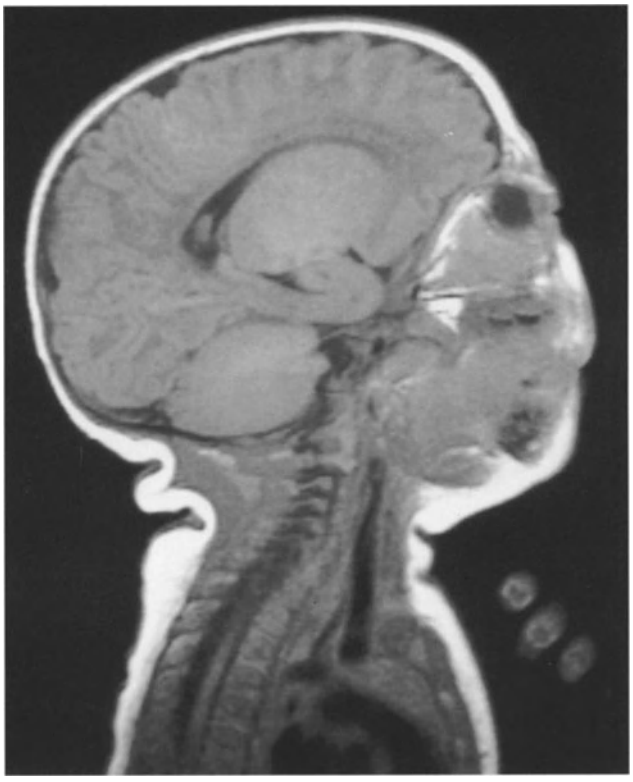


Figure 3.7. As we shall see in the following illustrations supraorbital and transfrontal approaches to the orbit permit the surgeon to operate effectively those tumors which occupy the superior half of the orbit, whether superior to the levator palpebrae or between the superior rectus and the optic nerve. However, diffusely infiltrating tumors, especially lymphangiomatous or angiomatous in nature, most often expand within the adnexa, displacing superiorly the superior rectus and the globe. In fact, the optic nerve often bows over the superior bulging of these tumors. Such an example is illustrated in this figure, an MRI study on a 1-month-old child with a progressively growing *lymphangioma*. Ideal access to these tumors, and certainly the simplest and safest since one may wish to limit oneself to a biopsy, is obtained either through the lateral orbitotomy (Krönlein approach) or the extended lateral orbitotomy of Jones.

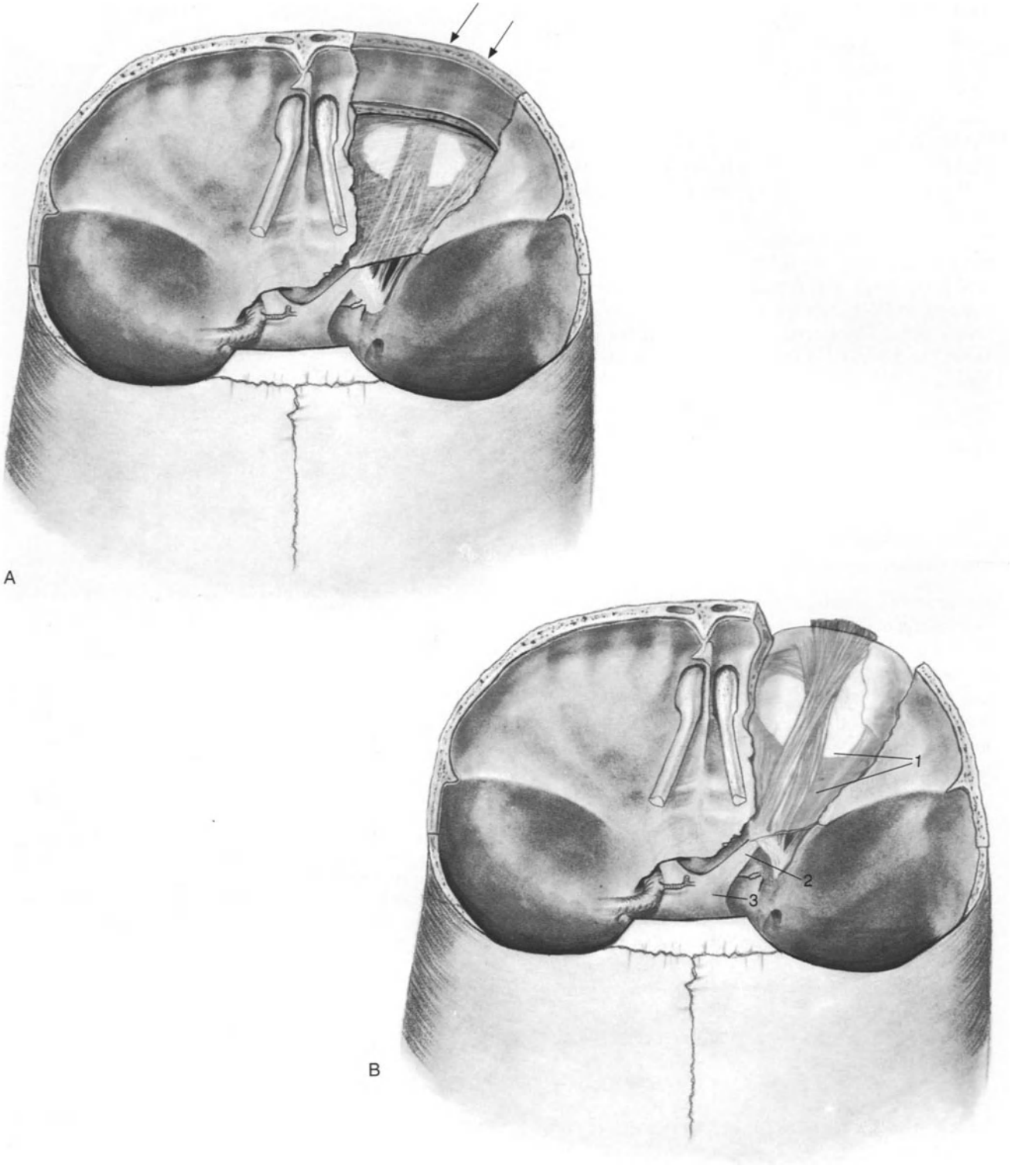


Figure 3.8. (A) The surgeon's view of the anterior fossa of a bifrontal craniotomy is followed by an extradural dissection of the orbital roof, with opening of the latter but leaving the supraorbital rim intact (*arrows*). (B) The supraorbital rim has been removed *en bloc*, after the dura was separated from the roof of the orbit during an approach down the lesser wing of the sphenoid (from the pterion to the anterior clinoid process). The roof of the orbit was then resected, using rongeurs posteriorly, to include the anterior clinoid process exposing the intact periorbita (*1*) and the optic nerve both within the optic canal (*2*) and at its entry into the optic chiasm (*3*). One must open the dura at the optic foramen if it is wished to follow the optic nerve into the optic chiasm. (C–D) see p. 87.

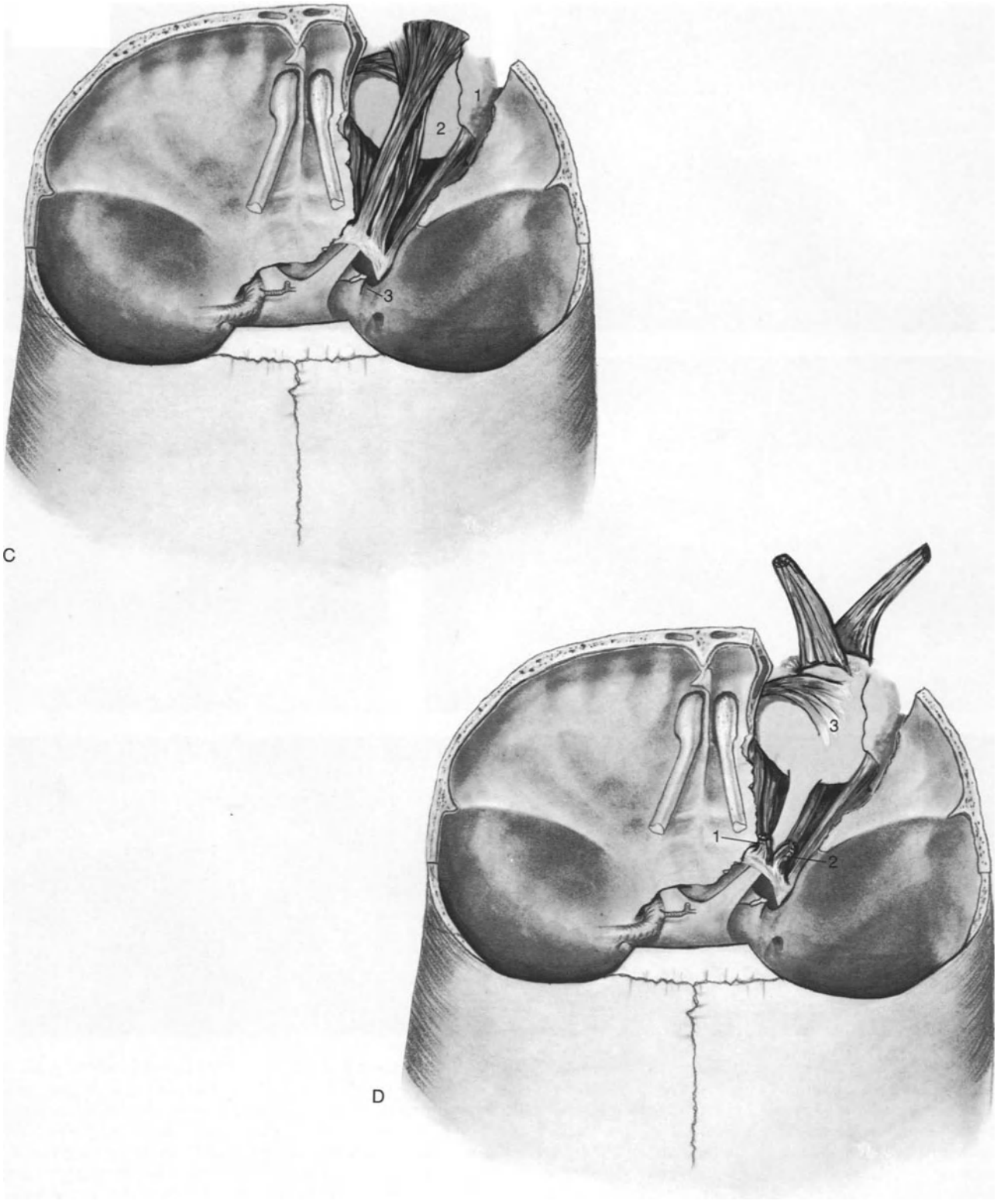


Figure 3.8. (C) The periorbita has been removed, bringing into relief the extraocular muscles, lacrymal gland (*1*) and medial wall of the orbit. This is the exposure one has of the globe (*2*), cone, optic canal, and the inferior orbital fissure (*3*) if the supraorbital rim is removed and the orbital roof is resected to the optic foramen posteromedially and the superior orbital fissure posterolaterally. (D) Both the levator palpebrae superioris (*1*) and the superior rectus (*2*) have been transected and reflected from the globe and muscular cone, exposing completely the intraconal contents, the exit of the optic nerve from the globe, and the insertion of the superior oblique muscle (*3*) onto the globe.

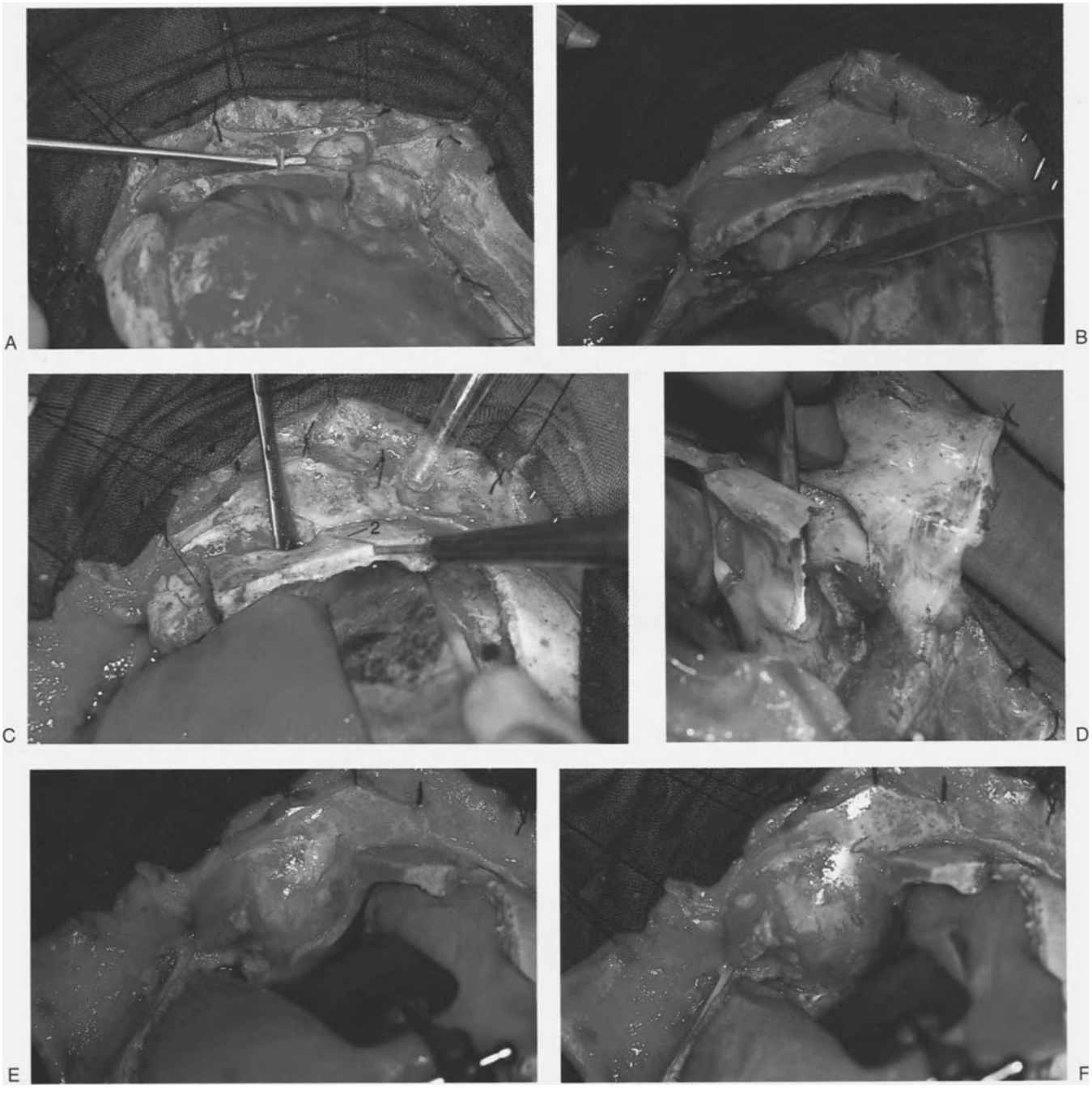
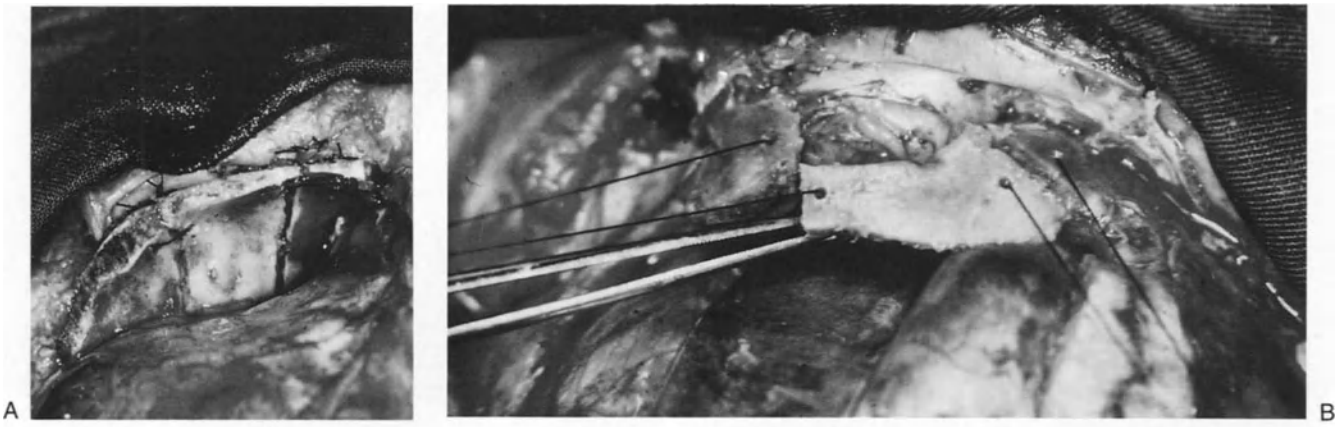
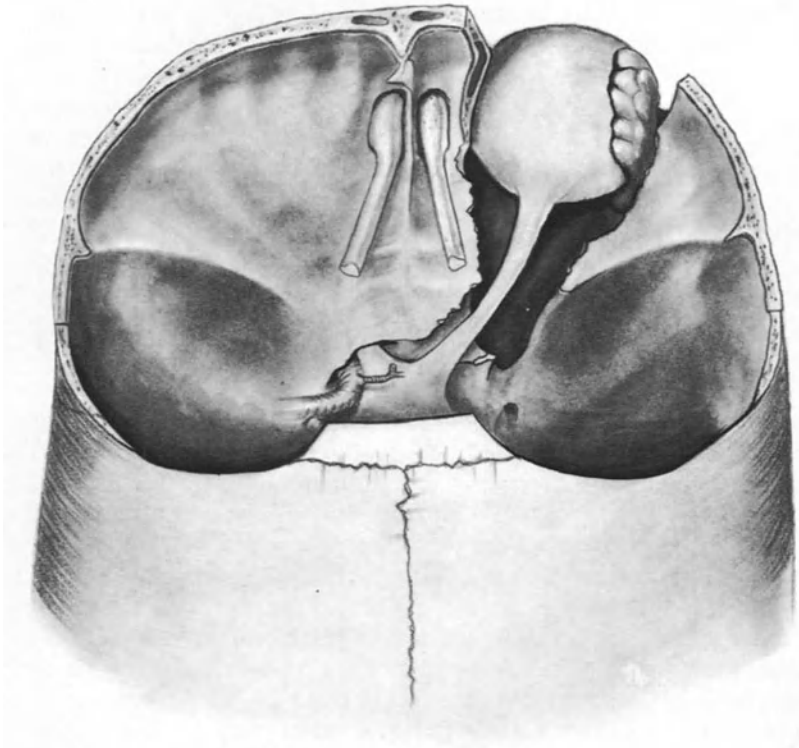


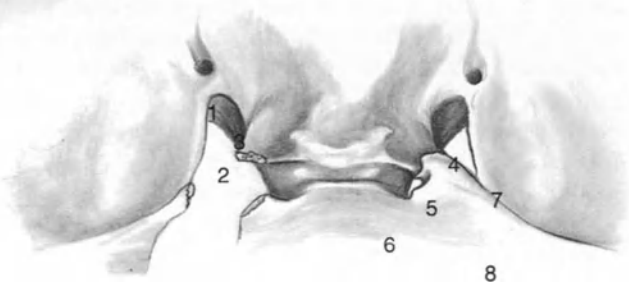
Figure 3.9. The technique for unroofing the supraorbital rim. (A) One notes the technique for preserving the frontal nerve. After the periosteum has been reflected onto the galea and sewn down, one identifies the supraorbital groove (sulcus or foramen) and then isolates the frontal nerve, taking care to separate it from the bone. Allow it to remain adherent to the periosteum peripherally. Respect its integrity at the point where it perforates the periorbita. (B) The periosteum has been stripped from the supraorbital rim, reflected anteriorly, and sewn to the galea of the frontal scalp flap so as to keep it stretched. The dura has been stripped from the inner surface of the frontal bone and roof of the orbit, isolating glabella, supraorbital rim, zygomatic process of frontal bone, and orbital roof. (C) Separation of the periorbita from the orbital surface of the orbital roof. A Penfield #4, or #2, dissector is quite adequate for performing this procedure. One should feel the bone of the roof of the orbit with the dissector at all times. This photograph illustrates the technique for osteotomizing the supraorbital rim. *On the left* (*1*), the osteotomy has already been performed with a high-speed drill. *On the right* (*2*) the high-speed drill is being used to cut the frontal bone. Subsequent to this, a Penfield #3 or 4 dissector is placed between periorbita and roof of the orbit, along the line of desired osteotomy in the roof of the orbit. This serves as both a guide and a protection against tearing the periorbita or orbital contents. (*Continued* on p. 89).



▲ **Figure 3.10.** At the end of the procedure one simply reapproximates the orbital roof and supraorbital rim (A) and then anchors it into place (B).



◀ **Figure 3.11.** Anatomical representation of the relative size, location, and course of the optic nerve, diagrammatically exposed by removing the extraocular muscles and adnexa.



◀ **Figure 3.9.** (D) Once the osteotomy has been performed, the Penfield dissector is swept posteriorly, stripping all of the periorbita from the orbital surface of the roof. At this time the supraorbital rim may be lifted away *en bloc*. (E) One then notes some remaining orbital roof posteriorly. (F) This is bitten away with a Leksell rongeur. At this time, one has excellent visualization of the intraorbital contents, still covered by the periorbita.

Figure 3.12. Compare resected side (*left*) with intact side (*right*). Anatomical representation of unroofing of the most posterior portion of the orbit (*1*), removing the anterior clinoid process (*2*) with the lateral strut of the optic canal (*3*). This opens completely the superior orbital fissure (*4*). Note that unroofing of the orbit brings the surgeon into the superior orbital fissure. This latter structure and the inferior orbital fissure (*5*) are in direct continuity with one another along the lateral surface of the body of the sphenoid (*6*) and the medial surface of the greater wing of the sphenoid (*7*). The area of the foramen rotundum (*8*) is also shown.

After the orbital roof has been exposed, one may choose only to unroof the orbit if one is certain that the intraorbital tumor is located posteriorly, at the apex of the *cone*, or to remove the supraorbital rim if more complete access to the globe and medial portion of the orbital cone is desired. Leaving the supraorbital rim in place limits considerably the exposure of the intra- and extraconal contents, does not permit exposure of either the lacrymal gland or the trochlear region, and precludes rolling the globe over itself to inspect the region where the inferior oblique and inferior rectus muscles insert onto it. When the periorbita is reflected to either side of the midline, opening it exactly as one would the dura mater but remembering that it is only half as thick (since it is the periosteal layer of the dura mater), fat insinuates itself quickly through the incisional line. Anchoring of the reflected periorbita is advisable after it is opened, since it allows the surgeon to work freely within the orbit, without having flaps of periorbita repeatedly falling back into the field.

One now has a complete view of all of the orbit as well as the globe and its adnexa. If it is desirable, for example, when beginning resection of a nerve sheath tumor, an angioma, and so on, one may expose the apex of the cone by transecting the levator palpebrae superioris and the superior rectus muscles, sewing them out of the way, and proceeding to work within the base and the apex of the cone. At the end of the procedure, these muscles may be brought back into the field and the cut trunks of each sewn individually to one another, restoring anatomical continuity and function. The orbital roof and supraorbital rim are now replaced and anchored.

On occasion, when dealing, for example, with optic nerve tumors that involve the intraorbital and intracranial portions of the optic nerve but spare the optic chiasm, one may choose to perform a combined extradural approach to the orbit (in order to resect the intraorbital portion of the tumor) and an intradural approach to the parasellar area (so as to follow the tumor through the optic foramen and resect it at the point of entrance of the involved optic nerve into the optic chiasm). Figure 3.11 illustrates the relative anatomical positions of the globe, intraorbital optic nerve, intracranial optic nerve, and optic chiasm, after the extraocular muscles have been removed. A view of the unroofed orbit, with the osteotomy extending into the superior orbital fissure, and with the anterior clinoid removed and optic canal opened, is illustrated in Fig. 3.12. This allows the reader to understand that unroofing the orbit posteromedially, that is, resecting the lesser wing of the sphenoid, is to remove the superior border of the superior orbital fissure and expose the inferior orbital fissure [4].

The opening of the optic canal is accomplished by taking the anterior clinoid from the superior surface of the internal carotid artery and the optic nerve, and then resecting the lateral orbital strut so as to expose completely the optic nerve.

Parietal Flap (Figs. 3.13–3.16)

The parietal bone flap may be penta- or quadrilateral, depending upon the convexity of the skull, the specific location of the flap with regard to its extension across the coronal suture, or its limitation entirely to the parietal bone. The pentalateral flap permits a craniotomy that extends well anterior to the coronal suture and, consequently, one with greater curvature. The quadrilateral flap is almost completely limited to the parietal bone so that the curvature is less and the need for the fifth bur hole does not exist. Parietal flaps allow access to the convexity of the parietal lobe or to the SSS and falx cerebri. Also, one may continue the dissection down the falx cerebri to the corpus callosum for tumors within this structure, or split the corpus callosum for access to tumors (almost invariably pineal tumors) between it and the roof of the III ventricle. Because of the presence of the internal cerebral veins in the roof of the III ventricle, this approach along the falx cerebri for entrance into the III ventricle is not recommended. The parasagittal approach to the corpus callosum offers relatively immediate access to the entirety of this commissure from the genu to the splenium. Although the dissection around the medial surface of the parietal lobe is tedious, owing to the presence of bridging cortical veins, it is not as difficult as it is in adults. Children do not have adherent arachnoidal granulations binding the arachnoid to the dura, the bridging cortical veins, and the SSS.

Immediately beneath the body of the corpus callosum one finds the roof of the III ventricle, and beneath the splenium of the corpus callosum one encounters the suprapineal recess, pineal gland, and collicular plate. Therefore, the parietal flap may be used for pineal tumors which extend between the roof of the III ventricle and the undersurface of the corpus callosum, or for arteriovenous malformations of the galenic system in which the tributary vessels enter this anomaly along its superior surface.

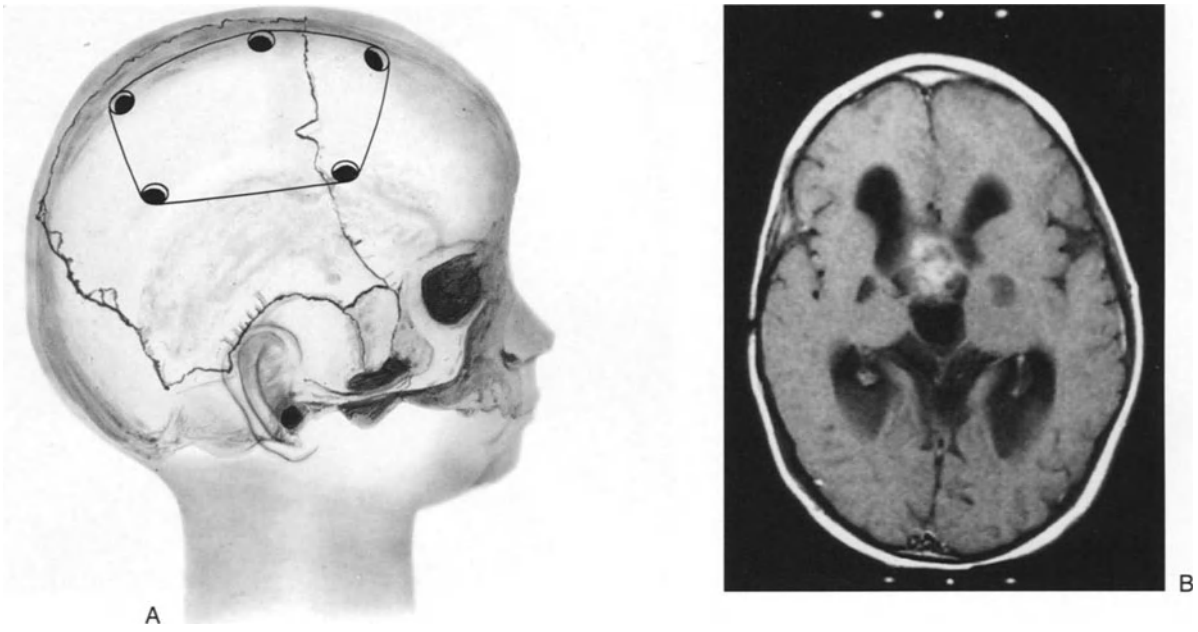


Figure 3.13. (A) This figure illustrates a pentalateral parietal flap, with the most anterior limb extending across the coronal suture, into the frontal bone. (B) The pentalateral flap is an ideal optional for *subependymal glioma* of the III ventricle, in this case occluding both foramina of Monro because of its very anterior location, and probably resting upon the lamina terminalis. Consequently a frontoparietal pentalateral flap permits one access to the homolateral foramen of Monro, the septum pellucidum, and a parasagittal descent in the event a transcallosal approach is also desired.

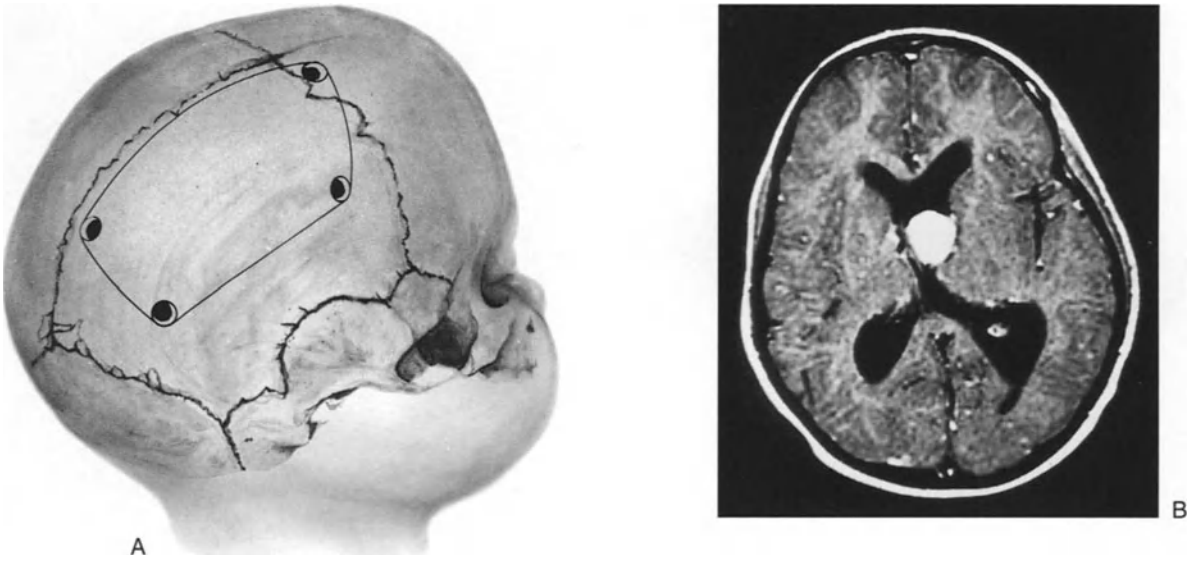


Figure 3.14. (A) A quadrilateral parietal flap offers exposure of the parietal convexity, parasagittal area, corpus callosum, and III ventricle. (B) This MRI scan is slightly different from that illustrated in the previous figure in that the subependymal glioma is located superior and posterior to the foramen of Monro, only partially obstructing that *on the right*, and extending posteroinferiorly to obstruct the aqueduct of Sylvius. Therefore, a large frontal component to the parietal flap is not indicated, a quadrilateral parietal flap suffices to permit the surgeon entrance into the homolateral ventricle opening of the septum pellucidum and the peeling away of the choroid plexus with the fornix from the region of the lamina affixa of the thalamus for complete exposure of the III ventricle.

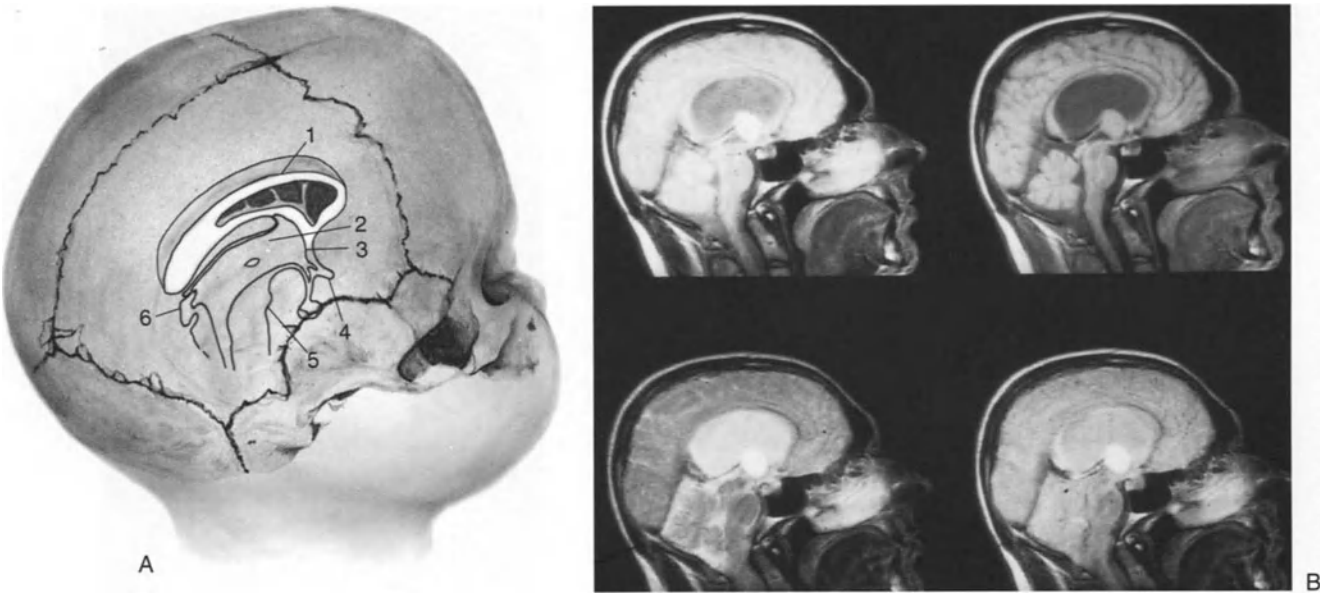


Figure 3.15. (A) This is an anatomical representation, depicting diagrammatically, in three dimensions, the access a quadrilateral bone flap affords to the corpus callosum (*1*), III ventricle (*2*), septum pellucidum (*3*), optic chiasm (*4*), midbrain (*5*), and pineal gland (*6*). (B) By referring to the schematic drawing superimposed upon the skull as illustrated in (A), one may place anatomically this *colloid cyst* of the III ventricle in a 5-year-old child. For its resection, the pentalateral parietal flap illustrated in Fig. 3.13 is preferable.

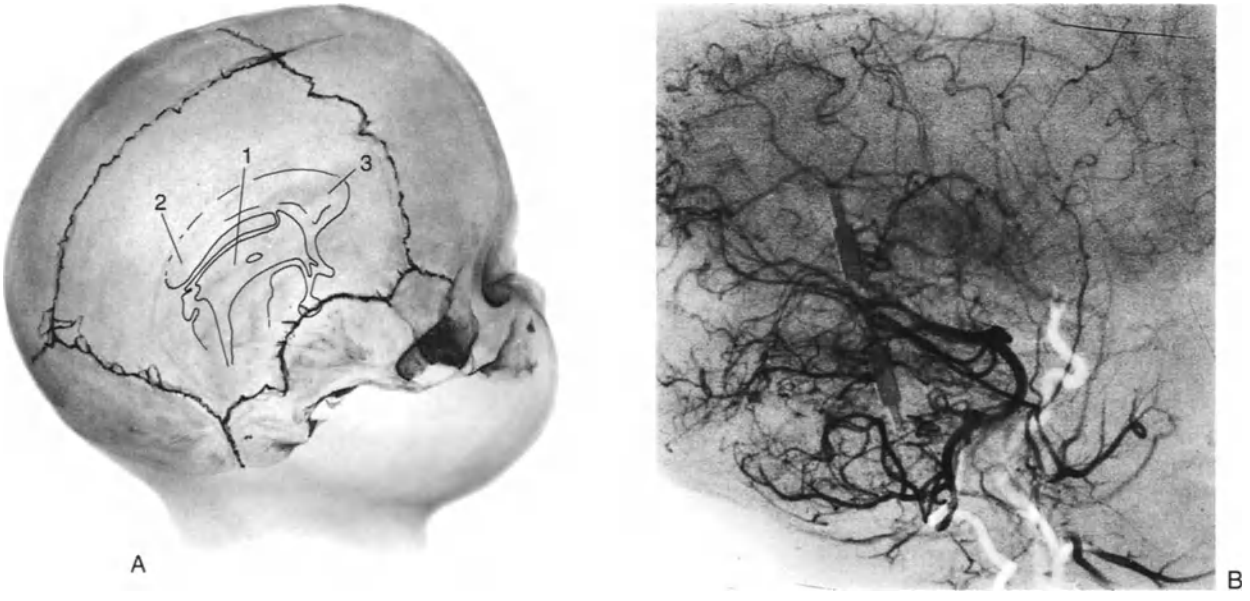


Figure 3.16. The III ventricle (*1*) is projected into its anatomical position in this three-dimensional drawing to illustrate access to it through a parietal flap. Either transventricular or parasagittal approaches may be used. The splenium (*2*) and genu (*3*) are shown at the posterior and anterior surfaces of the III ventricle. (B) The *chorioid plexus papilloma* of the III ventricle drained directly into the internal cerebral veins in the roof of the III ventricle, providing clear indications for a transcallosal approach: the transforaminal approach does not permit one to isolate microsurgically the internal cerebral veins from the dome of the papilloma.

Parietotemporal Flap (Figs. 3.17, 3.18)

The parietal flap may easily be extended into a parietotemporal flap, if the parietal lesion also invades the superior or the middle temporal convolutions posteriorly (in correspondence to the supramarginal and angular gyri). Similarly, one may place the bur holes more laterally on the convexity, exposing only the inferior portion of the parietal lobe and, with this, the posterior superior portions of the temporal lobe. This entails placing the superior bur holes at (1) the parietal eminence, (2) the coronal suture, (3) the pterion, and (4) just above the base of the mastoid bone.

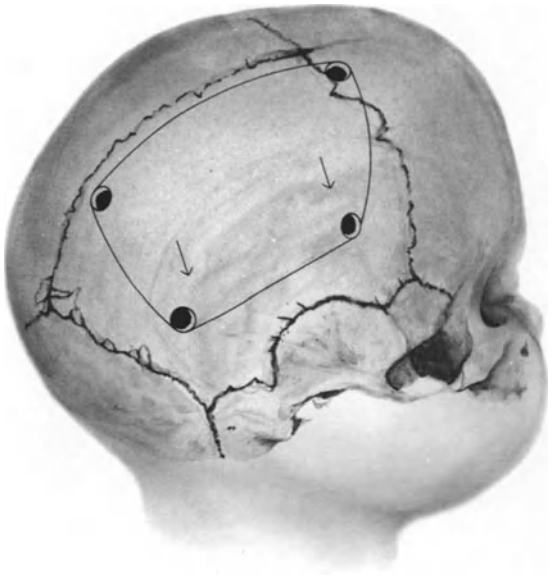


Figure 3.17. One may gain access to the supramarginal and angular gyri as well as the superior temporal convolution through a superior parietotemporal flap, by placing the inferior bur holes below the superior temporal line (*arrows*).

Biparietal Craniotomy (Figs. 3.19, 3.20)

The biparietal craniotomy is used for resection of the sagittal suture (really a craniectomy), to gain access to the SSS or both sides of the falx cerebri, to attain complete exposure of the great vein of Galen for arteriovenous fistulae, and to lower the SSS for chronic subdural hematoma.

Depending upon whether the target area is located entirely behind the coronal suture or whether, for example, when approaching lacerations of the SSS or wishing to approach the genu of the corpus callosum, it extends anterior to the coronal suture and into the frontal bone, one may reflect either quadrilateral or pentalateral flaps. Although the skull may be lifted from the SSS safely, especially in infants with sagittal synostosis, reflecting two separate flaps is preferable: it permits more protection to the SSS both during and after the surgery. If, however, the biparietal flap is reflected in a single piece, then great care must be taken to use the Gigli saw so as to assure solid seating of the bone flap at the time of closure. Sinking of a biparietal flap may cause compression or occlusion of the SSS.

The intraoperative precautions to be taken when reflecting a biparietal flap in a single piece of bone are (1) to assure beveled edges by using the Gigli saw, and (2) to suspend the parasagittal dura anteriorly and posteriorly, immediately after the bone has been removed from over the SSS, on either side, to minimize the risk of intraoperative kinking of the SSS.

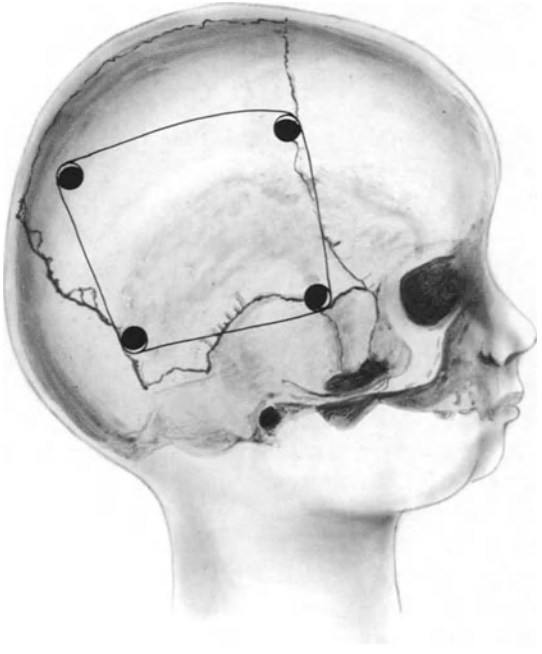


Figure 3.18. Inferior parietotemporal bone flap.

Temporal Flaps

Temporal bone flaps may be placed in three basically different positions, depending upon whether the surgeon wishes access to the temporal tip, the posterior portion of the temporal lobe, or the entirety of the temporal lobe and the sylvian fissure.

Figure 3.19. Biparietal quadrilateral bone flaps, placed one on either side of the sagittal plane, leaving the bone over the superior sagittal sinus (SSS) intact. In the event access to the SSS becomes desirable, this strip of bone may be quickly removed.

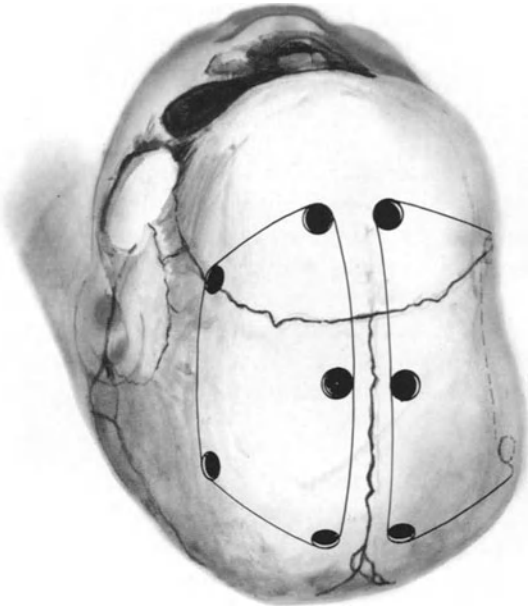


Figure 3.20. Bilateral frontoparietal (pentalateral) bone flaps may be extended across the coronal suture, leaving bone over the superior sagittal sinus.

Anterior Temporal Flaps (Figs. 3.21, 3.22)

The *anteroinferior bur hole* is placed at the pterion so that the surgeon may have immediate access to the temporal pole. Dissection of the temporalis muscle should be extensive, exposing the greater wing of the sphenoid and the squamous portion of the temporal bone. This allows for craniectomy of these two latter bones in the event one wishes to gain access to the floor of the temporal fossa. Since this is a common site for dural sarcoma, aplasia of the temporal lobe, arachnoidal cysts of the sylvian fissure, and dermoid tumors, it is of importance to take care to assure access to the inferior portion of the temporal fossa during the preparation of the craniotomy for an anterior temporal approach. The *superior bur holes* are placed immediately beneath the superior temporal line, the *posteroinferior bur hole* above the base of the mastoid process.

Temporal flaps (anterior, posterior, middle) permit access to varying portions of the temporal lobe, the entirety of the sylvian fissure, the temporal horn and trigone, the lateral portion of the circle of Willis, the posterior cerebral and superior cerebellar arteries, the tentorial edge and ambient cistern, and the surface of the tentorium along the petrous apex. One may choose to reflect an osteoplastic (from the temporalis muscle) rather than a free bone flap for all temporal exposures (anterior, posterior, mid), since the closure provides much more solid seating of the bone.

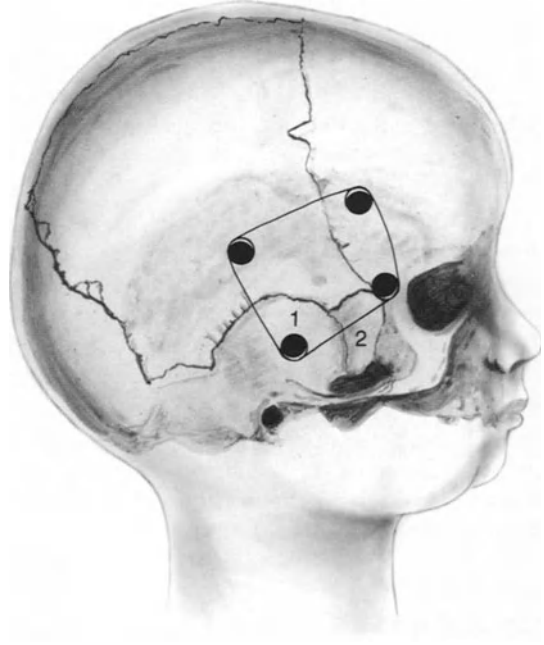


Figure 3.21. Anterior temporal craniotomy. The anteroinferior bur hole is placed at the key hole so as to assure exposure of the temporal pole. Note that the inferior craniotomy line extends through the squamous temporal (*1*) and greater wing of the sphenoid (*2*). Rongeuring these bones gives access to the floor of the middle fossa.

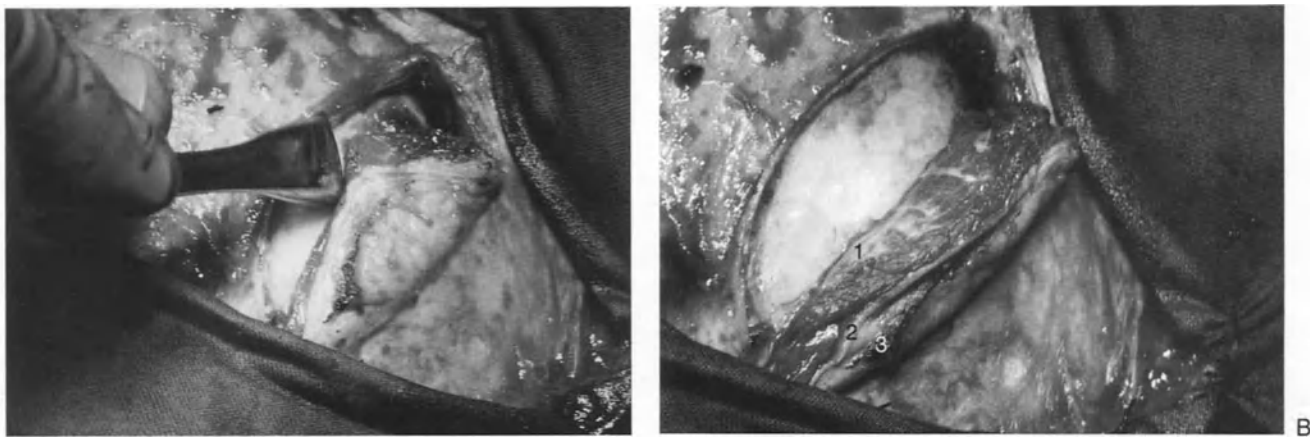


Figure 3.22. (A) Suggested technique for use of periosteal elevator to strip periosteum from the greater wing of the sphenoid and squamous temporal bones, so as to avoid fraying the periosteum: cutting edge to be held firmly and run parallel to muscle insertion. (B) The bipenniform temporalis muscle has been dissected from bone, preserving its three layers: (*1*) deep, (*2*) intermediate, (*3*) superficial.

Posterior Temporal Flaps (Fig. 3.23)

Posterior temporal flaps do not necessitate anteroinferior dissection into the bulk of the temporalis muscle and are, consequently, much easier to reflect. Similarly, since the posterior temporal lobe is considerably higher than the temporal pole, it is not necessary to dissect the temporalis muscle from the squamous temporal and greater wing of the sphenoid bones. The superior bur hole is placed at the superior temporal line, directly above the external auditory canal, and the inferior bur hole is placed at the base of the mastoid process. The anterior and posterior bur holes are placed within the parietal bone, posterosuperior to the pterion and directly inferior to the parietal eminence, respectively. This flap does not permit access to the floor of the middle fossa. Rather, it is for exposure of the temporal lobe (from the temporal operculum posteriorly) or for access to the trigone of the lateral ventricle.

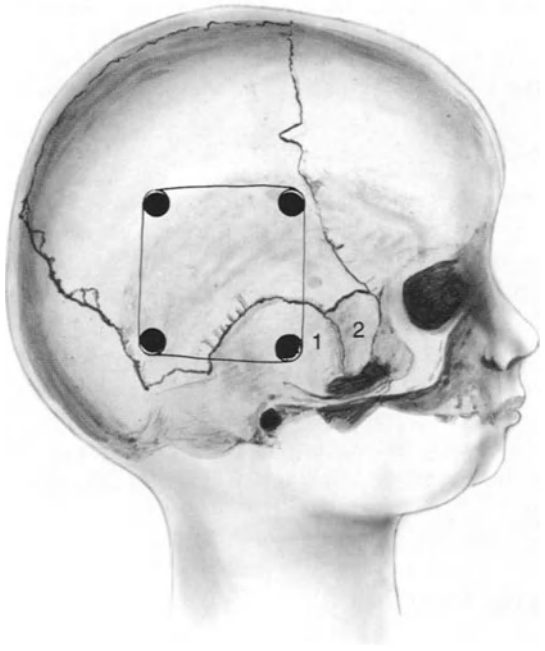


Figure 3.23. Posterior temporal flap, illustrating placement of bur holes. Note that no bur hole is placed at the pterion. Access to the anterior portion of the squamous temporal (*1*) and to the greater wing of the sphenoid (*2*) is not possible.

Mid-temporal Flap (Fig. 3.24)

The mid-temporal flap incorporates the anterior and posterior temporal bur holes so as to permit access to the entire temporal lobe, from the temporal pole as far posteriorly as the temporo-occipital area, and from the floor of the middle fossa as far superiorly as the frontal and parietal operculae anteriorly and the angular gyrus posteriorly. Tumors of the middle fossa, arteriovenous malformations of the temporal horn, the lateral portion of the circle of Willis, the tentorial edge and ambient cistern, and the hippocampus may all be safely approached through this flap. Also, one may approach, in the infant and the toddler, the superior cerebellar and posterior cerebral arteries, in their course around the midbrain, for occlusion of inferior tributaries to an arteriovenous malformation of the galenic system, brainstem, superior surface of the cerebellum, or medial surface of the occipitotemporal lobes.

The mid-temporal flap is generally pentalateral. The anteroinferior bur hole is placed at the pterion, the anterosuperior bur hole direcly above it at the superior temporal line. The posterosuperior bur hole is placed

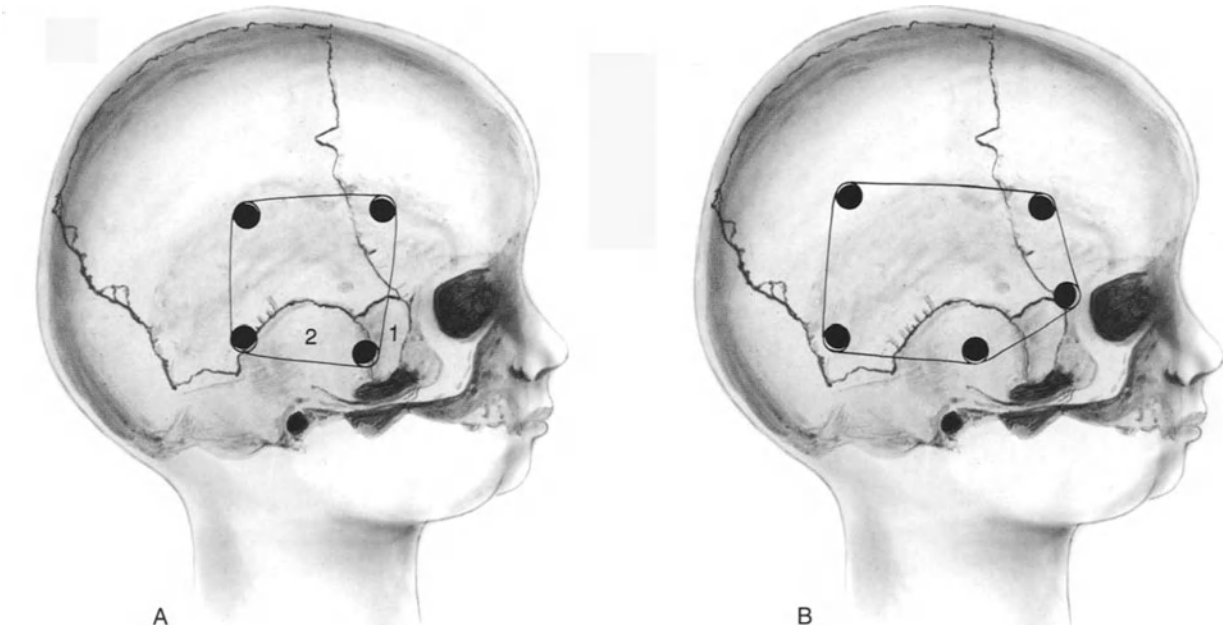


Figure 3.24. (A) Midtemporal flap showing the quadrilateral form of the bone flap and illustrating the greater wing of the sphenoid (*1*) and the squamous temporal bones (*2*). The superior bur holes are all just beneath the superior temporal line; the anteroinferior bur hole is at the pterion. One may, with this flap, have exposure permitted by both anterior and posterior temporal flaps, reflecting a pentalateral craniotomy. (B) Enlarged temporal bone flap.

immediately beneath the parietal eminence, and the posteroinferior bur hole just anterior to the lamboidal suture in a line above the digastric groove. The inferointermediate bur hole is placed at the floor of the middle fossa within the squamous portion of the temporal bone. Stripping the temporalis muscle (as just described) from the greater wing of the sphenoid and the squamous temporal bones permits the added margin of safety of being able to craniectomize portions of these bones for access to the floor of the middle fossa anteriorly.

Occipital Flaps (Fig. 3.25)

Medial Occipital Flap

A medial occipital flap permits access to the convexity, tentorial and falx surfaces of the occipital lobe. The occipital pole presents at the inferomedial aspect of this flap, at the tentorial junction. The culmen monticuli of the cerebellar vermis may also be exposed through this flap (after the tentorium has been incised and its cut edges reflected medially and laterally), as may the quadrigeminal cistern, the pineal region, and the torcular Herophili.

This approach to the pineal region is ideal for tumors expanding within the quadrigeminal cistern and the posterior portion of the III ventricle (as indicated angiographically by elevation of the posterior portion of the internal cerebral vein). It allows the surgeon control of the supraculminate vein and the great vein of Galen, as well as the possibility of separating the III ventricular portion of the tumor from the inferior surfaces of the internal cerebral veins.

The most important bur hole for free occipital bone flaps is the inferomedial one, which should be placed over the lateral portion of the torcular Herophili, at the junction of the superior sagittal and transverse sinuses. It is essential to have access both to the falx and the tentorium if one wishes to expose the quadrigeminal cistern and the pineal region. If, however, the convexity of the occipital lobe is the target area, then the inferomedial bur hole need not – and probably should not – be placed in such a critical area. Setting it slightly superiorly and laterally, a few millimeters from the superior sagittal and transverse sinuses, respectively, is quite adequate. Similarly, for access to the falx cerebri, the superomedial bur hole must be placed over the superior sagittal sinus, with its medial surface just touching the sagittal suture. The superolateral and inferolateral bur holes are placed, respectively, just behind the parietal eminence and over the lambdoidal suture (only slightly above the point where the transverse sinus turns inferiorly to extend into the sigmoid sinus).

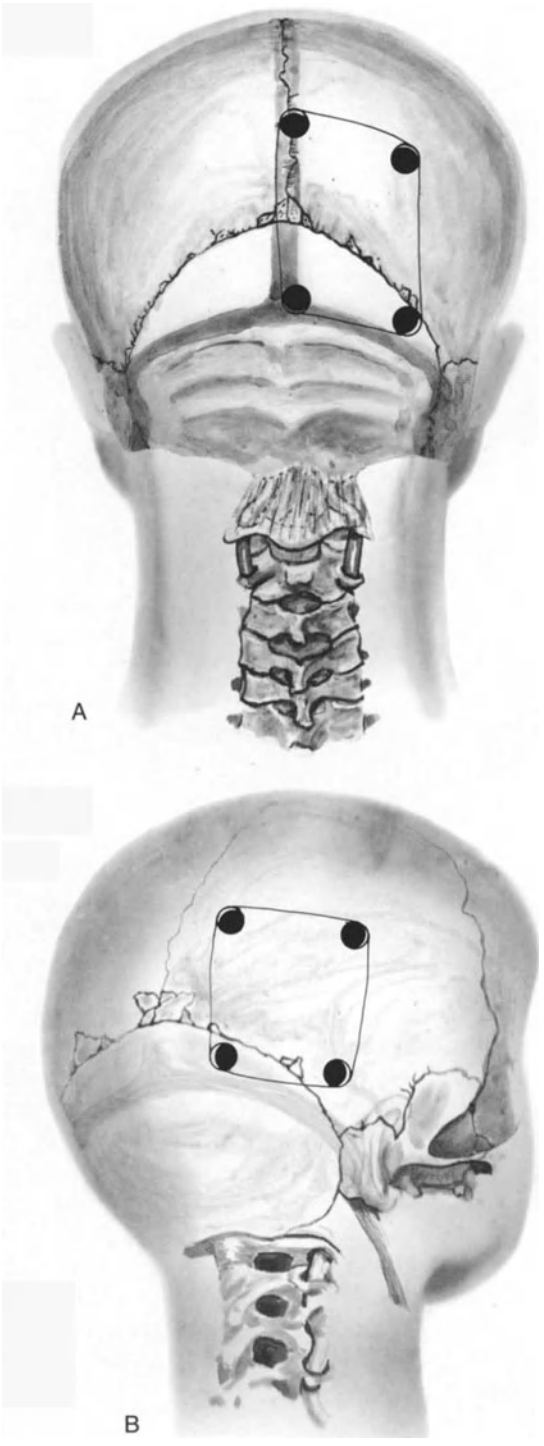


Figure 3.25. (A) Medial occipital bone flap, illustrating location of bur holes and osteotomy lines, as viewed by the surgeon, in a straight posteroanterior view. Note that the lambdoidal suture is within the flap, and that the medial osteotomy follows the sagittal suture. The inferior osteotomy line should be at, or below, the transverse sinus if one wishes to work along the medial and the lateral occipitotemporal gyri or within the pineal region. (B) Lateral occipital bone flap as viewed in a postero-oblique line, illustrating the location of the lateral (superior and inferior) bur holes.

Lateral Occipital Flap

The lateral occipital bone flap has the inferomedial bur hole just superolateral to the torcular Herophili, the inferolateral hole above the sigmoid sinus. The superior holes are placed medial and lateral to the parietal eminence.

Suboccipital Flaps

Suboccipital bone flaps may be midline or lateral.

Midline Suboccipital Craniotomy

The midline suboccipital craniotomy may be superior or inferior, depending upon whether the surgeon wishes to expose the superior or the inferior cerebellar triangles (Figs. 3.26, 3.27).

The *superior cerebellar triangle* has the horizontal fissure of the cerebellum as its base, the culmen monticuli its apex, and the (inferolateral coursing) superior surfaces of the cerebellar hemispheres its sides. The apex of the superior cerebellar triangle points upward.

The *inferior cerebellar triangle* also has the great horizontal fissure as its base, but its apex, the interval between the tips of the two cerebellar tonsils, points downward. The lateral surfaces of the inferior cerebellar triangle consist of the (superolateral coursing) cerebellar hemispheres.

Lesions within the superior triangle are tumors of the culmen monticuli and culmen declive, pineal tumors, and those arteriovenous malformations of the galenic system (whose tributaries enter the great vein of Galen posteriorly). Inferior triangle lesions include medulloblastoma, cerebellar hemisphere astrocytoma, ependymoma of the IV ventricle, foramen magnum tumors, arachnoid cysts of the cisterna magna, and other space-occupying lesions of the inferior vermis or cerebellar hemisphere.

Suboccipital Craniotomy Versus Craniectomy

The suboccipital craniotomy is preferable to the craniectomy! It permits the repositioning of a solid bone flap over the closed dura (Fig. 3.28), giving the child an anatomical reconstruction of the suboccipital area, not entrusting protection of the contents of the posterior fossa to the very thin muscle layers at the base of the skull. It eliminates completely the all-too-common suboccipital bulge observed in children who have had a posterior fossa craniectomy, a bulge holding herniated cerebellum (hence stretched cerebellar peduncles) and cerebrospinal fluid.

The practice of performing a suboccipital craniectomy and of not closing the dura should be avoided!

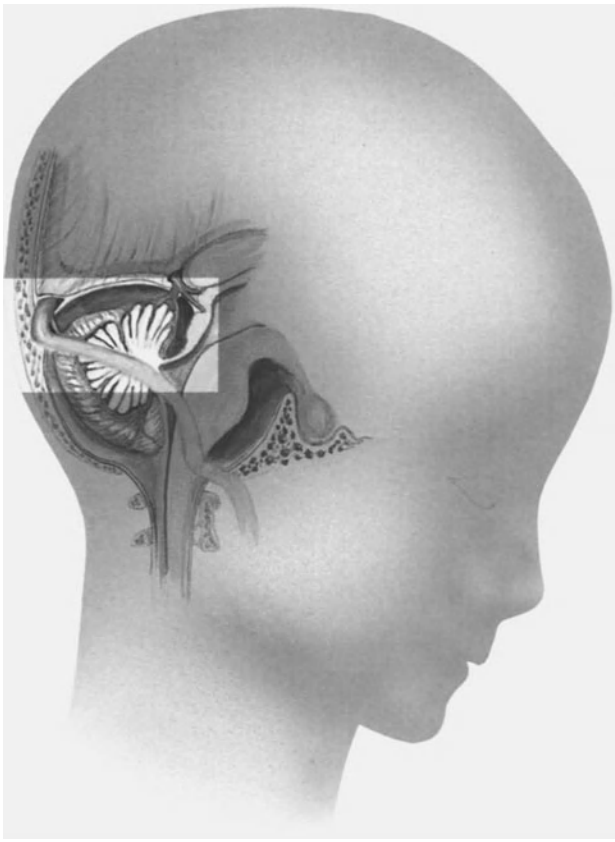


Figure 3.26. Superior cerebellar triangle.

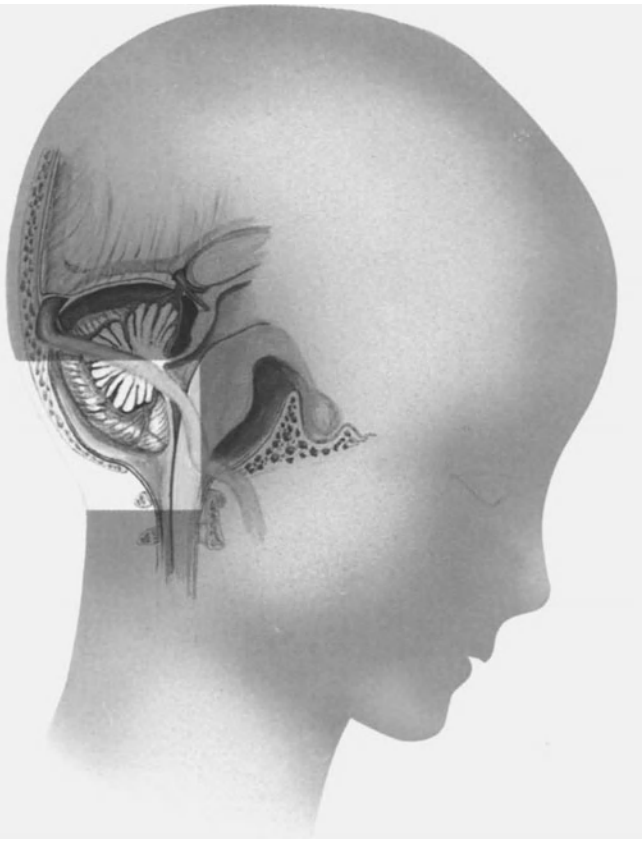


Figure 3.27. Inferior cerebellar triangle.

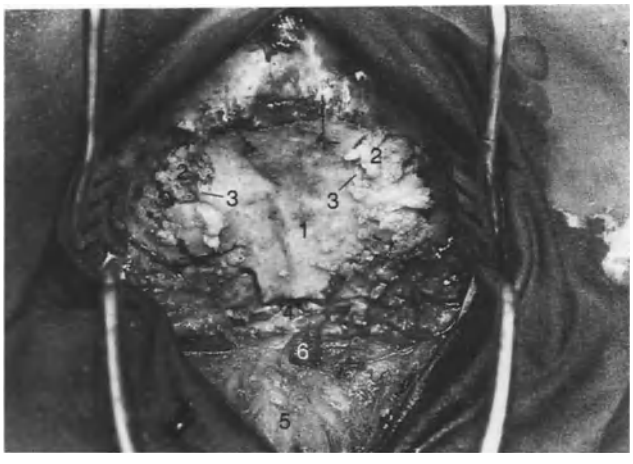


Figure 3.28. Suboccipital craniotomy at time of closure, illustrating replacement of the bone flap (*1*) and filling of the bur holes with bone plugs (*2*) and bone chips (*3*). One notes an intact arch of C-1 (*4*) and erector cervicis muscles (*5*) still attached to C-2 (*6*).

Failure to close the dura mater results in herniation of the posterior fossa contents into the dead space beneath the erector capitis and trapezius muscles, with resultant adhesions of the cerebellar surface to muscular tissue and prolonged postoperative morbidity. The performance of craniectomy precludes replacement of the bone flap, resulting in the formation of a dense scar tissue between muscle and dura, and in a high incidence of suboccipital bulging. This leaves the child with a weakened area over one of the most vital portions of the brain. Craniectomy should be performed only when the craniotomy, for technical reasons, proves impossible to perform.

Lateral Suboccipital Craniotomy

The lateral suboccipital craniotomy is used for access to the most lateral portion of the cerebellar hemisphere, the pontocerebellar angle, the clivus, jugular foramen, posterior inferior cerebellar artery, and the region of the IX, X, and XI cranial nerves.

As has already been described, dissection of the soft tissue for exposure of the squamous portion of the occipital bone entails stripping of the periosteum

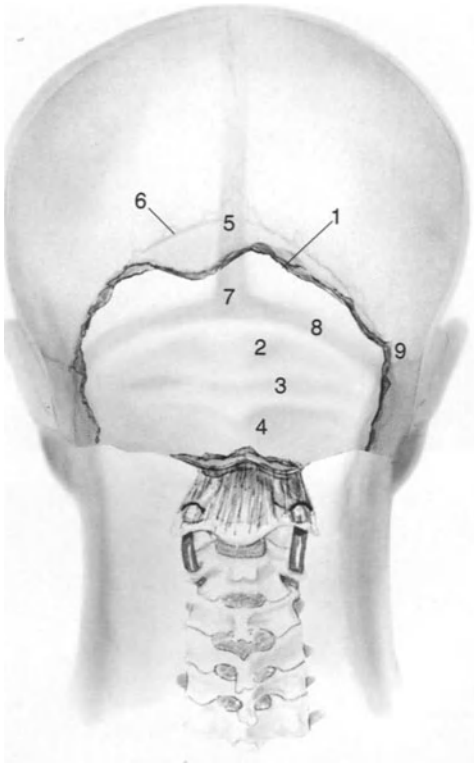


Figure 3.29. Periosteal dissection for suboccipital craniotomy. The periosteal dissection (*1*) has been brought above the highest nuchal line (*2*). This drawing shows both the intermediate (*3*) and lowest (*4*) nuchal lines. Lambda (*5*) and the squamosal suture (*6*) are superior to the torcular Herophili (*7*) and the transverse sinus (*8*) medially, but the squamosal suture is at the same level as the sigmoid sinus (*9*) laterally.

(Fig. 3.29). However, the stripping is not complete because it is limited to the highest and lowest nuchal lines, the insertion of the erector capitis and trapezius muscles. Above and below the highest and lowest nuchal lines the periosteum may be preserved. In planning either a superior or an inferior midline occipital flap, one must remember that the lambdoidal suture runs well superior to the transverse sinus medially, but that it becomes superimposed on the venous sinuses at the point where the transverse sinus passes into the sigmoid sinus. This is also the area where the parieto-occipital (lambdoidal), occipitomastoid, and parietomastoid sutures meet.

Midline Suboccipital Craniotomies

Inferior Suboccipital Craniotomy (Figs. 3.30, 3.31)

Suboccipital craniotomy for access to the inferior cerebellar triangle consists of reflecting a triangular bone flap whose base is located beneath the transverse sinus along the highest nuchal line, and whose (flat) apex is the rim of the foramen magnum.

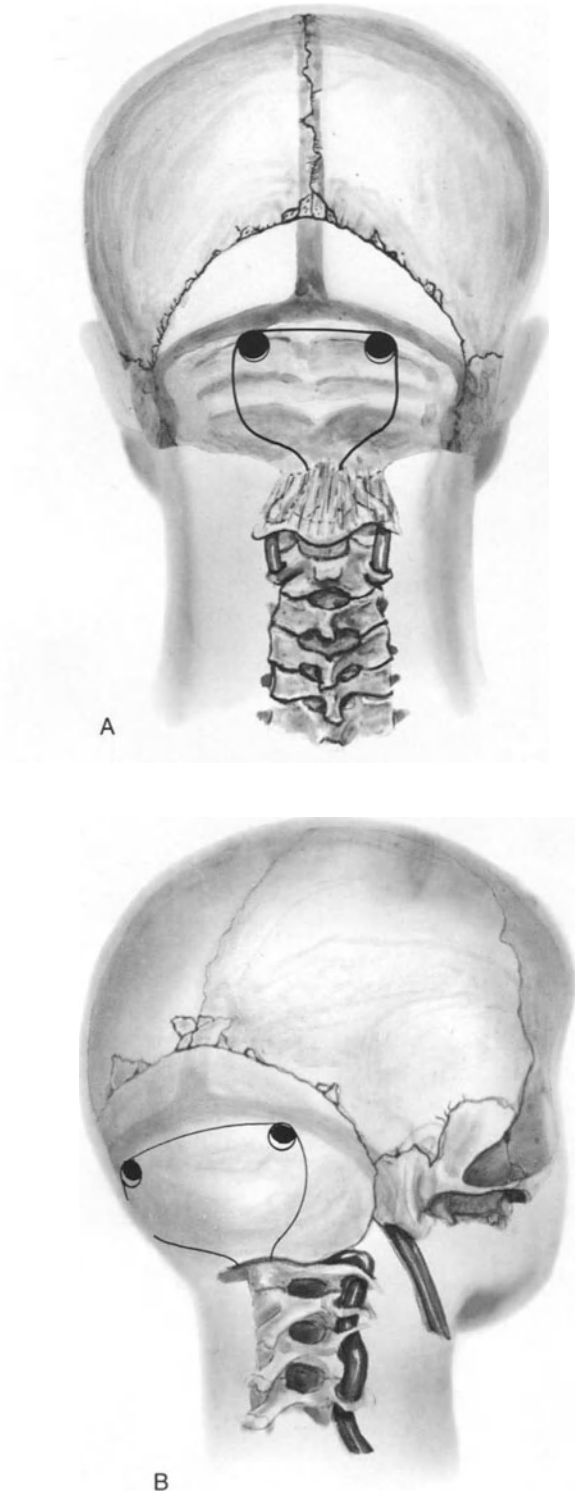


Figure 3.30. (A) Inferior suboccipital craniotomy as viewed in a straight posteroanterior line, illustrating the bur holes and osteotomy lines outlining the free (squamous occipital) bone flap. (B) Inferior suboccipital craniotomy as viewed in a postero-oblique line.

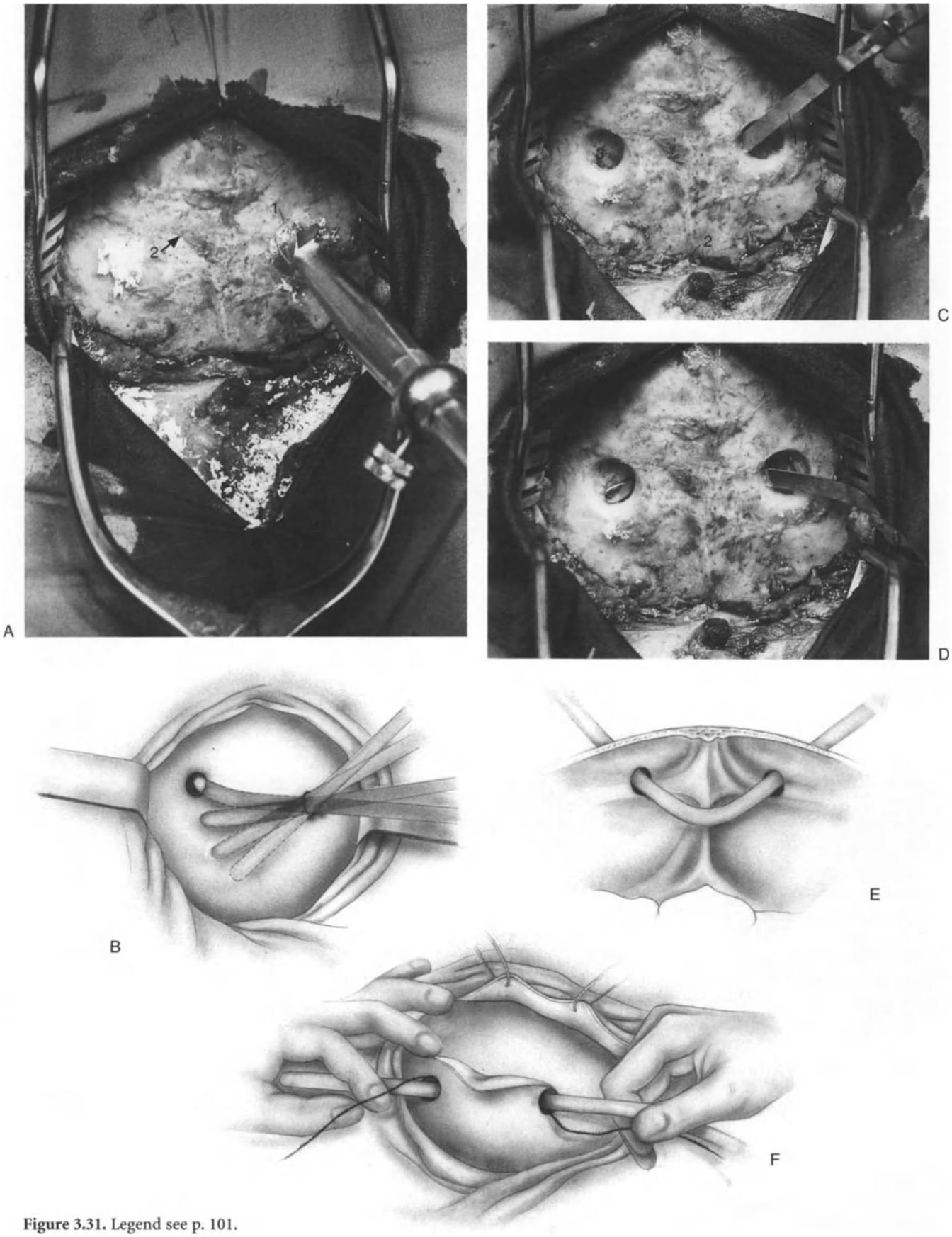


Figure 3.31. Legend see p. 101.

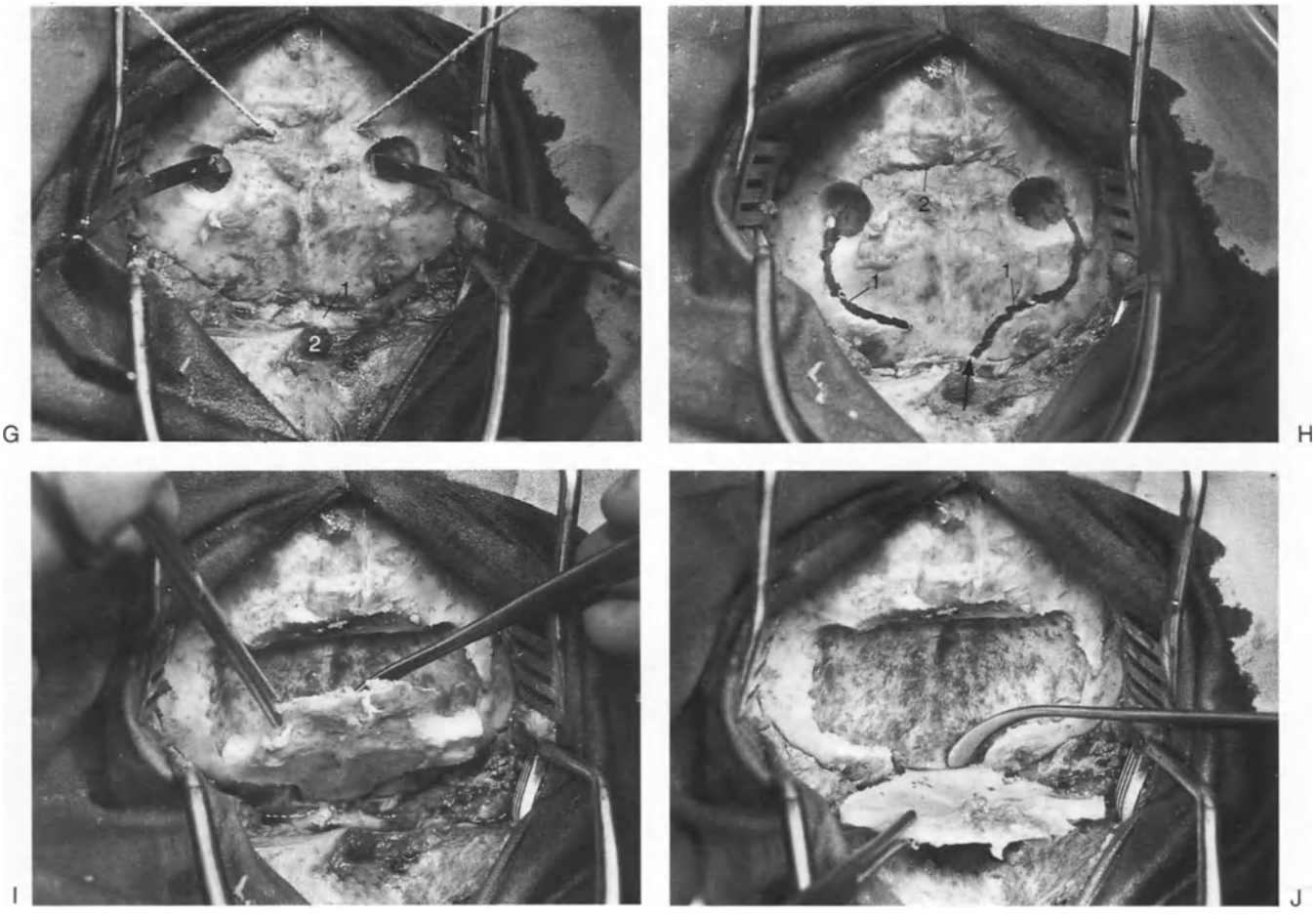


Figure 3.31. (A) Placement of bur hole (*1*) just at the highest nuchal line (*2*). (B) This drawing illustrates insertion of the Gigli saw guide into the epidural space and then rotating it around the bur-hole axis to separate the dura from the inner surface of the skull, especially along the midline where there is very often a keel-like bony protuberance extending from the line of the transverse sinus inferiorly to the opistion of the foramen magnum (C) In passing the Gilgi saw guide (*1*) from one bur hole to another, across the midline, it is advantageous to direct it inferiorly, towards the rim of the foramen magnum (*2*), first, and then to sweep it superiorly toward the opposite bur hole (*3*). This facilitates separating dura from the internal occipital crest and free dura from the inner surface of the flap area. (D) Completion of passage of Gigli saw guide across the midline. (E) The Gigli saw guide has been brought through the subdural space from one superior bur hole to the other. This drawing permits one to identify the "keel-like" bony protuberance in the midline extending from the region of the torcula Herophili internally down to the opistion. (F) The Gigli saw, hooked into the guide, has been passed through the epidural space from one bur hole to another and is now being freed from the guide. *On the reader's left* the saw has already been brought to the superior arch of the bur hole and *to the right* is still resting inferiorly. Once it is brought to the superior arch of the bur hole it is in position for the sawing. (G) The Gigli saw guide is left in place, and the Gigli saw is used to make the superior (horizontal) osteotomy, beveling superiorly *only after* the saw is fully within bone so as to avoid damaging the torcular Herophili or transverse sinus. Note intact C-1 (*1*) and undisturbed attachment of erector cervicis muscles to laminae and spinous process of C-2 (*2*). (H) After the superior osteotomy has been completed, the craniotome is used to make the lateral osteotomies, bringing them *to* (*arrow*), *not across*, the rim of the foramen magnum. The lateral osteotomy *on the right* has been completed; the one *on the left* has yet to be brought to the rim of the foramen magnum. Note the wide gutter made by the craniotome (*1*), and compare it to the beveled cut which the Gigli saw achieves (*2*). (I) Holding the squamous occipital bone flap with a Kocker and gradually lowering it facilitates dissection of dura and prevents the flap from falling. As the flap is depressed, it should be pulled from the annular (marginal) sinus so as to avoid jamming the bony posterior rim of the foramen magnum into the sinus. (J) The final dissection stage consists of freeing the dura/periosteum from the rim of the foramen magnum, a point of danger where caution must be taken not to open the annular sinus. A Penfield dissector, or #15 blade, may be used, always working from the bony surface of the rim. Note the sunken dura, a result of having inserted a ventriculoperitoneal shunt (in this child with a medulloblastoma and severe triventricular hydrocephalus) 10 days before the midline suboccipital craniotomy.

Care should be taken to dissect the periosteum from the outer (*posterior*) rim of the foramen magnum, but *not to extend the dissection around the rim of the foramen magnum, since use of periosteal elevators in this area may cause damage to the annular (marginal) sinus, causing profuse venous bleeding or air emboli.*

The Gigli saw guide is passed horizontally from one bur hole to the other, and the Gigli saw used to connect the bur holes with the linear osteotomy. Passage of the saw guide horizontally presents no difficulties, though care must be taken at the midline, where there is often a spine of occipital bone extending into the dural groove at the fissure between the cerebellar hemispheres. Molding of the tip of the Gigli saw guide, if obstruction is encountered at the midline, and dissecting first from the right and then from the left, always feeling the tip of the guide against bone and dura, eases the guide across the midline into the contralateral bur hole. If it proves to be difficult to pass the guide directly horizontally from one bur hole to the other, the tip should be directed inferiorly, toward the rim of the foramen magnum (where the inner occipital spine is least prominent) and then to sweep it superiorly once the tip has crossed the midline.

The saw guide is then used to dissect the dura from the inner surface of the occipital bone toward the foramen magnum, by advancing it in that direction, remembering that the opening of the posterior rim of the foramen magnum is extraordinary small (measuring 2.0 cm) and that its lateral surfaces consist of thick bony struts which lead to the occipital condyles. Therefore, an attempt to pass the Gigli saw guide directly inferiorly from the bur holes will result in the guide encountering the bone struts along the superior surface of the occipital condyles, and then being deflected medialward.

It is not advisable to attempt to pass the saw guide from a bur hole downward across the rim of the foramen magnum, in the epidural space, since this, too, puts the annular (marginal) sinus at risk. Rather, the guide should be advanced inferiorly as far as the rim of the foramen magnum, removing it from time to time to measure the length of guide inserted (to be certain that one is at the foramen magnum). No attempt should be made to strip the outer layer of the dura matter from the rim of the foramen magnum: it adheres tenaciously to the rim, the point at which it is continuous with the periosteum. In fact, the outer layer of the dura mater (which is, indeed, the inner periosteum of the skull) is continuous with the periosteum (pericranium) at all foramina of the skull. This dural duplication within the foramen magnum forms the annular sinus.

A craniotome is used to connect the lateral surface of each bur hole to the rim of the foramen magnum *but it must not be brought all the way through the bone lest it tear the annular sinus.* The triangular, free bone flap is separated from the dura by use of a Penfield #3 dissector or an Oldberg periosteal elevator, gradually lowering the base, as the dissection proceeds, until the dural-periosteal transition point at the rim of the foramen magnum is visualized. Now the dura-periosteum may be dissected from the rim of foramen magnum within the free flap, under direct visual control, either with a sharp periosteal elevator or by cutting it from the bone with a #15 blade. This exposes the point at which the dura mater, atlanto-occipital membrane and periosteum/pericranium join. This now redundant mass of connective tissue, measuring approximately 3 × 5 mm, may be dissected from the inner layer of the dura mater as it continues inferiorly over the craniocervical junction. If one wishes a bit more lateral exposure at the rim of the foramen magnum, a rongeur may be used to nibble away 2 or 3 mm of bone, extending the bites toward the occipital condyles.

Superior Suboccipital Craniotomy (Fig. 3.32)

The superior suboccipital craniotomy is one performed by placing four bur holes in a quadrilateral fashion, the upper two along the highest and the lower two along the lowest nuchal lines. The same precautions as for the

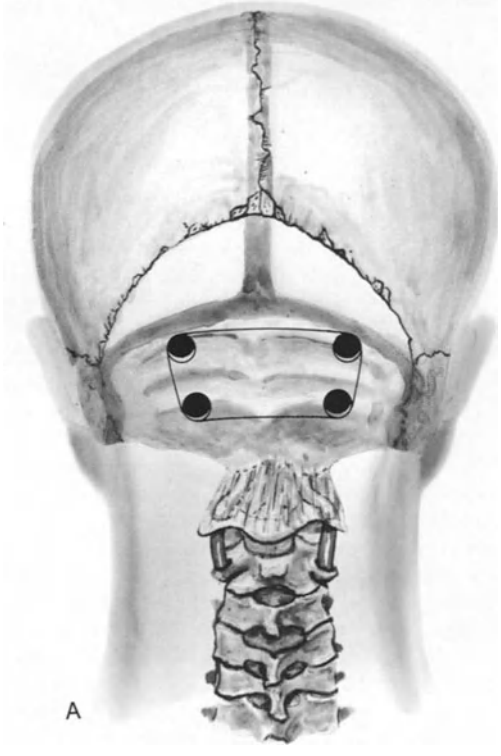
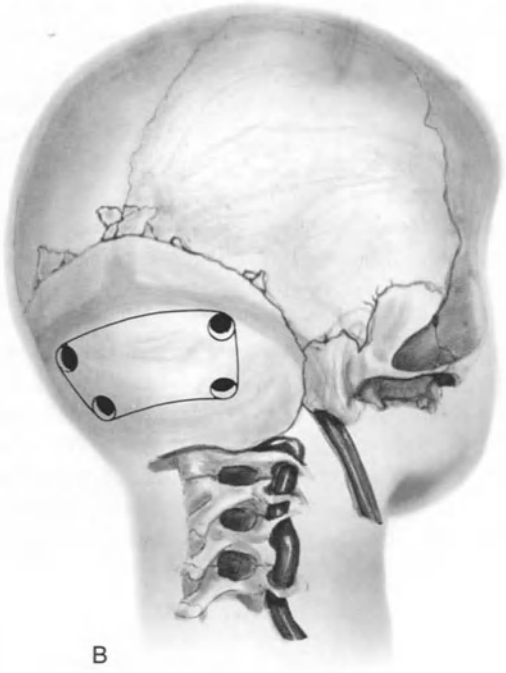
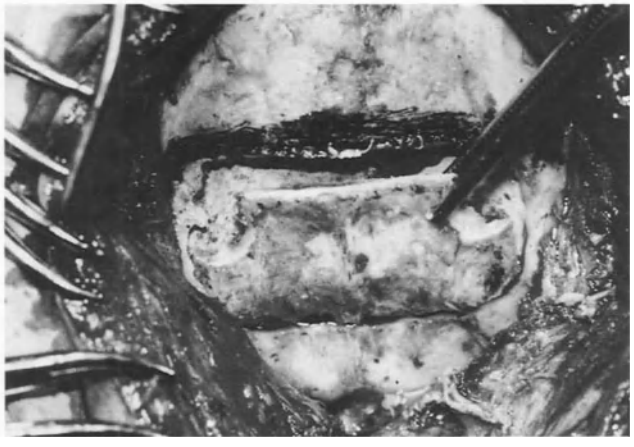


Figure 3.32. (A) Superior suboccipital craniotomy as viewed from the straight posteroanterior line. The upper bur holes are inserted along the highest nuchal line, the bottom two along the lowest nuchal line. The middle nuchal line, when present, runs across the center of the free flap. (B–E) see p. 103.





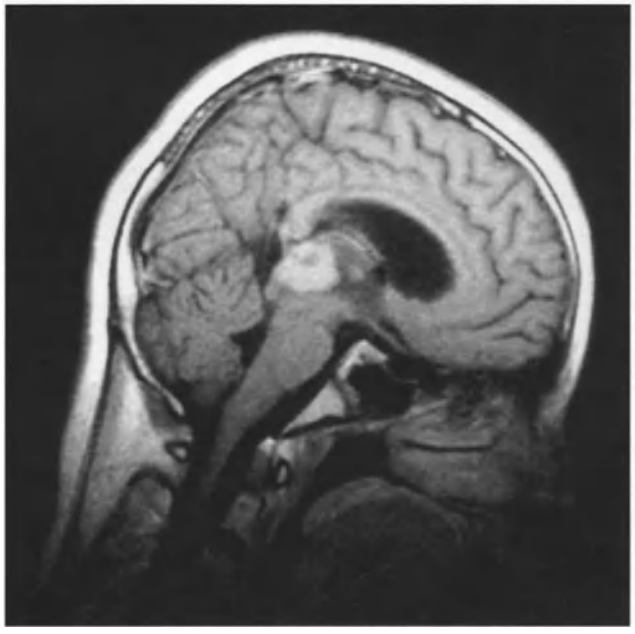


Figure 3.32. (B) Superior suboccipital craniotomy as viewed from a postero-oblique line. (C) The osteotomies have been completed, the free flap is being lifted away. (D) Photograph of the suboccipital area, showing dura after superior suboccipital craniotomy, and the craniovertebral junction. Note that the erector cervicis muscles remain attached to the vertebrae. (E) *Pineal region tumors* extending into the third ventricle are very well resected through a superior suboccipital craniotomy as illustrated diagramatically in (A) and (B), and photographically from surgery in (C) and (D). Studying this MRI scan permits one to see how such a flap allows the surgeon to work through the space between the top of the cerebellum and tentorium, looking directly into the cavity of the III ventricle after the tumor has been removed.

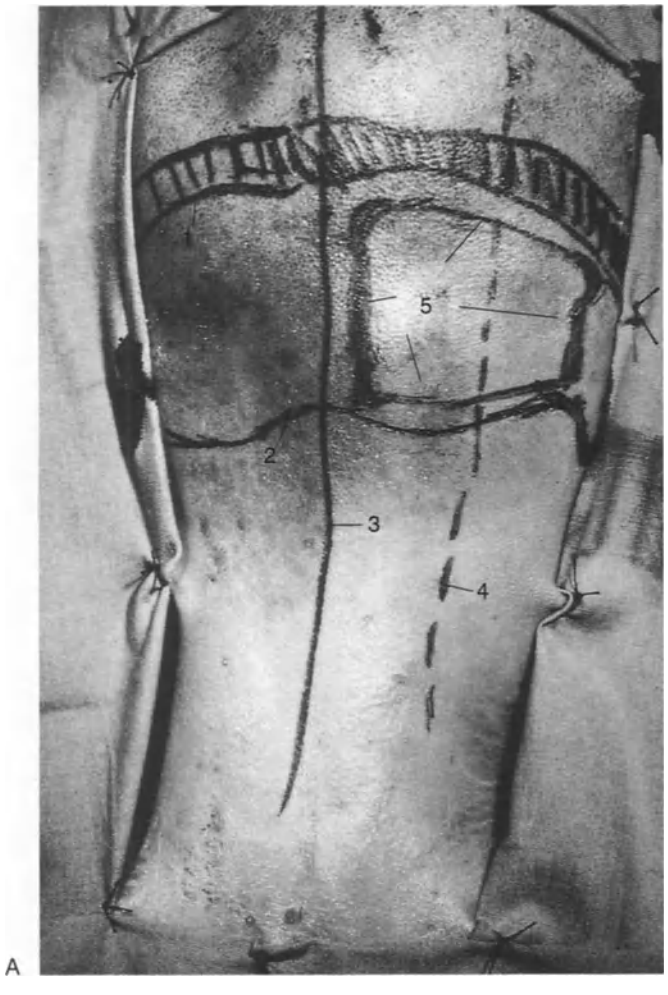
inferior suboccipital craniotomy are taken in passage of the Gigli saw guide across the midline. The Gigli saw is recommended for connecting the bur holes to one another, since one will not be crossing the foramen magnum. Once the Gigli saw guide has been passed horizontally across the midline, it may be swept in an arc from the opposite superior to the opposite inferior bur hole in one direction first, then in the opposite direction. This frees all of the dura, and permits safe osteotomies.

Lateral Suboccipital Craniotomy (Fig 3.33)

The lateral suboccipital opening is placed entirely within the squamous portion of the occipital bone, at its most extreme lateral portion (immediately inferior to the transverse sinus and posteromedial to the jugular bulb).

Consequently, the occipitomastoid suture and the diagastric groove are at the anterior edge of the bone flap, and the lambdoidal suture is considerably superior to it. The access it offers to the lateral surface of the cerebellar hemisphere is excellent. It is the only flap that permits one to work effectively in the pontocerebellar angle, the jugular foramen, or along the lateral surface of the medulla oblongata and pons. A craniectomy is neither necessary nor advisable.

Because these flaps are very small, measuring approximately 4 × 4 cm in surface area, one should use a single bur hole. This is placed in the squamous occipital bone, immediately beneath the point at which the transverse sinus passes into the sigmoid sinus. The linear osteotomy is then performed with a craniotome, since it is not possible to use the Gigli saw properly for such a small flap. Also, the beveling of the squamous occipital bone as it passes from its vertical to its horizontal portions at the base of the skull, thickening remarkably both medially at the inner surface of the occipital condyles and laterally at the mastoid base, render attempted passage of the Gigli saw guide dangerous.



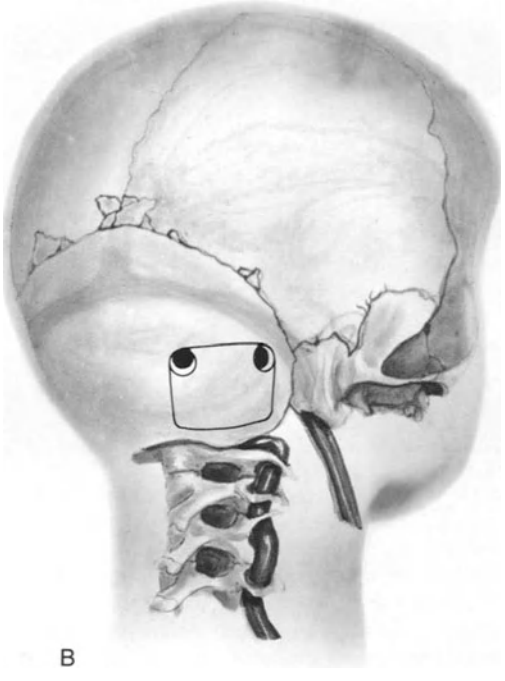


Figure 3.33. (A) Lateral suboccipital craniotomy for removal of a solid cerebellar hemisphere astrocytoma. Transverse sinus (*1*), foramen magnum (*2*), midline (*3*), and incision line (*4*) have been marked off. Subsequent to this, the area for the suboccipital craniotomy (*5*) was drawn on the skin. (B) Lateral suboccipital craniotomy as viewed along a posterior oblique line, showing the flap in relationship to the transverse sinus superiorly, the sigmoid sinus anteriorly, the point at which the squamous occipital bone passes from a vertical to a horizontal structure inferiorly. Note that the flap does not extend to the midline. At times, one may find it advantageous to place two bur holes, as illustrated here.

Supra- and Infratentorial Craniotomy (Fig. 3.34)

A lateral suboccipital and parietotemporal craniotomy are preferable for a combined approach, irrespective of whether the supra- and infratentorial lesion begins at the pontocerebellar angle or the tentorial opening, or grows through the dura (dumbbell fasion). A wider or narrower strip of bone may be left over the transverse sinus, or the bone flap may be lifted in a single piece from over the transverse sinus, occipital lobe, and cerebellar hemisphere.

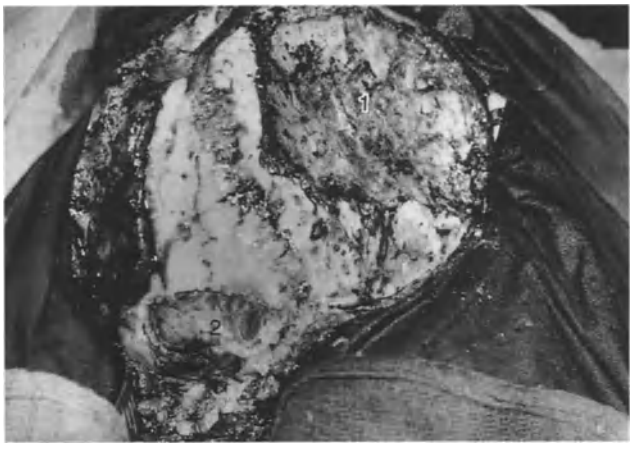


Figure 3.34. Supra- and infratentorial craniotomies. The parietotemporo-occipital (*1*) and lateral suboccipital (*2*) bone flaps have been reflected.

Hemispherical Craniotomy (Fig. 3.35)

The hemispherical craniotomy is used primarily for hemispherectomy and, when bilateral, for lowering of the SSS. In the latter instance, the craniotomy does not come to the midline. The bone over the SSS is not removed. Rather, it is advanced in the sagittal plane so as to accomplish the lowering of the SSS onto the cerebellar hemispheres and to advance it, restoring normal anatomical relationships between bridging veins and the SSS. Maintaining the strip of bone over the SSS prevents kinking-obstruction of this channel. When the hemispherical craniotomy is being used for lowering the SSS, the medial bur holes are brought to the parasagittal plane with their medial surfaces bordering upon its lateral extremity. This allows for a median strip of bone and the anterior fontanelle to remain in the sagittal plane. The strip of bone is subsequently osteotomized at the inion and at the glabella, so that it may be lowered onto the underlying parasagittal surfaces of the cerebral hemispheres when the vault of the skull is reconstructed and molded over the atrophic hemispheres.

The placement of the bur holes for the hemispherical craniotomy is such as to permit access to the frontal, temporal, and occipital poles; to the internal carotid and posterior cerebral arteries; to the deep middle cerebral and cortical bridging veins; to the vein of Labbé and the SSS. Consequently, one hole is inserted at the glabella, two along the contralateral side of the sagittal suture, one just inferior and lateral to the point at which the sagittal and lambdoidal sutures meet, one along the parietosquamosal suture just superior to the external auditory canal, and one at the "keyhole."

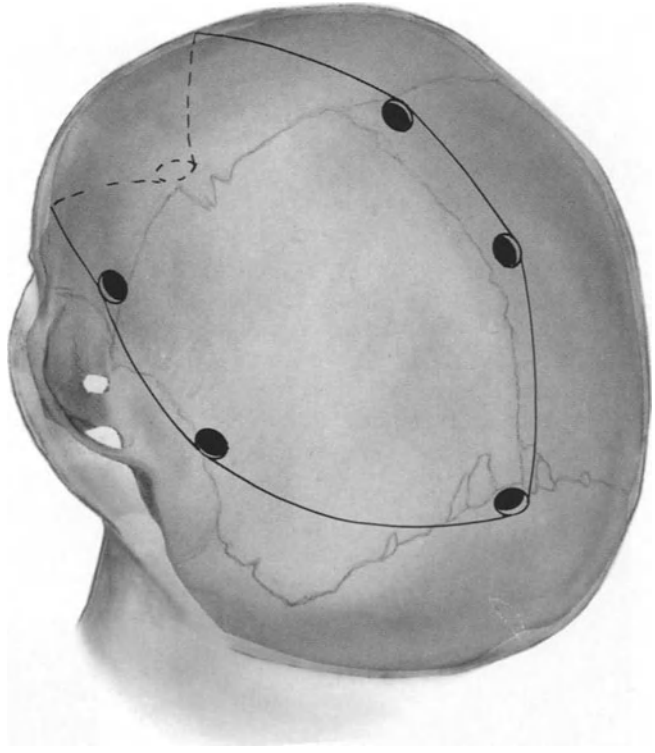


Figure 3.35. Placement of bur holes and osteotomy lines for hemispherical craniotomy are shown in this anatomical illustration. The placement of the bur holes permits access to all hemispherical vascular structures and to the entirety of the hemisphere. The superior holes are placed to the contralateral side of the sagittal suture so as to expose completely the SSS.

Laminotomy (Figs. 3.36–3.43)

Laminectomy is a destructive procedure that is indicated when there are intraspinal metastases compressing the cord or cauda equina. *When the intraspinal pathology is traumatic, benign neoplastic, or congenital malformation (e.g., diastematomyelia, diplomyelia, dermoid sinus tract), laminectomy may further weaken the spinal column.* Decompressive laminectomy for the drainage of epidural tuberculous abscess makes subsequent fusion, the treatment of choice, difficult or impossible. Whenever a limited or extensive laminectomy (two or more levels) is performed on a child, kyphosis and scoliosis may develop and become difficult clinical problems, necessitating spinal fusion. Kyphosis, anterior subluxation, and instability of the spine are postoperative complications of multiple-level laminectomies in children. The surgical procedure of multiple level-laminotomies is the preferable alternative.

Scoliosis and kyphosis were described following multiple-level laminectomies in children in 1965 by Tachdjian and Matson [5], and then confirmed in 1967 by Cattell and Clark [6]. In 1955 Bette and Englehardt [7] were the first to point out that anterior intervertebral body subluxation and kyphosis occurred following laminectomy. Since these changes have not been observed in adults, one must conclude that there is a fundamental anatomical (physioanatomical) difference between the fully developed and completely grown vertebral spine on the one hand, and the developing, nonossified spine of younger children (infants, toddlers, juveniles) on the other. In addition to this, one must take into consideration the completed development of the paraspinal muscle masses in the adult, and both the undeveloped and nonfunctional erector spinae masses in the infant.

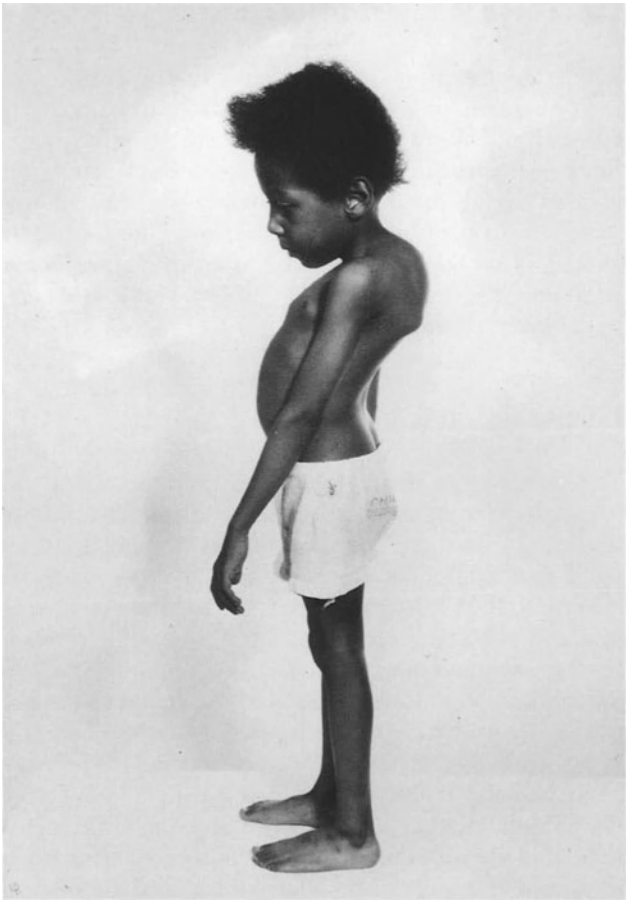
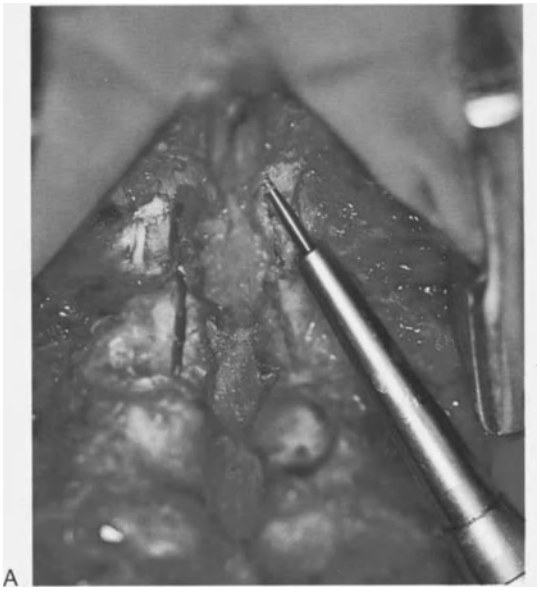


Figure 3.36. This child's kyphoscoliosis developed progressively over an 18-month period following multiple level thoracic laminectomy. None was present preoperatively.

Figure 3.37. (A) Laminar osteotomy being performed on the lumbar spine of a 9-year-old-boy. (B) Two laminae, with their spinous processes and ligamentous structures, after removal. ▼



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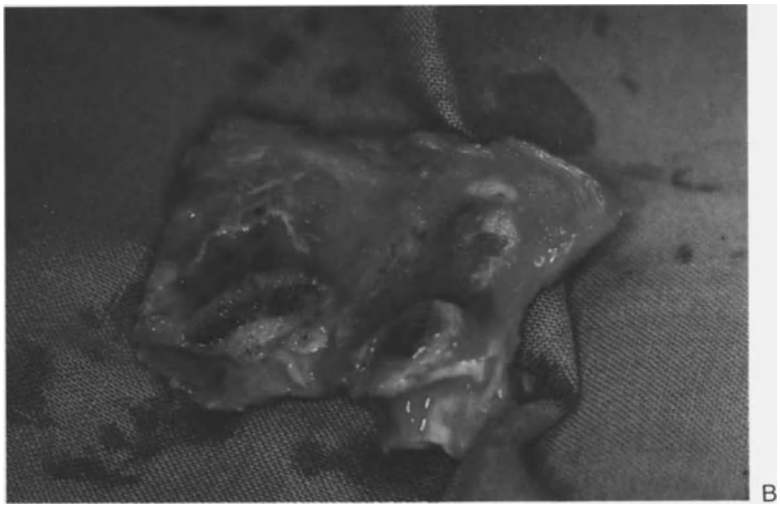

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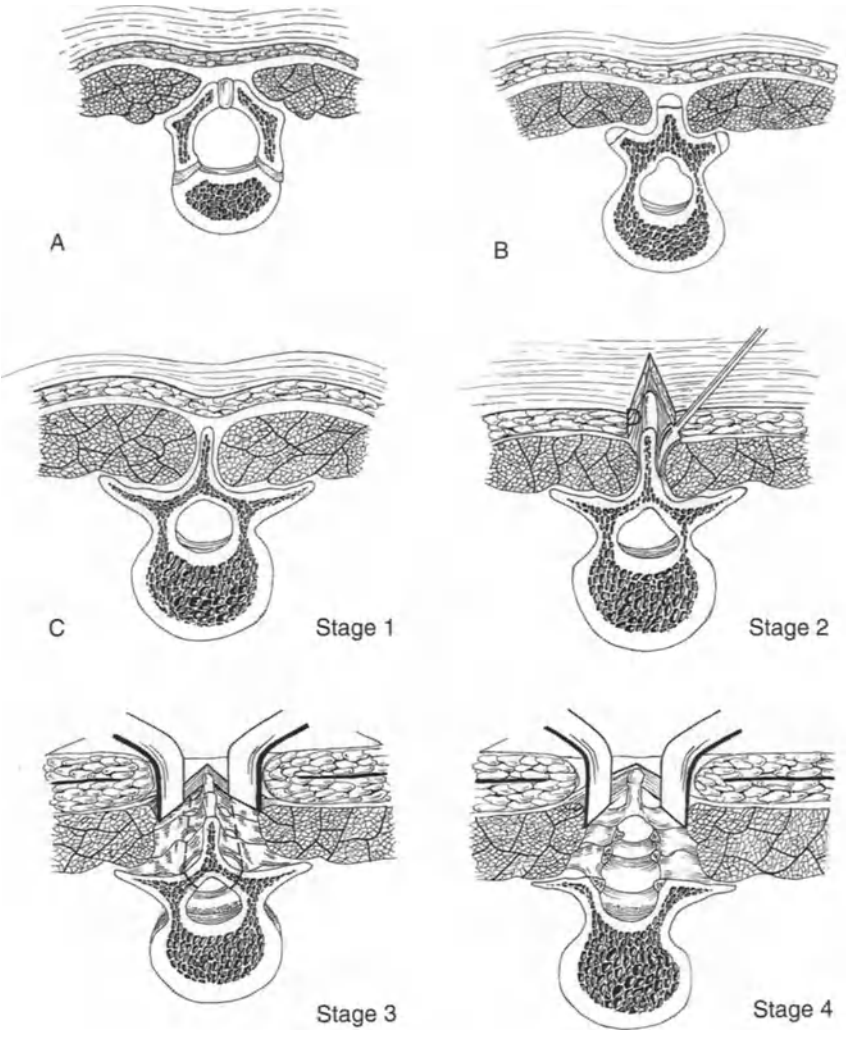
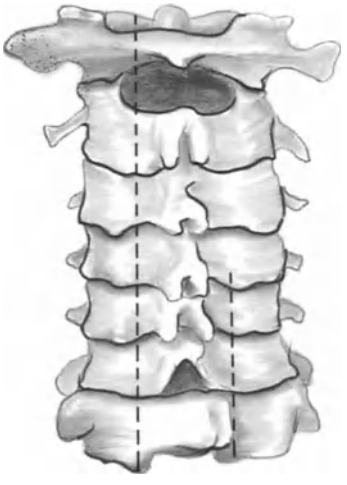


Figure 3.38. (A) Infant: note that there is almost no spinous process. The laminotomy is made at the same, paraspinous, position. (B) Toddler: the spinous process of the toddler is forming. (C) Juvenile: the four stages of exposure and osteotomy are illustrated in a juvenile to represent reflection of the skin flap and paraspinous muscles as well as the location and direction of the laminar osteotomy.

Figure 3.39. The general anatomical characteristics of the posterior spinous portion of the cervical canal are represented here as is the caudocephalad osteotomy.



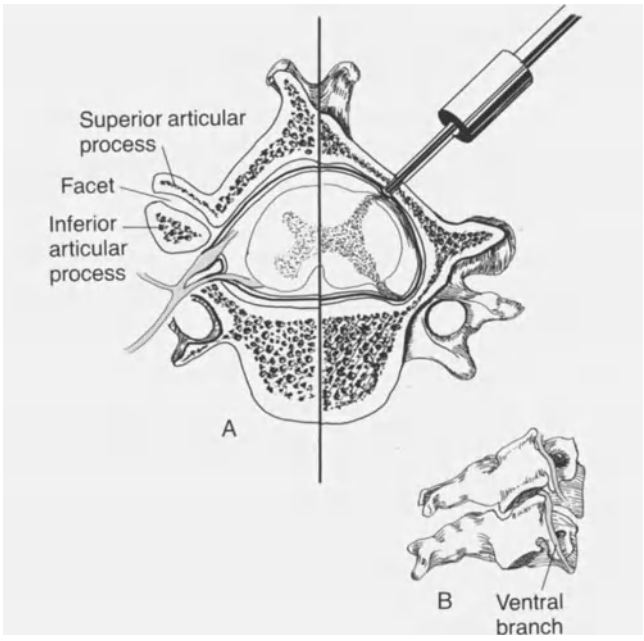
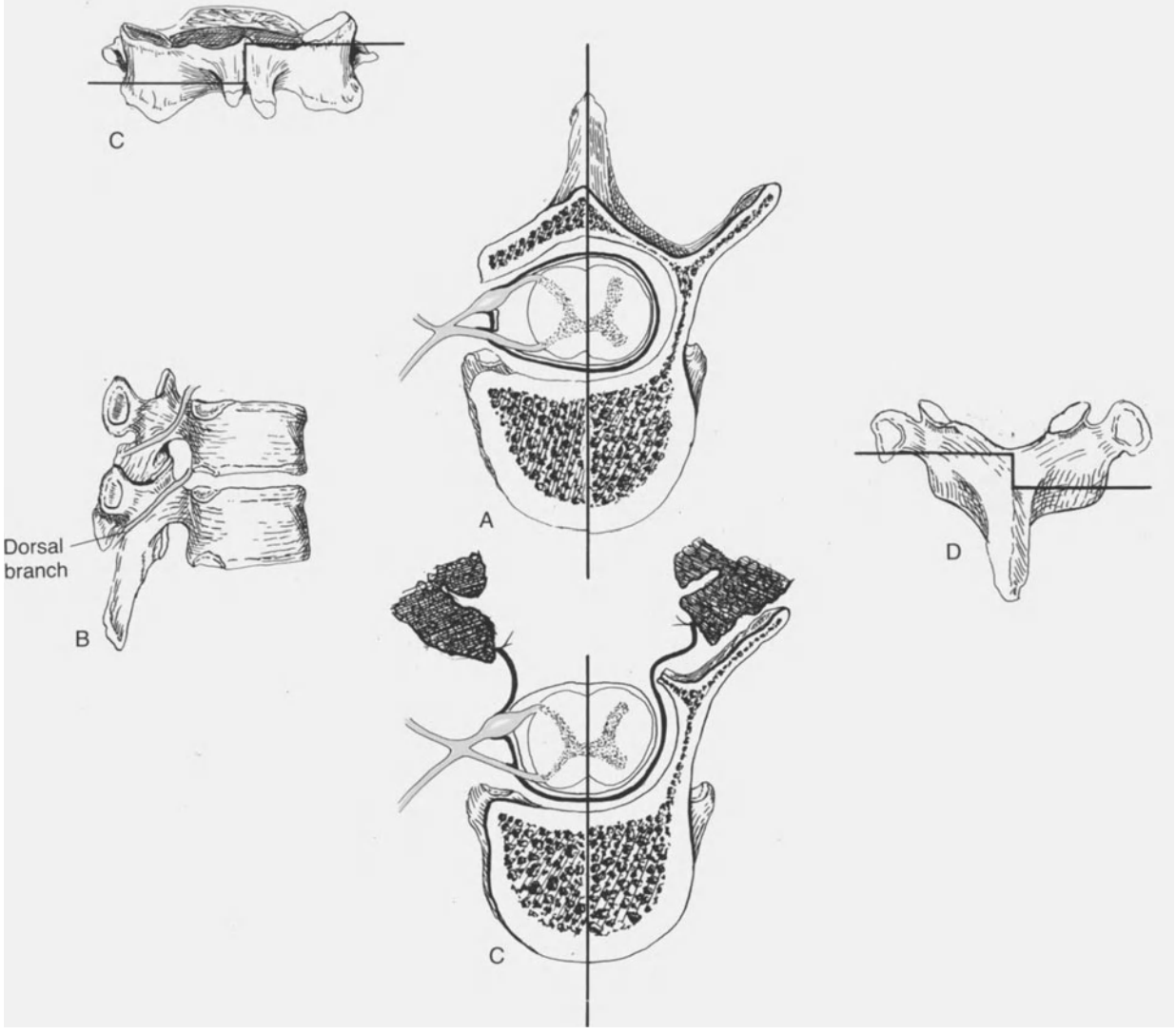


Figure 3.40. (A,B) The fourth cervical vertebra is (diagrammatically) shown cut at two levels, in horizontal section. (C) The line of section, not of recommended osteotomy. This line of section is used to illustrate the inferior level of the vertebra *on the left*, the superior level *on the right*.
◄

Figure 3.41. (A) The third thoracic vertebra is shown, diagrammatically cut at lower and higher levels to illustrate the different anatomical characteristics of the laminae and to illustrate shingling. (B) The course of the nerves in the lateral view is shown. (C) This illustrates the exposed spinal cord and arachnoid, the opened and reflected dura mater. (D) This depicts the lines of section, superior and inferior.



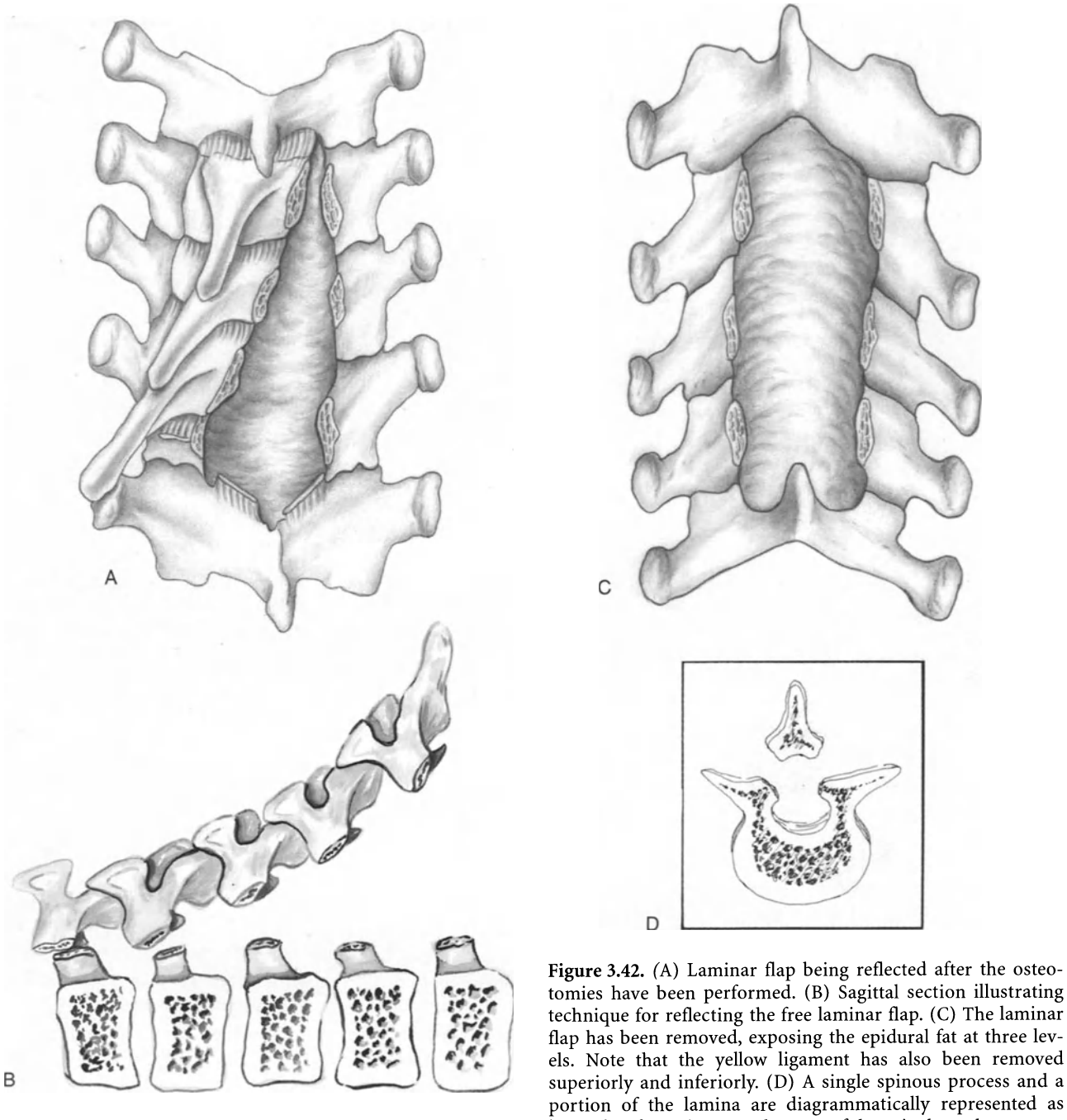


Figure 3.42. (A) Laminar flap being reflected after the osteotomies have been performed. (B) Sagittal section illustrating technique for reflecting the free laminar flap. (C) The laminar flap has been removed, exposing the epidural fat at three levels. Note that the yellow ligament has also been removed superiorly and inferiorly. (D) A single spinous process and a portion of the lamina are diagrammatically represented as being free from the ventral aspect of the spinal canal.

In their 1967 paper, Cattell and Clark [6] noted that Tachdjian and Matson [5] failed to comment on 24 patients of theirs with cervical cord lesions, concerning the development of cervical column instability, whereas they elaborated upon the onset of scoliosis and kyphosis at the thoracic and lumbar levels following multiple laminectomies in 115 children. This concerned Cattell and Clark, who were interested in the fact that the cervical spine, the most mobile segment of the vertebral column, is particularly subject to the destabilizing effects of laminectomy. They illustrated several cases, accentuating the fact that skeletal, ligamentous, neuromuscular, and progressive bony growth (with ossification of the centrum) are all, to a greater or lesser degree, responsible for vertebral column deformity following laminectomy. One of the most important points these authors make is that the vertebrae in children are dynamic, growing, and ossifying structures, which offer purchase to developing muscle masses. It is their conclusion that abnormal growth patterns and greater elasticity of mus-

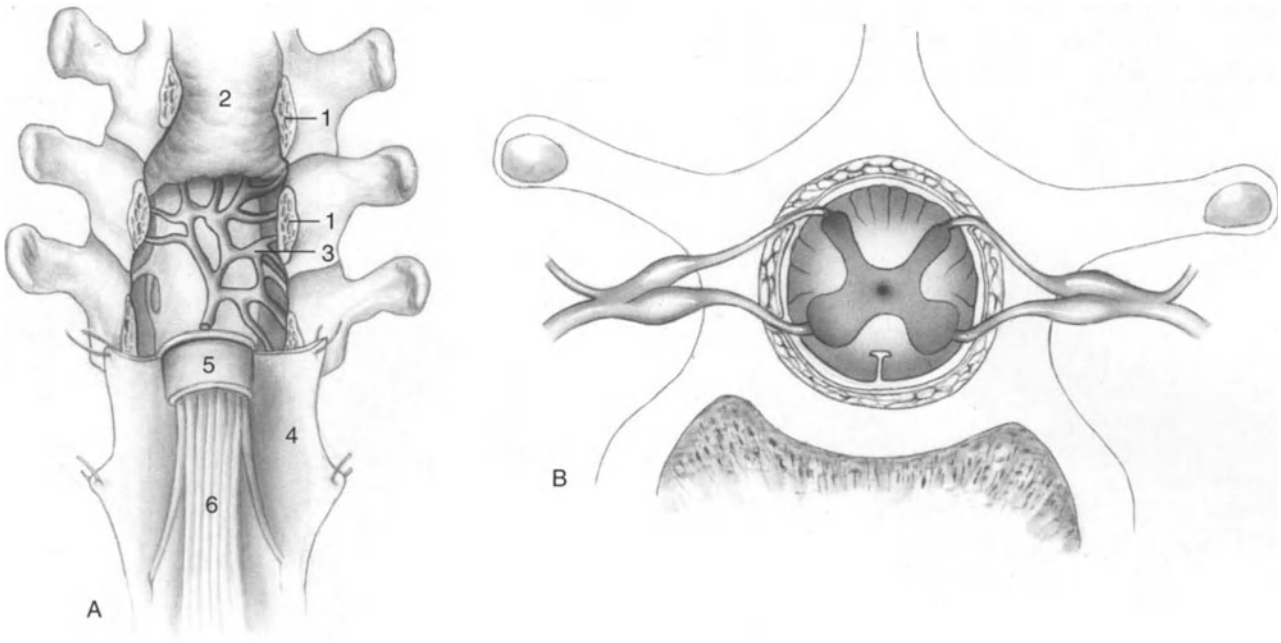


Figure 3.43. (A) An osteotomized thoracic spine is diagrammatically represented, illustrating the desired inclination of laminar cut (*1*); the epidural fat (*2*) and venous plexus (*3*), reflected dura mater (*4*), and, deeper, the enclosed arachnoid (*5*) and neural elements (*6*). (B) This axial drawing complements the vertical plane sectional drawing, with the former putting into relief the diffuse epidural venous plexus (in the spinal dura there is only one layer, that which corresponds to the inner layer of the dura over the surface of the brain, so that there are no dural sinuses). Also it makes clear the circumferential distribution of epidural fat, which is very abundant in children. One readily identifies the two concentric rings beneath the epidural fat, the outer one being the single layer of dura mater and the inner one being the membrane.

culoligamentous structures in children are responsible for the rapid and severe deformities of the vertebral column, especially the cervical column, which result following laminectomy.

Following the work by Raimondi and coworkers in 1976 [8], concluding that deformities (kyphosis, scoliosis, accentuated lordosis) of the vertebral column in childhood result from laminectomy, and recommending that laminotomy be substituted for laminectomy, Yasuoka and colleagues [9] in 1981, reported that "postlaminectomy spinal deformity can develop in children without irradiation or facet injury." It was their conclusion that the deformity results from a wedging transformation of the cartilaginous component of the vertebral bodies, and that the increased viscoelasticity of children's musculoligamentous structures is a significant contributing factor. Their attention was directed primarily to the treatment of postlaminectomy deformities of the vertebral column in childhood, not to abandoning the laminectomy and adapting laminotomy as the procedure of choice for access to the spinal canal in childhood.

An extremely interesting, and very important, contribution to the literature of this subject was made by Barbera and colleagues [10] in their 1978 paper concerning the "laminectomy membrane," previously described in 1974 by La Rocca and Macnab [11]. Specifically, the "laminectomy membrane" was found to be pathogenetic in producing or reproducing signs and symptoms of spinal cord compression following a laminectomy procedure. *The "laminectomy membrane" is scar tissue.* Barbera and his associates [10] recommended using either an acrylic plastic or kiel bone graft over the dura mater to prevent "expansion of the scar tissue inside the spinal canal and adhesions between the dura and the cicatricial overlying muscles." They concluded that this type of solid material, or tissue, is necessary to prevent the formation of the "laminectomy membrane."

Laminotomy, as herein described, consequently (1) restores bony protection to the spinal cord, (2) prevents or significantly diminishes postoperative spinal column deformity (kyphosis, accentuated lordosis, scoliosis), and (3) eliminates the formation of a "laminectomy membrane."

The criteria for performing a laminotomy include the extent of the surgical procedure, the age of the patient, and the nature of the lesion. In children under 1 year of age the surgeon should perform a laminotomy even if only one level is to be exposed; in children between 1 and 15 years for two or more levels; and in patients older than 15 years, when three or more levels are to be exposed. Independent of the age of the patient or

the extent of the intraspinal lesion, laminotomy should be performed in all patients with trauma, syringomyelia, hydromyelia, or tuberculosis. No attempt should be made to perform laminotomy in children with extensive epidural metastases.

Several structures provide for the stability of the spinal column: intervertebral joints, laminae, ligamentum flavum, spinous processes, interspinous and supraspinous ligaments, and paraspinal muscles. In the adult, stability depends mostly on the intervertebral joints, while the role of the other structures of the posterior arch is relatively less important. The vertebrae of the child are developing structures, for which balanced mechanical stimulations are necessary to assure normal growth. Spinal deformity and/or instability result from conditions in which bone, ligamentous deficiencies, or neuromuscular imbalance occur. Such a condition results from multiple laminectomies, which destroy growing bony structures (laminae and spinous processes), separate interlaminar and interspinous ligaments from adjoining vertebral arches, and substitute scar tissue for insertion of paraspinal muscle masses onto the laminae and spinous processes.

The reflection of a free laminar flap [12, 13] over the intraspinal pathology allows as complete access to the spinal canal as the most extensive laminectomy, since the lateral border of the laminotomy is at the medial surface of the pedicle. Multiple-level laminotomy flaps provide access to the entire spinal canal (C-1 through T-3, T-5 through L-3, L-2 through L-5), thereby allowing surgical removal of the most extensive lesions, without weakening permanently the vertebral column or destroying the growth center in the posterior portion of the spinal arch.

The removal of multiple laminae in a single laminar flap is a tedious procedure, and requires considerably more time than a laminectomy. It is not a more dangerous procedure than laminectomy, since magnification and high-speed drills permit one to separate the laminae and yellow ligaments with precision.

Performance of a laminotomy instead of a laminectomy permits complete reconstruction of the posterior arch of the spinal canal *and significantly diminishes* the complication of postlaminectomy scoliosis. It provides for complete anatomical reconstruction of the dura, the posterior arch of the spinal canal, and the muscular-bony relationships between the erector spinae muscles on one band, and the spinous processes and interspinous ligaments on the other. There is not as yet definite information concerning the incidence of postlaminectomy membrane formation as a pathogenetic entity.

Laminotomy Procedure

After the midline skin incision has been made, and extended the desired length, the skin is reflected laterally. The dissection is then carried along the midline, using the electrocautery knife (never the laser in young children!), taking care to remain within the ligamentous structures between one spinous process and another, until coming upon the tips of the spinous processes. Figure 3.38 illustrates the normal (juvenile) anatomical relationship between the skin, the ligamentous structures which bind the erector spinae muscles to the spinous processes, the laminae, and the transverse processes.

The exposure should extend from one full vertebra above through one full vertebra below the planned extent of the laminotomy. Thus, if a laminar flap is to be reflected from C-3 through T-4, one should expose the laminae from C-2 through T-5. Muscle and ligamentous attachments are separated from the vertebral arches, leaving the periosteum and interspinous ligaments intact. The dissection is carried laterally to just beyond the articular facets, with care being taken not to open into the joint or strip the capsular ligaments. The closure is facilitated if one leaves a ruffle of muscle and ligament on the spinous apophyses.

The younger the child, the smaller the spinous processes and the thinner the laminae. Similarly, the younger the child the thinner the erector spinae muscle mass. In fact, in the newborn and infant the spinous processes are almost nonexistent so that the laminae form a rather "domelike" structure. The infant is intermediate in muscle and bone development between the newborn and the toddler. Since the relative sizes of the laminae and yellow ligaments are equal, there is no shingling effect of the superior laminae overlapping the inferior laminae. This shingling occurs at approximately 6–10 years of age, when the muscle masses begin to develop. Once the spinous processes and laminae have been cleaned of adherent muscle and fascia, one may proceed to perform the laminotomy.

The laser is presently being used more and more in neurological surgery, and since it is ideal for dissecting erector spinae muscles from the spinous processes and the laminae in the adult, it deserves comment at this time. *Use of the laser in spinal cord injury and in children with spinal cord tumors is to be recommended when the child is over 10 years of age, but to be avoided completely when the child is under 5 years of age!* The exception is dissection of paraspinous muscles in spina bifida aperta children upon whom a kyphectomy is being performed. Its use in the 5- to 10-year range is to be decided upon only after careful review of the computed tomography (CT) scans and X-rays of the spinal column reveal that the spinous processes are completely formed and that the laminae are thick and overlap one

another. This care must be taken, since the laser beam may penetrate the yellow ligament and dura as the surgeon is dissecting the muscles from the laminae, with resultant risk of damaging spinal nerves or the spinal cord. Since the newborn and infant have yellow ligaments which are almost as wide as the laminae, one readily understands the risk. In the older child thick overlapping laminae protect completely the dura and cord.

The respective inferiormost and superiormost yellow ligaments are then incised from medial to lateral, bilaterally, prior to proceeding with the laminar osteotomies. A Penfield #3 dissector, or some similar instrument, may then be inserted beneath the incised yellow ligaments in a cephalad direction beginning at the level of the lowest laminae to be incorporated in the flap, so as to dissect the epidural fat from the spinal surfaces of the yellow ligament and laminae. This dissection is carried out from below (caudad) upward (cephalad).

Using power instruments and the finest drill blade available, one incises the laminae in a caudocephalad direction under the operating microscope, or loops, using a minimum of 3 × (preferably 10 ×) magnification, with constant but minimal irrigation and suction. The author uses a high-speed drill, not a craniotome. The osteotomy should be made in a dorsoventral direction, proceeding along a lateral medial plane so as to provide the maximum possibility of beveling, not with the expectation of obtaining an osteotomy which is wedge shaped, but with the hope of minimizing the size of the gutter and, thus, facilitating nestling it back into normal position at the time of closure. If one uses a very thin cutting blade on the power instrument (< 1 mm), bridging of the interval by bony tissue during the healing phases is greatly facilitated. (Some neurosurgeons use the craniotome footplate as a guide. performing the laminotomy as one would a craniotomy.)

The surgeon will both feel and see the penetration through the spinal surface of the laminar cortical bone if the osteotomy is performed by using brushlike strokes in precisely the same plane. It is as well to remember that the individual laminae are thinner caudally where the yellow ligament is thickest and on their ventral (spinal canal) surface, whereas they are thicker cephalad where the yellow ligament is thinnest and on the dorsal surface. The laminae are osteotomized in a caudocephalad direction, but the yellow ligaments are not incised until all laminae have been osteotomized and the laminar flap is being reflected. After one side of the planned flap is osteotomized in the caudocephalad direction, one returns to the contralateral, most inferior, lamina to be removed and repeats the procedure.

The laminar osteotomy is made using the high-speed drill along an imaginary line separating the pedicle from the lamina. Insertion of a curved dissector (Penfield #3) beneath the laminae assists the surgeon in identifying the medial surface of the pedicle, and may be used to protect the epidural vessels when the laminotomy is begun. One should use the drill in brushlike strokes along the surface of the lamina in the direction of the planned line, rather than as a perforator extending through the full thickness of the lamina each time. This latter technique is dangerous, the former is safe. A fine-tipped sucker (inserted into the laminotomy groove) and magnification allow the surgeon to see the full extent of his field. When the laminar incision is complete, the lamina may be easily moved by wedging a small dissector (Penfield #4) into the laminotomy groove and twisting it. This procedure is continued serially from one lamina to another along one side and then repeated on the other side.

One then incises the interspinous ligaments between the lowest spinous process to be reflected in the flap and the highest spinous process remaining, as well as the one between the highest spinous process to be reflected and the lowest one remaining. If possible, it is desirable to make the incisions in the interspinous ligaments midway between the two appropriate spinous processes so as to facilitate closure.

In freeing the laminar flap the yellow ligaments are cut individually, on each side, at each level, preferably with a #15 blade mounted on a long handle, with the direction of cut being ventrodorsal so as to minimize risk of damage to the epidural structures and the dura mater. The epidural fat is stroked away from the ventral surface of the laminae with either a Penfield #4 or #3 dissector, with care being taken not to compress the dura and underlying spinal cord and/or lesion. Bridging vessels are identified, coagulated individually with bipolar cautery, and then sectioned with microscissors. As each laminar segment is freed, the laminar flap is drawn dorsally and elevated slightly cephalad so as to avoid buckling at the fulcrum, thus eliminating the risk of compressing the underlying dura and cord.

Either freshly soaked fluffy cotton or pre-cut cottonoid patties may be placed upon the dura as one reflects the laminar flap. This affords maximal protection to the underlying structures and minimizes oozing. Once the laminar flap is completely removed, a fluffy cotton may be placed over the dura. The laminar flap is immediately put into normal saline where it is left until the intraspinal operative procedure is finished. Figure 3.43A illustrates the exposure one attains and visualization of epidural fat and venous plexus, dura and arachnoid, and cord or cauda equina.

Bone Closure

Craniotomy Closure (Figs. 3.44, 3.45)

As already discussed, when a craniotome is used to free the bone flap from the surrounding skull, a large gutter (varying in width from 2 to 4 mm!) is created, thereby rendering it impossible to attain bone to bone approximation when either a free or an osteoplastic bone flap is reinserted into position. It is disadvantageous to use power instruments to cut the bone between bur holes, since healing – bone to bone healing – seldom occurs (except in the newborn and very young infant) across these gutters. In fact, the flap sinks (Fig. 3.1A) varying depths beneath the level of the skull, fibrous tissue bridges across from skull to flap, and one has a moveable, albeit more or less limited, free bone flap. Use of the Gigli saw, on the other hand, permits immediate solid seating of the flap, like a cork in a bottle, since the cut may be beveled, and thus creates a wedge-shaped plug (Fig. 3.1B). Bony regrowth occurs across the osteotomy lines made by the Gigli saw. Although the vendors of power craniotomes may demonstrate techniques for obtaining a beveled cut, this is neither generally possible on an older child nor ever possible on an infant or young toddler (because of the thin skull).

In the supratentorial compartment reapproximation of the bone flap is followed by anchoring the flap into position with the use of periosteal sutures for medial frontal, parietal, and occipital flaps, and a combination of periosteal and fascial sutures for lateral frontal, bifrontal, and temporal flaps. If the bone flap is kept moist by wrapping it in a gauze sponge and then keeping it soaked throughout the time of the operative procedure, both the periosteum and fascial tissue adherent to it will be soft and compliant enough to give secure purchase to 4–0 and 3–0 suture material, respectively. Similarly, throughout the procedure, one must periodi-

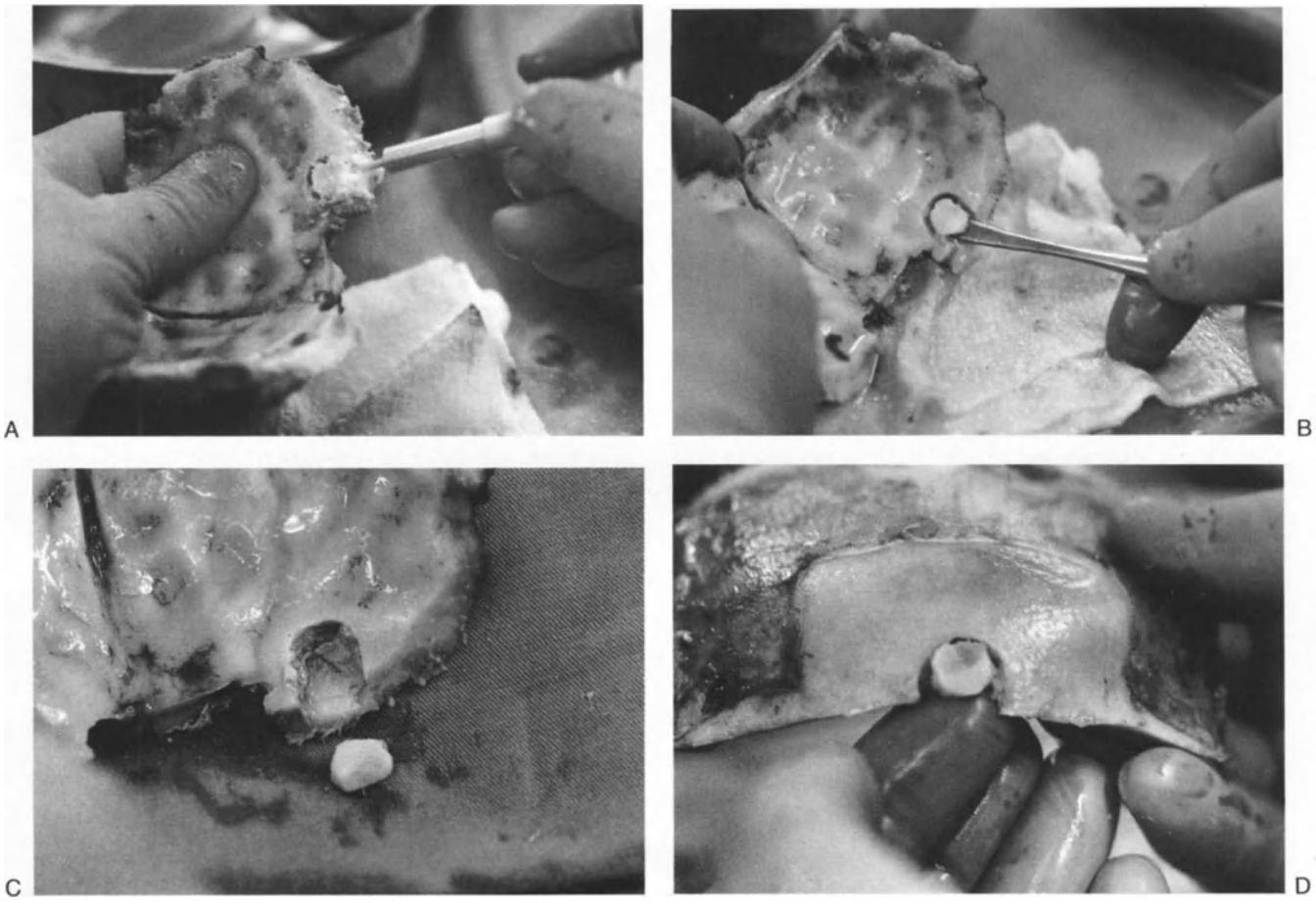


Figure 3.44. Preparation of the bone plug. (A) The high-speed drill is being used to fashion a plug from the inner table of the bone flap. (B) After the cut has been made through the inner table and into the diploic layer, a periosteal elevator is used to pry it free, taking care not to crack it. (C) This is a photograph of the freed plug and bone flap from which it was taken. (D) The bone plug is being held in the bur hole defect it will be used to close – in this case the glabellar hole for a bifrontal flap.

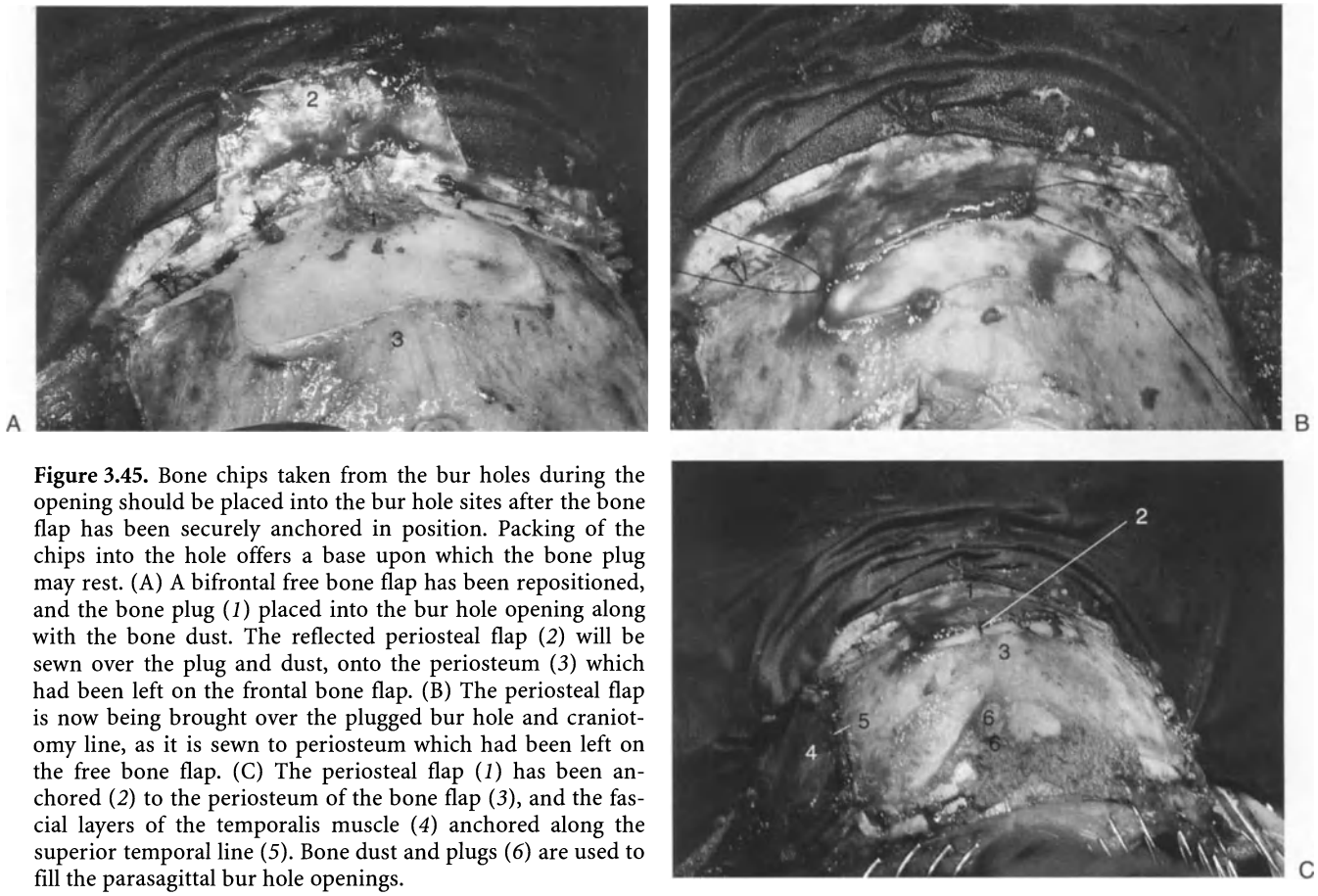


Figure 3.45. Bone chips taken from the bur holes during the opening should be placed into the bur hole sites after the bone flap has been securely anchored in position. Packing of the chips into the hole offers a base upon which the bone plug may rest. (A) A bifrontal free bone flap has been repositioned, and the bone plug (*1*) placed into the bur hole opening along with the bone dust. The reflected periosteal flap (*2*) will be sewn over the plug and dust, onto the periosteum (*3*) which had been left on the frontal bone flap. (B) The periosteal flap is now being brought over the plugged bur hole and craniotomy line, as it is sewn to periosteum which had been left on the free bone flap. (C) The periosteal flap (*1*) has been anchored (*2*) to the periosteum of the bone flap (*3*), and the fascial layers of the temporalis muscle (*4*) anchored along the superior temporal line (*5*). Bone dust and plugs (*6*) are used to fill the parasagittal bur hole openings.

cally irrigate the operative site so as to maintain adequate hydration of the periosteum and muscle.

Some surgeons choose to use a high-speed drill to perforate the borders of the bone flap and surrounding skull so as to pass suture material through the drill openings for the anchoring stitches. Wire suture is never indicated! Indeed, now that we are in the CT and magnetic resonance imaging (MRI) era, it is contraindicated: the metal produces artifact.

For the infratentorial craniotomy, unfortunately, the reapproximated bone flap must be anchored into position by the use of drill holes through the flap and surrounding squamous occipital bone, since it is not possible to preserve the periosteum, and there is no fascia adherent to the bone. The underlying, closed dura (the dura must *always* be closed, either directly or with the insertion of a fascia lata graft) offers a protective layer against: (1) compression of the cerebellum by the bone flap; (2) adhesions between the cerebellum, bone flap, periosteum. and muscle tissue; and (3) herniation of the posterior fossa contents into the deadspace between posterior fossa and muscle.

Bone chips taken from the bur holes during the opening should be placed into the bur hole sites after the bone flap has been securely anchored in position. Packing of the chips into the hole offers a base upon which a bone plug, fashioned from the inner table of the skull, may rest. This provides for complete bony refill of the bur holes. The regrowth of bone and its ossification is provided for by the underlying outer (periosteal) layer of the dura and the conserved periosteum which, at this stage of the procedure, lies over the bone plug. In the newborn or infant, fashioning of similar bone plugs and the use of bone chips are not necessary, since the skull regrows rapidly and completely. However, this is less true in the toddler, and not true in the juvenile or adolescent age groups. The plugs and chips are especially indicated for frontal or bifrontal flaps, to cover the bur hole defect at the glabella and behind the zygomatic process of the frontal bone ("keyhole").

Finally, the periosteum is brought over the bone plug and chips.

Laminar Closure (Figs. 3.46, 3.47)

The laminotomy flap is brought into the operative area, removing it from the moistened gauze sponge in which it was stored. If the laminar flap has been stored com-

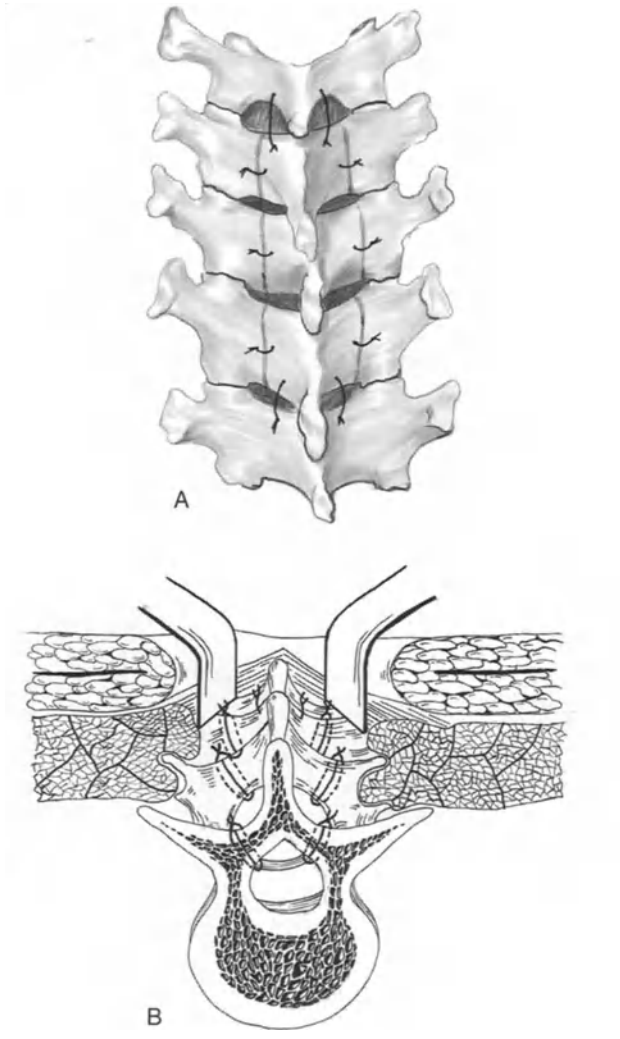


Figure 3.46. (A) Illustration of the suggested technique for replacing the free laminar flap by reapproximating it into its normal position and then anchoring the flap bilaterally, proceeding in a caudocephalad direction. (B) The anchoring sutures are shown penetrating the laminae through previously placed perforations.

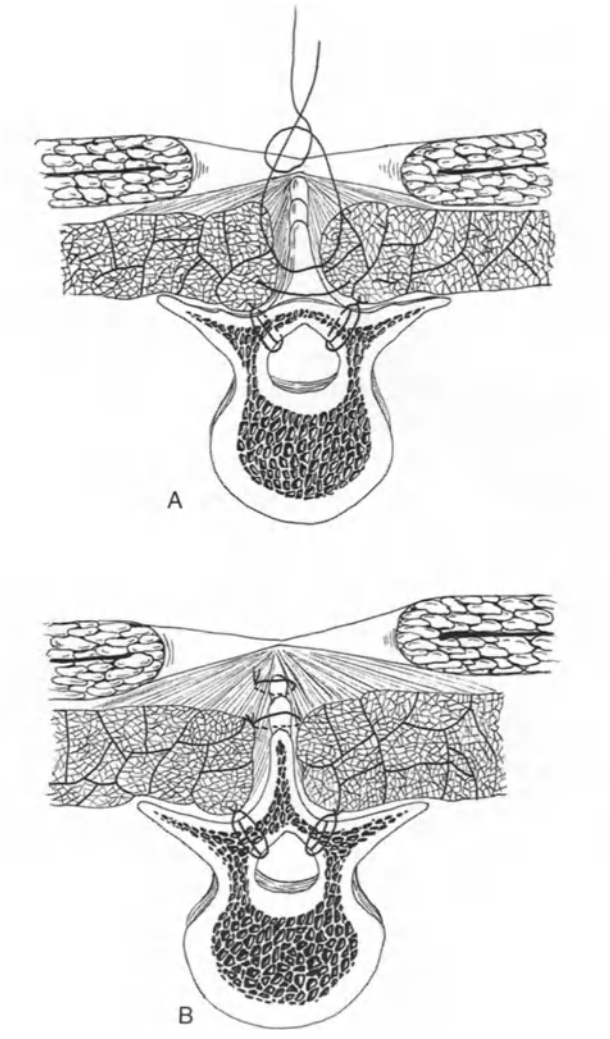


Figure 3.47. (A) The paraspinous muscle masses have been allowed to fall into their normal anatomical position, over the laminae and the spinous processes, and sutures placed so as to anchor these muscle masses, in two layers, to interspinous ligament. (B) Lastly, just before closing the skin, the fascia of the paraspinal muscle masses is sewn to the ruffle of ligament left on the supraspinous process.

pletely moistened, the interspinous ligaments will not have dried and shriveled. If it is stored dry, shrinkage occurs (rendering reapproximation of the laminar flap difficult or impossible).

A high-speed drill is used to perforate each of the laminae at the caudal and cranial ends of the laminar flap, and the portion of the laminae which remained in the vertebral body. With the flap brought back into its anatomical position, the surgeon passes sutures through the openings made in the most caudal laminae, and then ties them securely to one another, using 2–0 suture. This is done, from caudad to cephalad, at each level. These sutures are tied down individually at each level, first on one side then the other. The tying down of the laminar sutures one at a time from caudal to cephalad unfolds the flap as an open accordion, and brings each of the laminae to rest at its appropriate anatomical level.

Since it is not always possible to perform an osteotomy in a medial/lateral, dorsal/ventral, oblique line (which would allow the laminae to nestle into place without falling into the spinal canal), one must tie down the closing sutures snugly, so that the flap will not impinge upon the spinal canal. After the laminar flap is thus anchored into position, the interspinous ligament at the inferior and superior segments is tied to the fragments of the homonymous ligaments below and above. The paraspinous muscle masses are then allowed to fall

into their normal position by removing the self-retaining retractors. *They are sewn to the respective interspinous ligaments in two layers, deep and superficial, and the fascia over the paraspinuos muscle is sewn to the supraspinous ligments. It is our opinion that sewing the paraspinous muscle masses to their appropriate interspinous ligaments is the most important single factor in laminotomy as a prevention of kyphoscoliosis: it prevents uneven postoperative pull of muscles from one myotome on those of the other.* This brings muscle mass, spinous processes, and interspinous ligaments into anatomical juxtaposition, and prevents the laminar flap from moving or sinking into the spinal canal. The subcutaneous tissue and skin are then closed.

Postoperative Treatment and Follow-up of Laminotomy

The postoperative treatment for laminotomy consists of appropriate immobilization of the patient, which is obtained through a thoracic on lumbosacral corset for the corresponding spine segments, and a "four-poster" cervical collar or "halo" for the cervical spine. Serial X-ray controls are performed from the first days after surgery, and biweekly thereafter for 6 weeks. Once there is X-ray evidence of healing across the osteotomy site, no further X-rays are taken and the child may resume normal activity.

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4 Suturotomy for Various Flaps in the Newborn and Infant

"Nothing may fail like success."
VARÉ, Italian Diplomat

In the newborn and infant for reflection of frontal, bifrontal, parietal, and other flaps the use of the perforator and either Gigli saw or craniotome is unnecessary and potentially dangerous, since the skull thickness measures less than 3 mm, and each of the membranous bones rides freely over the underlying brain, suspended and moored, as it were, from the membranous sutures. Individual bones are separated from one another by open suture lines, and ossification is at least a year from being complete. The bones offer no resistance to the pressure of a perforator or bur (see Figs. 4.1–4.6).

The individual bones cover almost completely the respective lobes of the brain, so that one may expose the parietal, or frontal, lobes simply by cutting the appropriate bone from – and at – its junction with the surrounding suture. Reflection of a frontal flap necessitates use of heavy scissors to cut the squamous portion of the frontal bone from the supraorbital ridge, a line along which no suture exists. A suboccipital flap may not be reflected by sectioning a suture because the squamosal suture is located well above the transverse sinus.

The anterior fontanelle and the three sutures with which it is continuous (metopic, coronal, sagittal) offer the key area for reflecting frontal, bifrontal, parietal, or biparietal flaps. The metopic suture begins to ossify inferiorly at the glabella, a process that extends posterosuperiorly to the anterior fontanelle. Consequently, at the time of birth one may encounter bony union at the glabella.

The suturotomy is performed by stripping the periosteum from the bone edge at a point no more than 1 mm from the suture line. The periosteum is reflected from the bone edge, over a linear distance of approximately 1.5 cm. The highly vascularized bone and the interosseus portion of the suture are then exposed before using a sharp periosteal elevator, such as the Oldberg or a sharpened Penfield #4 dissector, to separate the suture line from the inner table of the skull. This permits one to separate completely the inner table of the skull from the outer layer of the dura by inserting a blunt Penfield #2 or #4 dissector and stripping the former from the latter. It is not possible to run the dissector across the suture line, so the surgeon separates the outer layer of the dura from the inner table of the skull on either side, bringing the separation up to the suture line. This is done through the small opening already described.

Heavy scissors, either curved or straight Mayo, are inserted so that one blade serves as a dissector, and then the bone is cut *along* the suture line, extending from superomedial to inferolateral, cutting the coronal suture. The direction of cut is from superior to inferior and then from the anterior fontanelle to the glabella if one wishes to open the metopic suture. To separate the parietal bone from the sagittal suture, it is best to proceed from the anterior fontanelle to the posterior fontanelle, taking care to cut along the junction of bone and suture, so as to avoid damage to the superior sagittal sinus or the bridging veins.

Reflection of the free or osteoplastic flap, after suturotomy, exposes the underlying dura and suture lines. One may choose to reflect any combination of unilateral frontal, bifrontal, biparietal, frontoparietal, and temporoparietal flaps for access to the desired area.

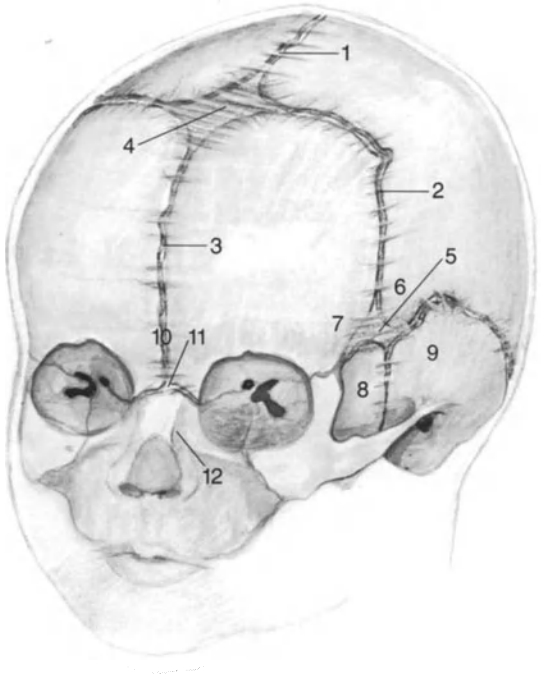

Figure 4.1. Anatomical drawing of the infant's neurocranium, illustrating the bones of the calvarium and the sutures continuous with it. The sagittal (*1*), coronal (*2*), and metopic (*3*) sutures are continuous with the anterior fontanelle (*4*). The coronal suture extends anteroinferiorly in the coronal plane, to the pterional area (*5*), where the parietal (*6*), frontal (*7*), sphenoid (*8*), and squamous temporal (*9*) bones fuse. The metopic suture extends anteroinferiorly, in the sagittal plane, past the glabella (*10*) to the frontonasal (*11*) sutures, where it is joined by the frontal processes of the maxillae (*12*).

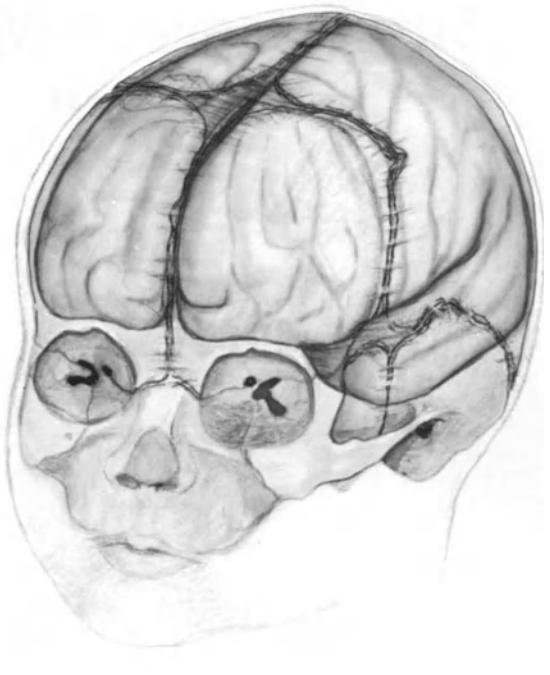

Figure 4.2. Membranous bony plates, seen in transparency, overlying the respective lobes (frontal, parietal, temporal) of the brain. Suturotomy around the appropriate bone, therefore, suffices to expose the underlying lobe of the hemisphere.

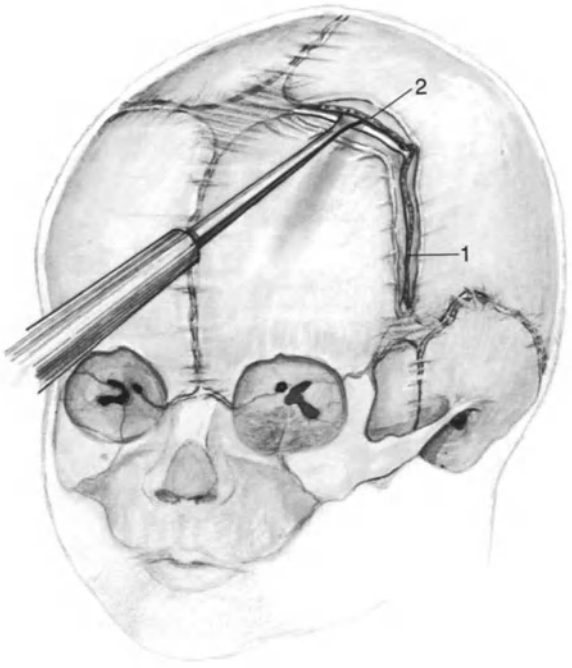

Figure 4.3. First stage of suturotomy, stripping of the periosteum across the suture line, and the second stage, separating the dura from the inner surface of the skull, are illustrated here. Inferiorly, along the coronal suture, one notes that the periosteum has been separated from the suture line (*1*) over a linear distance of approximately 1.5 cm and lateralward for approximately 2.5 mm. Superior to this, the technique for inserting a dissector between the inner table of the skull and the dura, extending distally from the suture line, is illustrated (*2*). The dissector is swept over the outer layer of the dura, within the epidural area, converting this from a potential to a virtual epidural space.

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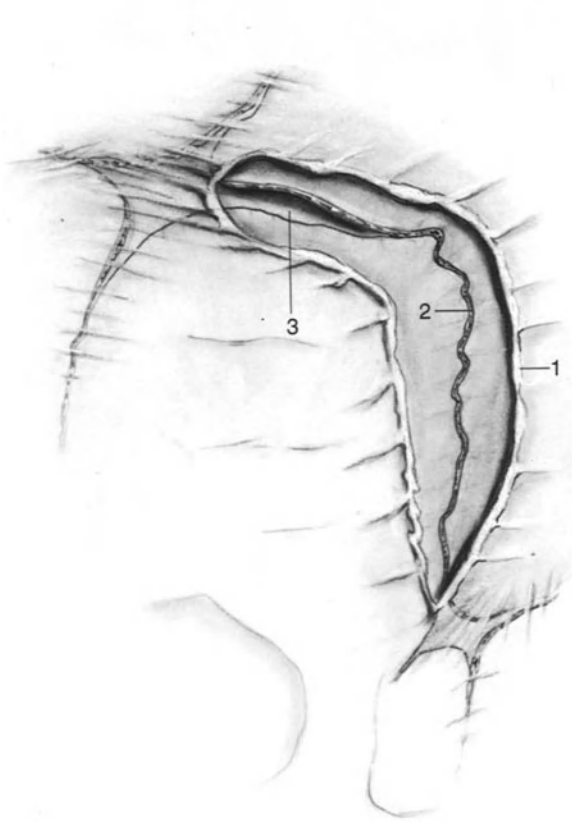


Figure 4.4. The periosteum (*1*) has been stripped from the coronal suture (*2*) and the outer layer of the dura (*3*) from the inner of the skull.

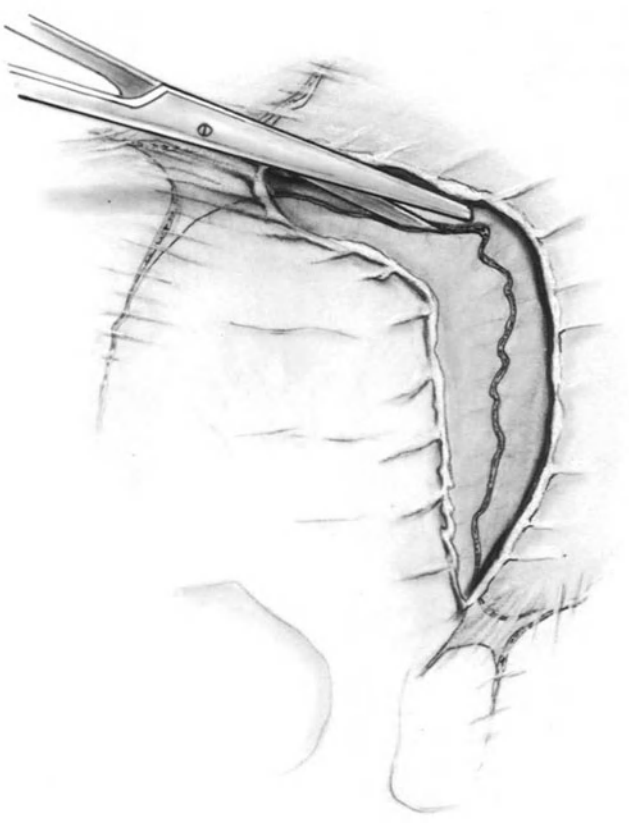


Figure 4.5. The scissors have been inserted so as to cut along the line of junction between the parietal bone and the coronal suture. The blade between the skull and the dura serves as a dissector.

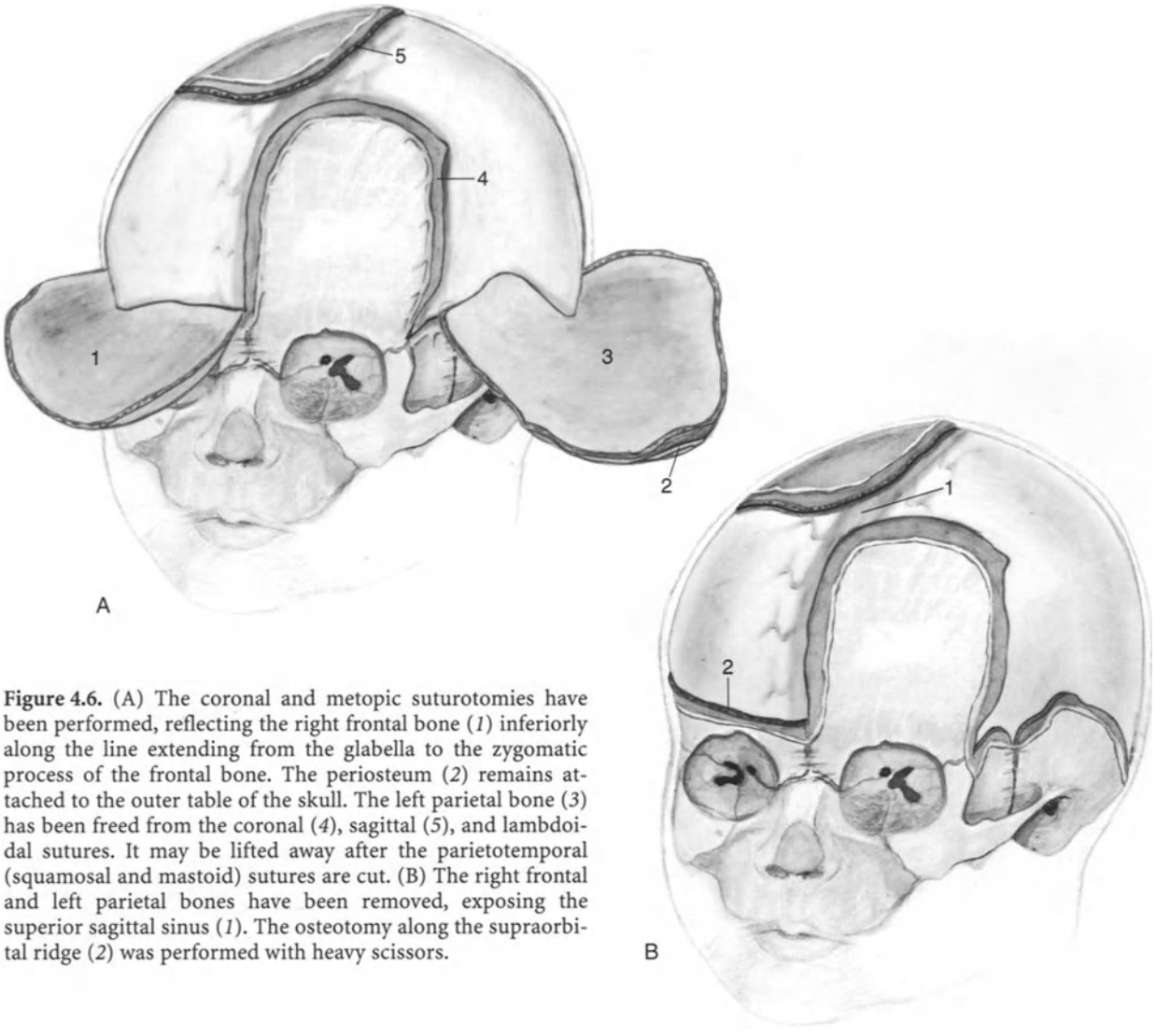


Figure 4.6. (A) The coronal and metopic suturotomies have been performed, reflecting the right frontal bone (*1*) inferiorly along the line extending from the glabella to the zygomatic process of the frontal bone. The periosteum (*2*) remains attached to the outer table of the skull. The left parietal bone (*3*) has been freed from the coronal (*4*), sagittal (*5*), and lambdoidal sutures. It may be lifted away after the parietotemporal (squamosal and mastoid) sutures are cut. (B) The right frontal and left parietal bones have been removed, exposing the superior sagittal sinus (*1*). The osteotomy along the supraorbital ridge (*2*) was performed with heavy scissors.

5 Dural Flaps

"There is no joy like the conquest of difficulties well within one's powers – she shook the dust of the place off her feet."

CHARLOTTE BRONTË, Jane Eyre

General Comments (Fig. 5.1)

After the craniotomy has been performed, and before the dura is opened, one should tack the dura to the periosteum of the skull. Because of the remarkably high degree of vascularization of the skull and the relatively loose adherence between outer layer of the dura and inner table of the skull, it is essential to sew the outer layer of the dura to the periosteum along the craniotomy line before proceeding to open the dura in order to minimize risks of epidural hematoma formation (4–0 sutures set at 3–4 cm from one another suffice to perform this). It is *not* necessary to perforate the skull to anchor these sutures because the periosteum is strong enough to offer adequate purchase, and it serves no purpose to bring sutures from the center of the dural flap through the center of the overlying bone flap (at the time of closure). However, one should place the dural needle through the periosteum along a line parallel to the cut edge of the periosteum (skull), not perpendicular to it, since this avoids fraying or tearing of the periosteum at the time the knot is tied. One need not sew the dura to the periosteum along the region of the anterior or posterior fontanelles, across a cranial suture, or at the superior sagittal or transverse sinuses, since at these points outer layer of dura, cranial suture, and periosteum are one continuous structure in the newborn and infant: there is no risk of dura stripping itself from the inner table of the skull.

A scleral hook is ideal for grasping the outer layer of the dura and elevating the intact dura from the surface of the underlying brain so that it may be incised with a #15 blade. Either Adson or Adson-Brown forceps are then used to grasp the dura mater, elevating this structure from the surface of the brain, before proceeding to cut it with tenotomy scissors.

For medial dural openings (frontal, parietal, occipital) care should be taken to look beneath the dura prior to extending the cut, lest a bridging cortical vein be opened. Indeed, it is strongly recommended that cuts along the parasagittal plane be avoided because of these bridging cortical veins. The dura should be opened in a horseshoe or trapdoor fashion, approaching the midline or the line of tentorial origin perpendicular to the sagittal or transverse sinuses, respectively.

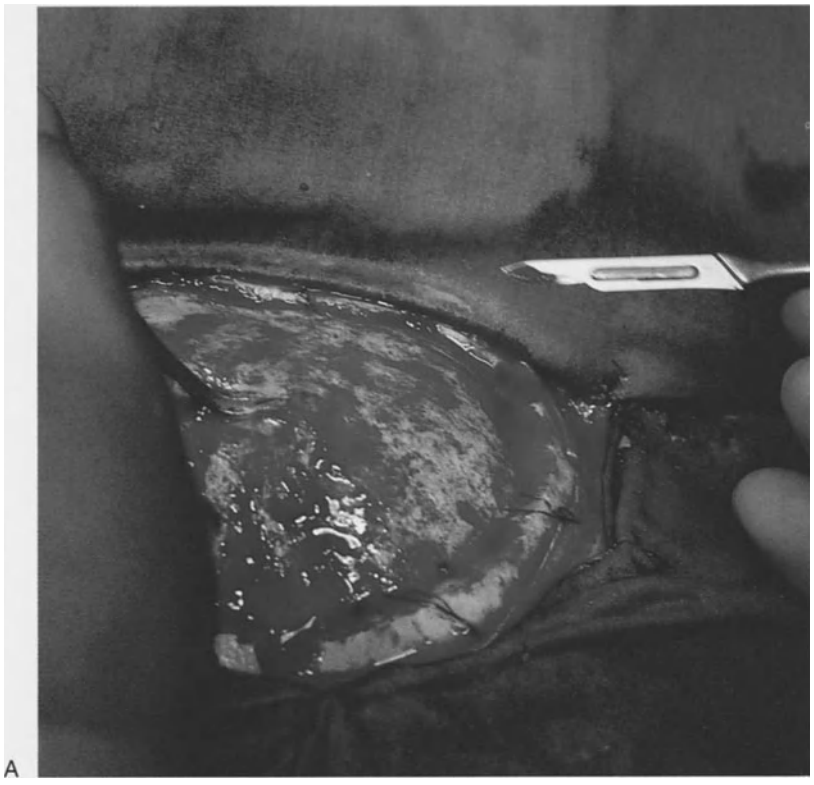
Dural Openings

The technique for opening the dura is the same, irrespective of where it is done. However, the incision line, the form of the opening (single trapdoor, double trapdoor, horseshoe, linear, etc.) varies with the location and underlying pathology.

Frontal Dural Openings

Medial Frontal Dural Opening (Fig. 5.2)

Dural opening and reflection of the medial dural flap for a medial frontal craniotomy is somewhat different from that for a lateral frontal craniotomy. In essence, reflecting the dura in a medial frontal craniotomy entails exposing the bridging cortical veins, something that is not done in a lateral frontal craniotomy, since the superior sagittal sinus is not exposed in this latter procedure. After the dura has been incised with a #15 blade, it is opened in a single trapdoor fashion, and then the dural flap is reflected medially over the osteotomy line and sewn down to the periosteum. This flap should be sewn out of the way, in a line that does not kink or stretch the bridging cortical veins. The cut anterior, inferior, and posterior edges of the dura are, in turn, sewn around the osteotomy line and to the periosteum. This prevents the frontal dural flaps from falling continuously into the operative field, and further minimizes the risk of stripping dura from the inner surface of the skull.



A

B

Figure 5.1. (A) The dura has been opened by using a scleral hook inserted into its outer layer, elevating it, and then nicking it with a #15 blade. Once both layers of the dura have been identified, a right-angled groove director is inserted between the arachnoid and inner layer of the dura. A #15 blade is used to start the dural opening. Here, 4–0 sutures (*arrows*) were sewn from the outer layer of the dura to the periosteum prior to opening the dura. (B) One notes arachnoid bulging through the dural opening, which is now being completed with the use of Adson-Brown forceps to elevate the dura and tenotomy scissors to cut it. Insertion of the scissors should be such as to use the blade placed between arachnoid and inner layer of the dura as a dissector, taking care to angulate the scissors in such a manner as to point the tip of the blade upward against the dura. This avoids cutting the underlying arachnoid or cortex.

Lateral Frontal Dural Opening

The dural opening for a lateral frontal craniotomy (Fig. 5.3) is different from that for the medial frontal craniotomy only in regard to exposure of the bridging cortical veins.

Bifrontal Dural Opening (Fig. 5.4)

The bifrontal (bifrontopterional) craniotomy exposes the superior sagittal sinus (SSS) and the intradural portion of the bridging dural veins bilaterally.

The dural opening differs from that used in either the medial or lateral frontal craniotomy in that trapdoor openings are not used. Rather, a single hockeystick incision is made on either side, extending from medial to lateral, along a plane parallel to the supraorbital ridge, with the angulation located at the zygomatic process of the frontal bone. The incision is then extended posteriorly to the pterion. The peripheral edge of the dura is incised to the base of the zygomatic process of the frontal bone. Subsequent to this, clips or sutures are placed across the most anteroinferior portion (beneath the glabella) of the SSS down to the falx cerebri, separated from one another by a distance of approximately 2 mm. The most anterior portion of the SSS is ligated before it is transected. The falx cerebri is

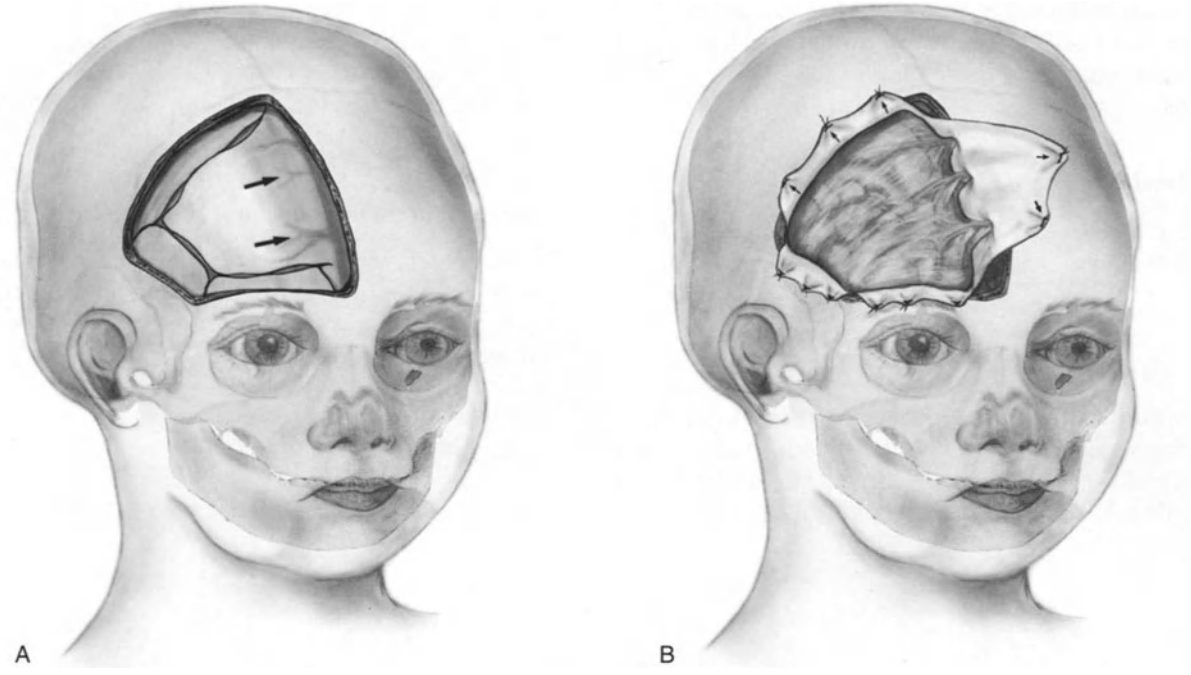


Figure 5.2. (A) Medial dural opening. The dura is opened (single trapdoor) and retracted along a line running parallel to the craniotomy, at a distance of about 1.0 cm (to facilitate closure), except along the parasagittal plane. This opening is fast, and minimizes damage to the bridging cortical veins (*arrows*). (B) Medial dural opening. The dural flaps are sewn (over the craniotomy edge) to the periosteum. Note (1) that the sutures are placed parallel (*arrows*) to the cut surfaces of the dura and periosteum, avoiding the suture tearing through these structures when the knot is tied; (2) the entrance of the bridging cortical veins into the dura; and (3) the tacking down of each to prevent the flap from following into the field and to hold it fully stretched throughout the procedure.

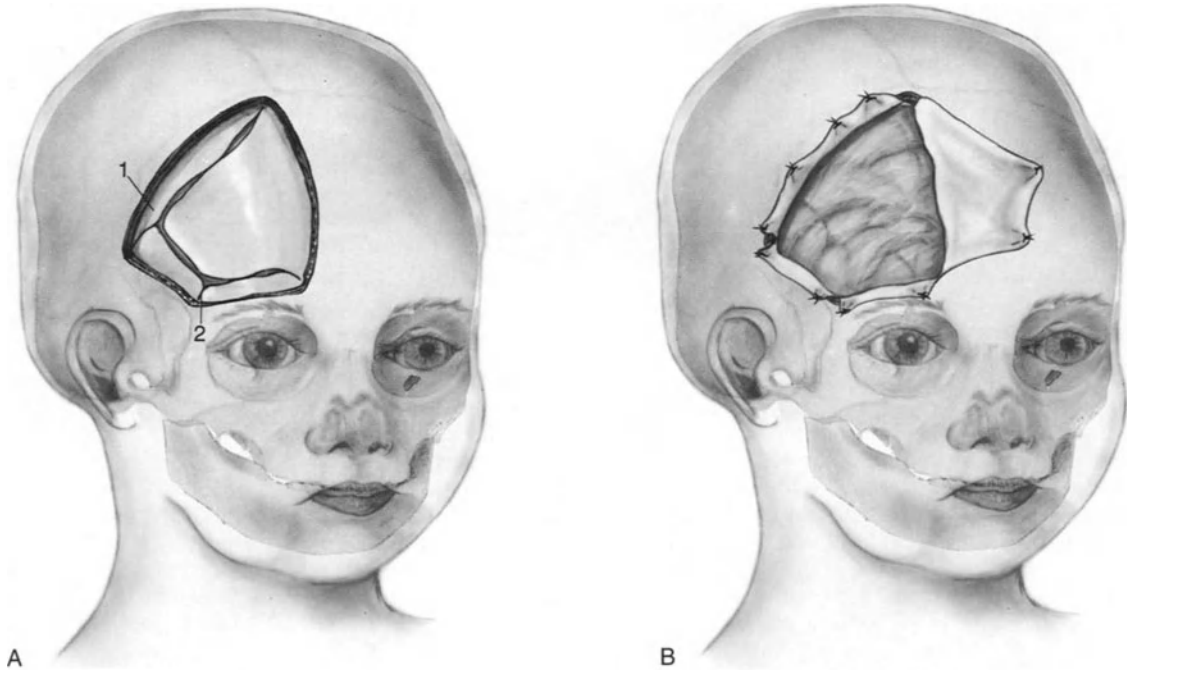


Figure 5.3. (A) Lateral dural opening. The dura is incised in a single trapdoor fashion, and cuts are made at the posterolateral (*1*) and anterolateral (*2*) edges. The craniotomy is well lateral to the superior sagittal sinus, so the intradural portion of the bridging veins is not visible. (B) Lateral dural opening. The single trapdoor and the three peripheral edges of dura have been sewn to the periosteum, exposing the brain and securing (and stretching) the dura out of the field.

cut from its insertion onto the crista galli, allowing the dura mater to be retracted with the frontal lobes. This prevents stretching or kinking of the bridging cortical veins.

Parietal Dural Opening (Figs. 5.5–5.8)

The parietal dural openings may be superior parietal, parietotemporal (inferior parietal), biparietal, and parietofrontal.

Superior Parietal Dural Opening

Superior parietal flaps are reflected so as to offer access to the hemispherical convexity, superior sagittal sinus, falx, and corpus callosum. Consequently, an asymmetrical, double trapdoor dural opening is fashioned. This provides adequate exposure of the parietal convexity and sagittal structures, and allows the option of uncovering the lateral surface of the parietal lobe if needs must. The anterior and posterior limbs of the dural opening permit full utilization of the craniotomy.

Parietotemporal Dural Opening

The parietotemporal dural opening is also effected with a double trapdoor, again to expose only the desired area and to permit the option of a more extensive cerebral exposure.

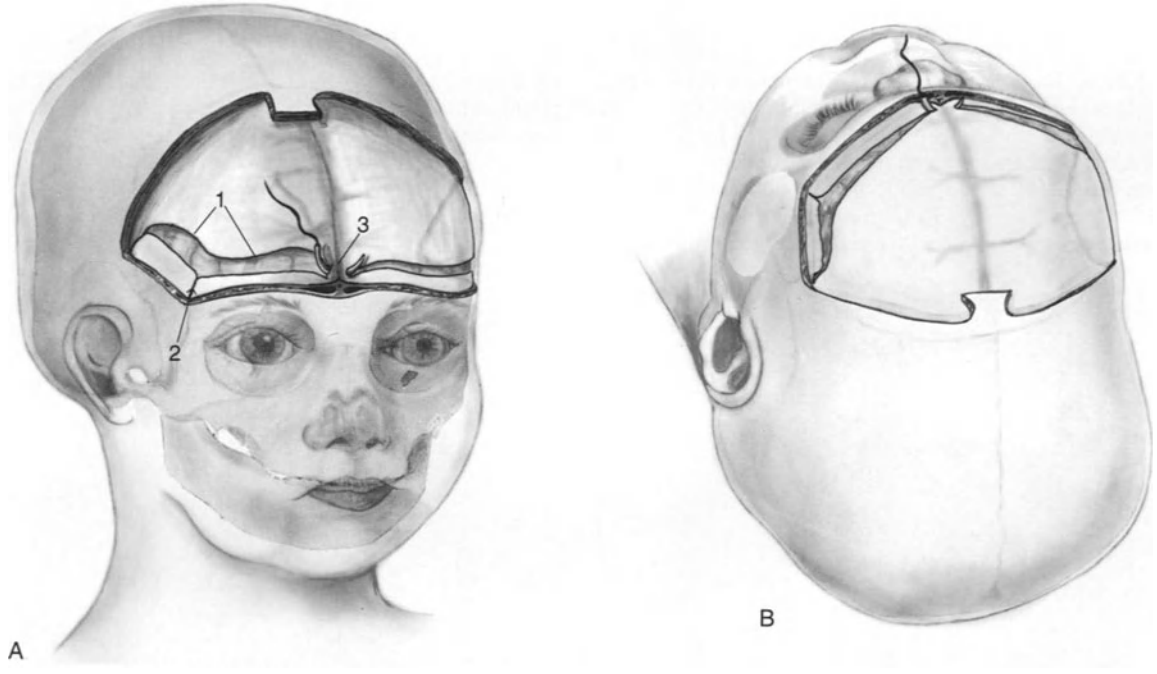
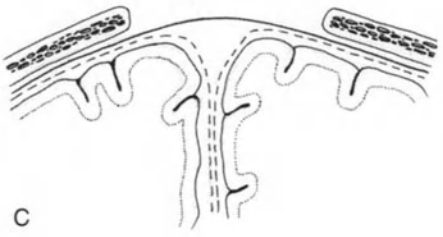
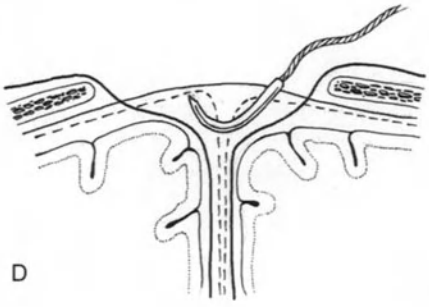
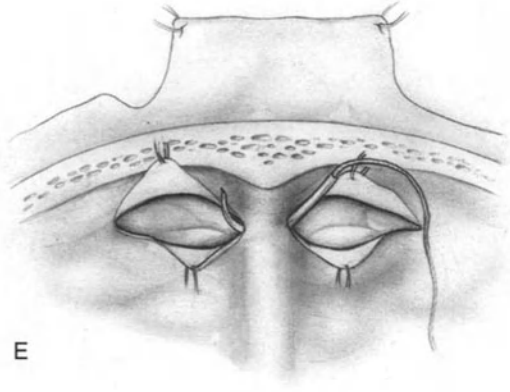
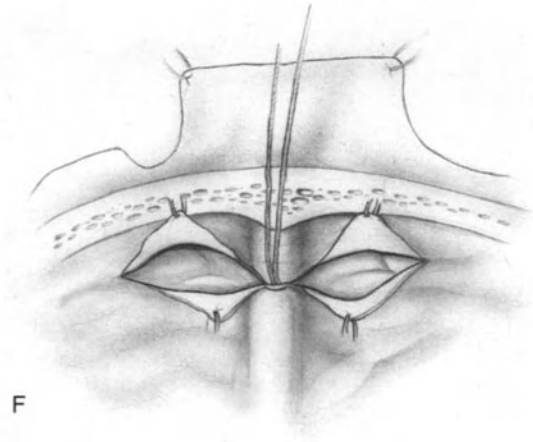


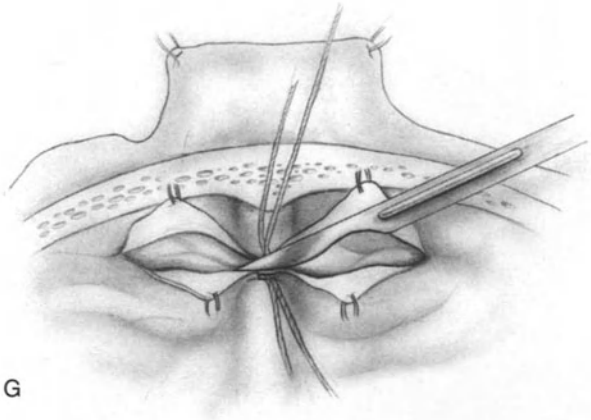
Figure 5.4. (A) The bilateral hockey-stick incisions (*1*) have been made and the bordering edge cut at the zygomatic process of the frontal bone (*2*). Suture, preferable to clips, is being passed through the falx (*3*), to ligate the SSS. The child is in the anatomical position. (B) With the child supine, one notes the hockey-stick incisions, and the suture being passed through the falx cerebri. The head has been positioned obliquely to illustrate the full extent of the hockey-stick dural incision. (C) This is a schematic representation of a sagittal cross section at the region of the glabella, illustrating the bone defect produced by the craniotomy, the underline anterior sagittal sinus with the outer layer of the dura represented as a *continuous line* and the inner layer represented as a *broken line*. (D) Telfa, the *heavy continuous line* passing from the surface of the skull across the dura and then between the falx and the cerebral cortex, lain in place to protect the brain. (E) The curved needle is passed from one medial extremity of the dural opening through the falx to the other medial extremity, well below the superior sagittal sinus. (F) The ligature is tied down slowly using a two-handed knot. (G) A second ligature has been brought into place and tied down, and a #15 blade is being used to cut across the superior sagittal sinus between the two ligatures. (H) This drawing illustrates the transected anteroinferiormost portion of the superior sagittal sinus completed. (I) Supine position. This illustration permits one to appreciate how the frontal lobes (*1*) and dura (*2*) fall away, together, from the floor of the anterior fossa (*3*) as the head is lowered. Also, one notes that the edges of dura anchored to the periosteum of the orbital rim (*4*) and to that over temporosphenoidal bones (*5*) are thereby prevented from falling continuously into the operative field. (C–I) see p. 125.

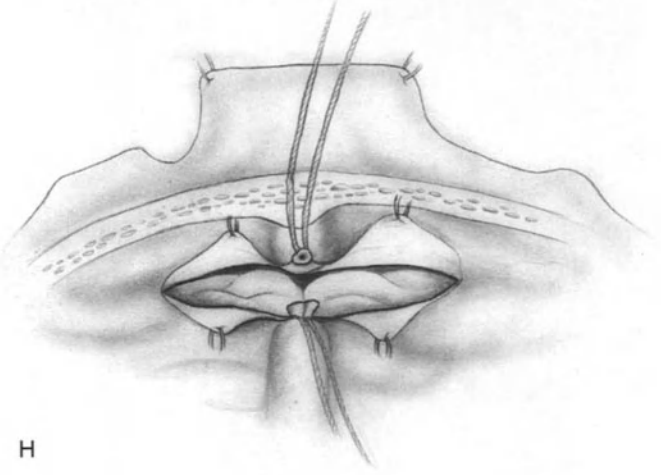












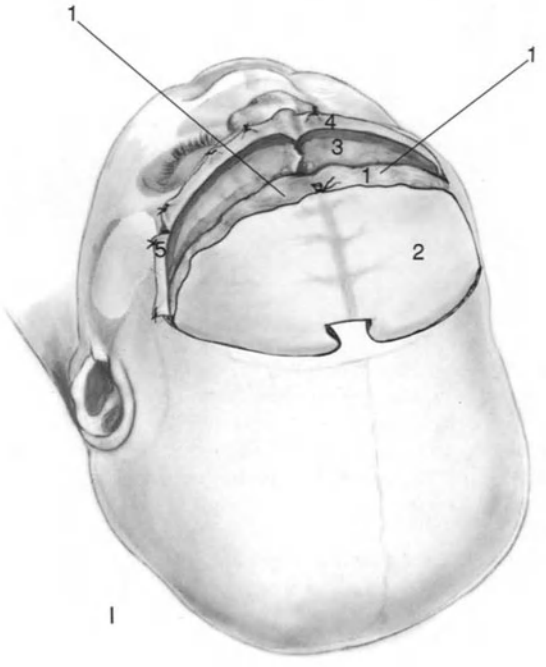


Figure 5.4. Legend see p. 124.

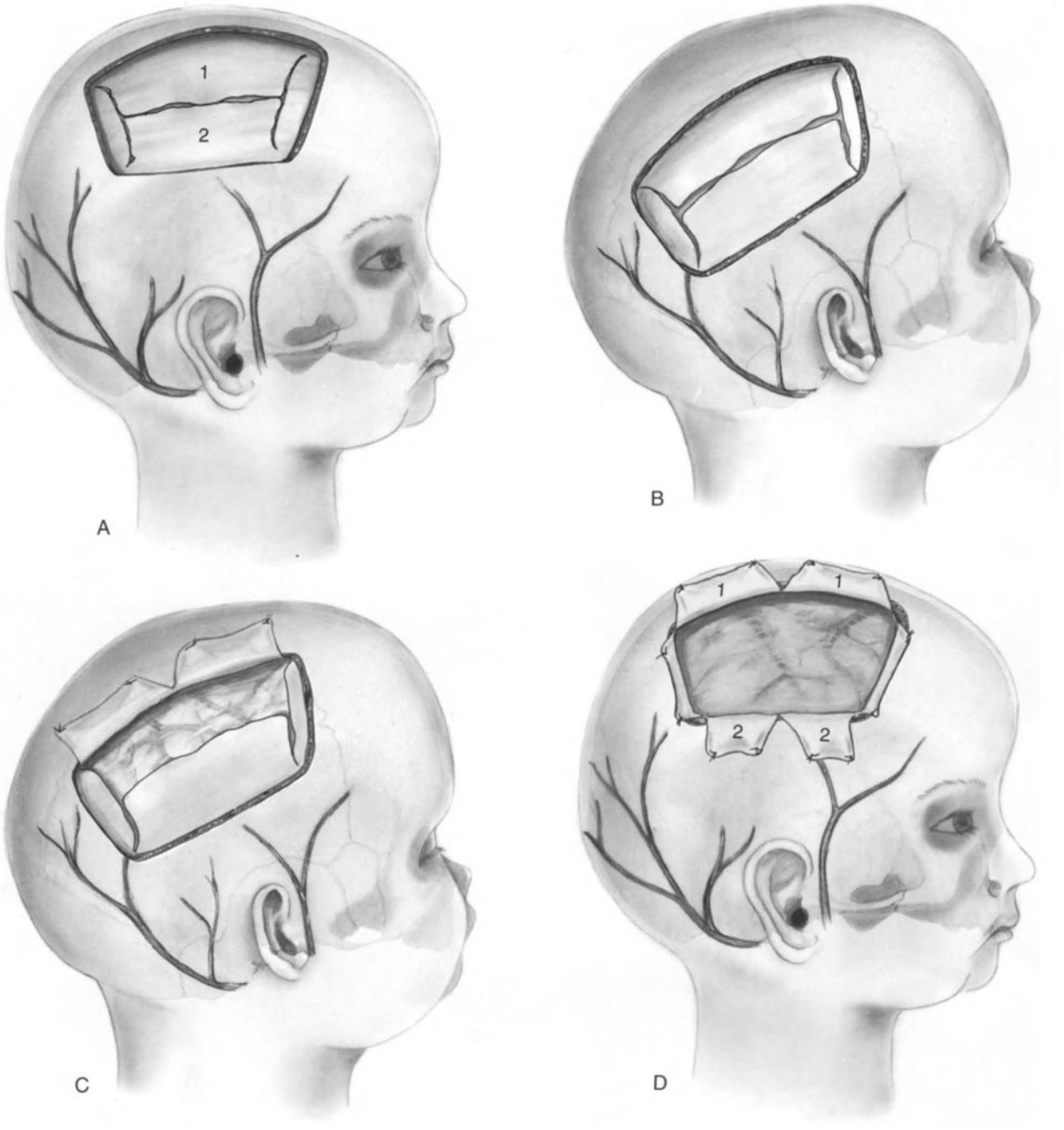


Figure 5.5. (A) Superior quadrilateral parietal flap as seen from a straight lateral perspective, in order to visualize the relationship between the posterior extremity of the flap and the parietal eminence. The dura has been incised but not reflected. Note that only the superior portion of the dura (*1*) may be reflected over the superior sagittal sinus without opening the inferior trapdoor (*2*), or the inferior trapdoor may be opened for greater access to the inferior parietal lobe. (B) Postero-oblique view of the dural opening in a quadrilateral bone flap to illustrate access to the parietal convexity. (C) Postero-oblique view, with the split superior trapdoor cleft and sewn over the superior sagittal sinus and anchored to the periosteum. (D) The dura beneath this quadrilateral parietal flap is reflected at all four corners and sewn into position after it has been stretched. The medial (*1*) and lateral (*2*) flaps have been cleft at their center to avoid dural buckling and to provide full visualization of the sagittal, parietal, and temporoparietal areas.

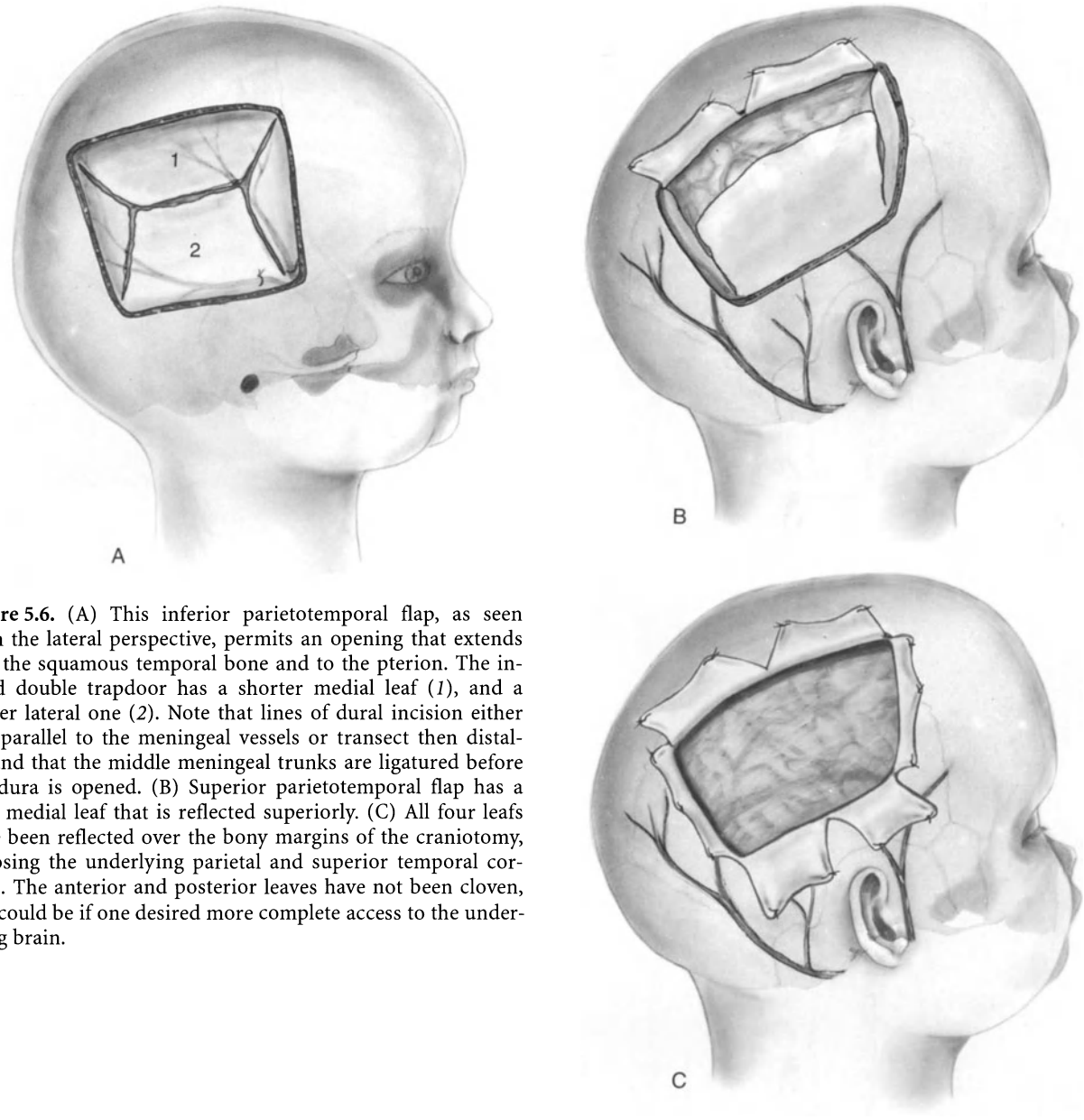


Figure 5.6. (A) This inferior parietotemporal flap, as seen from the lateral perspective, permits an opening that extends into the squamous temporal bone and to the pterion. The incised double trapdoor has a shorter medial leaf (*1*), and a longer lateral one (*2*). Note that lines of dural incision either run parallel to the meningeal vessels or transect then distally...and that the middle meningeal trunks are ligatured before the dura is opened. (B) Superior parietotemporal flap has a cleft medial leaf that is reflected superiorly. (C) All four leafs have been reflected over the bony margins of the craniotomy, exposing the underlying parietal and superior temporal cortices. The anterior and posterior leaves have not been cloven, but could be if one desired more complete access to the underlying brain.

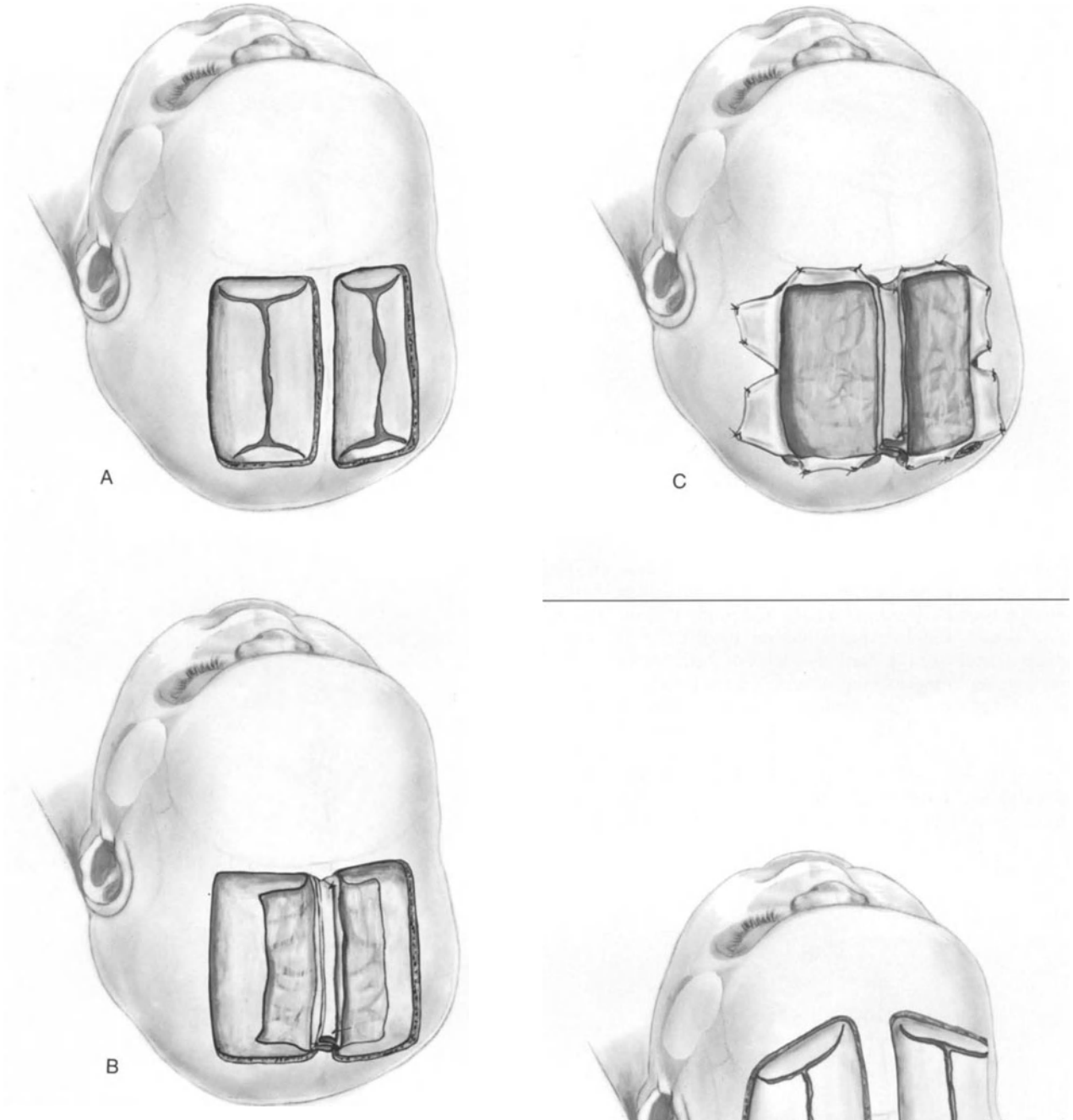


Figure 5.7. Biparietal dural flaps with the dura completely incised. (A) The durotomies have been made in double trapdoor fashion, so that one may expose the medial, lateral, or entire parietal lobe on both sides. (B) Opening only the medial trapdoors gives access to the sagittal area and medial parietal lobes, while affording dural protection to the lateral parietal surface. The medial flaps are sewn over the strip of bone bordering the sagittal suture. (C) The dura has been reflected over the free edges of the parietal craniotomy and anchored into position. The medial trapdoors have each been brought over the midline and anchored to one another, keeping them stretched over one another when the sagittal strip of bone has not been removed in reduction cranioplasty, and sewing both to one another and brought forward to suspend and protect the SSS sinus when it has.

Figure 5.8. (A) Parietofrontal craniotomy. The durotomies have been made in double trapdoor fashion, permitting exposure of the entirety of underlying cerebral and vascular structures. (B, C) see p. 129.

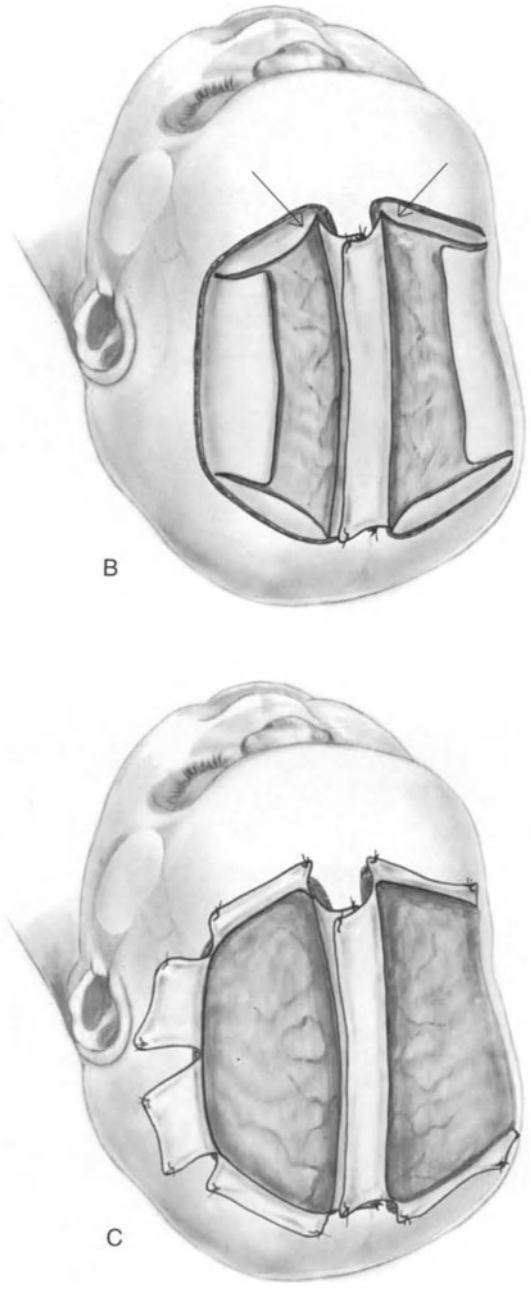


Figure 5.8. (B) Bilateral parietofrontal opening. Only the medial trapdoors have been reflected over the midsagittal plane, exposing the parietal lobes and the posterior portions of the frontal lobes. The dural opening extends anteromedially (*arrow*) so as to put into view the anteromedial bridging cortical veins. (C) The inferior trapdoor has been opened and cut in two places so as to afford maximum exposure of the frontoparietal cortex.

Biparietal Dural Opening

The biparietal dural openings may be either parietal or parietofrontal. They permit access to the convexities of both parietal (or parietofrontal) lobes, to both sides of the falx cerebri, and to the splenium of the corpus callosum. The parietofrontal flap permits access to the genu, as well as the body and splenium of the corpus callosum. It also allows the surgeon to perform independent cerebrotomies in the frontal and parietotemporal areas for access to the frontal horn and trigone, respectively, for those choroid plexus papillomas extending from the glomus to, and through, the foramen of Monro.

Temporal Dural Openings (Fig. 5.9)

Anterior Temporal Dural Opening

Particular attention is given to studying the anterior opening of the dural flap, as it extends inferiorly across the posterior branch of the middle meningeal artery but remaining posterior to the middle meningeal proper. This opening must be reflected over the lesser wing of the sphenoid in two or more folds, to avoid tenting and, consequently, converting the dural opening into a tube.

Middle Temporal Dural Opening

The middle opening reflects the dura over the craniectomized squamous temporal and greater wing of the sphenoid bones, giving the surgeon a straight line of vision to the floor of the middle fossa, unimpeded by a ledge of temporal squama. This minimizes, or eliminates, the need to elevate significantly the temporal lobe for access to the tentorial edge and ambient cistern.

Posterior Temporal Dural Opening

The posterior opening is designed for access to the superior and middle temporal convolutions, the terminal portion of the sylvian fissure. The angular and supramarginal gyri are most conveniently approached through a parietotemporal flap.

Enlarged Temporal Dural Opening

An enlarged temporal opening is no more than an incorporation of anterior and posterior openings: it provides access to the entire temporal lobe, tentorial surface and edge, and ambient cistern.

One may choose to cut the dura so as to avoid the middle meningeal artery, but I discourage this because it serves no purpose, permits continuous, nagging bleeding from the dural edges, and increases greatly the

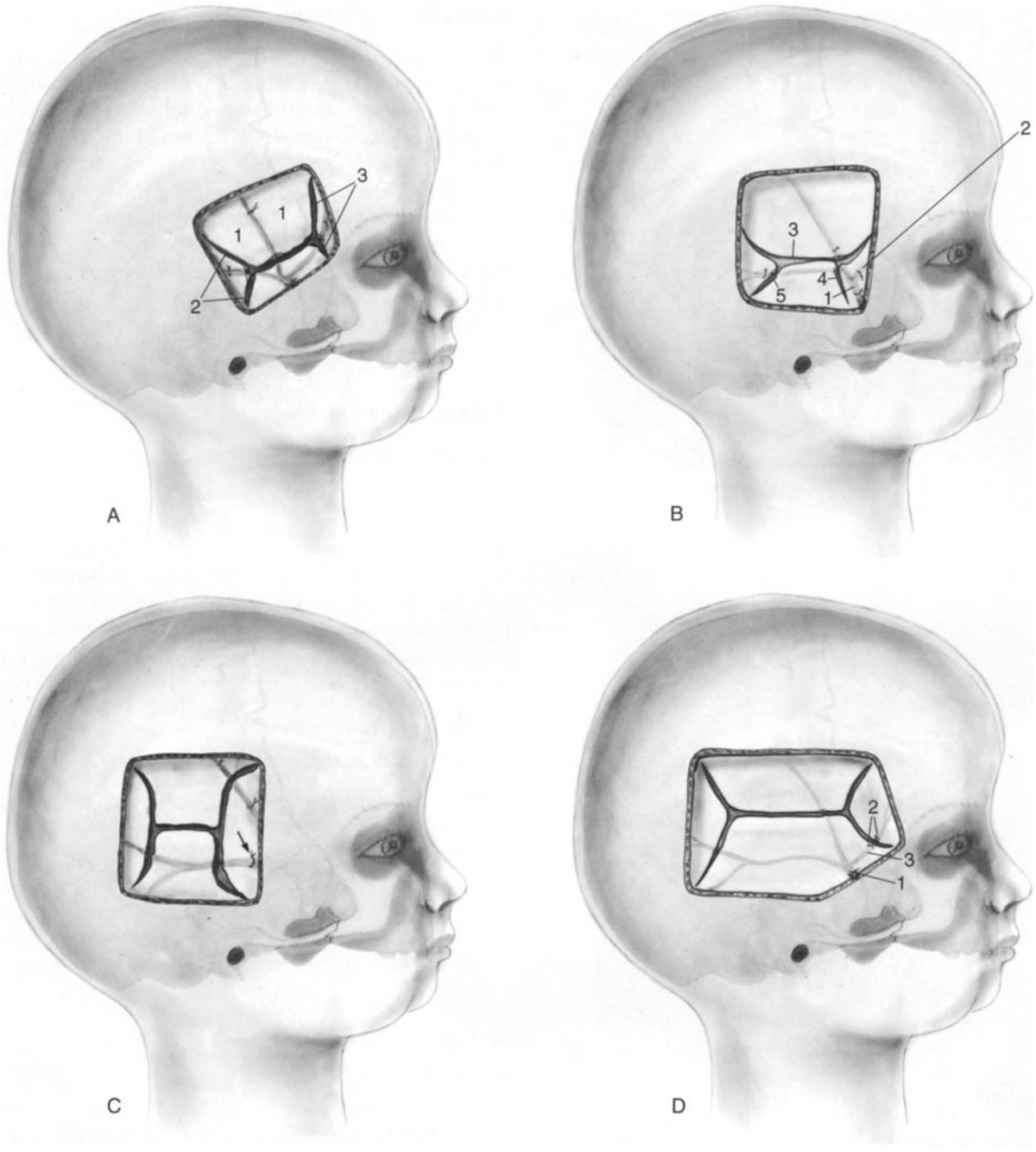
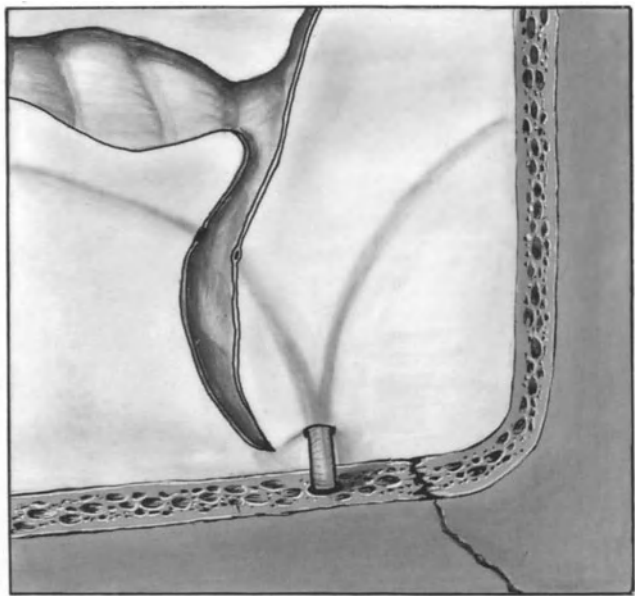


Figure 5.9. (A) The anterior temporal dural incision and reflections are illustrated. The middle meningeal artery is ligated by passing a 4–0 suture through both layers of the dura and then tying them down securely. This prevents bleeding from the dural edges. The dura is then opened by making a semilunar incision (*1*), convexity downward, and perpendicular incisions posteriorly (*2*) and anteriorly (*3*). This opening minimizes bleeding and maximizes exposure. (B) Midtemporal dural incision and reflections. In this craniotomy, the middle meningeal artery (*1*) is located at the anterior extremity of the bone flap, dural exposure. It is essential that it be identified and securely ligatured before proceeding to open the dura. A ligature should also be put around its anterior branch (*2*). The semilunar durotomy is then made (*3*) and perpendicular cuts extended anteriorly (*4*) and posteriorly (*5*). The terminal posterior branch(es) may be ligated, or coagulated, since bleeding is only retrograde: the middle meningeal will already have been occluded. (C) Posterior dural incision and reflections. A single ligature around the main trunk of the posterior branch of the middle meningeal artery (*arrow*) suffices to prevent all bleeding from the cut dural edges. The dura is then opened in a double trapdoor fashion. (D) The enlarged temporal dural incisions are illustrated in this drawing. The middle meningeal artery (*1*) is generally exposed at the junction of the anterior and posterior two-thirds of the dural exposure. After it is ligated, two additional ties (*2*) should be placed around its anterior branch (*3*). The dura is then opened in a double trapdoor fashion. (E) see p. 131.



E

Figure 5.9. (E) The middle meningeal artery is illustrated exiting from the diploë, inferior to the pterion, and bridging the epidural space to enter the dura mater. The surgeon may choose not to transect it electively, but to extend the durotomy posterior to the middle meningeal proper. This is not recommended.

risk of an intraoperative epidural hematoma (which may result from tearing the middle meningeal artery as the dura falls from the inner table of the skull).

Occipital Dural Openings (Figs. 5.10, 5.11)

The medial and lateral occipital dural openings permit one, respectively, to work along the junction of the falx and the tentorium to reach the culmen monticuli and the pineal area, and to gain full access to the occipital pole.

Medial Occipital Dural Opening

In the medial occipital craniotomy the dura is opened in a double trapdoor fashion, bringing the line of incision parallel to the superior sagittal sinus, at a distance of approximately 3 cm from it. The incision is extended inferiorly to approximately 5 mm above the transverse sinus, the cut inferomedially to approximately 2 mm lateral to the torcular Herophili. This allows one to reflect the medial trapdoor over the transverse sinus. One may then approach the pineal region either along the falx or the tentorium, with the option to retract the occipital lobe superolaterally beneath the unopened lateral dural trapdoor. This affords it protection, under a dural covering, and keeps it from bulging through the craniotomy as retraction in the depths exposes the lesion.

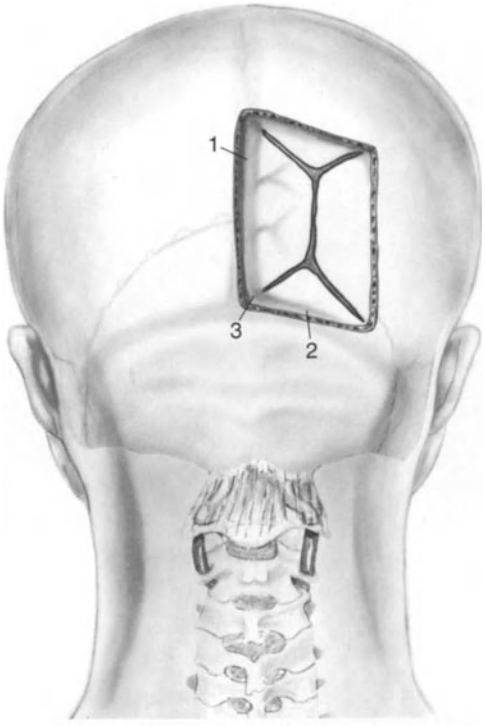


Figure 5.10. The dural opening is marked off, permitting reflection of a double trapdoor within the medial occipital craniotomy lines. Note that the superior sagittal (*1*) and transverse (*2*) sinuses are within the craniotomy, as is the torcular Herophili (*3*).

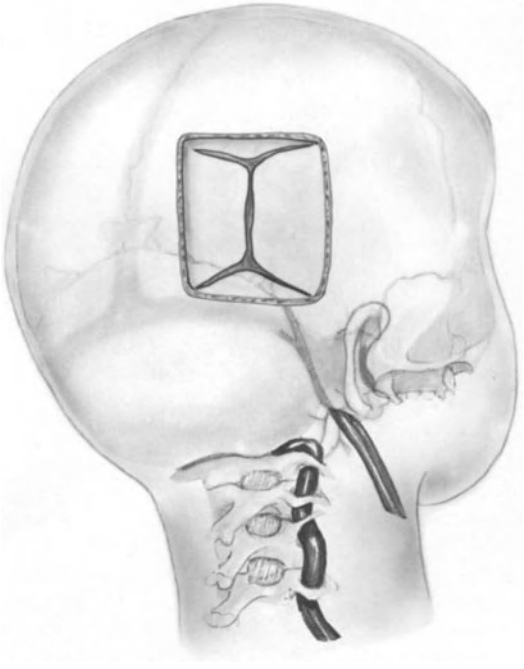
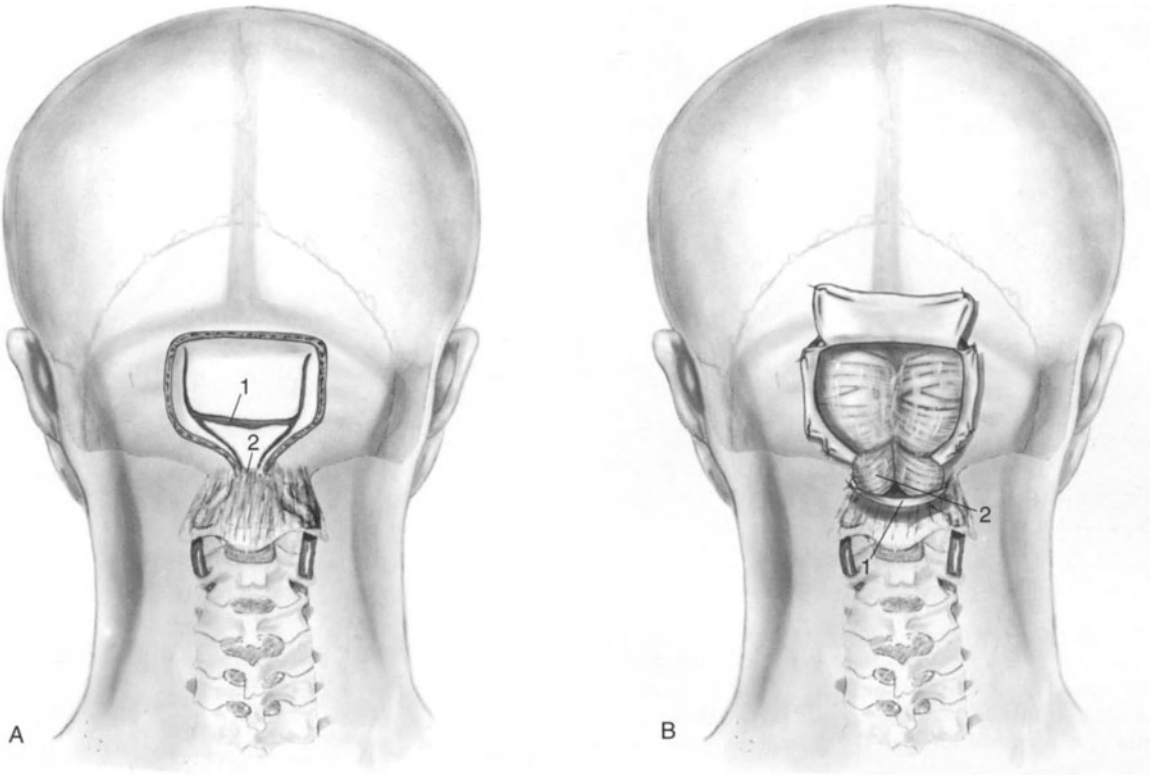


Figure 5.11. The lateral occipital craniotomy exposes dura but neither sinus nor, in most instances, major bridging cortical veins. One must, however, be cautions for there is occasionally a draining vein that goes from the tentorial surface of the occipital lobe into either the tentorium or the transverse sinus.



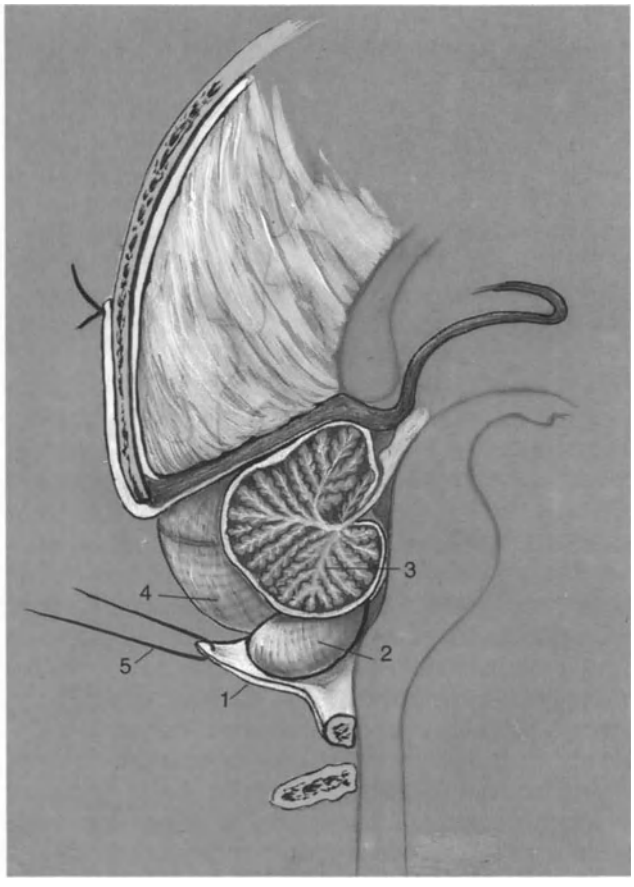


Figure 5.12. (A) Dural flaps for access to the inferior triangle. The lines of the two U-shaped dural flaps are indicated. This permits opening the dura so as to fashion a sling to suspend the cerebellar tonsils and minimize traction on the cerebellar peduncles. The horizontal dural cut (*1*) is placed well above the rim of the foramen magnum (*2*). (B) The dura is reflected laterally over each side and superiorly over the base of the triangular craniotomy. The sling (*1*) is shown supporting the tonsils (*2*) in the posterior fossa. This latter point is illustrated to much better advantage in C. (C) The sling (*1*) gives support to the tonsils (*2*) and, consequently, to the inferior cerebellar vermis (*3*) and medial portions of the cerebellar hemisphere (*4*). The ligatures suspending it (*5*) are sewn to the erector capiti muscle fascia.

Lateral Occipital Dural Opening

The lateral occipital flap is designed for access to the convex surface of the occipital lobe, not to the tentorium, falx, or either the transverse or superior sagittal sinuses. It is not adequate for medial occipital pole lesions.

Posterior Fossa: Suboccipital Dural Openings

The opening of the dura in the posterior fossa is both tedious and delicate because the cerebellar hemispheres rest snugly upon it when the child is in the lounging position. Also, dural sinuses are in the midline and immediately beneath the rim of the foramen magnum. They are variable in size and location. The dura covering the inferior cerebellar hemispheres may be converted into one sinus (especially in Chiari II children). There are medial and lateral suboccipital dural openings.

Medial (Midline) Suboccipital Dural Opening

(Figs. 5.12–5.14)

Medial suboccipital openings permit exposure of either the inferior or superior cerebellar triangles, with the former being used either for inferior vermian/IV ventricle lesions or for lesions within the foramen magnum, and the latter for superior vermian and pineal region tumors.

Inferior Cerebellar Triangle Dural Opening

Inferior cerebellar triangle exposure may be used for lesions within the IV ventricle and inferior cerebellar vermis, or for those extending across the foramen magnum. This distinction (inferior and superior cerebellar triangles) is of value since IV ventricle and vermian lesions, which are generally very large at the time of surgery, may be removed without opening the dura across the annular sinus and without extending the durotomy into the atlanto-occipital membrane. Thus, for inferior triangle lesions (within the inferior vermis or IV ventricle), careful placing of the durotomy incision allows the surgeon to fashion a sling from the inferior portion of the dura. Such a sling serves to suspend the inferior cerebellar hemispheres and tonsils, preventing them from herniating into the operative field, and to avoid potentially damaging traction on the brainstem.

The dura is cut inferomedially from the point of opening (which is best made at approximately the junction of the superior and inferior thirds of the dura exposed by the suboccipital craniotomy). A cut across the midline from right to left, connecting the two inferomedially coursing durotomy incisions, is made with tenotomy scissors. The cut across the midline may have to be made with heavier scissors because of the dural reduplication at the annular (marginal) and cerebellar sinuses.

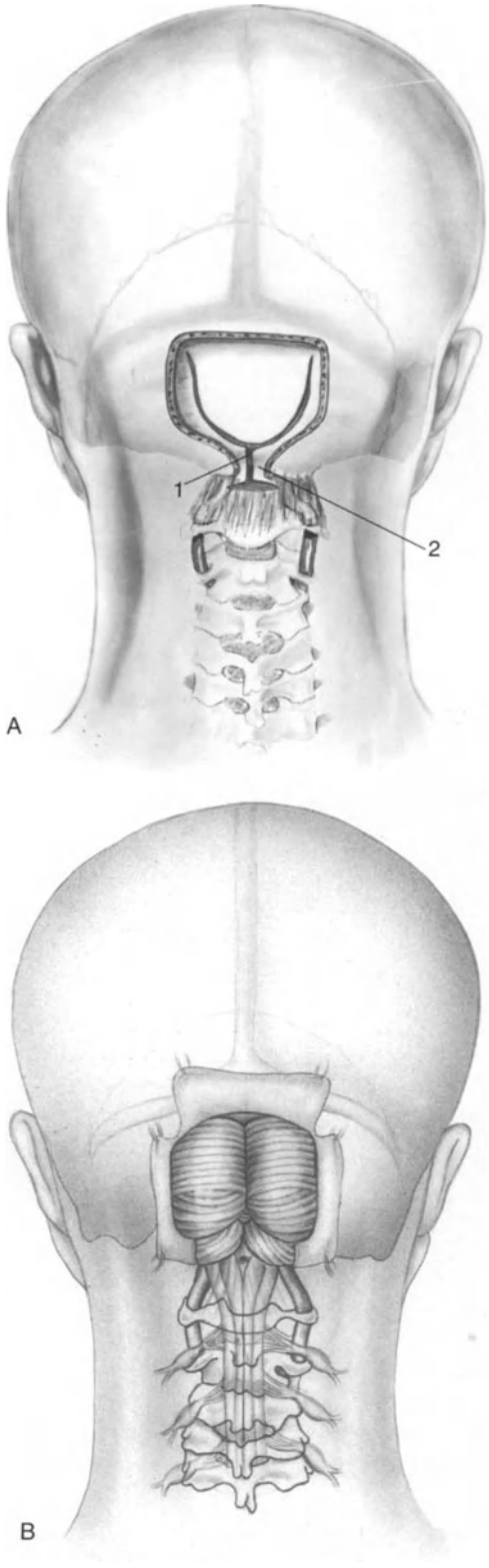


Figure 5.13. (A) This is the recommended durotomy for lesions within, or extending into, the foramen magnum. There is a vertical durotomy (*1*) across the level of the foramen magnum (*2*). (B) The dura is reflected superiorly over the craniotomy edges and inferiorly over the arch of C-1. This exposes completely the tonsils, inferior vermis, and cisterna magna.

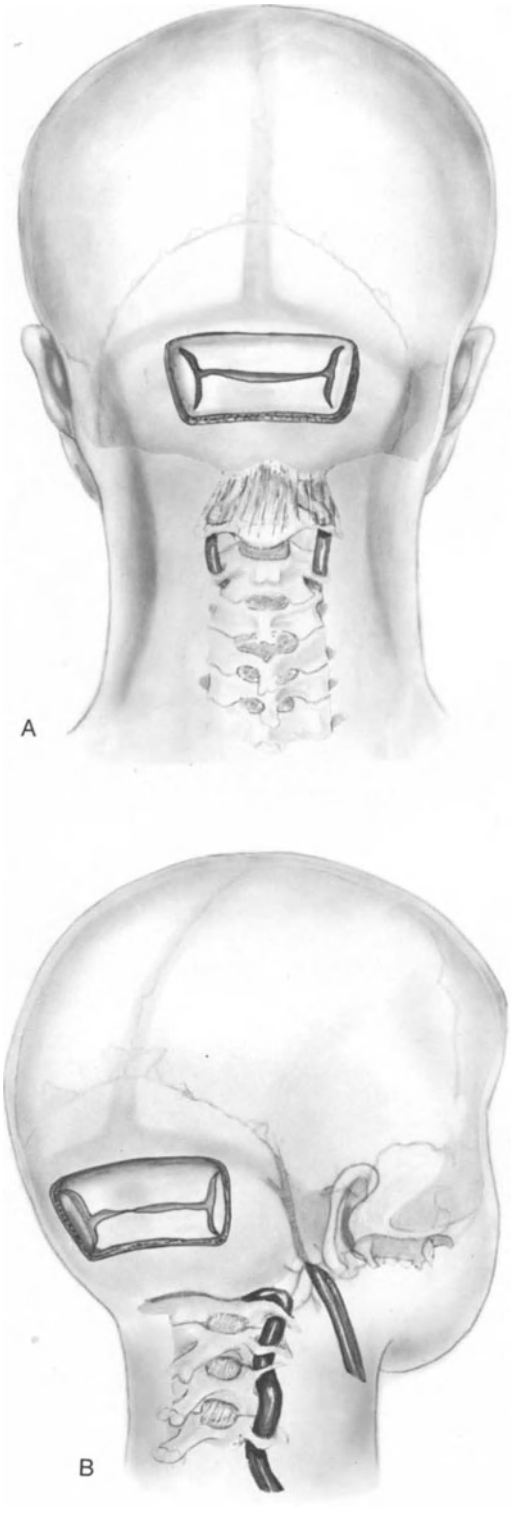


Figure 5.14. The double trapdoor durotomy for access to the superior cerebellar triangle permits the surgeon the option of reflecting only the superior trapdoor, or both. (A) The durotomy lines are depicted for complete exposure of the superior cerebellar triangle. (B) Postero-oblique perspective of the double trapdoor durotomy.

If a dural sinus is encountered it may either be coagulated with the bipolar forceps or clipped. This latter technique is discouraged, since the clips may come dislodged at the time of closure. The result may be bleeding into the posterior fossa, which, unfortunately, may occur without the surgeon being able to identify the source. Also, and of more immediate danger, opening dural sinuses subjects the child to air embolism. Incomplete closure (often the case when clips are used) of these dural sinuses may present continued risk of air embolism throughout the operative procedure.

Opening of the dura over *the craniovertebral junction for inferior triangle lesions at the foramen magnum* entails fashioning a U-shaped dural flap and convexity pointing inferiorly, beginning at the superolateral border on each side of the craniotomy. It should be extended inferomedially to approximately 0.5 cm above the dural fold located just beneath the posterior arch of the foramen magnum, bilaterally. Then, a midline durotomy is cut vertically across the foramen magnum, coagulating the annular sinus with a bipolar forceps on either side of the cut. The spinal dura (there is only one layer) is then cut horizontally, first to one side and then to the other, immediately above the arch of C-1. (It is almost never necessary to remove the arch of C-1 in order to gain complete access to the structures within the foramen magnum. In rare circumstances the tumor extends beneath the level of C-1, at which time an osteotomy at either lateral extremity of the arch of C-1 allows the surgeon to displace this arch inferiorly and, thus, extend the dural incision almost to the level of C-2). The bilaterally osteotomized but otherwise very much intact arch of C-1 may then be anchored back into anatomical position at the end of the procedure, set back simply into its periosteal envelope.

Superior Cerebellar Triangle Dural Opening

Superior cerebellar triangle exposure and access to the pineal region, through a quadrilateral craniotomy flap, is attained by a double trapdoor durotomy. This permits one to reflect the upper trapdoor superiorly, if the culmen monticuli and pineal region are the target area(s), and the lower trapdoor inferiorly, if the superior portion of the cerebellar hemispheres is the target area.

Lateral Suboccipital Opening (Fig. 5.15)

For access to the lateral cerebellar hemisphere or the pontocerebellar angle, a double trapdoor durotomy, with the superior trapdoor being larger than the inferior, is reflected. The advantages of a larger superior segment rest in ease of identification of the superior cerebellar veins and the lateral sinus to which they are tributary.

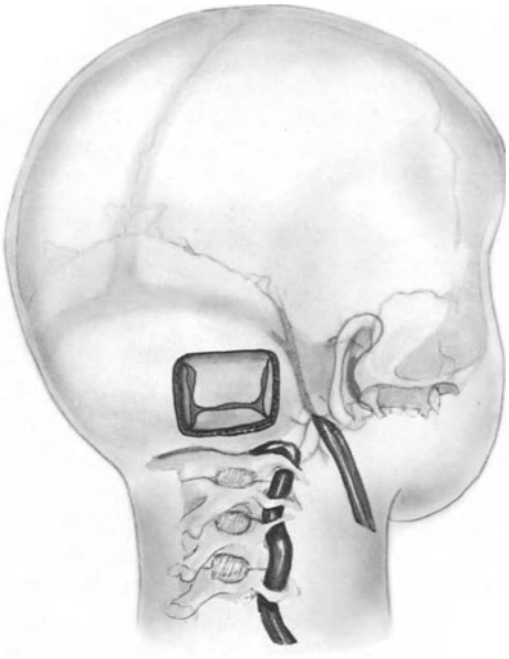


Figure 5.15. This is an asymmetrical double trapdoor durotomy flap. The transverse sinus has not been exposed.

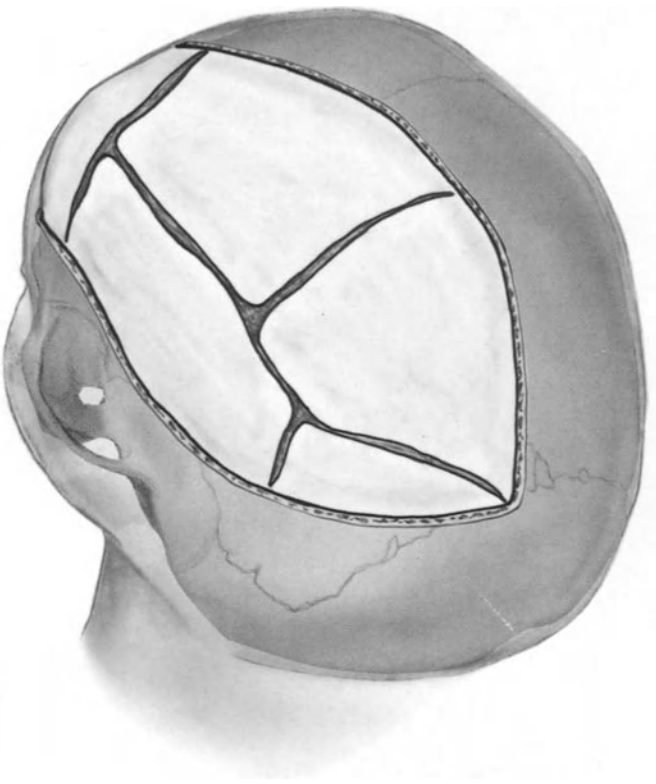


Figure 5.16. Hemispherical dural opening.

Hemispherical Dural Opening (Fig. 5.16)

The dural opening for hemispherectomy must be such as to permit access to the superior sagittal sinus, the torcular Herophili and the transverse sinus, the tentorial surface and greater wing of the sphenoid (floor of the middle fossa), and the orbital roof.

Spinal Dural Openings (Figs. 5.17–5.20)

The spinal dura mater differs from the cranial dura mater in having:

1. A layer of epidural fat between the dura and the bony portion of the spinal canal
2. A large plexus of epidural veins (Batson's plexus) freely communicating with the intra-adbominal and intrathoracic venous systems, extending the full length of the spinal cord
3. The dura mater composed of only one layer, since the periosteal component of the outer layer of the cranial dura mater becomes the periosteum of the external table of the skull as it remains adherent to the skull at all cranial foramina, canals, and fissures
4. A single, enormous cistern, rather than compartmentalized subarachnoid-trabecular compartments, surrounding the spinal cord from the cisterna magna to the spinal cul-de-sac.

Of singular importance – as an hydraulically buffered CSF pressure maintenance system – is the interconnecting venous plexus from the sacral canal inferiorly to the jugular foramen superolaterally and the foramen magnum superomedially (Batson's plexus), especially in light of the extensive plexiform anastomoses between this plexus and the retropleural and retroperitoneal...-major...and minor veins. Coughing, straining at stool, the erect posture, sighs, gastrointestinal distention, constipation, inferior vena cava or portal system occlusion, superior vena cava or jugular vein occlusion, all cause dilation of Batson's plexus first and increased spinal CSF pressure secondarily. There is a reciprocal pressure interplay between intra-abdominal/intrathoracic pressure and spinal subarachnoid CSF pressure. This assures particular importance with regard to intraventricular CSF and intra-abdominal pressure differences in patients with V-P shunts.

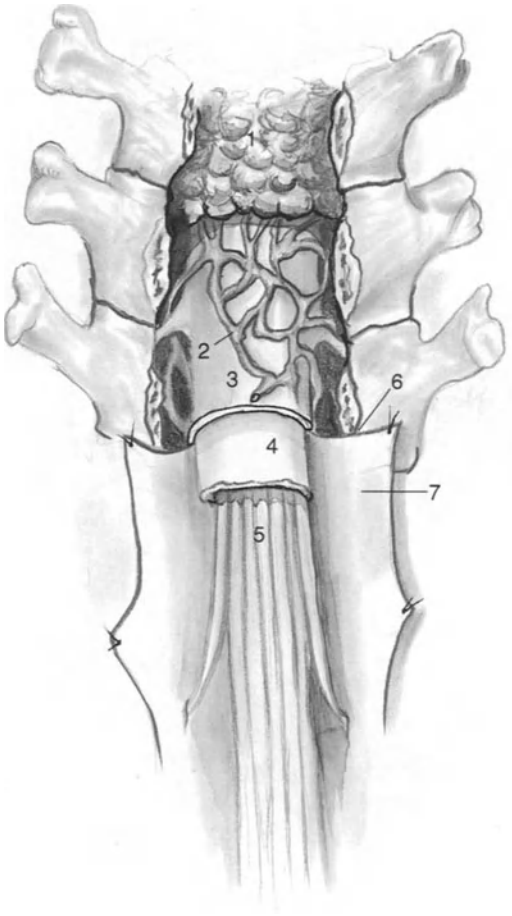


Figure 5.17. The spinal canal contains, from peripheral to central, epidural fat (*1*), epidural venous plexus (*2*), a single layer of dura (*3*), a single arachnoid membrane without compartmentalized subarachnoid chambers (*4*), and then either the spinal cord or the cauda equina (*5*). The dura mater has been cut in the midsagittal plane and then circumferentially perpendicular to this (*6*) before reflecting the dural flap (*7*).

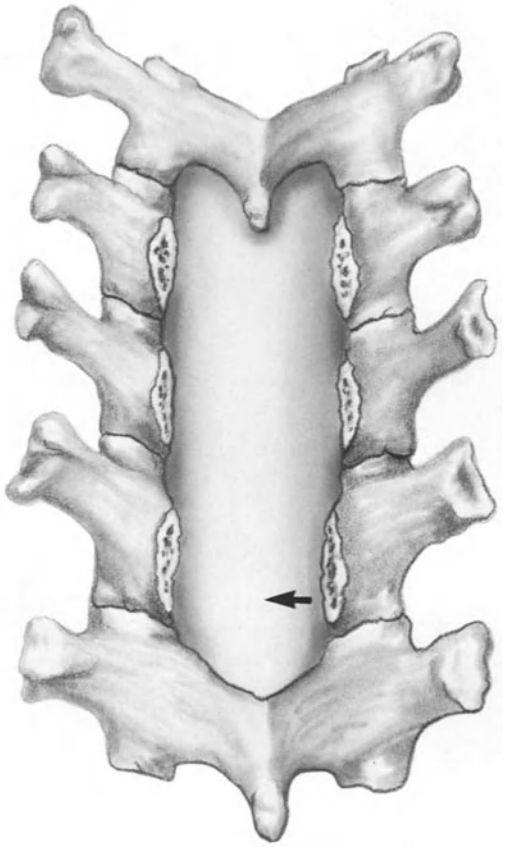


Figure 5.18. The epidural compartment has been cleared of fat and the venous plexus. Expansion of the intradural contents distends the (single layer) dural sac (*arrow*) to fill the spinal canal after the epidural veins and fat have been removed.

After the laminotomy has been performed, the surgeon comes upon the layers of epidural fat and veins. These are taken away by coagulation, cutting, and brushing movements with a wet fluffy cotton.

Once the epidural fat and veins have been cleared, the dura is noted to be relatively pale in appearance and to expand to fill the entire spinal canal, taking up the space left by the dissected fat and veins. This, no doubt, is an expression of the reciprocal volumetric relationship between the subarachnoid fluid and the epidural venous system.

The exposed dura is ready for incision. The scleral hook is used to pick up the dura, a #15 blade to incise it. It is recomended that the dura *not be opened only in the midsagittal plane*, but rather that cuts perpendicular to the midsagittal opening be extended from both the superior and inferior extremities of the durotomy. This technique assures minimal damage to the spinal cord. *In essence, if a simple linear, sagittal, incision is made in the dura and the dura is then either sewn back or retracted, it will suspend the spinal cord, elevating it from its resting place within the ventral portion of the bony spinal canal.* On the other hand, if perpendicular cuts are put in the dura at the upper and lower extremities of the sagittal durotomy, the result is a bilateral trapdoor dural opening. No suspension of the spinal cord or elevation of it from its bed at the ventral aspect of the bony spinal canal results.

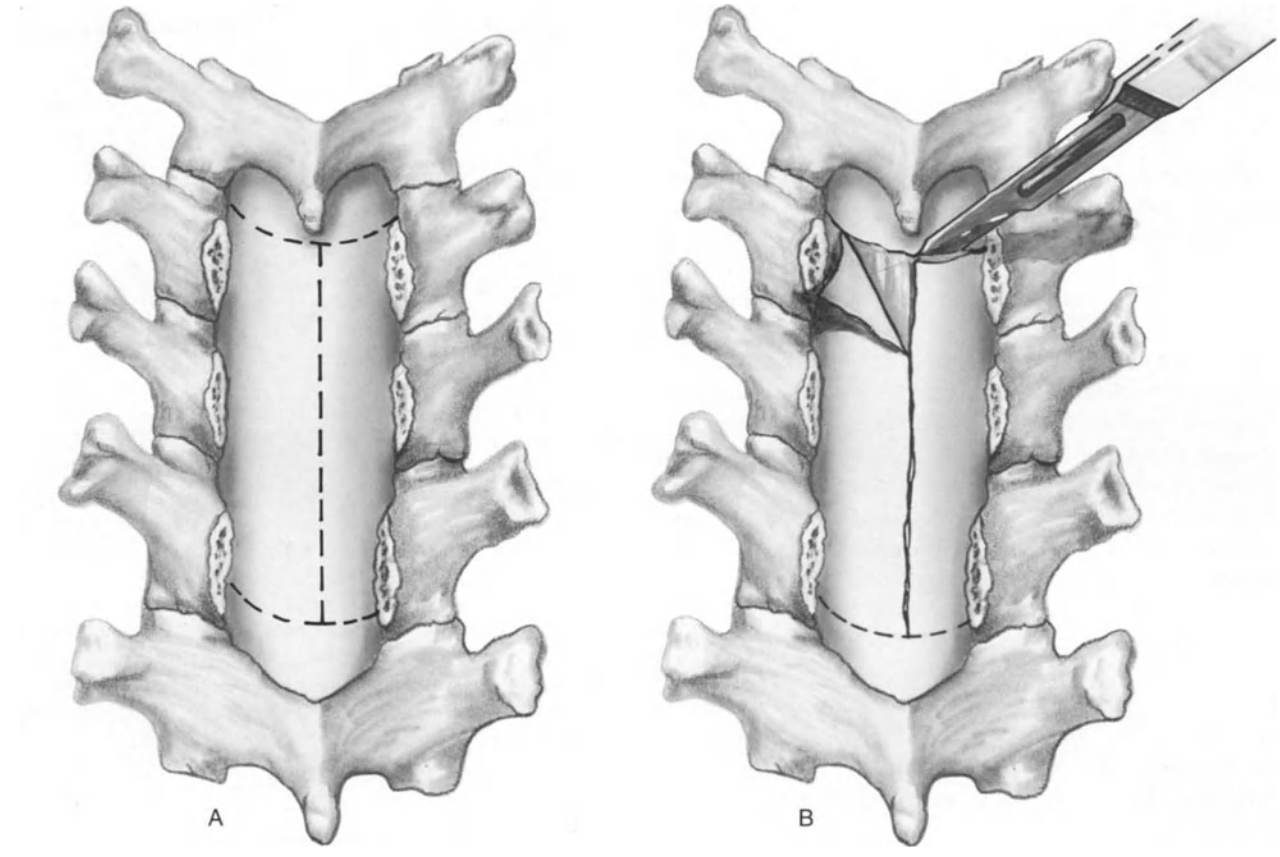


Figure 5.19. (A) The area of dural incision is marked off to show the durotomy lines. (B) The sagittal durotomy has been made and the perpendicular cuts are being extended. One may use either a #15 blade or a tenotomy scissors. Note the lateral reflection of the trapdoor flap *on the reader's left*, a technique that avoids "suspension" and possible damage to the spinal cord.

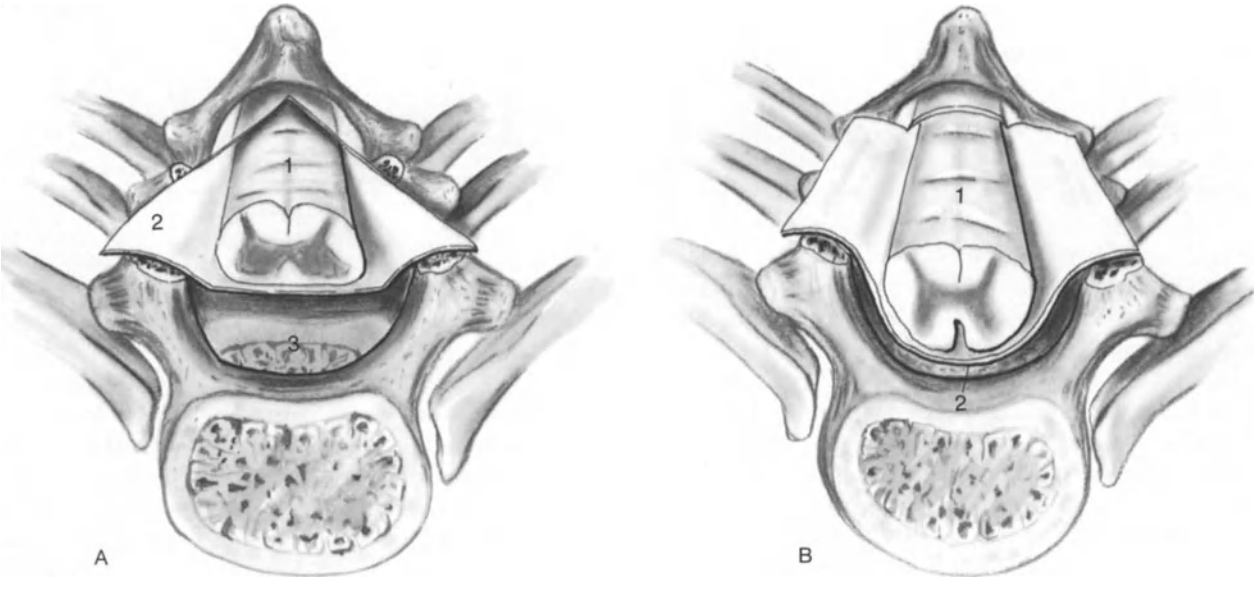


Figure 5.20. (A) Midsagittal durotomy only. A horizontal cut across the ventral aspect of the bony spinal canal, illustrating the suspension of the spinal cord (*1*) by the tented dura (*2*), thus lifting it from the ventral portion of the spinal canal (*3*). This is a potential cause of spinal cord damage. (B) With a double trapdoor durotomy incision and flap, as illustrated, the spinal cord (*1*) rests within its bed in the ventral aspect of the bony spinal canal; (*2*) durotomies made on both the right and left.

Closure

Cranial Closure

Dural Closure (Figs. 5.21, 5.22)

The dural closure is greatly facilitated when the dura has been kept stretched – by having sewn it back tautly – throughout the operative procedure (*from the moment of the dural opening*), and by keeping it moist so that it does not shrivel during the hours of intracranial surgery.

The dura is closed with 4–0, interrupted, nonabsorbable sutures. Absorbable sutures could theoretically increase the risk of cortical scarring. Continuous stitches, if used, should be locked to favor a "watertight" closure (however, some surgeons argue that noninterlocking sutures more reliably assure a watertight closure, sustaining that the overlapping of the dural edges provides a greater surface area for wound healing). One should place sutures, alternatively, in each limb of the dural flap, rather than beginning at one limb and working around to the other. This assures stretching the dural flap evenly between the two limbs. Also, if the sutures are put into the anchored end of the dural flap at 5-mm intervals and then into the free end of the flap at 4-mm intervals, the free end will be stretched gradually into place.

Beginning the closure with anchoring sutures at the corners, or at the center of the flap, is to be avoided because it puts the dura on an extreme stretch suddenly and permits the brain to bulge through the intervening openings.

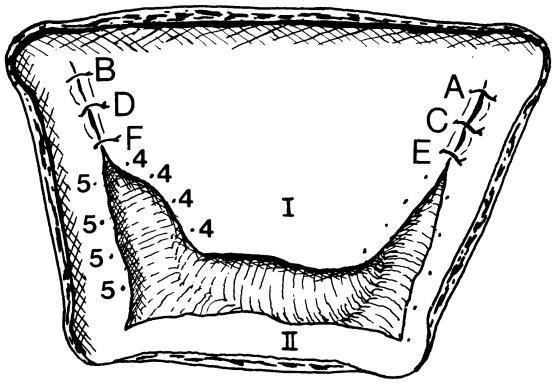


Figure 5.21. Opposite borders of the dural defect are used as initial anchoring sites. The free edge of the dural flap (*I*) is that which is medial to the dural opening; the anchored edge (*II*) is between the dural opening and the bone. Sewing the dura short, along the free edge and long on the anchored edge, with stitches alternating from limb to limb, stretches it out evenly and gradually, assuring uniform closure. In this drawing *the letters* indicate sequence of suture placement, *the arabic numerals* indicate number of millimeters between sutures on each side. This allows one to envision a gradual ironing out of the free dural edge as the closure proceeds with sutures at A, B, C, D, etc., and at 5-mm intervals along the anchored edge, but with 4-mm intervals along the free edge.

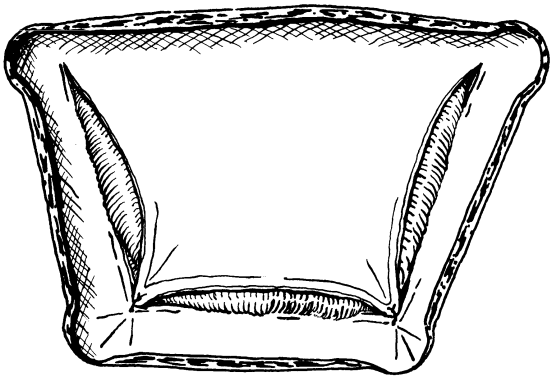


Figure 5.22. Beginning the dural closure by placing sutures at the corners as illustrated here is to be avoided, except in those rare instances when there is an abdundant subdural space and the dura is, for all intents and purposes, redundant.

Use of the Periosteum and Fascia to Reconstruct the Dura (Fig. 5.23)

When it is not possible to close the dura, or doing it demands inordinate stretching, insertion of a periosteal graft is preferable. The periosteal graft is taken from the periosteum of the bone flap for intracranial dural repair; fascia lata is used to repair spinal dura. For this reason, when the cranial bone flap is reflected, care should be taken to keep the periosteum and the bone moist. One should moisten the dural flap periodically throughout the surgery. Abdominal fascia lata or erector spinae muscle fascia suffice as donor sources for spinal dura repair.

After the intracranial periosteal graft has been fashioned, it is removed from the skull simply by lifting it away with an Adson-Brown forceps and using a Penfield #4 dissector to separate it cleanly and uniformly from the skull. It should be taken in one piece, anchored at two ends to the opposite borders of the dural defect, and then tied down at either end. Sutures are used to bring the dural graft into apposition with the remainder of the edges of the dural defect.

In the posterior fossa it is not often possible to find an adequate amount of periosteum, so a graft from the abdominal fascia lata or erector capitae muscles is used. The procedure for fashioning, anchoring, and attaining a watertight closure is the same. One is advised to take care to bring the fascial surface of the graft to border on the surface of the cerebellum or the cisterna magna, since the muscular surface may form adhesions between the dura and the cerebellum. The same technique and precautions apply to the use of fascial grafts for repair of the spinal dura [1–3].

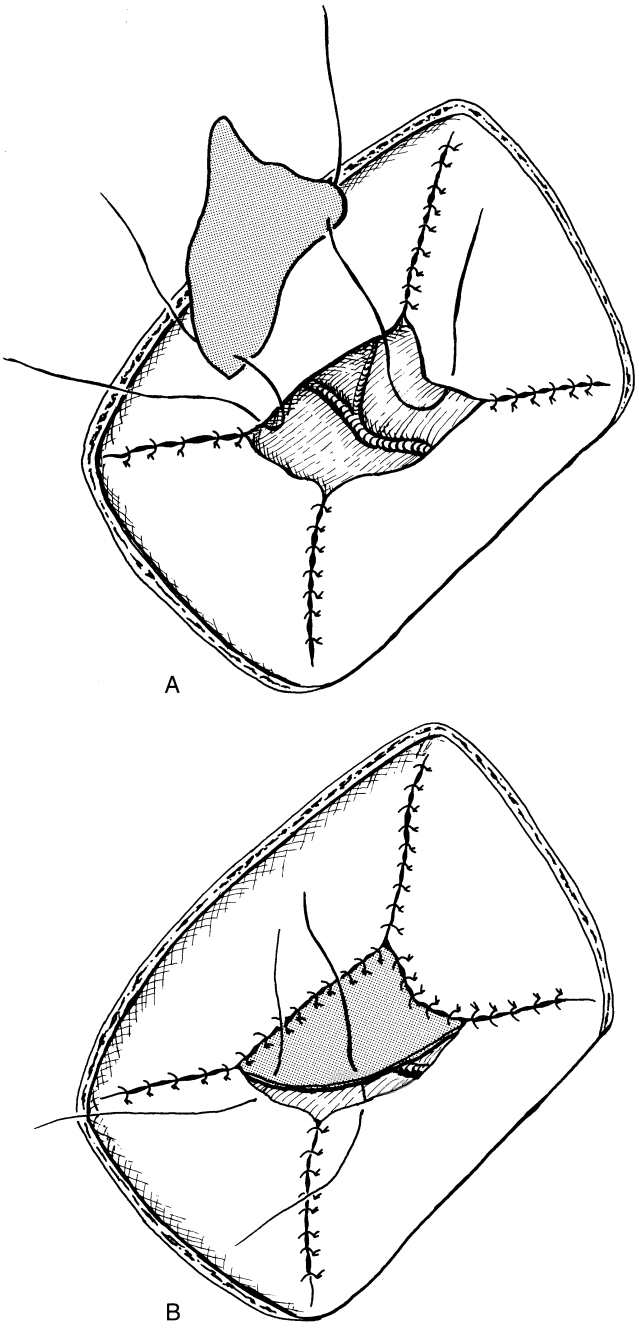


Figure 5.23. (A) Periosteal graft to cover dural defect. Opposite borders of the dural defect are used as initial anchoring sites. (B) 4–0 interrupted sutures are used to attain a watertight closure between graft and dural edges.

Spinal Closure

Arachnoid Closure

The closure of the arachnoid [1] is recommended in spinal cord tumor surgery, and is essential for syringomyelia when the syrinx is marsupialized to the spinal subarachnoid space. It may be closed with either interrupted or continuous 6–0 sutures.

Adequate closure of the arachnoid [1] is guaranteed at the time of opening, since at this time the surgeon has the opportunity to identify it clearly, before placing guide sutures, 6–0 or 7–0, so as to tent it. If the sutures are lain in at 4- or 5-mm intervals on either side of the planned line of arachnoid incision, one may make the line of incision while drawing on these guide sutures, using them for retraction sutures after the arachnoid has been opened. They may then be brought medialward and tied to one another at the beginning of the arachnoid closure.

Dural Closure

The dura is closed with interrupted 4–0 sutures in the same manner as the dura over the brain, but remembering that the spinal cord dura [2, 3] consists of only one layer and, consequently, that it is more easily frayed. If, as is so often the case, one does not get an adequate, watertight closure, it is advisable to insert a fascial patch graft. Dural cerebrospinal fluid leaks may compromise wound healing either by resulting in a collection of subcutaneous cerebrospinal fluid (seroma), or by leaking through the skin incision. Both necessitate reoperation, which increases the risk of infection. Consequently, dural grafts should be inserted without hesitation, at the slightest suspicion that the closure may not be watertight. Surgicel or glues are not reliable if used alone.

References

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2. Raimondi AJ, Gutierrez FA (1979) Reconstruction of the posterior vertebral arch and laminotomy for intraspinal surgery. In: Ransohoff J (ed) Modern technics in surgery – neurosurgery. Futura, Mount Kisco, NY, pp 10–11
3. Raimondi AJ, Gutierrez FA, DiRocco C (1976) Laminotomy and total reconstruction of the posterior spinal arch for spinal canal surgery in childhood. J Neurosurg 45:555–560

6 Cerebral Retraction

"What is history, but a fable agreed upon!"
NAPOLEON BONAPARTE, Comments

Cistern Openings (Figs. 6.1–6.13)

Approximately 1300 years passed between the time that Galen first described the cerebral cisterns, concluding that they represented an interval between the pachymeninges and the leptomeninges, and the time Varoli first described the arachnoid membrane. Another 100 years were to pass before this membrane was named (Vesalius, ca. 1670) and another 145 years before Cotugno (ca. 1815) discovered the cerebrospinal fluid and identified the subarachnoid spaces. Magendie, in 1827 [1], described cerebral cisterns, which he called confluents. The identification of subdural spaces, however, was not made until Key and Retzius [2] published their work in 1875.

Since the pia is densely adherent to the glia, there is no subpial space. Conversely, since the arachnoid rests upon (but is not adherent to!) the dura mater, and is connected to the pia by trabeculae, there is a subarachnoid space which varies in presence and volume: where arachnoid is adjacent to pia, as over the hemispherical gyri and cerebellar folia, there is a microscopic subarachnoid space, but where the arachnoid and pia are separated by lesser or greater distances (the sulci and intervals between lobes or over the brainstem) cisterns are formed. The cisterns and subarachnoid spaces are in free communication with one another.

The subarachnoid spaces are not named, but there is a nomenclature of the cerebral cisterns, taken from the major anatomical structure upon which they border. These are, from anterosuperior to posteroinferior and then around the cerebellum and corpus callosum: sylvian, lamina terminalis, chiasmatic, ambient, interpeduncular, pontine, medullary, magna, superior cerebellar, quadrigeminal, cerebellopontine, transverse fissure, and pericallosal.

Exposure, opening, of the cistern, consequently, is attained by cutting the arachnoid with a pair of microscissors, or "burning" one's way through the arachnoidal membrane with microbipolar forceps. Once the arachnoid has been breached, cerebrospinal fluid emits, and one is within the cistern. The remainder of the cisternal wall, invariably the arachnoidal membrane, is cut open with microscissors, taking care not to damage the cisternal arteries or veins. It is not important to cut the arachnoidal membrane along the border of the cistern, where it becomes adjacent to the pia. One should take the arachnoid from the subarachnoid vessels to which it may be adherent, since pulling upon it during the operative procedure may damage these vascular structures. It is important to remember that all of the cranial nerves course within cisterns and that, consequently, these should be identified and protected by covering them with Telfa or fluffy cotton immediately the cistern is opened. This prevents inadvertent damage to the cranial nerve by mechanical compression or aspiration into the sucker.

Use of Gravity

The least traumatic and most efficient technique for achieving cerebral retraction is the use of gravity (to allow the cerebrum to fall away from the desired area), assisting this by giving egress to cerebrospinal fluid from sulci or cisterns. This technique is employed when operating on the parasellar or sylvian fissures, the ambient cistern, and ventricular, pineal, or posterior fossa areas.

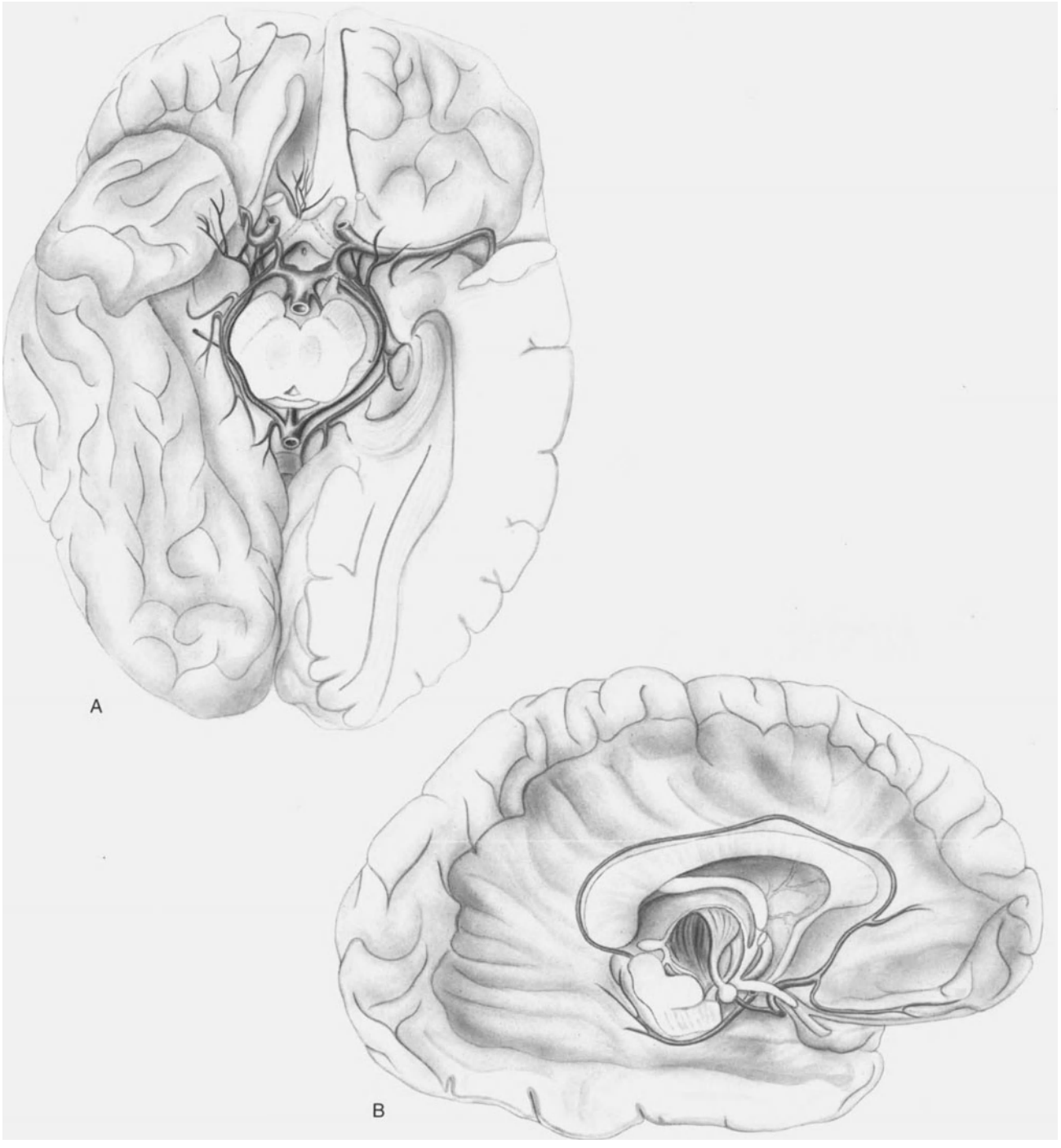
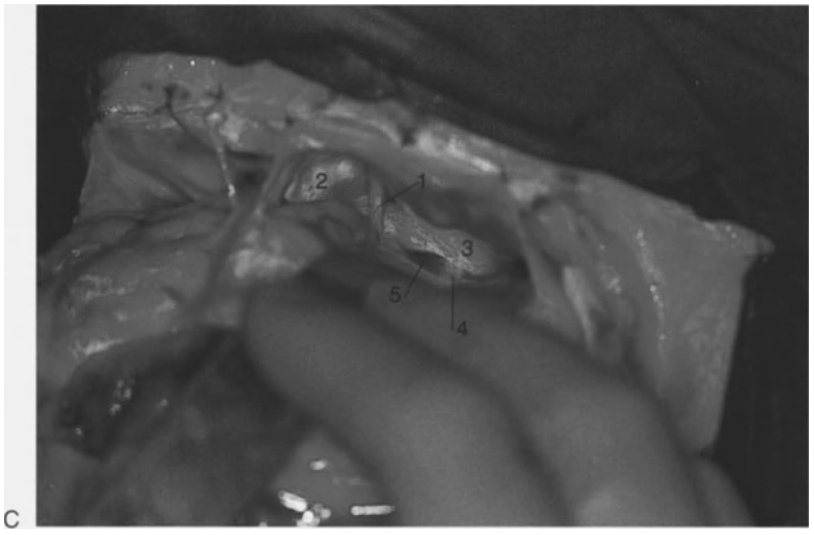


Figure 6.1. (A) This drawing of the surfaces of the cerebral hemispheres which abut upon the anterior fossa, the middle fossa, and the tentorium, extending from the petrous ridges to the occipital bones, places into relief the relative positions of the first two cranial nerves vis-à-vis one another, the frontal lobe, the inferior surface of the frontal lobe and the medial surface of the temporal lobe, the anterior perforated substance, and the *full* circle of Willis. Parasellar surgery must take into consideration the posterior communicating/posterior cerebral basilar/posterior cerebral and origin of vein of Rosenthal systems. For the purposes of this section, emphasis is put upon the olfactory nerve and trigone, the anterior perforating substance, the special relationships between the latter and the bifurcation of the internal carotid artery, and the anatomical juxtapositioning of the trunk of the olfactory nerve and the optic chiasm. (B) Sagittal drawing with olfactory nerve/frontobasal artery, optic chiasm/A-1, A-2/subcallosal gyrus of the frontal lobe, which is contiguous with the lamina terminalis. (C, D) see p. 143.



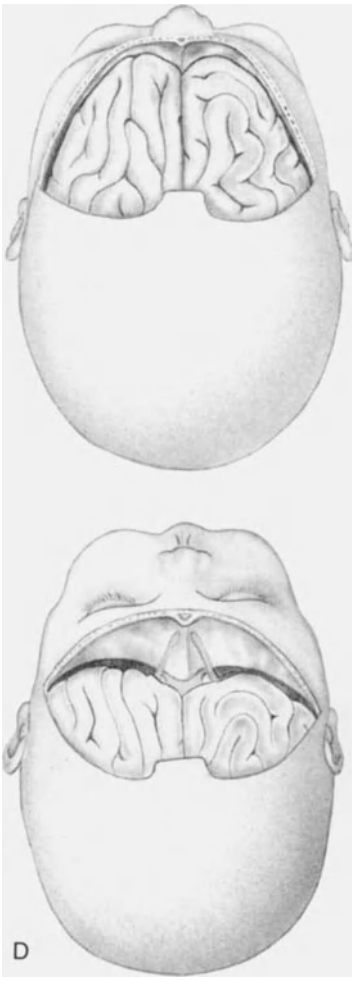


Figure 6.1. (C) Olfactory, prechiasmatic, and optic cisterns. The dura has been opened and the right frontal lobe is being retracted, exposing the right olfactory nerve (*1*) after the cistern has been opened with bipolar forceps and the arachnoid incised. The crista galli (*2*), anterior clinoid (*3*), and optic nerve (*4*) are exposed. The *olfactory cistern*, around the olfactory nerve, has been opened. The *prechiasmatic cistern* (*5*) is still closed, as is the *optic cistern*. (D) The remarkably unlimited perspective the surgeon has of the olfactory, optic, and arterial structures when the frontal lobes have been allowed to fall away from the inner surface of the frontal bone.

Parasellar Area

With the child's head hyperextended, through a bifrontal craniotomy for approach to the parasellar area, the cerebral hemispheres gradually settle inferiorly and slightly posteriorly, exposing the roofs of the orbits first and then, with time, the lesser wings of the sphenoid. The frontal lobes in the newborn, infant, and even the toddler are quite small; consequently, the anterior fossa is shallow and the distance from the supraorbital ridge to the lesser wing of the sphenoid is not nearly as great as it is in the adolescent or adult. Transection of the olfactory nerve(s) and opening of the prechiasmatic and sylvian cisterns allows egress of cerebrospinal fluid. This is gradually aspirated as the downward displacement of the brain is increased, giving the surgeon an excellent view of the entire parasellar area.

Sylvian Fissure

The use of gravity and drainage of cerebrospinal fluid for exposure of structures in and around the *sylvian fissure* is accomplished by proper positioning of temporal flaps. The head should be on a higher horizontal plane than the heart, but with the sagittal plane of the skull in the coronal plane of the body. The vertex should be dropped slightly so as to produce an approximately 10° angle of inclination from the mandible to the vertex, with the latter being lower. This provides for the hemisphere to gravitate toward the vertex, bringing the sylvian fissure into the surgeon's view. Opening of the sylvian fissure with microdissection of the arachnoid that borders it allows egress of the cerebrospinal fluid and augments the exposure of lesions in this area. For access to lesions medial to the trifurcation of the middle cerebral artery, this fissure should be opened from lateral to medial; for lesions at the trifurcation, from medial to lateral. This use of direction of cistern opening always keeps the normal anatomy proximal to the pathology.

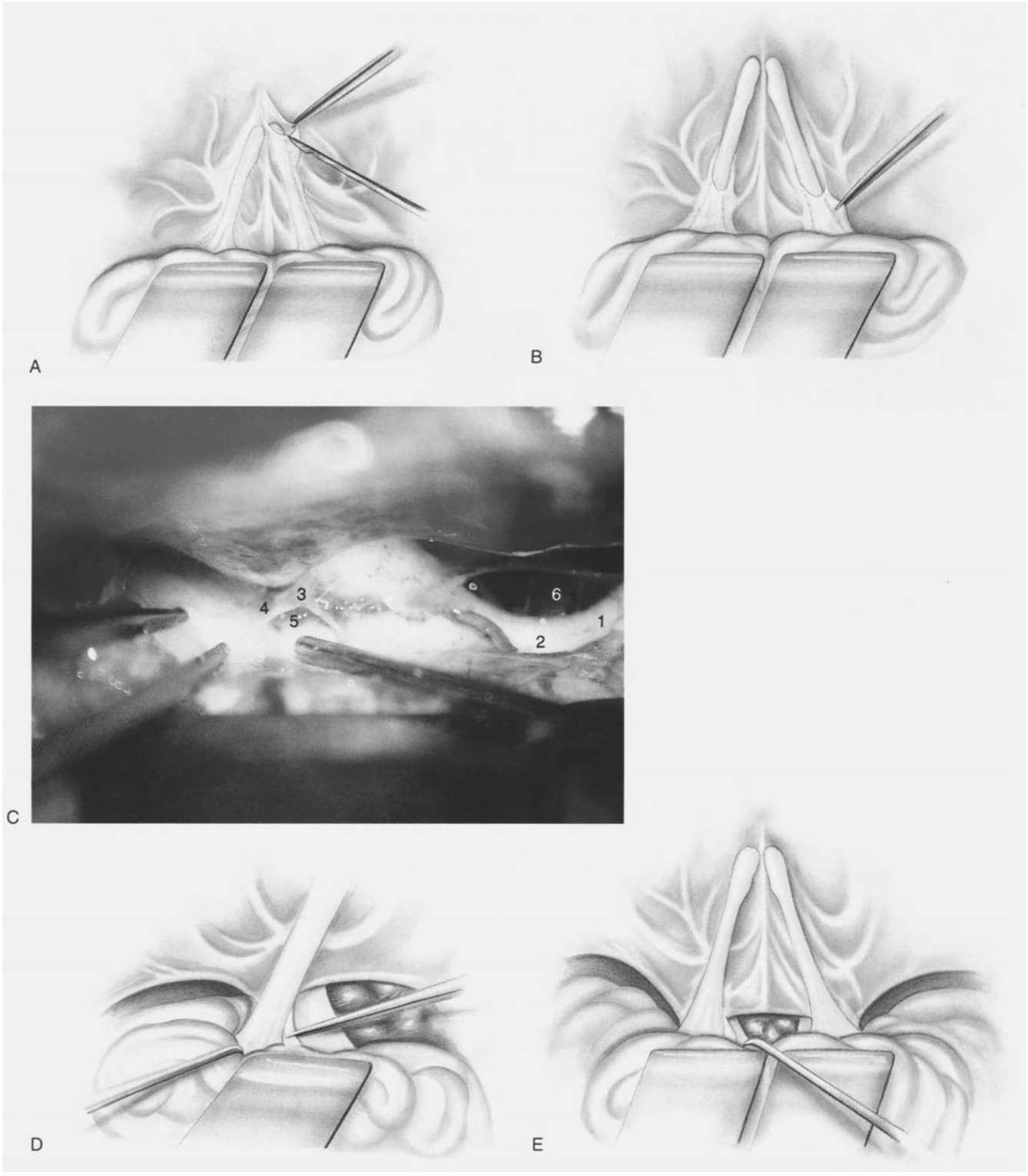
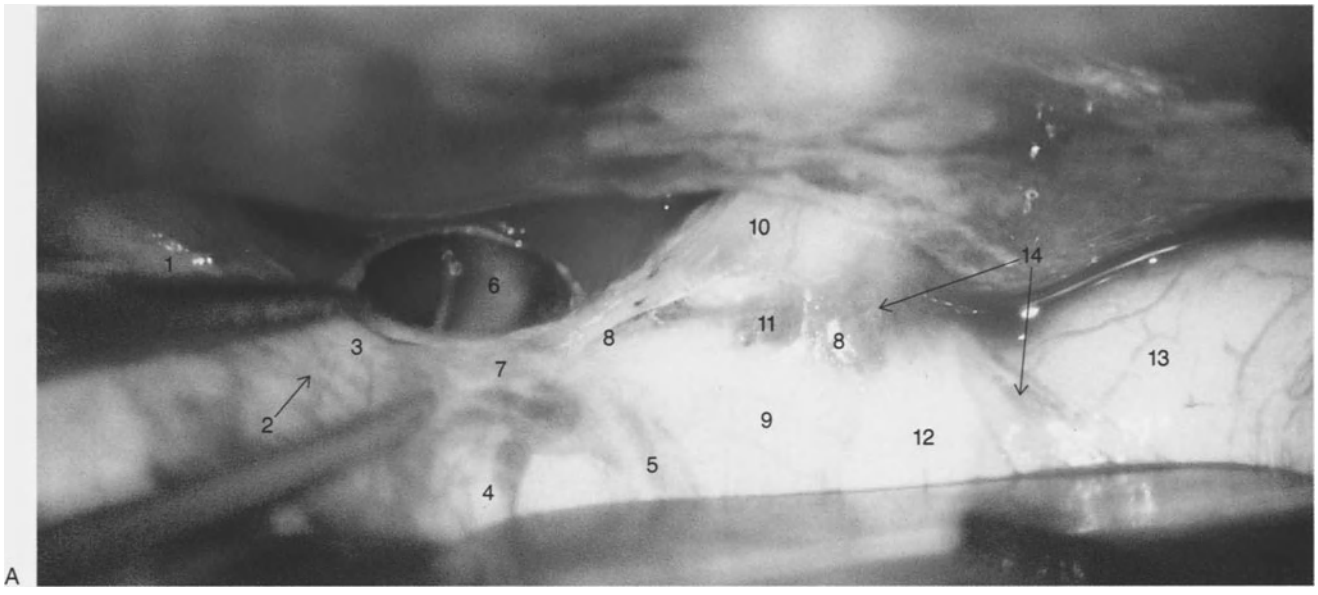


Figure 6.2. The series of drawings and the single intraoperative photograph in this figure illustrate the dissection used by some surgeons to conserve the olfactory nerve. I have performed this dissection many times, conserving the nerves anatomically and without evident signs of devascularization. However, I have not been successful in conserving olfactory function. In (A) the arachnoid from around the olfactory bulb is open with microdissection. The bulbs are opened one at a time. (B) illustrates dissection of the bulbs to the line of the posterior third of the olfactory nerve, a rather easy dissection, and one which proceeds rapidly up to this point. (*Continued* on p. 145).

Figure 6.3. Interopticocarotid cistern. The planum sphenoidale (*1*), anterior clinoid (*2*), and sphenoparietal vein (*3*) are shown. The arachnoid from over the *prechiasmatic cistern* (*4*) has been opened (5), permitting removal of the arachnoid from over the right optic nerve (*6*) and the internal carotid artery (*7*). This allows one to separate the optic nerve from the internal carotid artery, entering the *interopticocarotid cistern* (*8*).

◀ **Figure 6.2.** From here on, however, the dissection is quite difficult in that one must not only open the lateral and medial surfaces of the olfactory cistern, but also separate the arachnoid, which runs along the dorsal surface of the olfactory nerve from the arachnoid of the basal surface of the frontal lobe. In doing this, it is best to remain within the olfactory cistern, allowing the frontal lobe to fall away gradually as one proceeds inferolaterally. (C) The right optic nerve (*1*), optic chiasm (*2*), and arachnoid covering the left optic nerve (*3*) are well shown, as is the arachnoid bridging from the left optic nerve to the left temporal lobe (*4*). This arachnoid, along the interval between the optic nerve and the temporal lobe, has been coagulated with bipolar forceps opening into the subarachnoid space (*5*), thus providing entry for one of the blades of the microscissors so that the *optic cistern* may be opened, exposing the underlying optic nerve and the internal carotid artery immediately beneath it. The prechiasmatic cistern (*6*) has been opened, exposing the pituitary stalk. (D) After the arachnoid of the olfactory cistern has been separated from the lateral and superior surfaces of the olfactory nerve, to the olfactory trigone, the arachnoid from the medial surface of this nerve is then cut away, so as to expose the cistern of the optic nerve and then the interval between the olfactory and optic nerves. (E) With the dissection completed on both sides, one has a complete view of the interoptical component of a prechiasmatic craniopharyngioma, or access to the infrachiasmatic compartment. Access to the lamina terminalis is never a problem. However, unimpeded visualization and a work area between the ventral surface of the olfactory nerve and the opticocarotid space do not exist, so that conserving anatomically the olfactory nerves in effect is an exchange for not using the interopticocarotid space and not having full access to the posterior circle.

Figure 6.4. Chiasmatic cistern. Both optic nerves (*1, 2*) are exposed after the *prechiasmatic and optic cisterns* have been opened, allowing one to expose the optic chiasm (*3*) after lifting the arachnoid from over it.



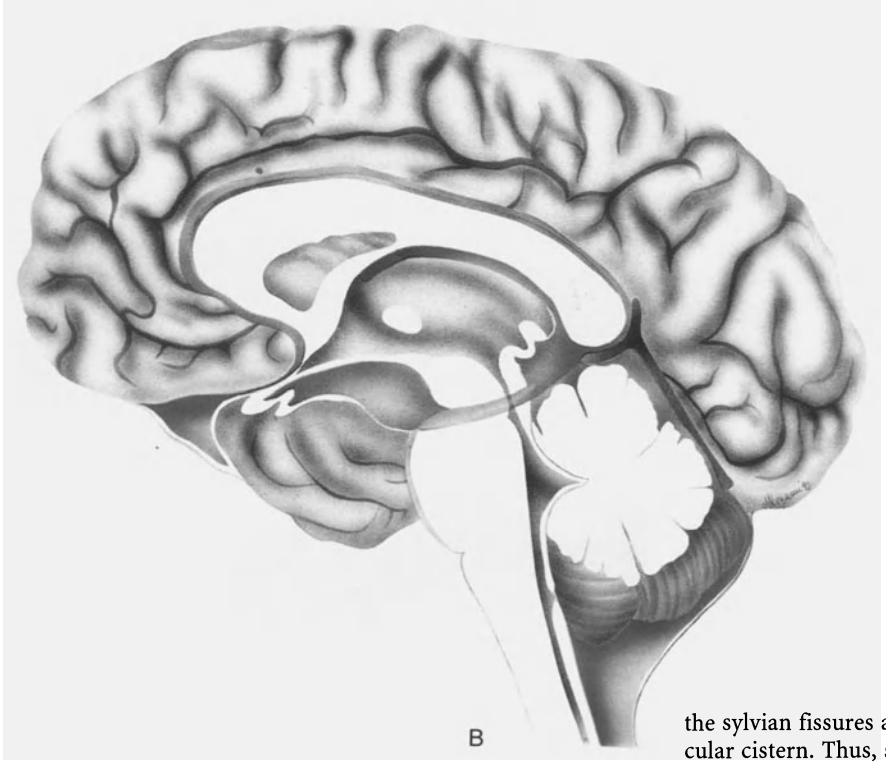


Figure 6.5. (A) The left optic nerve (*1*), olfactory nerve (*2*), and gyrus rectus (*3*) are exposed, as are the right gyrus rectus (*4*) and right olfactory nerve (*5*). The *prechiasmatic cistern* has been opened, exposing the pituitary stalk (*6*). The arachnoid bridging from the left gyrus rectus to the right gyrus rectus (*7*), at the tip of the sucker, is enclosing the *interhemispherical cistern*. Arachnoid (*8*) stretches from the posterior orbital gyrus (*9*) to the optic nerve (*10*), enclosing the *optic cistern* (*11*), and from the lateral orbital gyrus (*12*) to the temporal lobe (*13*), enclosing the *deep sylvian cistern* (*14*). (B) The parasellar cisterns as illustrated in (A), whether optic, prechiasmatic, chiasmatic, infrachiasmatic, or olfactory, are in communication laterally with the most medial extension of the sylvian fissures and posteriomedially with the interpeduncular cistern. Thus, since the interpeduncular cistern communicates directly with the pontine and bulbar cisterns along the midline and with the precerebellar and collicular cisterns posterosuperiorly through the ambiens cistern, one understands that the entirety of the basal cisterns are in free and open communication with one another. With particular reference, then, to the parasellar area (B), communications laterally through and from the entirety of the cerebral convexity occur via the medial, transverse, and anteroposterior portions of the sylvian fissure. Posterior communications are via the ambient, precerebellar, collicular cisterns. Flow along the midline occurs along the route chiasmatic, lamina terminalis, pericallosal cisterns. As an aside, it is of value at this time to comment on the significance of the cisterna magna both as a CSF volume reservoir and as a hydrostatic supportive force.

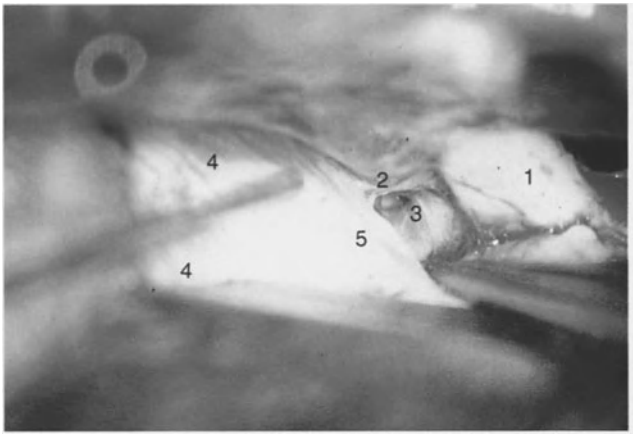


Figure 6.6. The left optic nerve (*1*) and anterior clinoid (*2*) are visible. The arachnoid has been taken from the lateral surface of the left internal carotid artery (*3*) but not from the posterior orbital gyrus of the left frontal lobe (*4*). One sees the arachnoid over the *sylvian fissure* (*5*) lateral to the posterior clinoid process. The arachnoid of the medial third of the sylvian fissure has been opened, so that one may see the internal carotid artery.

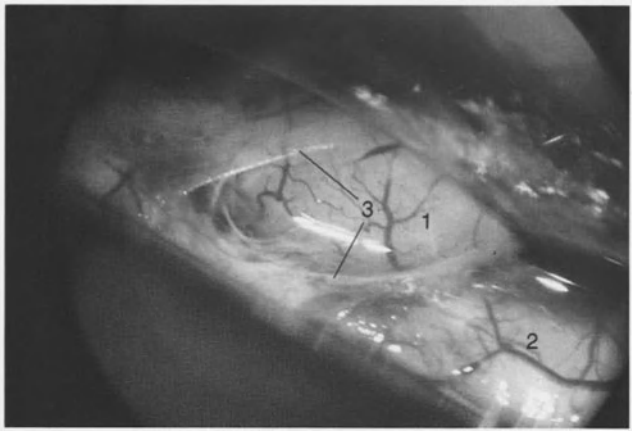


Figure 6.7. The lateral sylvian fissure. The superior medial surface of the left temporal lobe (*1*) is not covered by arachnoid, but the inferior lateral surface of the left frontal lobe (*2*) is. The arachnoid covering the lateral third of the sylvian fissure has been opened so one may see its edges (*3*).

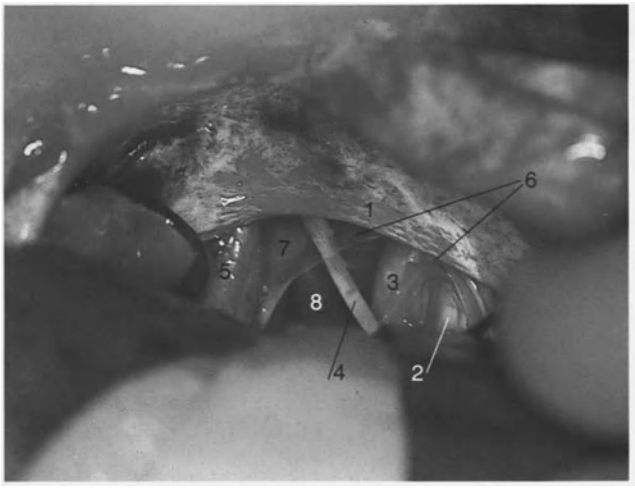


Figure 6.8. Ambient and interpenducular cisterns. The tentorial edge (*1*), pons (*2*), basilar artery (*3*), III cranial nerve (*4*), and internal carotid artery (*5*) are well visualized. The *ambient cistern* runs along the tentorial edge (*6*). Lillequist's membrane (*7*) has been opened, allowing one to look into the *interpeduncular cistern* (*8*).

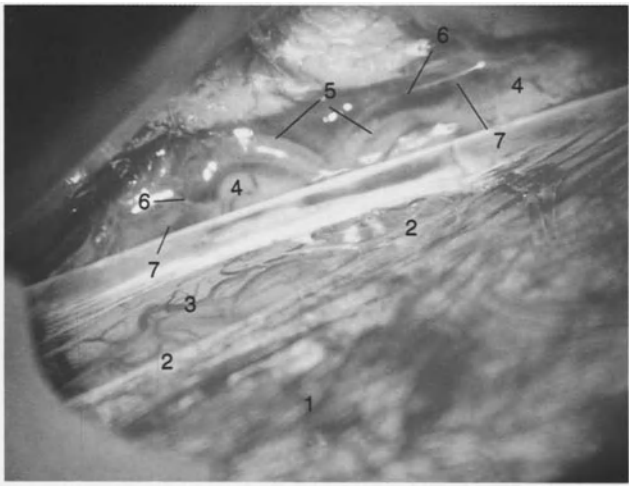
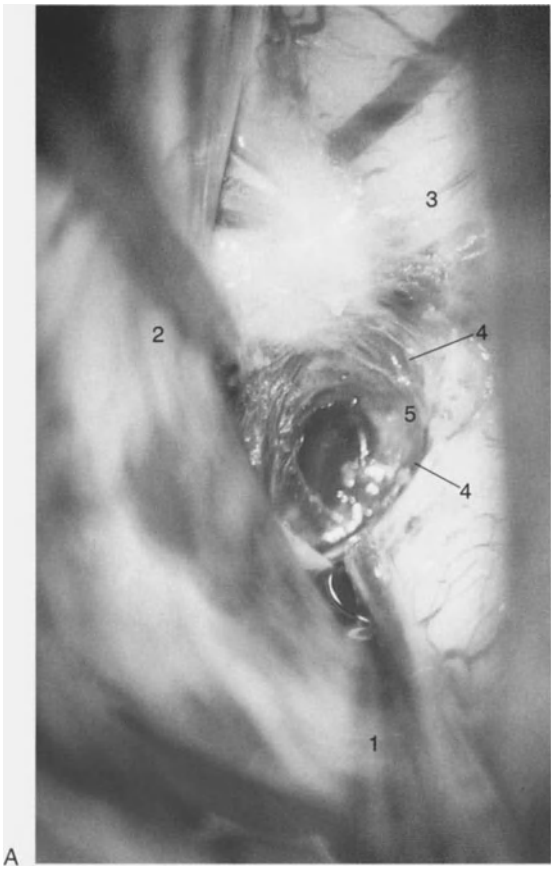


Figure 6.9. Pericallosal cistern. The falx cerebri (*1*), inferior longitudinal sinus (*2*), supracallosal gyrus (*3*), corpus callosum (*4*), and pericallosal artery (*5*) are all identifiable. The *pericallosal cistern* (*6*) is located between the corpus callosum and the arachnoid (*7*), and contains the pericallosal artery.



◀ **Figure 6.10.** (A) Supracerebellar cistern. This child has a pineal tumor. The tentorium (*1*), falx cerebri (*2*), and isthmus of the hippocampus (*3*) have been exposed, and the archnoid (*4*) along the posterior surface of the *supracerebellar cistern* opened, exposing the tumor (*5*) which occupied the entirety of this cistern. (B) In this photograph one notes the cerebellar hemispheres (*1*), the tentorial opening (*2*), the superior cerebellar vermis (*3*), and the arachnoid bordering the posterior portion of the superior cerebellar cistern (*4*).

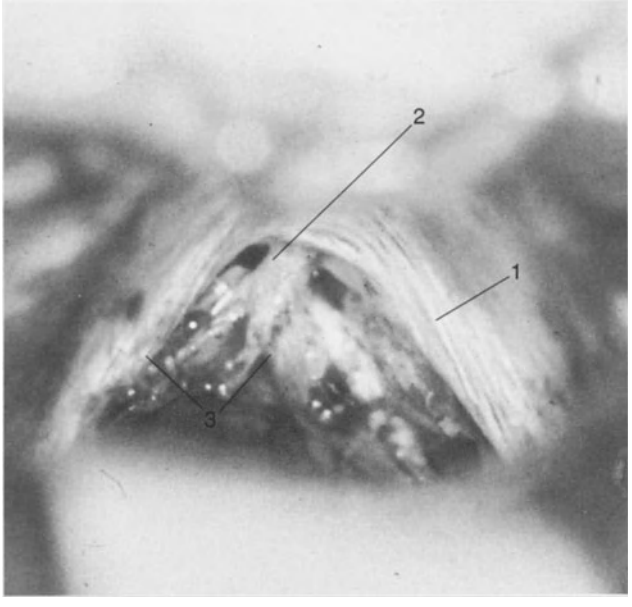


Figure 6.11. Quadrigeminal cistern. The tentorial opening (*1*) has been exposed and the culmen monticuli of the cerebellar vermis retracted downward, after the posterior and anterior arachnoidal membranes of the *supracerebellar cistern* had been coagulated and opened. This exposed the great vein of Galen (*2*), and both veins of Rosenthal (*3*) within the *quadrigeminal cistern.*

Ambient Cistern Lesions

Ambient cistern lesions (medial temporo-occipital arteriovenous malformations, tentorial edge tumors, etc.) are exposed by opening first the sylvian fissure, then entering into the ambient cistern at its confluence with the interpenduncular cistern. This allows drainage of all cerebrospinal fluid entering the ambient cistern and subsequent exposure of the hippocampal gyrus, brainstem, tentorial edge, and neurovascular structures within the cistern. Lowering the vertex of the skull beneath the level of the base, as described above, facilitates exposure.

Pineal Lesions

For pineal lesions, the use of gravity is to be avoided when one approaches the tumor through a parietal flap and along the parasagittal route! If one approaches the corpus callosum with the cerebral hemisphere inferior and the falx cerebri superior, one gains great exposure advantages in that the cerebral hemisphere follows gravity. A remarkable space opens between it and the falx cerebri, eliminating the need for self-retaining retractors. However, *this is extraordinarily disadvantageous because it puts stretch on the parietal* (anterior and posterior cortical) *bridging veins*, stagnating venous blood within them and the parietal lobe (Fig. 6.14). This increases the risk of venous infarct.

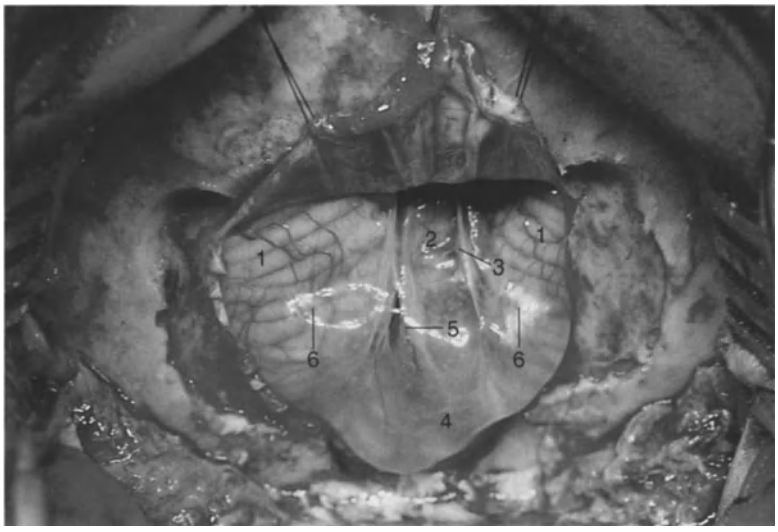


Figure 6.12. Cisterna magna. The cerebellar hemispheres (*1*) and cerebellar vermis (*2*), as well as the inferior vermian veins (*3*), may be seen beneath the arachnoid of the *cisterna magna* (*4*). This cistern has been opened (*5*) but most of the cerebrospinal fluid remains within it, permitting one to appreciate how much fluid the cisterna magna may contain. One may also see the point at which the arachnoid of the cisterna magna becomes adherent to the cerebellar hemispheres and vermis (*6*).

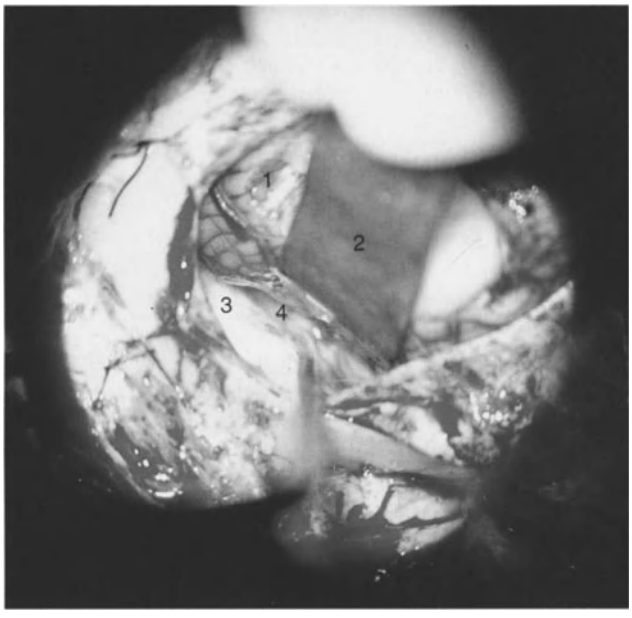


Figure 6.13. Pontocerebellar cistern. The left cerebellar hemisphere (*1*) has been elevated superomedially by a spatula (*2*), exposing the dura over the petrous apex (*3*) and the left *pontocerebellar cistern* (*4*) at the point of entry of the lateral recess of the IV ventricle.

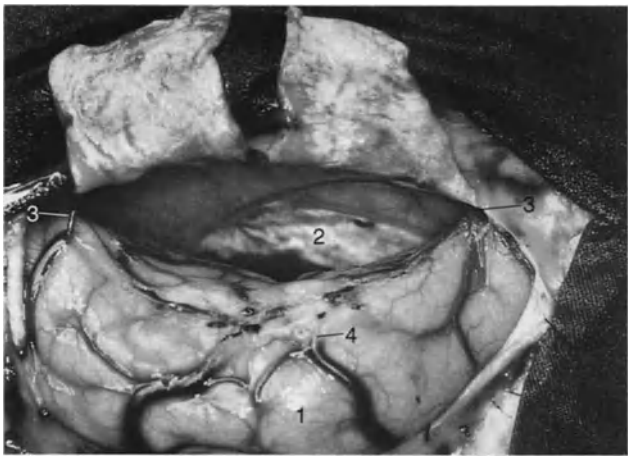


Figure 6.14. Disadvantages of gravitational retraction of the parietal lobe for pineal tumors. Notice the stretching of bridging veins. This child's pineal tumor was operated along the parasagittal route, with the child's head positioned so that the right parietal lobe (*1*) was recumbent. The retraction, of course, is greatly facilitated by the recumbency of the parietal lobe, permitting excellent visualization of the pericallosal cistern (*2*) but, unfortunately, stretching unacceptably major cortical bridging veins (*3*), which in this child resulted in a transient contralateral hemiparesis. This stretching of bridging cortical veins by a pendant parietal lobe is to be avoided, in light of the fact that one must sacrifice at least a single bridging cortical vein (*4*) in order to approach the corpus callosum safely.

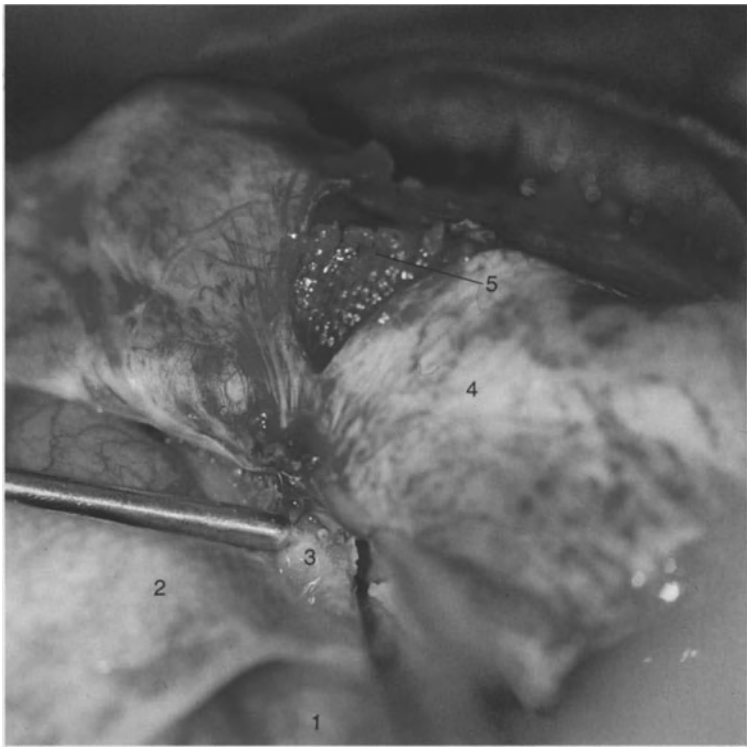


Figure 6.15. Separation of parietal lobe from arachnoidal granulations. This child, who also had a right parasagittal approach to his pineal tumor, was positioned with his left parietal lobe recumbent. Consequently, retraction of the right parietal lobe entailed elevating it, thus eliminating unwanted stretching of the bridging cortical veins at either end of the opening between retracted parietal lobe and falx cerebri. The right parietal lobe (*1*) is being elevated with use of a fluffy cotton (*2*), exposing the pacchionian granules (*3*) in this 14-year-old boy. Note that the dura (*4*) has been bivalved and sewn over a Surgicel roll (*5*), protecting the SSS from compressive occlusion.

It is preferable to position the patient so that the surgeon's interval of vision is between the falx cerebri *inferiorly* and the retracted parietal lobe *superiorly*, taking care and time to separate the parietal lobe from any arachnoidal granulations that may be present (Fig. 6.15). *The younger the child, the fewer the arachnoidal granulations.* Therefore, dissection over the parasagittal surface of the convexity of the parietal lobe and along the superior sagittal sinus and the falx cerebri is easier and safer. The rigid falx remains inferior. It offers support to Telfa and fluffy cottons used to elevate the parietal lobe slowly and gradually. Minuscule arachnoid bands bridging to the dura along the edges of the sinus are cut. The hairpin curves of the bridging veins are gently unfolded, placing soaked fluffy cottons over them for protection against injury and desiccation. Ever-increasing sizes of soaked fluffies are lain over the falx at the angles, to displace gently the parietal lobe. Progressively larger Telfas are lain along the falcian surface of the parietal lobe as this latter is lifted with a wide spatula, exposing gradually the parietal lobe convexity and medial surface, inferior sagittal sinus, pericallosal cistern, pericallosal artery, and corpus callosum. The spatula blade of the self-retaining retractor is locked into position repeatedly and progressively, avoiding any attempt to accomplish complete exposure with one insertion.

Self-retaining retractors (using broad blades) placed over the Telfa thus *elevate* the parietal lobe and allow the surgeon to monitor carefully the amount of stretch and narrowing of bridging cortical veins. By angulating, or molding, wide-bladed retractors as the pericallosal cistern is approached, one may limit stretching and risk of damaging the bridging veins to an absolute minimum. When the cistern is opened there is an outpouring of cerebrospinal fluid, rendering the retraction easier, often allowing the surgeon to permit the retracted parietal lobe to approximate a bit more the falx. After splitting the splenium, or body, of the corpus callosum, the quadrigeminal and/or superior cerebellar cisterns are opened. The exposure of the pineal region is now completed. One must not split more of the corpus callosum than is absolutely necessary and never more than one-third of its length. Take care not to damage the body and crus of the fonices!

Once the retractors have been set, inspect their edges to ascertain that they are not cutting into the brain and that no vessel is precariously stretched. The tendency is

to attain more retraction than is necessary. The cortical bridging veins are covered with arachnoid. They leave the convexity of the cerebral cortex in an anterior superior direction, and then penetrate the inner layer of dura mater, becoming dural sinuses as their covering changes from arachnoid to dura and they are contained between the outer and inner layers of the dura. Some cortical bridging veins do not penetrate the dura but enter the superior sagittal sinus directly. These vessels course anteriorly within the subdural space, and then turn back on themselves (hairpin fashion) to enter the sinus so that the flow of blood is in the same direction as within the sinus.

Intraventricular surgery is greatly facilitated if the surgeon carefully reviews the imaging studies to identify encysted ventricles, the result of an obstructive lesion either at the trigone (obstructing the occipital and/or temporal horns) or the foramen of Monro (obstructing one or both lateral ventricles). If an encysted ventricle is identified, the surgeon may puncture it while performing the cerebrotomy, attaining excellent exposure, and minimizing any risk of brain shift resulting from the combined pressure vectors of the space-occupying lesion and the encysted ventricle(s). In instances when an intraventricular (III ventricle) tumor occludes one or both foramina of Monro, the surgeon attains his "retraction" preoperatively by inserting bilateral ventriculoperitoneal shunts. A single ventriculoperitoneal shunt is sufficient if the obstruction is at the aqueduct of Sylvius within the posterior III ventricle.

Hydrocephalus is present in 100% of the children with pineal tumors, 90% of the children with medulloblastoma, 75% of children with astrocytoma, and 35% of those with brainstem glioma. Consequently, preoperative shunting provides the best form of decompression and the best assurances that cerebral retraction will not be necessary. Entrance into the cisterna magna for midline tumors (either surgically for inferior cerebellar triangle lesions, or by puncturing the cisterna magna through the exposed atlanto-occipital membrane for superior triangle lesions) offers even more exposure. Lateral suboccipital craniotomy permits the surgeon to decompress the basal cisterns either by opening the medullary or pontine cisterns medially or opening the pontocerebellar cistern superolaterally.

Use of Cotton Fluffies and Telfa

(Figs. 6.16–6.18)

Cotton fluffies, prepared simply by soaking absorbent cotton in saline after it has been fashioned into the desired size and form, provide excellent protection during the exposure of such vital and delicate structures as the cranial nerves, normal or pathological vasculature, the intraventricular (ependymal) surfaces, or the nuclei in the floor of the III or IV ventricles.

Figure 6.16. Cotton fluffies should be pulled into the desired form and then soaked immediately before applying them to the brain or a bleeding vessel. They should not be left soaking in a basin prior to use, for this makes them dense.

It is preferable to keep the fluffies wet while dissecting and exposing the desired structure, not to suck them dry when teasing loose adhesions from them. Dense adhesions must be cut!

Telfa, cut to the desired form and length, moistened, and then lain over the surface of the brain, provides an excellent means by which proper retraction may be attained and the brain protected either from compression or inadvertent glancing blows or cuts. The Telfa is applied directly to the surface of the brain. Sliding it, moist or dry, over the brain has to be avoided because it irritates the cortex and tears small cortical vessels. Rather, it is preferable either to place the Telfa over the surface of the brain and then to unroll the redundant segment (as one would unfold a rug) in advancing it over cortical surfaces, which either are out of view or facing dura, or to slide the redundant segment over the dura. After the Telfa has been applied to the desired brain surface, the self-retaining retractor may be brought into position. Moisten the Telfa periodically to prevent desiccation of the cortical cells and damaging adherence to the cerebral surface.

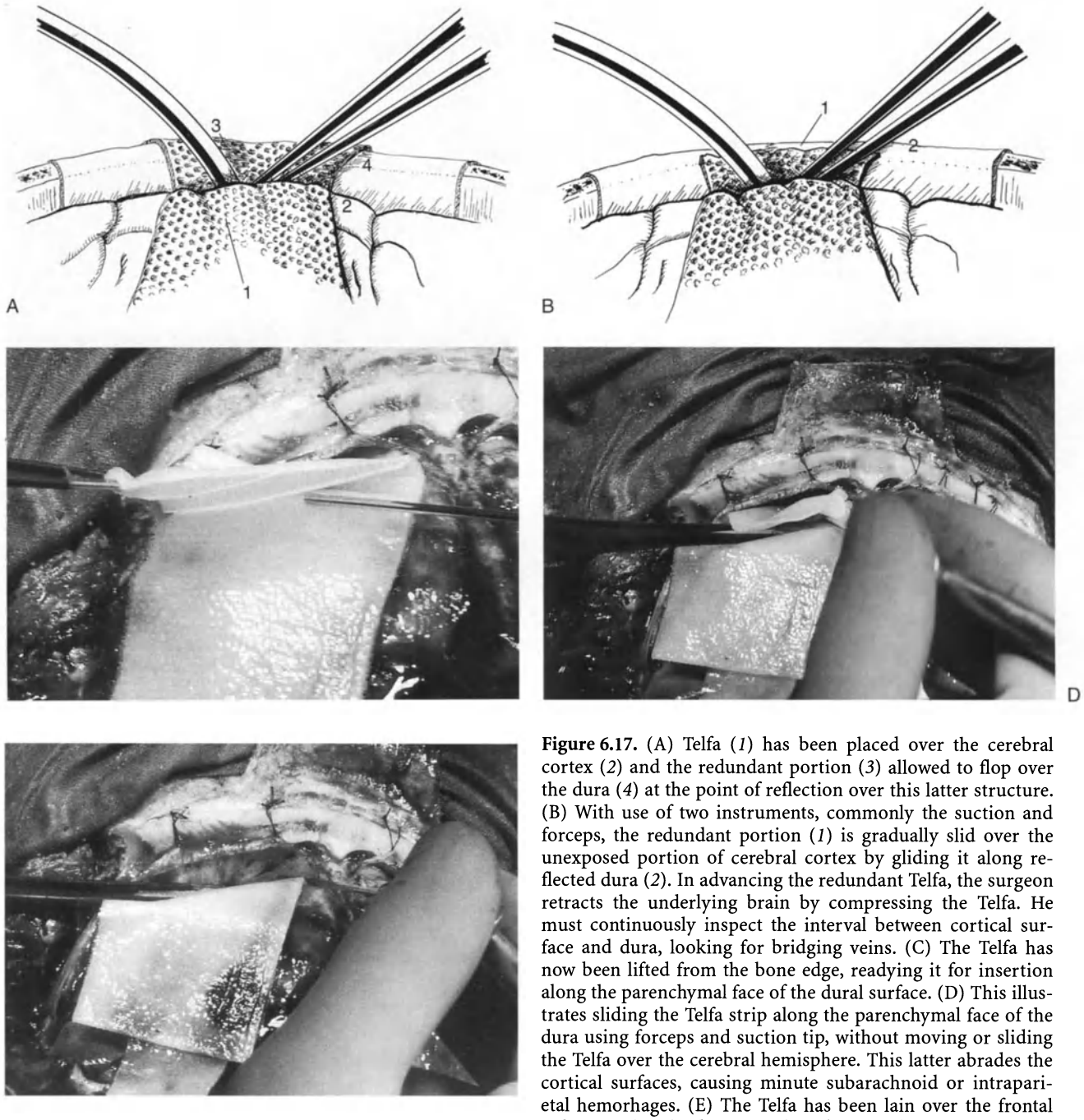


Figure 6.17. (A) Telfa (*1*) has been placed over the cerebral cortex (*2*) and the redundant portion (*3*) allowed to flop over the dura (*4*) at the point of reflection over this latter structure. (B) With use of two instruments, commonly the suction and forceps, the redundant portion (*1*) is gradually slid over the unexposed portion of cerebral cortex by gliding it along reflected dura (*2*). In advancing the redundant Telfa, the surgeon retracts the underlying brain by compressing the Telfa. He must continuously inspect the interval between cortical surface and dura, looking for bridging veins. (C) The Telfa has now been lifted from the bone edge, readying it for insertion along the parenchymal face of the dural surface. (D) This illustrates sliding the Telfa strip along the parenchymal face of the dura using forceps and suction tip, without moving or sliding the Telfa over the cerebral hemisphere. This latter abrades the cortical surfaces, causing minute subarachnoid or intraparietal hemorhages. (E) The Telfa has been lain over the frontal pole, exposing the dura over the orbital roof.

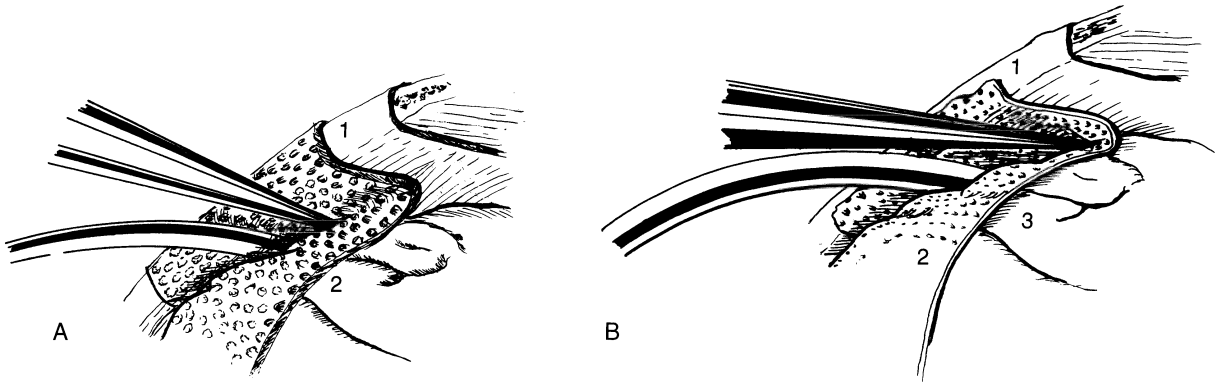


Figure 6.18. (A) In this drawing, the line of vision is oblique so as to allow one to appreciate the advancement of Telfa by sliding it along the dura (*1*) and *not* over the surface of the cortex (*2*). (B) Later stage in advancement of the Telfa, to allow the reader to appreciate advancement of Telfa along the surface of reflected dura (*1*), as the portion (*2*) over the brain (*3*) is held in place.

Self-Retaining Retractors

Self-retaining retractors are essential to the proper perfomance of many neurosurgical procedures. They allow the surgeon to perform microsurgical procedures without the need for an assistant. It is physically impossible for an assistant to hold them steadily because of the extremely small working area and the extraordinary length of time a microsurgical procedure takes. Also, self-retaining retractors assure a minimum of cerebral damage in that there is no jiggling, repositioning, pushing, and so on that the human hand cannot avoid. The retractors remain where they are placed, continue to exert the force at which they were set, and may be reset repeatedly as one proceeds with the dissection.

Because of the fact that retraction of the brain exerts a physical force that may result in destruction of gray and white matter, one is to retract the brain with the least force possible, and for the briefest time necessary to accomplish the intended procedure. Investigators have applied strain gauges to cerebral retractors in order to quantify time/force factors. Forces of 350 mmH_2O [3], 20 torr [4], and 30 *g* continuous [5] have been reported. Many have noted that the effects of brain retraction are different from what results when the brain is compressed by other expansile masses [6–11]. Of no mean significance is the fact that there is a real difference in damaging effects resulting from continuous as compared to intermittent retraction, *intermittent retraction being much less damaging*. "The brain tolerates about 70% more intermittent retraction than continuous retraction from a morphological standpoint, while electrophysiologically the difference is about 40%".

Self-retaining retractors which take purchase from the skull (as the De Martell retractor) are of no value in the newborn and the younger infant. Fortunately, the retraction provided by gravity and cisternal opening in these age categories is more than adequate for the procedures that would call for self-retaining retractors: arteriovenous malformations of the galenic system, intraventricular tumors, and posterior fossa tumors. The craniopharyngioma is almost unknown in patients under 2 years of age. If the need for self-retaining retractors in operating on a newborn or infant were to arise, one could use the type (Greenberg) which is mounted to the operating table.

The blades for self-retaining retractors are applied flatly over the Telfa, not slid forward on it. In order to bring the blades deeper, one may use two blades in tandem, setting one first and then bringing the other parallel to it, but slightly deeper, before locking it. One then returns to advance the first retractor still deeper and closer to the target area, and so on. Bending the blades to mold their form to fit the anatomical contour of the brain is generally less favorable than slightly angulating them: the brain assumes a rectilinear course as it is retracted. Once the desired exposure of the target area is attained, the locking nuts for the self-retaining retractors should be released so that there is no compression of the brain and the retractor serves merely to protect the surface of the brain! If additional retraction becomes necessary, as it very well may because the pulsations of the brain re-expand it, one may simply do so, and tighten the lock-nut once more for a brief period of time.

Take a great deal of time to position the self-retaining retractors. This minimizes cerebral compression and the risk of tearing (by stretch) a bridging cortical vein, a potentially disastrous event. If it is necessary to keep the lock-nuts fastened to maintain exposure, release the retractor for 3–5 min every 5–10 min.

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7 Cerebrotomy

"Then out spoke brave Horatius,
The captain of the Gate:
'To every man upon this earth
Death cometh soon or late.
And how can man die better
Than facing fearful odds.
For the ashes of his fathers
And the temples of his Gods,'..."

THOMAS BABINGTON MACAULAY, Horatius.
Lays of Ancient Rome

Exposure of intraparenchymal or intraventricular tumors necessitates opening the cerebrum (cerebrotomy). Classically, there are two techniques for perfoming cerebrotomy: (1) going through a gyrus and (2) going through a sulcus. Proponents of each sustain that their particular procedure of choice affords easier access to the cerebral substance or ventricle, and causes less damage. This, indeed, is a moot point, since cortical tissue at the gyral and sulcal levels is equally damaged when cut. Retraction of a cortical cerebrotomy may result in extension of the opening and, consequently, increase the risk of cerebral damage. For matters purely of habit, the author prefers the sulcal cerebrotomy, but herein describes both. Also, the use of laser to incise cerebral substance is described, as is the technique for surface thermocoagulation and parenchymal spreading. I have doubts concerning the value of laser for cortical or white matter incision.

Whichever of the two cerebrotomy procedures, cortical or sulcal, is used, it is essential for the surgeon to coagulate, with bipolar forceps or laser, the pia-arachnoid and underlying vessels prior to advancing through gray matter, and then using spatulae to separate the bundles of axons.

Microforceps are used for bipolar coagulation, with the surface coagulation extending the full length of the desired cerebrotomy incision. This coagulates the cortical vessels and "burns" an opening through the arachnoid. Generally, one attains coagulation of the arachnoid in some places, its destruction in other places. All vessels along the line of cerebrotomy must first be coagulated and then cut, with no attempt being made to dissect them to either one side or the other since these vessels receive, and give off, a multitude of perpendicular collaterals.

Attempts to work around them are futile, lengthen the procedure, and result very often in the tearing of perpendicular collaterals relatively remote to the site of the cerebrotomy. This amplification necessitates extending the area of cerebral damage in order to coagulate the retracted, torn vessel. Every effort should be made to keep the line of coagulation as narrow as possible! *The finest bipolar blades available are preferable to the CO_2 laser.*

If the laser is available it provides an opportunity to perform a cortical cerebrotomy of acceptable width, sealing those vessels less than 1 mm in diameter, as well as the arachnoid, to the cortex, as it cuts. Larger caliber vessels, of course, must be coagulated with the bipolar before they are transected with the laser. For surface vaporization of the pia-arachnoid and underlying cortical microvasculature, a defocused beam of 5 W, in the continuous mode, is used. It is best to use the micromanipulator to obtain the thinnest and straightest line of coagulation possible: a line of 2 mm in width. After the pia-arachnoid and cortical surface have been coagulated, one passes to using a focused beam, without changing wattage or mode, to incise the cortical surface, stopping to coagulate opened vessels when bleeding starts: the use of the CO_2 laser in a bloody field accomplishes nothing. After the cortical mantle has been crossed and the white matter entered, the laser may still be used, but it is both time-consuming and inefficient. One reaches the desired depth more quickly and efficiently by using two spatulae as described later in this chapter.

Gyral Cerebrotomy (Fig. 7.1)

The microforceps are used to coagulate the arachnoid and underlying cortical vessels along (a line on the gyral surface) where there is minimal vasculature. Once the entire cerebrotomy line has been coagulated, the forceps are used to continue the coagulation in a given point to "burn" an opening in the arachnoid. Then they are so placed as to hold the arachnoid between the blades, which are extended horizontally, parallel to the arachnoid and cortex, sealing the arachnoid and any microvessels adherent to it. The completely coagulated arachnoid is then cut with a microscissors and the underlying cortex inspected (using tiny wet fluffy cottons to dry the field) for uncoagulated microvasculature.

Pia-arachnoid and cortical vessels, arteries or veins, running perpendicular to the line of cerebrotomy should be coagulated and cut. This technique is for transecting vessels only partially, an event which results in bleeding that is particularly difficult to stop. It also assures transecting a vessel completely, so that in the event it has not been fully coagulated one may stop the bleeding quickly by taking the full circumference of the vessel in the forceps, constricting it, and then coagulating. This is followed along the line of desired cerebrotomy. One then brings the dissection through the full thickness of the cortex, by coagulating cortical surface between microbipolar forceps held with the blades 1 mm apart, but not into the underlying white matter: there is no need to coagulate the white matter, since it is avascular. Small cortical vessels present no problems if one stops to coagulate and transect each as it is identified.

Sulcal Cerebrotomy (Fig. 7.2)

If the surgeon opts to use the sulcus as the point of entry for performance of the cerebrotomy, the procedure for coagulating the arachnoid and its adherent microvasculature is similar, but not identical, to that used in the gyral cerebrotomy. One may find that a sulcus may be identified immediately adjacent to a relatively large vessel and that, consequently, this is the best sulcus to enter. However, relatively large veins may cross the sulci. The arachnoid bridging the sulcus, stretched from one gyrus to another, is coagulated with the microbipolar forceps in one spot until it is perforated, at which time the blade of the forceps is applied to either surface of the bridging arachnoid and the line of coauglation is extended alongside major sulcal vessels, taking care not to occlude them. The coagulated arachnoid is then cut, laying open the sulcus.

Small Vessels at the Depth of the Sulcus or Gyrus

The small vessels at the very depth of the sulcus or gyrus are coauglated individually. Generally, they withdraw and may be pulled apart with the bipolar forceps since they are tiny. Occasionally, however, it may become necessary to cut them with the microscissors. In either event, one should coagulate completely all vessels at the depth of the sulcus. Then, a long, thin fluffy cotton may be inserted into the now created gutter. Rolling this fluffy from one end to the other permits one to identify vessels in the path of the cerebrotomy, so that they may be coagulated before proceeding. Once the gutter of the sulcus has been completely cleared of bridging vessels, one may either continue the dissection through the cortex with bipolar forceps and fluffy cotton or, preferably, a narrow spatula.

Cerebrotomy Through White Matter (Fig. 7.3)

In extending the cerebrotomy down to either a deep seated lesion or the intraventricular compartment, one need no longer worry about bleeding: there is no (surgically significant) vasculature within the white matter. Consequently, it is as well to proceed directly with the dissection by extending the already placed spatula from the junction between cortex and white matter to the desired depth, preferably ranging from 3 to 5 mm. This is extended longitudinally from one end of the cerebrotomy to the other, rather than proceeding directly to a greater depth, an undesirable technique that results in creating a cone rather than the desired cylindrical opening. After this has been done on one side, a spatula is inserted on the opposite side and the process repeated. Each insertion should extend to a 5- or 6-mm increase in depth of the cerebrotomy. As the performance of the cerebrotomy through the white matter proceeds, one may spread open the exposed area to inspect its depth before proceeding to another level, ascertaining that the cerebrotomy is of equal depth along its full length, avoiding a conical opening (wider at the cortical surface, narrower at the base) which narrows the working area. Finally, the positioning of the blades is changed, rotating them 90°, so that each is at an edge of the cerebrotomy and set upon either a cotton strip or Telfa. They are inserted either into the ventricle or over the surface of an intraparenchymal lesion.

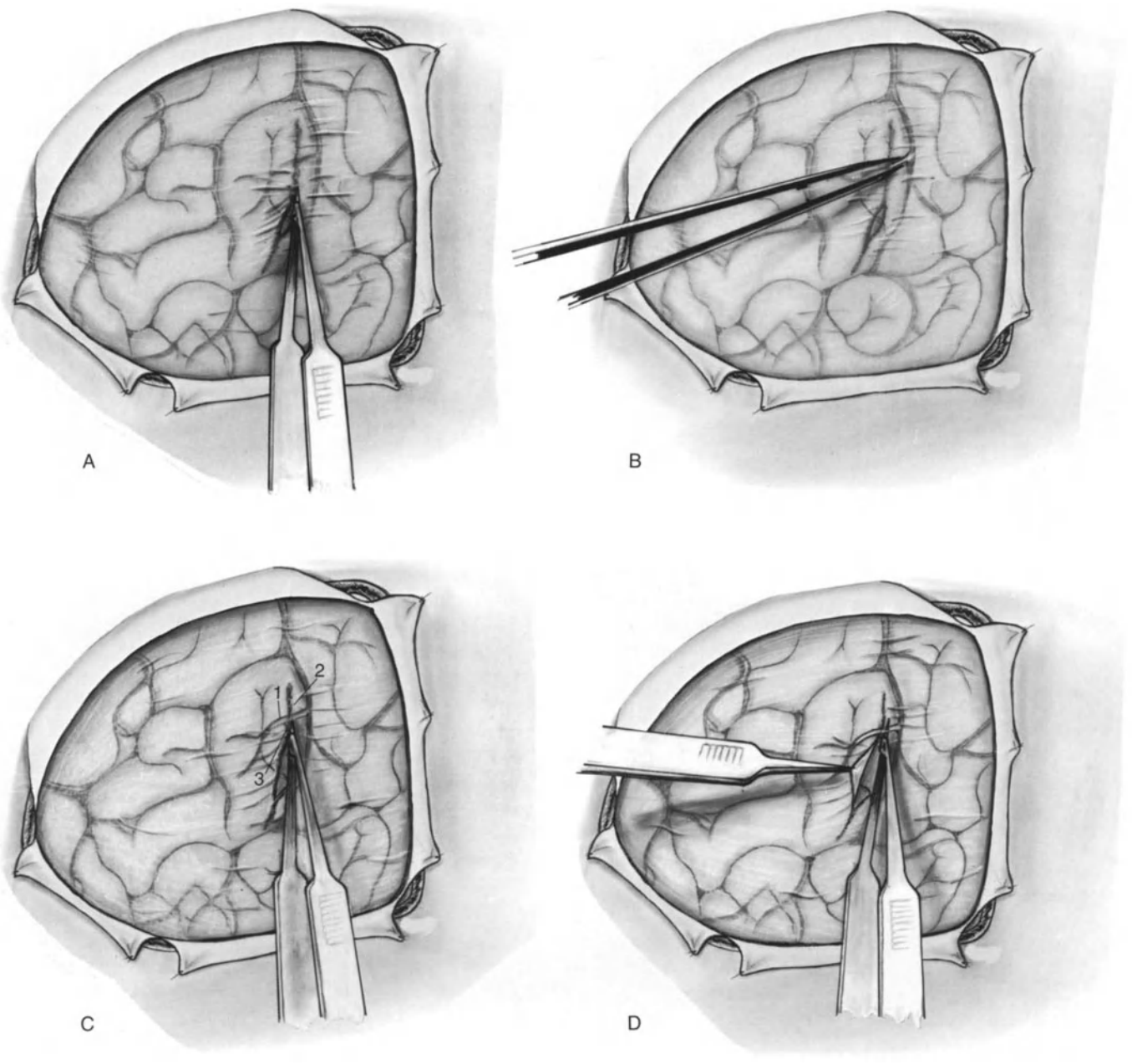

Figure 7.1 Gyral cerebrotomy. (A) After the arachnoid and adjacent microvasculature have been coagulated over the full extent of the cerebrotomy incision, and bipolar forceps have been used to "burn" an opening in the arachnoid, microscissors are inserted into the opening so as to take only the arachnoid and its adherent microvasculature between the blades. (B) As the cutting of the arachnoid and its adherent microvasculature proceeds, one may identify larger cortical vessels, arteries and veins, running perpendicular to the line of desired cerebrotomy. At this time, the vessel should be taken in the microforceps by applying the blades to its sides and coagulating, adding a drop of water. This technique prevents the coagulated vessel from adhering to the microbipolar forceps. (C) Once the vessels running perpendicular to the line of cerebrotomy are coagulated (*1*) over a distance of approximately 4 mm, the cutting of coagulated arachnoid (*2*) and adjacent microvasculature is extended first to the very edge of the coagulated vessel (*3*). (D) Then, the scissors are advanced so as to take the vessel fully in its blades at the time of the cut.

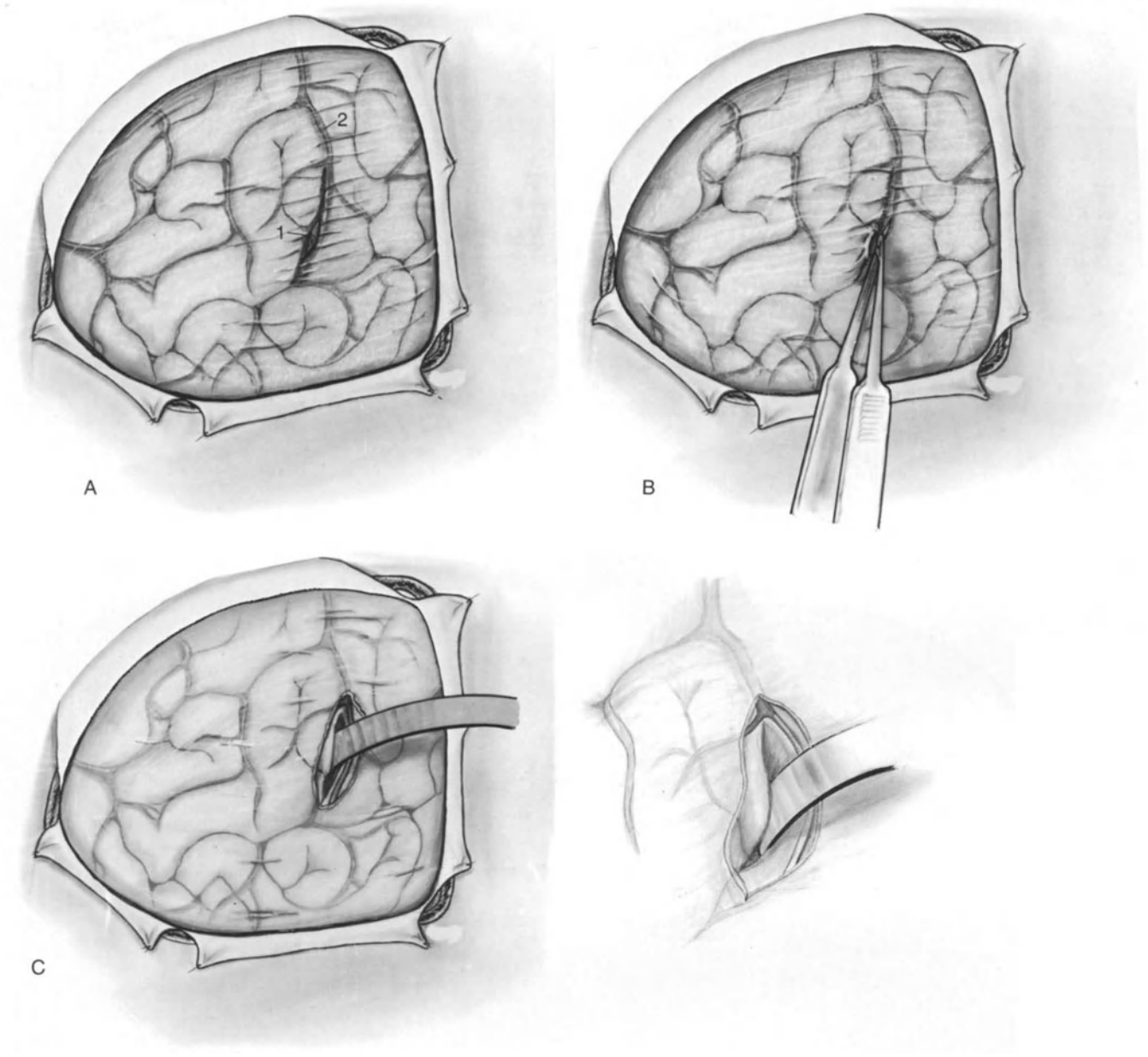

Figure 7.2. Sulcal cerebrotomy. (A) The identified sulcus (*1*), generally along one side or the other of a sulcal vessel (*2*), is entered by first coagulating the arachnoid, taking care not to coagulate the adjacent vessel! It is quite easy to coagulate the arachnoid over the sulcus because there is cerebrospinal fluid beneath it. Similarly, one may insert the blades of the bipolar forceps through the opening in the arachnoid, and extend them along the sulcus, coagulating adherent vessels as one proceeds. (B) The surgeon may cut along the sides of large sulcal vessels. (C) After all the sulcal microvasculature has been coagulated and cut, the malleable thin-bladed spatula is inserted to perform the dissection down to the white matter.

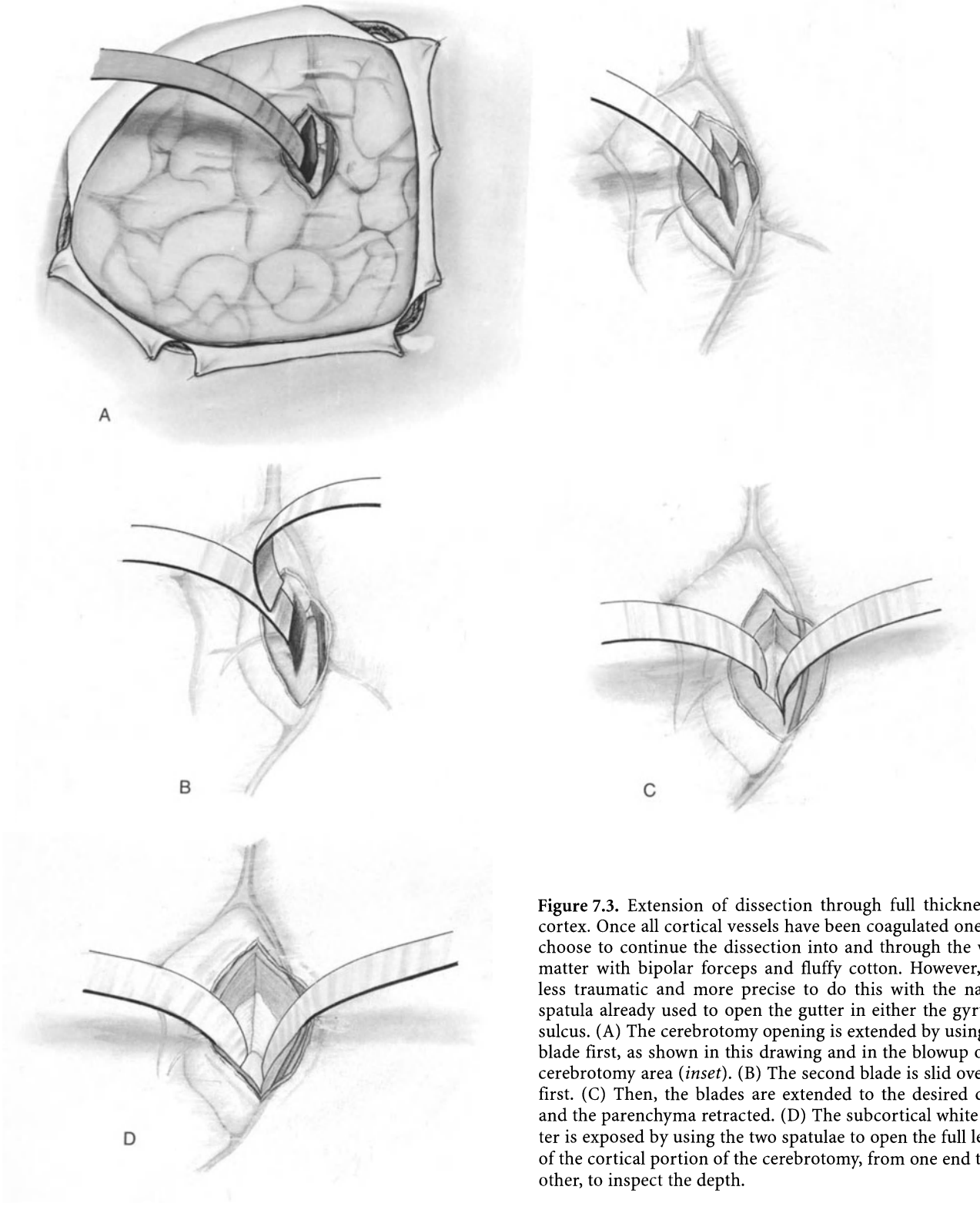

Figure 7.3. Extension of dissection through full thickness of cortex. Once all cortical vessels have been coagulated one may choose to continue the dissection into and through the white matter with bipolar forceps and fluffy cotton. However, it is less traumatic and more precise to do this with the narrow spatula already used to open the gutter in either the gyrus or sulcus. (A) The cerebrotomy opening is extended by using one blade first, as shown in this drawing and in the blowup of the cerebrotomy area (*inset*). (B) The second blade is slid over the first. (C) Then, the blades are extended to the desired depth and the parenchyma retracted. (D) The subcortical white matter is exposed by using the two spatulae to open the full length of the cortical portion of the cerebrotomy, from one end to the other, to inspect the depth.

8 Cerebral Resection

"Hew down the bridge, Sir Consul,
With all the speed ye may:
I, with two more to help me,
Will hold the foe in play.
In yon strait path a thousand
May well be stopped by three.
Now who will stand on either hand,
And keep the bridge with me?"

THOMAS BABINGTON MACAULAY, Horatius.
Lays of Ancient Rome

Biopsy

An adequate brain biopsy consists of approximately a 1-mm^2 surface area and a full thickness plug of cerebral parenchyma containing cortex and white matter, generally 1.5 or 2 cm in length. This gives the pathologist between 1.30 and 1.45 cc of tissue.

The biopsy should be taken from a gyrus. The pia-arachnoid over the gyrus is coagulated circumferentially, leaving intact leptomeninges, vascular structures, and cortex at the center. A #11 blade is then inserted through leptomeninges, cortex, and white matter, precisely along the arch of coagulation, and a plug of cortex and white matter cut from the surrounding cerebral parenchyma. The freed plug is lifted away with saline and the field with a fluffy cotton. The bleeding is stopped with bipolar cautery, Avitene, or fluffy cotton and aspiration. One should wait approximately 10 min after all of the bleeding has been stopped before proceeding with the closure.

Lobectomy

Lobectomy is a procedure which may be used for the treatment of epilepsy (temporal), subtotal resection of glioma (frontal, temporal, occipital, cerebellar), or, *very* rarely, arteriovenous malformations. Parietal lobectomy is not described because the resultant neurological deficit, irrespective of the operative indications or pathological condition, is so severe as to preclude its consideration. The technique for lobectomy consists of exposing the desired lobe adequately so as to be able to isolate the arterial and venous systems completely. Consequently, one must have access to the main trunk arteries, going to the desired lobe, and to the cortical bridging veins both at their points of *exit* from the cerebral convexity and *entry* into the sinus.

After the vascular structures have all been exposed and coagulated, one may proceed to the cerebrotomy stage of the operation, extending it from inferior to superior, because arterial supply to the frontal, temporal, occipital, and inferior triangle of the cerebellar lobes extends from inferior to superior. For example, the frontal lobe is fed by branches of the *anterior and middle cerebral arteries,* extending from the bifurcation of the interval carotid arteries upward. Similarly, the temporal lobe is fed primarily by branches of the *middle cerebral* artery, but also by the *anterior inferior temporal branch of the posterior cerebral artery,* both systems extending from inferior to superior. The occipital lobe, nourished exclusively by the *terminal branches of the posterior cerebral artery*, also follows this rule, as does the inferior cerebellar triangle, being fed by the *anterior* and *inferior cerebellar arteries.* The pericallosal artery runs a superior course around the corpus callosum. Almost all (with the one exception being the *frontobasal artery*) of its branches run superiorly over the pericallosal and medial frontal gyri, to drape over the cerebral convexity. Though the *sylvian branches* of the middle cerebral artery follow a superoinferior course as they loop over the temporal operculum and superior temporal gyrus, their main direction is from inferior to superior, since their insular course is no more than a hairpin curve around the overlapping temporal lobe. Only the superior cerebellar triangle is fed by an arterial system that courses from above downward (*the three branches of the superior cerebellar arteries*).

It is, consequently, best to place the line of incision from inferior to superior, proceeding to the level of the white matter along the entire lobectomy line, just as one does for a cerebrotomy. This assures complete occlusion of vascular supply to the lobe, and provides the surgeon an opportunity to coagulate and transect those venous structures which he encounters as he proceeds.

The major draining veins are identified, coagulated, and cut individually, immediately after the major feeding arteries have been transected, before the cerebrotomy stage.

The white matter of the lobe is transected with spatulae, as described under cerebrotomy. The laser may also be used to perform the cerebrotomy stages of cerebral resection. However, it is not as anatomical as separating white matter axonal bundles with spatulae; it is not as reliable as the bipolar to coagulate cortical vessels less than 1 mm in caliber. *Dissection of the white matter with laser is much more time-consuming than with spatulae, and it does not preclude the use of spatulae, since one must retract the already incised parenchyma lest it flop back into the field.*

Frontal Lobectomy (Fig. 8.1)

The bifurcation of the internal carotid artery is identified by retracting posteriorly the frontal lobe from the ortbital roof, following this dissection posteriorly to the olfactory trigone. The olfactory nerve remains intact from the olfactory bulb to the trigone. The suprachiasmatic and optic cisterns are entered and the optic nerve is dissected from the internal carotid artery, opening the interopticocarotid space and stripping the arachnoid membrane from these structures. The dissection then proceeds along the lateral surface of the internal carotid artery, as far posterosuperiorly as its bifurcation

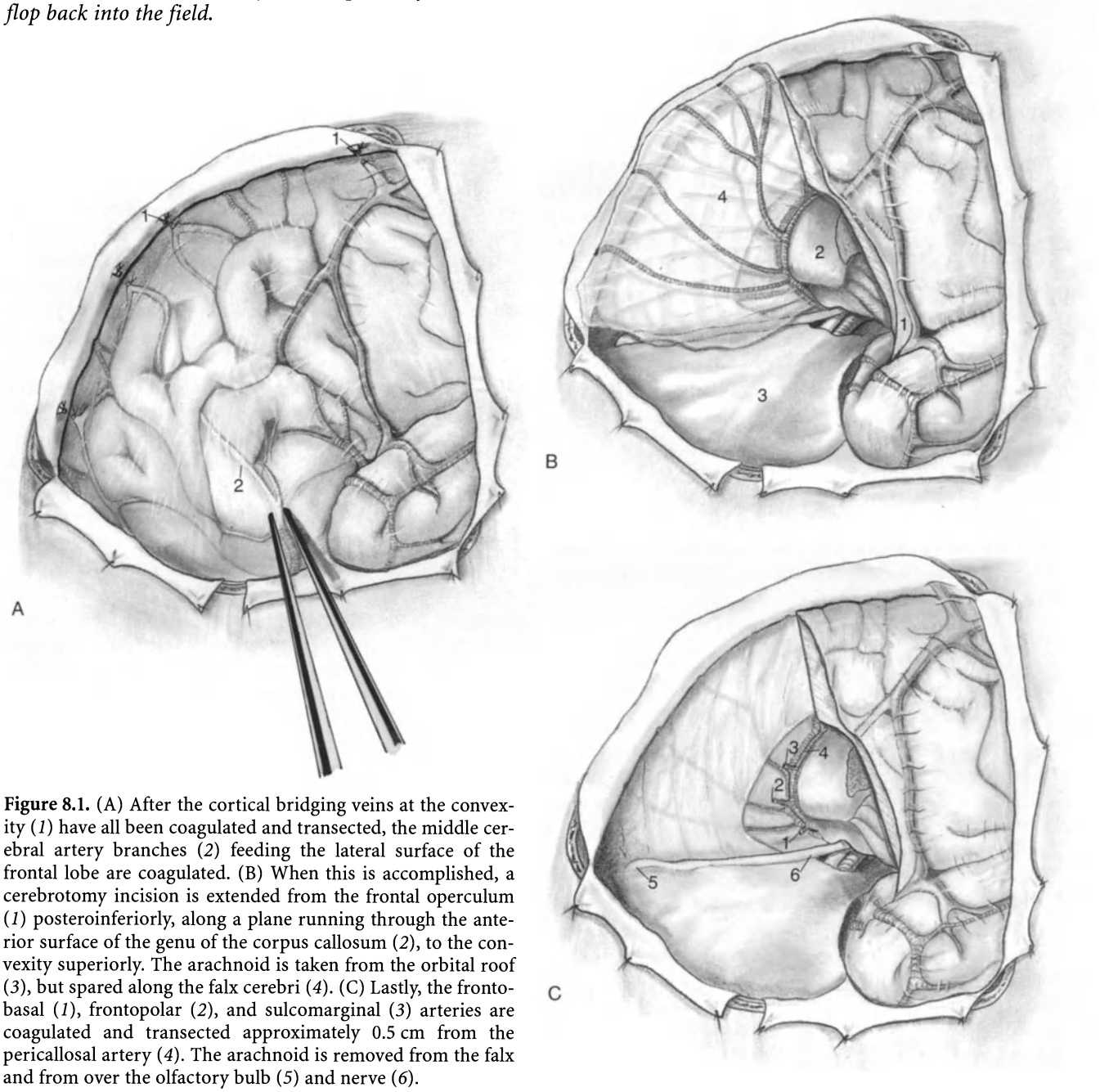

Figure 8.1. (A) After the cortical bridging veins at the convexity (*1*) have all been coagulated and transected, the middle cerebral artery branches (*2*) feeding the lateral surface of the frontal lobe are coagulated. (B) When this is accomplished, a cerebrotomy incision is extended from the frontal operculum (*1*) posteroinferiorly, along a plane running through the anterior surface of the genu of the corpus callosum (*2*), to the convexity superiorly. The arachnoid is taken from the orbital roof (*3*), but spared along the falx cerebri (*4*). (C) Lastly, the frontobasal (*1*), frontopolar (*2*), and sulcomarginal (*3*) arteries are coagulated and transected approximately 0.5 cm from the pericallosal artery (*4*). The arachnoid is removed from the falx and from over the olfactory bulb (*5*) and nerve (*6*).

into the anteromedially coursing A-1 segment of the anterior cerebral artery, and posteromedially coursing M-1 segment of the middle cerebral artery. Care must be taken not to put traction on the perforating branches coming from the bifurcation, for these arteries nourish the anterior perforated substance, basal ganglia, and anterior limb of the internal capsule. The A-1 segment is then followed anteromedially, taking care to avoid damage to the vessels going from A-1 to the chiasm, as far as the anterior communicating artery.

The surgeon then passes to the parasagittal surface of the frontal lobe at its convexity (extending from anterior to posterior along the border of the frontal lobe and superior sagittal sinus), coagulating and transecting the branches of the anterior cerebral artery at the frontal convexity, and the bridging cortical veins, as he proceeds. This separates the frontal lobe from the superior sagittal sinus and occludes all anterior cerebral artery vessels feeding it. Those segments at the frontobasal, frontopolar, and sulcomarginal arteries between the point of transection and their origin from the A-2 segment of the anterior cerebral artery are left within the subarachnoid space, to be dealt with in the last stages of the procedure.

One now proceeds to the region of the triangular operculum, and begins to coagulate and transect the cortical arteries, one at a time, as they are identified. This allows entrance into that portion of the insular cortex beneath the olfactory, triangular, and frontal operculae (all segments of the frontal lobe), permitting identification of those branches of the sylvian system (middle cerebral artery) which nourish the frontal lobe. The dissection into the region of the operculum, and then into the sylvian fissure, should be carried out with the surgeon proceeding anteromedially until he identifies the main trunk of the middle cerebral artery, taking care not to damage those branches of the sylvian system which nourish the insula, the parietal operculum, and the temporal lobe. Only by identifying the main trunk of the middle cerebral artery may one attain this degree of precision. Take care not to run the lobectomy incision through the central sulcus. This results in a contralateral hemiplegia and subjects the child to a risk of loss of parietal sensory perception. Rather, the line of cerebrotomy should run parallel and approximately 1.5–2.0 cm anterior to the fissure of Rolando. This line of cut is brought through the sulcus, separating the frontal from the triangular operculae, and then into the limbus of the insula on the right.

Once this has been accomplished, one may proceed with the cerebrotomy, proceeding from the frontal operculum inferiorly, along a line approximately 1.5–2.0 cm anterior to the motor cortex, to the convexity superiorly.

At this stage all major arterial trunks and cortical draining veins have been coagulated and severed. Individual bleeding vessels are dealt with in the usual manner. The dissection is followed inferiorly to the roof of the orbit and medially to the arachnoid adjacent to the falx cerebri, identifying the frontobasal, frontopolar, and sulcomarginal arteries. The line of cut brings the surgeon to a plane which runs along the anterior surface of the genu of the corpus callosum, exposing at the base the intact olfactory trigone, the optic nerve, and the suprasellar portion of the internal carotid artery.

Finally, approximately 0.5 cm distal to the origin of the pericallosal artery, one coagulates the frontobasal, frontopolar, and sulcomarginal arteries. They are then transected, cutting the arachnoid with them.

Temporal Lobectomy (Fig. 8.2)

The temporal lobectomy is described here as a full temporal lobectomy, recognizing that the surgeon may choose to amputate only the temporal pole, the anterior two-thirds of the temporal lobe, or the entire lobe, depending upon the specific indications for the procedure. The vascular supply to the temporal lobe comes primarily from the middle cerebral artery, whose sylvian branches course through the sylvian fissure and the limen of the insula to the mid portion of the insular cortex. They then splay out over the long and short insular gyri, curving around the temporal operculum to descend immediately, passing posteroinferiorly and sending some branches posterosuperiorly to the posterior portion of the temporal lobe. This supplies all of the convex surface of the temporal lobe. The tentorial and medial surfaces of the temporal lobe, however, are nourished by the anterior and posterior inferior temporal arteries (branches of the posterior cerebral artery).

In essence, the surgery for a temporal lobectomy entails identifying and coagulating these vessels, and then proceeding with the cerebrotomy along the planned anatomical line. There is little venous drainage from the temporal lobe into the deep middle cerebral vein (sphenoparietal sinus), somewhat more into the superficial temporal vein. The primary drainage is into the vein of Labbé.

After the temporal lobe, the opercular portions of the frontal and parietal lobes, and the angular gyrus have been exposed, one proceeds to coagulate the arachnoid from the sylvian fissure. As this is opened, retracting the temporal operculum inferiorly and slightly posteriorly, the leash of sylvian vessels coming from the main trunk of the middle cerebral artery is unfolded. One may identify readily those branches going to the temporal lobe, and separate them from the branches coursing over the insular cortex to nourish the frontal and parietal lobes. Working posteriorly from the temporal pole which borders upon the parietal lobe, one coagulates and transects the superior temporal

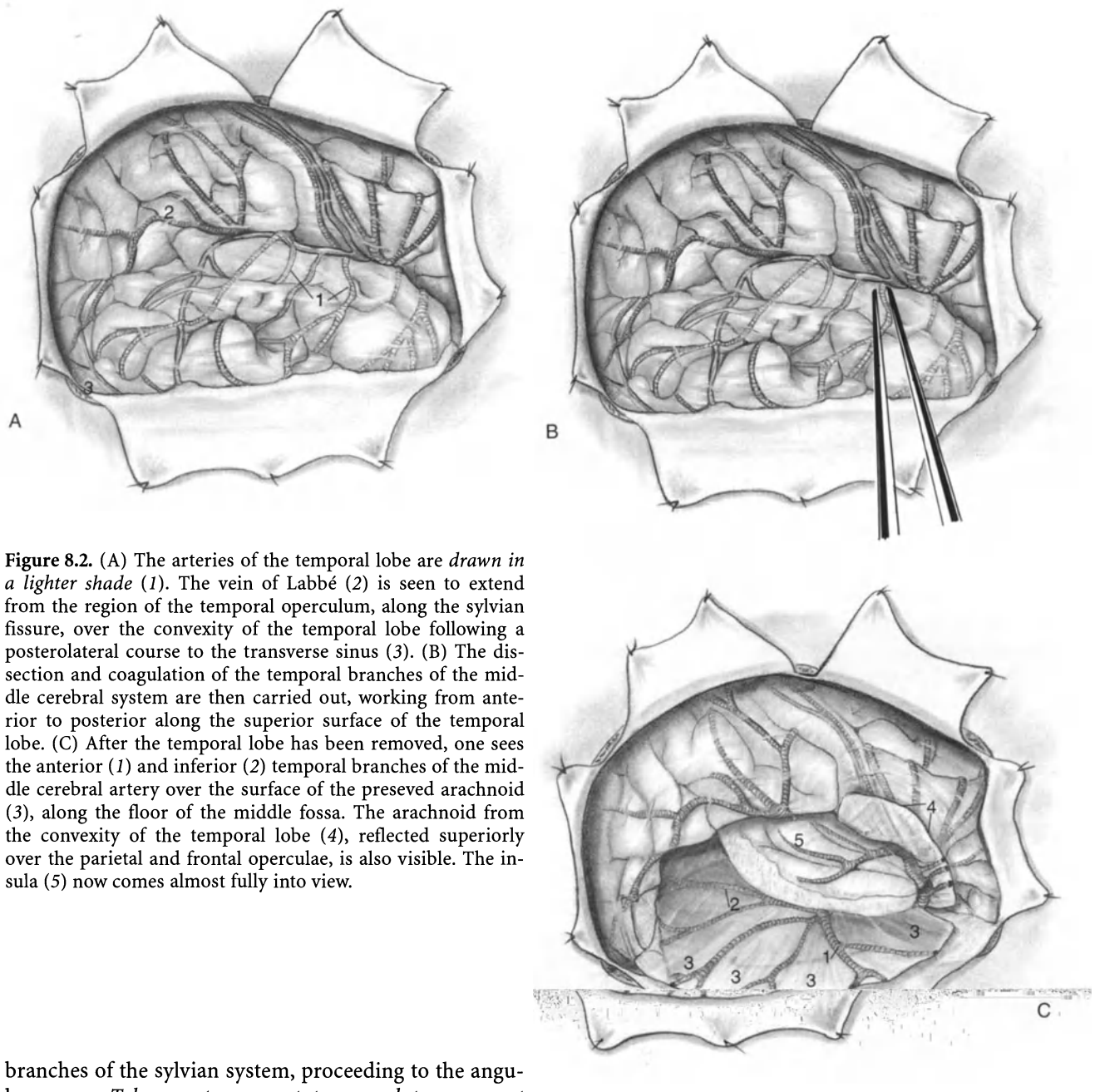

Figure 8.2. (A) The arteries of the temporal lobe are *drawn in a lighter shade* (*1*). The vein of Labbé (*2*) is seen to extend from the region of the temporal operculum, along the sylvian fissure, over the convexity of the temporal lobe following a posterolateral course to the transverse sinus (*3*). (B) The dissection and coagulation of the temporal branches of the middle cerebral system are then carried out, working from anterior to posterior along the superior surface of the temporal lobe. (C) After the temporal lobe has been removed, one sees the anterior (*1*) and inferior (*2*) temporal branches of the middle cerebral artery over the surface of the preseved arachnoid (*3*), along the floor of the middle fossa. The arachnoid from the convexity of the temporal lobe (*4*), reflected superiorly over the parietal and frontal operculae, is also visible. The insula (*5*) now comes almost fully into view.

branches of the sylvian system, proceeding to the angular gyrus. *Take great care not to coagulate recurrent branches to the insular cortex.* The line of coagulation and transection, once the temporal operculum is reached, should extend along the inferior portion of the circular sulcus of the insular cortex, placing a Telfa over the short and long gyri of the insula, protecting them from damage.

The superficial middle cerebral vein may be coagulated and transected when it is encountered, as may the vein of Labbé at its origin. When the latter is accomplished, the surgeon should pass over the convexity of the posterior portion of the temporal lobe, identify the terminal portion of the vein of Labbé and coagulate it at the cortical level rather than adjacent to the transverse sinus.

The cerebrotomy is then begun at the limen of the insula, and continued posteriorly along the inferior arc of the circular sulcus, extending from lateral to medial, separating the temporal lobe first from the amygdala and then retracting the hippocampal gyrus and isthmus of the hippocampus from the proximal portion of the cerebral hemisphere. Continue posteriorly to the angular gyrus before turning posteroinferiorly along the posterior border of the temporal lobe. The cerebrotomy should be extended through the entirety of the temporal lobe up to, but not across, the arachnoid, along its medial surface, leaving the arachnoid intact because of the branches coming from the posterior cerebral artery.

Once the cerebrotomy has entered the temporal horn (at its exit from the trigone), the chorioid plexus resting within the chorioidal fissure is identified. It is coagulated at its junction with the glomus and transected, entering the ambient cistern. The coagulation of the plexus is then extended anteriorly along the chorioidal fissure until the anterior chorioidal artery is identified, penetrating the most anterior portion of the temporal horn through the chorioidal fissure. This artery is coagulated and transected.

The cerebrotomized temporal lobe is now rolled inferiorly and laterally, dissecting it from the arachnoid. The terminal branches of the anterior and posterior inferior temporal arteries are coagulated along the most lateral and inferior surfaces of the temporal lobe, *en bloc*, from its bed in the middle fossa, leaving the arachnoid with the temporal branches of the posterior cerebral artery. Then, the arachnoid is coagulated and transected, proceeding from anterior to posterior. The same is done to the posterior cerebral branches along the inferomedial border of the circular sulcus of the insula, until the origin of the posterior cerebral branches to the temporal lobe are identified. At this time they are coagulated and transected, approximately 1.0 mm from their origin. When the entire temporal lobe and its covering arachnoid have been resected, one views the inferior half of the trigone, looking at the glomus of the chorioid plexus.

Occipital Lobectomy (Fig. 8.3)

Exposure for an occipital lobectomy entails a craniotomy, which permits access to the superior sagittal sinus medially, the torcular Herophili inferomedially, the transverse sinus inferiorly. The flap must extend laterally around the convexity of the posterior aspects of the skull, allowing the surgeon access to the occipital pole. The cortical arteries and veins are then coagulated, one

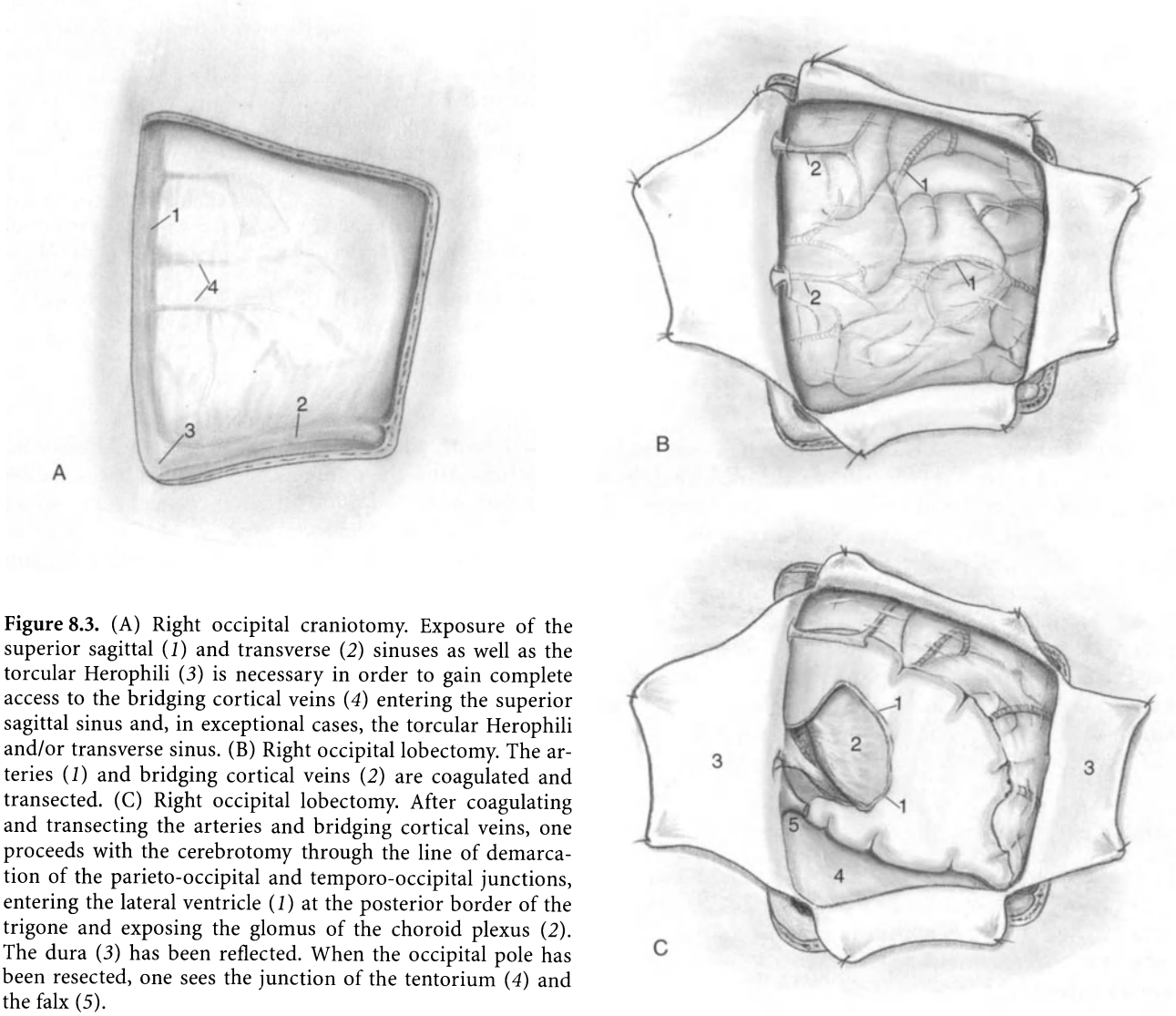

Figure 8.3. (A) Right occipital craniotomy. Exposure of the superior sagittal (*1*) and transverse (*2*) sinuses as well as the torcular Herophili (*3*) is necessary in order to gain complete access to the bridging cortical veins (*4*) entering the superior sagittal sinus and, in exceptional cases, the torcular Herophili and/or transverse sinus. (B) Right occipital lobectomy. The arteries (*1*) and bridging cortical veins (*2*) are coagulated and transected. (C) Right occipital lobectomy. After coagulating and transecting the arteries and bridging cortical veins, one proceeds with the cerebrotomy through the line of demarcation of the parieto-occipital and temporo-occipital junctions, entering the lateral ventricle (*1*) at the posterior border of the trigone and exposing the glomus of the choroid plexus (*2*). The dura (*3*) has been reflected. When the occipital pole has been resected, one sees the junction of the tentorium (*4*) and the falx (*5*).

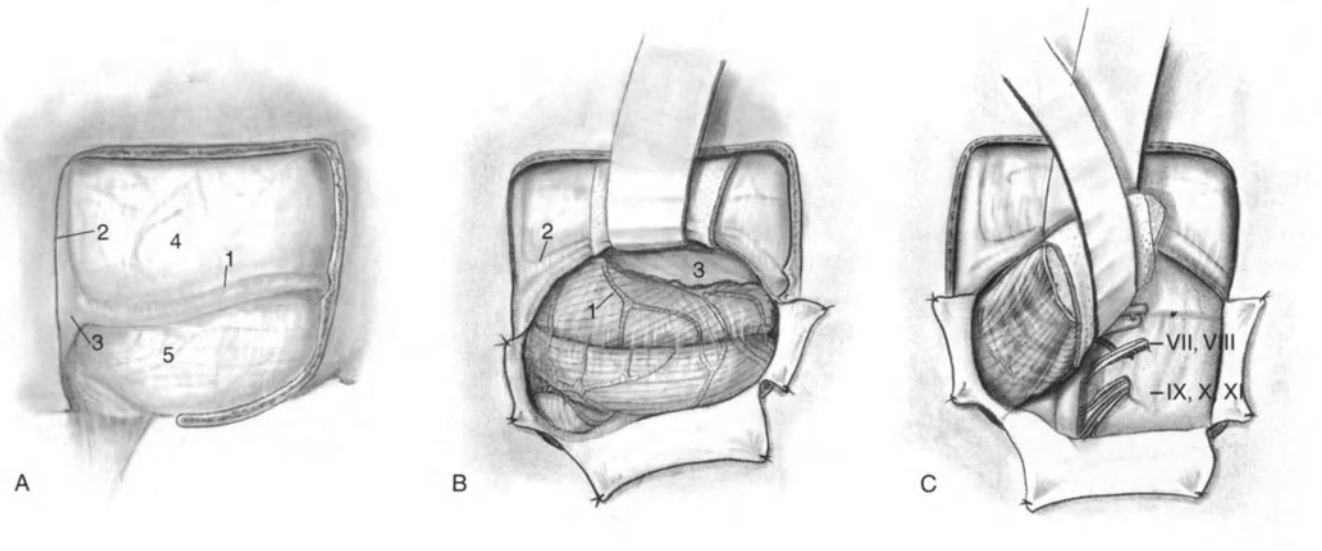

Figure 8.4. Cerebellar lobectomy. (A) The craniotomy should permit visualization of the transverse sinus (*1*), the superior sagittal sinus (*2*), the torcular Herophili (*3*), the occipital lobe (*4*), and the cerebellar hemisphere (*5*). (B) After the superior bridging veins (*1*) coursing from the superior cerebellar triangle to the transverse sinus (*2*) and tentorium (*3*) have been coagulated and transected, the cerebellum falls into the horizontal portion of the squamous occipital bone. (C) The surgeon then proceeds to coagulate the arachnoid along with the superior and inferior cerebellar arteries. Once these arteries are transected the cerebellar lobectomy is completed, allowing one to see the VII, VIII, IX, X, and XI cranial nerves (if only the most lateral tip of the cerebellar hemisphere is resected). (D) These nerves, plus the lateral surface of the pons (*1*) and the medulla oblongata (*2*), are exposed if the entire cerebellar hemisphere is resected. In either case, the undersurface of the tentorium (*3*) is also exposed.

at a time, extending first from inferolateral to superolateral and then from superolateral to superomedial, before proceeding inferiorly along the sagittal aspect of the occipital pole. The draining veins at the cortical surface are coagulated as they are encountered. Finally, the arteries along the tentorial surface of the occipital lobe, in the coronal plane of the cerebrotomy, are coagulated, proceeding from lateral to medial. The cerebrotomy is completed by passing through the most posterior portion of the trigone of the lateral ventricle. The occipital horn, when present, is included in the occipital lobectomy. It is not necessary to coagulate the glomus.

Cerebellar Lobectomy (Fig. 8.4)

It is safest to reflect an occipital-suboccipital bone flap for a cerebellar lobectomy in order to have access to the entirety, supratentorial and infratentorial, of the transverse sinus. The inferior portion of the craniotomy should extend to the point at which the squamous occipital bone passes from a vertical to an horizontal structure. After the dura has been opened in the double trapdoor fashion and the bridging cortical veins, which extend from the superior portion of the cerebellar hemisphere to the inferior sagittal sinus and tentorium, have been coagulated and transected, the cerebellar hemisphere falls into the horizontal portion of the squamous occipital bone. This exposes the superior surface of the cerebellum. One may now proceed from anterior to posterior in the coagulation and transection of the medial and lateral branches of the superior cerebellar artery. The medial branches of this artery course over the vermis. They need not be exposed. The surgeon may now begin the lobectomy at the desired point, proceeding to coagulate the superior and inferior cerebellar arteries, dealing first with the superior cerebellar arteries, as he proceeds from posterior to anterior. The inferior cerebellar arteries are dealt with subsequently. These vessels should be coagulated over the surface of the cerebellum as the arachnoid is coagulated and

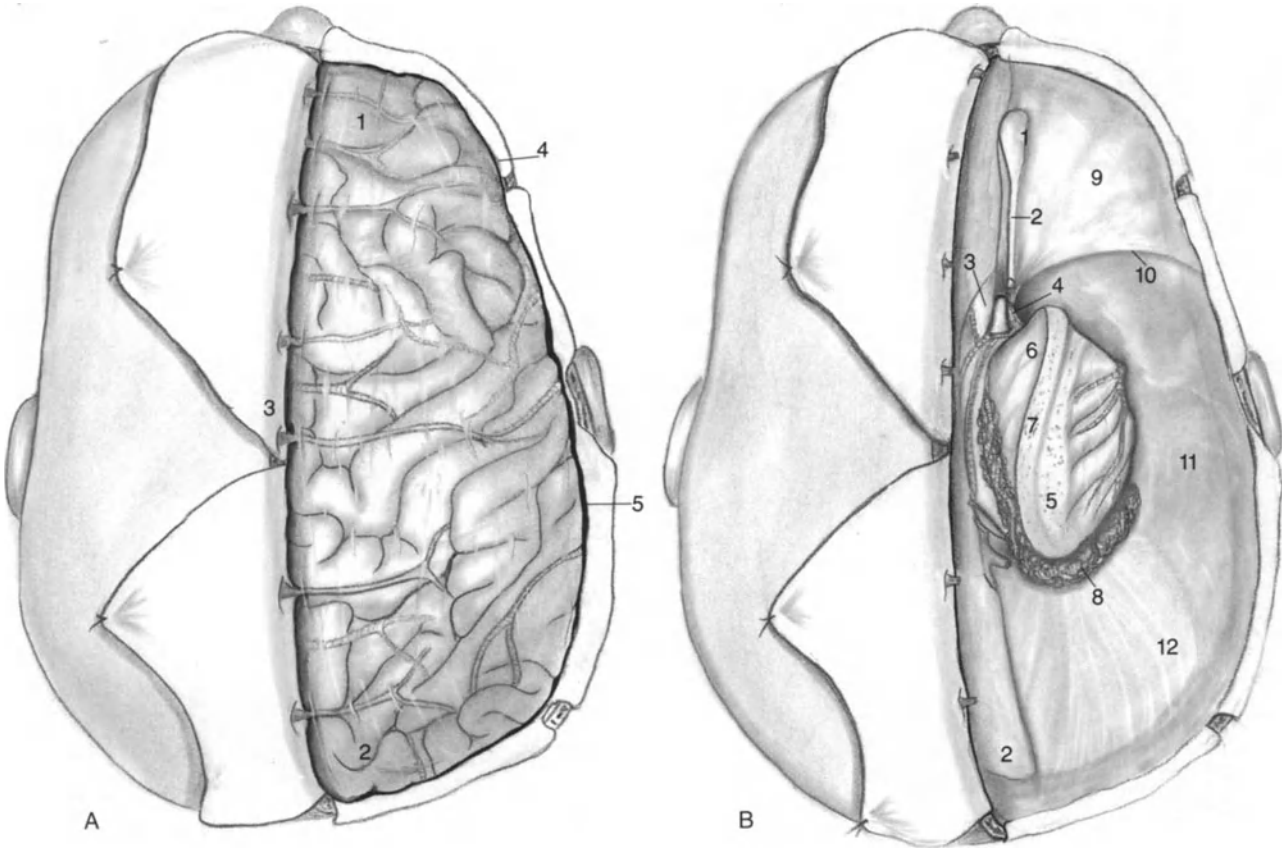

Figure 8.5. (A) When the craniotomy for a hemispherectomy has been completed, one has full visualization of the hemisphere from the frontal (*1*) to the occipital (*2*) poles, and from the superior sagittal sinus (*3*) to the lateral surfaces of the anterior (*4*) and middle fossae (*5*). (B) After removal of the hemisphere, one has a view of the olfactory bulb (*1*) and nerve (*2*), the optic chiasm (*3*), and the internal carotid artery (*4*). The thalamus (*5*), head (*6*) and body (*7*) of the caudate nucleus, and choroid plexus (*8*) are all in view. This is the time to coagulate the choroid plexus. The anterior fossa (*9*), lesser wing of the sphenoid (*10*), middle fossa (*11*), and tentorium (*12*) are exposed.

transected. Once this is done the cerebellectomy is carried out and the desired amount of lobectomy completed.

Because of the uniformly small arteries and veins over the surface of the cerebellar hemispheres (and vermis), block resection of a portion of the cerebellum may most effectively be performed with use of the laser. This may be done with great ease using the hand-held CO_2 laser, or the micromanipulator if one has already brought the operating microscope into the field.

Hemispherectomy (Fig. 8.5)

Exposure of the entire hemisphere is essential for the perfomance of a hemispherectomy, since one must have access to the anterior, middle, and posterior cerebral arteries, as well as the superior sagittal sinus, the vein of Labbé, the vein of Trolard, the deep middle cerebral vein, the sphenoparietal sinus, and all of the bridging cortical veins going to the SSS.

It is preferable to identify and coagulate the arteries and veins by proceeding from the frontal pole, dealing with the vessels of the frontal lobe in exactly the same manner as in the perfomance of a frontal lobectomy, over the sagittal surface of the parietal lobe and the occipital lobe to the torcular Herophili. Then, by dissecting along the lesser wing of the sphenoid, one may expose the sphenoparietal sinus and the main trunk of the middle cerebral artery, coagulating and transecting first the artery and then the sphenoparietal sinus.

The bifurcation of the internal carotid artery is identified. The A-1 segment of the anterior cerebral artery is followed over the optic chiasm to the anterior communicating artery, where the A-2 segment of this artery courses superiorly, at which point it is coagulated and transected. The main trunks of the frontobasal, frontopolar, and sulcomarginal arteries are identified, coagulated, and transected, approximately 0.5 cm from their point of origin from the pericallosal artery. The inferior temporal branches of the posterior cerebral and the anterior chorioidal arteries are the only remaining ar-

terial structures. The vein of Labbé is the only remaining venous structure. The anterior choroidal artery is identified at the ambient cistern (where it perforates the chorioidal fissure), coagulated, and transected.

The dissection is extended posteriorly, by following the tentorial edge and retracting the hemisphere superiorly and slightly laterally so as to expose the inferior temporal branches of the posterior cerebral artery. They are coagulated and transected approximately 0.5 cm from their individual points of origin from the posterior cerebral artery. Lastly, the vein of Labbé is coagulated and transected on the cortical surface from whence it bridges to enter the transverse sinus. This completely devascularized the cerebral hemisphere, which is then allowed to flop within the hemicranium, exposing the corpus callosum from the genu to the splenium. The cerebrotomy is extended through the forceps minor and then along the parasagittal plane of the body of the corpus callosum posteriorly to, and through, the forceps major. This brings the surgeon into the right lateral ventricle, which also falls away, leaving the hemisphere attached to the brainstem anteriorly at the amygdala, and posteriorly at the isthmus of the hippocampus. The cerebrotomy is extended posteroanteriorly from the lateral surface of the amygdala (coagulating and transecting the arachnoid at the choroidal fissure) to isthmus of the hippocampus at the posterolateral surface of the thalamus. The freed cerebral hemisphere may be lifted from the right hemicranium.

9 Epilepsy

"I'm going out to fetch the little calf
That's standing by the mother. It's so young
It totters when she licks it with her tongue.
I shan't be gone long – You come too."

ROBERT FROST, The Pasture

"Crito, We owe a cock to Aesculapius."

SOCRATES, Dying Words

Introduction

In this chapter we consider the possibilities and conditions available to a multidisciplinary group charged with the responsibility of managing *drug-resistant partial epilepsies.*

One may consider epilepsy to be *drug resistant* when the patient continues to suffer seizures after 2 years of medical treatment with different drug combinations. It is advisable to consider the available surgical procedures when faced with a patient with drug-resistant partial epileptic crises. In such an event the patient should be sent to a center, where the appropriate history, physical, electrophysiological, neuropsychological, and structural/functional imaging studies may be performed preparatory to surgery.

The prevalence of epilepsy in pediatric ages is about 0.4%, the incidence about 0.6–0.7‰ [1].

Pediatric epilepsy surgery is a field of growing interest, because:

1. The majority of seizures begin in childhood [2].
2. Intractable epilepsies in children very often determine devastating sequelae, such as serious behavioral disturbances, cognitive impairment, regression in intelligence, and major neurological deficits.
3. Most lesional pediatric epilepsies are due to very slow or nonevolutive structural lesions, such as low-grade gliomas, hamartomas, dysembryogenic neuroectodermal tumors (DNETs), and cortical areas of dysplasia, which very often do not greatly limit life expectancy.

The result of the coexistence of these factors is that many epileptologists tend to indicate surgery as soon as possible, trying to avoid, or at least to minimize, neurological sequelae. Nevertheless the delay from the diagnosis of partial drug-resistant epilepsy to surgery is still too high (3–15 years, with a mean of 7–8 years). This latter may be due to the high percentage of spontaneous remission in childhood epilepsies. However, most infantile epileptic syndromes may be traced to a lesional cause.

Benign infantile epileptic syndromes, which require a waiting strategy, are:

1. Benign rolandic epilepsy
2. Childhood epilepsy with occipital paroxysms
3. Benign psychomotor epilepsy

Because of the frequently good prognosis, these syndromes very often are treated only with pharmacological measures. In contrast the pediatric epileptic syndromes which may more correctly be considered for surgical treatment are:

1. Cortical migrational defects
2. Neurofibromatosis
3. Tuberous sclerosis
4. Rasmussen's encephalitis
5. Sturge-Weber syndrome
6. Lesional infantile spasms

This is only a schematic list of the syndromes generally considered characteristic of the pediatric age, although almost all the different kinds of epileptogenic lesions may be found in both adults and children.

If the epileptogenic zone does not lie within the motor strips or other eloquent areas, one may plan a cortical resection with or without a lesionectomy. This procedure, and its various modifications, is without a doubt the most commonly used operation for the treatment of drug-resistant epilepsy. However, other such surgical procedures as multiple subpial resections, hemispherectomy, callosotomy, and chronic stimulation of the vagus nerve may be indicated and afford excellent clinical results.

Results

The results of epilepsy surgery in children, considered as *control of the seizures*, are not very different from those in adults. Maehara [3], in 1996, compared the results in adult and pediatric ages, with the percentages of seizure-free patients shown in Table 9.1.

However, the control of seizures is not the only goal of early surgery. The quality of life is another important element of evaluation. Unfortunately, in this field, there are really dyshomogeneous methods of evaluation, and the results are very often not comparable. The trend is to operate earlier and earlier, to avoid or to limit neuropsychological and cognitive impairment, but real superiority of early surgery has not yet been demonstrated [4].

Nevertheless, the longer the epilepsy, the poorer the prognosis of seizure control, and this is sufficient to justify surgery as early as possible, especially when a symptomatogenic lesion is evident. The superiority in quality of life has not been demonstrated, but it seems improbable that the opposite would be true.

In conclusion, pediatric epilepsy surgery is a reality, which may definitively cure many children, freeing them from seizures, drugs, and psychosocial problems, with a permanent morbidity risk significantly lower than in adults.

Table 9.1. Comparison of epilepsy surgery in adults and children

	Temporal	Extratemporal
Adults	60/90 (66.6%)	7/28 (25%)
Children	6/9 (66.6%)	4/12 (33.3%)

Definitions

The *epileptogenic zone* is the *"area of cortex that is necessary and sufficient for initiating seizures and whose removal (or disconnection) is necessary for complete abolition of seizures."* This definition, originally stated by Tailarach et al. [5] in 1974, was reported by Luders, Engel and Munari [6] to introduce the general principles essential to an understanding of epilepsy surgery. In this chapter we use terms which are necessary to express clarity of thought and meaning.

1. *Irritative zone:* "Area of cortex that generates interictal spikes": may be delineated by electrophysiological investigations [6].
2. *Ictal onset zone:* "Area of cortex where seizures are generated, including areas of propagation under certain circumstances": may be delineated by electrophysiological investigations [6].
3. *Epileptogenic lesion:* "Structural abnormality of the brain that is the direct cause of the epileptic seizures": may be delineated by neuroimaging and pathology [6].
4. *Symptomatogenic zone:* "Area of brain that produces initial clinical symptomatology": may be delineated by behavioral observation and clinical reports [6].
5. *Functional deficit zone:* "Cortical area of nonepileptic dysfunction": may be delineated by neurologic examination, neurospychological testing, EEG, positron emission tomography (PET), and single photon emission computed tomography (SPECT) [6].

The epileptogenic zone comes from the intersection of the above-defined zones: it is not an anatomical zone, nor a functional or a lesional one, *but* it is a conceptual zone. Nothing other than the postsurgical recovery can confirm that the delineated epileptogenic zone is the correct one: this makes it evident that the presurgical evaluation of the patient must be as exhaustive as possible.

Epidemiology

Ranking second only to cerebrovascular accidents, epilepsy is among the most frequent forms of cerebral pathology. The epidimiologic definition of *prevalence*, as put forth by the WHO in 1966, is "the number of cases or of patients, in a given population, without any distinction between new or old cases. It may be expressed in total numbers or, more often, as a fraction of the total population. *Prevalence* is always reported for a given year. Throughout the world it has been calculated to range between 4.9% and 14.5%, as presented in Table 9.2.

Incidence is defined by the WHO as the "number of cases of a given disease diagnosed, or of patients afflicted by the given disease, in a specified year and population." Hauser [12] estimated that in the United States there are somewhere between 70,000 and 130,000 new cases per year. However, in the light of this book, the important consideration is neither *prevalence* nor *incidence. It is that number (fraction) of the epileptic*

Table 9.2. Prevalence of epilepsy

Authors	Population sample	Year	Value
Haerer et al. [7]	Copiah County, Mississippi (USA)	1978	6.7%
Joensen [8]	Faroes (Denmark)	1980	7.6%
Ishida [9]	Okayama (Japan)	1975	14.5%
Bondestam et al. [10]	Zanzibar	1988	4.9%
Hauser et al. [11]	Rochester, Minnesota (USA)	1980	6.6%

population which may be considered candidates for surgery. In 1983 Ward [13] estimated that the number of people in the United States who have drug-resistant epilepsy amounted to 360,000. He estimated that at least 54,000 of them could benefit from surgical treatment. Uthman [14] calculated in 1990 that the number of surgical procedures performed for epilepsy in the United States was approximately 400/year. Furthemore, a recent study [15] revealed that the total costs for medical care of an epileptic patient who is cured of the disease early add up to $472.00, but amount to $138,602 for those patients with medically uncontrollable epilepsy. Antiepileptic drugs were the major components of the total costs.

Classification of Epileptic Seizures

Since the general term *epilepsy* is used to describe a multitude of clinical manifestations and syndromes, it is best to outline the most widely used classification of epileptic seizures.

This is not a book on epilepsy, so the reader with particular interest must consult specific texts. The classification we use here (Table 9.3) was proposed in 1981 by the Commission on Classification and Terminology of the International League Against Epilepsy [16]. The individual epileptic types are grouped according to prevalently clinical criteria.

Patient Selection

There is, unfortunately, a great discrepancy between the number of epileptics potentially susceptible to surgical treatment and the fraction of these who are referred to treatment centers for evaluation. The number and complexity of diagnostic modalities and specialized personnel are such that these evaluations may only be conducted in centers, and this is certainly true for invasive techniques.

Noninvasive Methodologies

These consist of history, physical, extracranial EEG evaluation, long-term video-EEG monitoring, structural and functional imaging studies, and neuropsychological and psychiatric studies.

History and Physical

Concerning general information, specific attention is given to such causes as birth injury, febrile convulsions, trauma, and early left-handedness which may have been

Table 9.3. Classification proposed by the Commission on Classification and Terminology of the International League Against Epilepsy

A. Partial seizures
  1. Simple partial seizures
    a) With motor components
      (1) Motor signs without a march
      (2) Motor signs with a march (jacksonian)
      (3) Versive
      (4) Postural
      (5) Phonic (either vocalization or an arrest of speech)
    b) With sensory manifestations
      (1) Somatosensory
      (2) Visual
      (3) Auditory
      (4) Olfactory
      (5) Gustatory
      (6) Vertiginous
    c) With autonomic manifestations
    d) With psychic manifestations
      (1) Dysphasia
      (2) Dysmnesia
      (3) Cognitive
      (4) Affective
      (5) Illusory
      (6) Hallucinatory
  2. Complex partial seizures
    a) With simple onset and then a loss of consciousness
      (1) Without automatisms
      (2) With automatisms
    b) With loss of consciousness from the beginning
      (1) Without automatisms
      (2) With automatisms
  3. Partial seizures secondarily generalized
    a) Simple partial seizures followed by generalized seizures
    b) Complex partial seizures followed by generalized seizures
    c) Complex partial seizures followed by complex partial seizures and in turn by generalized seizures
B. Generalized seizures
  1. Absences
    a) Typical
      (1) Simple loss of consciousness
      (2) With slight clonic component
      (3) With slight atonic manifestations
      (4) With slight tonic manifestations
      (5) With automatisms
      (6) With autonomic components
    b) Atypical
      (1) With more intense manifestations
      (2) With progressive onset and/or resolution
  2. Myoclonic seizures
  3. Clonic seizures
  4. Tonic seizures
  5. Tonicoclonic seizures
  6. Atonic seizures
C. Nonclassified seizures

subsequently suppressed. Also, one should enquire in detail about the individual particulars of the seizure itself from both the patient and the family. Similarly, details concerning individual drugs used and results are most important. Careful notes concerning the critical phases of the seizures, such as events over time and in response to individual medication, round out the initial history. Such a history, in fact, may already lead the clinician to suspect whether the seizures may or may not be surgically treatable: temporal monomorphism, absence of early signs referable to eloquent areas of the brain, seizure polymorphism, early dysphasia.

A careful description of the ictal semiology, delineating the symptomatogenic zone (together with video-EEG), is of great value in defining the epileptogenic zone and programming the individual strategies to plan the intracranial recordings.

EEG and Video-EEG

Of course, scalp EEG is the point of departure. Interictal EEG has two main roles:

1. To define the irritative zone
2. To define the functional deficit zone

Interictal spikes are the distinctive element of the irritative zone: their identification may be useful in determining the side and the localization of the epileptogenic zone, but one must remember that the epileptogenic cortex must be removed, not the spikes! However, continuous and constant interictal EEG alterations in an anatomical area identified clinically on the basis of seizure phenomenology should be considered very important.

Interictal EEG, together with neuropsychological testing and functional imaging exams, is important in the definition of the functional deficit zone, showing abnormalities other than spikes.

Long-term video-EEG monitoring is certainly one of the most significant noninvasive diagnostic techniques: simultaneous examination of the ictal scalp EEG recording and the ictal behavior of the patient is very useful to define the boundaries of the symptomatogenic zone and the ictal onset zone: sometimes (and maybe even "very often") extracranial video-EEG, together with the other noninvasive examinations, may lead to a surgical intervention, without any invasive diagnostic procedure, especially in the case of temporal lobe epilepsy. It is stressed that the patient must be tested during the seizure: this is the only way to avoid dangerous misunderstandings (how does one determine whether the patient can speak without stimulating him or her?)

Diagnosis by Imaging Studies

The three primary goals of imaging are:

1. Structural analysis to define an epileptogenic lesion
2. Morphofunctional analysis to define the functional deficit zone
3. Morphological analysis to plan the surgical procedure and to evaluate its results

The value of magnetic resonance imaging (MRI) is such that any comment upon it at this time is trivial. One may be sure that the continuous improvements in technology and versatility are such as to permit us to expect that in the near future there will be a reduction to less than 10% of the cases of idiopathic epilepsy.

PET and SPECT are based upon concepts which permit the quantification of blood flow or tissue metabolism. The interictal use of these techniques may reveal areas of cortical hypometabolism, a good index of lateralization.

Peri-ictal use of SPECT is one of the most recent applications of this technique, and it seems to be very useful in delineating the epileptogenic zone [17].

Invasive Methodologies

Wada Test

The intracarotid injection of sodium amobarbital is a very useful, often indispensable, neuropsychological test which has two goals:

1. To determine language lateralization
2. To determine the memory supportive capacity of mesiotemporal structures contralateral to a planned resection

The dispensability of a defined epileptogenic zone depends (only!) upon the results of this test, introduced by Wada in 1949 [18].

Intracranial Recordings

If we sum the information provided by the history, physical, neuropsychological, neuroimaging, and neurophysiological (EEG and video-EEG) evaluations, we should be able to identify between 22% and 40% of those cases which have solid criteria indicative of surgical treatment [19]. In some cases, however, and especially in the more deep-seated ones, surface neurophysiological studies fall short of localizing correctly the ictal onset zone and the irritative zone: the voltage registered at any given distance from its source is inversely proportional to the square of the distance [20]. Hence, for the present there may be indications for intracranial recordings. These may be classified as invasive or semi-invasive.

Table 9.4. Semi-invasive electrodes

	Nasopharyngeal	Sphenoidal	Foramen ovale	Epidural peg electrodes
Location	Basifrontal, mesial and inferior temporal lobes	Basifrontal, mesial and inferior temporal lobes	Mesial temporal structures	Cerebral convexity
Advantages	Ease of placement	Relatively risk-free placement	Intracranial location	Intracranial location
Disadvantages	Poorly tolerated, many artifacts	Extracranial location	More invasive	More invasive
Complications	Nasal congestion	Infection	Infections, hemorrhage	Infections, hemorrhage

Table 9.5. Invasive electrodes

	Intercritical recording	Ictal recording	Lateralizing	Stereotaxic	Stimulation	Insertion
Intracerebral	Yes	Yes	Yes	Yes	Yes	Bur hole
Subdural strips	Yes	Yes	Yes	No	Yes	Bur hole
Subdural grids	Yes	Yes	No	No	Yes	Craniotomy
Corticography	Yes	No	No	No	Yes	Craniotomy

Semi-invasive Electrodes (Table 9.4)

The signature of these electrodes is that they are brought to the immediate vicinity of the cerebral region to be studied, but that they are not put in direct contact with the brain: nasopharyngeal, sphenoidal, foramen ovale, and epidural peg electrodes. In children, it is quite difficult to use these effectively.

Invasive Electrodes (Table 9.5)

These may be placed directly within the brain substance, in the subdural or epidural spaces, or along the surface of an operative field. Thus, they may be described as intracerebral electrodes, sub- or epidural electrodes, or intraoperative electrocorticographic electrodes.

Intracerebral electrodes must be positioned stereotactically, and because they are multicontact (5–18), it is possible to record electrical activity from the cerebral neocortex, white matter, deep structures, and intracranial lesions. When they are implanted with the main goal of lateralizing the ictal onset zone, with a near-standard strategy, it is correct to define the technique as "depth electrodes" (DE); when the implant strategy is strictly individualized, it is better to name it "stereo-*e*lectro*e*ncephalography" (SEEG) [21].

Subdural and epidural grids must be implanted via craniotomy, the strips via multiple large bur holes. The main disadvantages of sub- or epidural placements are craniotomy and infections [22] of the intracranial electrodes, the enormous technical implications and possibilities of intracerebral hemorrhage, and the fact that intraoperative electrocorticography (the oldest "direct" recording) is able only partially to delineate the irritative zone [23, 24].

These electrodes are then used for long-term intracranial video-EEG: this is the best way to delineate the ictal onset zone in the light of precise intracranial recordings and patient ictal behavior. A majority of well-selected patients may so be proposed for surgical treatment.

Operative Procedures

There are many surgical alternatives to the treatment of drug-resistant partial epilepsies. The most commonly used are shown in Table 9.6. The majority of operations performed at the present time consist of cortical resection. These procedures are performed both on patients with documented epileptogenic lesions and on those with cryptogenic epilepsy. Of course, the presence of an identifiable lesion polarizes the clinician's attention and orients him toward resection of the lesion and the surrounding epileptogenic parenchyma.

Table 9.6. Operative procedures

A. Resective surgery
  I. Unilobar resections
    1. Temporal resections
      a) Lesionectomy
      b) Tailored cortical resection
      c) Standard cortical resections
        "En bloc" anterior temporal lobectomy
        Anteromedial temporal lobectomy
        Amygdalohippocampectomy
          Trans-sylvian
          Transcortical
        Lesionectomy + tailored or standard cortical resection
    2. Extratemporal resections
      a) Lesionectomy
      b) Tailored cortical resection
      c) Lesionectomy + tailored cortical resection
  II. Multilobar resections
    1. Lesionectomy
    2. Tailored cortical resection
    3. Lesionectomy + tailored cortical resection
  III. Hemispherectomy
B. Disconnection surgery
  I. Callosotomies
    1. Anterior callosotomy
    2. Total callosotomy
C. Alternative surgery
  I. Multiple subpial transections
  II. Chronic intermittent vagal stimulation
  III. Thalamic stimulation

Resective Surgery

Cortical resection consists of removal of the epileptogenic zone and may be performed if:

1. This zone has been clearly identified preoperatively.
2. This zone does not include eloquent cortical areas.
3. The natural history of the underlying disease is not such as to outweigh the undertaking of surgery.

Temporal Lobe Resection

Temporal lobectomy deserves very special consideration. Partial epileptic seizures of temporal lobe origin are the most common drug-resistant seizures, and this operation is the one most commonly performed [25, 26]. One must, however, know that the term "temporal lobectomy" is used loosely for many operations performed on the temporal lobe. These different procedures may be more or less aggressive, consist of resecting neocortex or paleocortex, and have different topographic schemata.

"En Bloc" Anterior Temporal Lobectomy

Murray Falconer, in 1972, was one of the first to describe this procedure [27]. His technique consisted of removing the temporal lobe with its mesial structures (uncus, amygdala, hippocampus) but sparing the superior temporal gyrus. The posterior extent of the resection was 5.5 cm from the temporal pole on the dominant hemisphere and 6 cm on the nondominant side. Crandall's procedure is identical, except that the resection is only carried posteriorly 4.5 cm on the dominant side [28].

Anteromedial Temporal Lobectomy

Kraemer and Spencer are of the opinion that the superior temporal gyrus should be spared, and that the middle and inferior temporal gyri should be resected only as far posteriorly as 3.0–3.5 cm, independent of the side. In addition, the large part of the amygdala along with 3.0–4.0 cm of the hippocampus and parahippocampal gyrus, and the entirety of the uncus, are removed [29].

Amygdalohippocampectomy

A far less extensive procedure is one that involves resecting only the mesial temporal structures. In this case, access to the uncus, amygdala, parahippocampus, and hippocampus may be trans-sylvian, transcortical, or stereotaxic. The trans-sylvian approach was proposed by Wieser and Yasargil in 1982 [30], whereas the transcortical approaches were proposed by Olivier [31], who went through the superior temporal gyrus, and by Niemeyer [32], who went through the middle temporal gyrus. Shimizu [33] recommended an approach through the inferior temporal gyrus.

Technique

Temporal lobectomy as a treatment for partial epilepsy that originates in the temporal lobe is performed along the following lines:

Exposure

The patient is positioned supine, with the head rotated 80° and a pillow placed under the shoulder (so as to facilitate jugular venous drainage). For the head position, refer back to Chap. 1 ("Positioning"); for the skin flap, dural opening, and craniotomy, refer to the appropriate chapters in this text.

Lateral Resection

"En bloc" resection of the lateral portion of the temporal lobe permits a detailed and anatomically oriented histopathological study of the resected tissue. For this same reason it is also advisable to resect the medial temporal structures. Subpial dissection is preferred. It diminishes scarring and limits deafferentation, and assists in avoiding damage to vessels that pass to other cortical areas. Blunt dissection, using a Penfield #4, is often possible though an ultrasonic aspirator may become necessary. The blunt dissection is possible if one follows the fissure lines where a redoubling of arachnoid facilitates the maneuver [25].

Resection of neocortex is begun with an incision through the pia mater after the surface is coagulated, and then the cerebrotomy is effected from either the superior or middle temporal gyrus (depending upon the surgeon's plan) and extended inferomedially until the insertion of the tentorial edge onto the anterior clinoid is reached. Posteriorly, the cerebrotomy is extended to the root of the temporal lobe. After the lateral portion of the temporal lobe is resected, one proceeds to the medial lobectomy.

Medial Resection

The key to the medial resection is the identification and opening of the temporal horn.

Brief Anatomical Survey of the Temporal Horn

The temporal horn describes a curve whose concavity faces anteromedially as it passes posterior to the thalamus to extend towards the temporal pole, ending 2.5 cm from its tip. The projection of the lateral surface of the temporal horn is marked rather precisely by the superior temporal sulcus. The roof of the temporal horn is concave towards the cavity and bordered, from lateral to medial, by the tapetum, the tail of the caudate nucleus, and the optic tract. Superior to these structures are the claustrum, the basal ganglia, and the internal capsule. The tail of the caudate nucleus (within the roof of the temporal horn) ends at the amygdala. This latter, therefore, is located anterosuperiorly to the tip of the temporal horn. The floor of the temporal horn is convex towards the cavity, a result of the bulges determined medially by the hippocampus and laterally by the collateral sulcus. The very membranous medial surface consists of the choroidal fissure.

Surgical Procedure

The line of transcortical entrance into the temporal horn should be determined on a case by case basis: the variation in anterior extent of the tip of the temporal horn is very variable in the different age categories, especially among the varying pathological conditions. Temporal lobe gliomas deform and displace it, atrophic lesions permit dilation, etc. It is not wise to set arbitrary coordinates. The same applies to the choice of which gyrus (superior, middle, inferior) to traverse. Personalize the coordinates on the basis of high-resolution MRI scans, and know the anatomy of the temporal lobe, amygdala, hippocampal complex, etc.

Preferably, these rhine-encephalic structures targeted for resection should be taken out "en bloc," performing first a subpial resection, proceeding posteriorly along the hippocampus to the coronal plane, passing to an imaginary line running through the colliculi (posterior surface of the mid-brain).

Complications

The specific neurologic deficits which may result from temporal lobectomy are paralysis, field defects, cognitive or psychologic disturbances, and dysphasia.

1. Hemiplegia may result either from direct internal capsule damage or indirectly from spasm or occlusion of the middle cerebral artery, the anterolateral or anteromedial perforators, and the medial choroidal artery.
2. Field defects: whereas homonymous quadrantanopsia is generally not perceived by the patient, a homonymous hemianopsia may be quite disturbing. Generally, the patient adapts to the deficit psychologically, but is at risk during sports, cycling, driving a motorcar, etc. This deficit, as the quadrantic cut, may result either from direct sectioning of the optic tract, or damage to the posterior cerebral artery.
3. Dysphasia: the dramatic impact which aphasia, or even dysphasia, may have on a growing child or adolescent is indescribable. Hence, the importance of identifying the dominant hemisphere, plotting cautiously the posterior line of the temporal lobe resection, and at times performing intraoperative electrocorticography (*very* rarely possible in children!). Some authors on surgical epilepsy state that some degree of nominal dysphasia temporarily occurs when the dominant hemisphere is operated on.
4. Memory deficits: If there is already present contralateral (to the operated side) hippocampal damage, postoperative memory disturbances may occur.
5. Cognitive and psychological deficits: Similar to the memory disturbances, but less frequent, these may occur when mesial temporal structures are resected. The patients may behave as uninhibited personalities when the limbic system is ablated. This, fortunately, is generally temporary.

Results

The results, quite naturally, may be considered an expression of the degree and quality of preoperative studies, the extent of mesial and/or lateral resection, and the selection of cases.

It is of value to cite a recent report by Wyler [34] on 70 patients operated on for temporal lobe epilepsy, none of whom had an MRI identifiable lesion and all of whom were studied with the full battery of pre-epilepsy surgery tests. Only 38% of the 34 patients on whom an anterior temporal lobectomy (through the line of the anterior surface of the brainstem) were totally cured, *but 69% of the 36 patients in whom the temporal lobectomy was brought as far posteriorly as the collicular plate plane were cured.* The work by Nayel [35] corroborates this conclusion: in 90 patients, only 20% of those with an anterior resection were cured whereas 75% of those with a posterior resection enjoyed a cure.

Comparing simple lesionectomy with temporal lobectomy, Jooma et al. [36] report that only 20% of the former but 93% of the latter are cured. The group from Grenoble [37] support these data with their series of highly individually treated patients: 76% of those with lesions and 100% of those without a lesion (82% overall cure rate) were cured.

Extratemporal Resection

Resections of cortical areas beyond the limits of the temporal lobe are planned on the basis of the same battery of clinical, imaging, and physiologic studies. It is always necessary to identify the epileptogenic zone and to outline the parameters of the corticectomy along the lines of objectively obtained information.

Extratemporal epileptogenic zones are more varied in clinical manifestations, regarding origin and diffusion. Thus, it is rare to identify an operative plan without invasive electrophysiological studies. Extratemporal epilepsy surgery requires extensive structural and human resources. The surgery itself is more involved, since the involved cortex must be resected, without damaging the underlying (association fibers) of white matter. Angiography and MRI assist one in planning the surgery, and intraoperative sparing of juxtaposed pial surfaces helps to protect the microvasculature while performing the subpial gray matter resection [38]. Though cortical mapping is a reality in adults, and very helpful in resecting extratemporal cortical foci, in children intraoperative corticography – which entails local anesthesia – is not realistic.

Hemispherectomy

This procedure is the only form of surgical treatment of epilepsy performed exclusively in children. The criteria for its use are:

1. Unilateral hemispherical disease
2. Preexisting contralateral hemiplegia
3. Clearly lateralized drug-resistant partial seizures

The most common underlying pathological processes are:

1. Rasmussen's encephalitis
2. Infantile hemiplegia
3. Porencephalic cysts
4. Hemimegalencephaly
5. Sturge-Weber encephalotrigeminal angiomatosis
6. Diffuse cortical dysplasia

When the proper indications are present, the surgical results are quite satisfactory. The technique put forward in 1950 by Krynauw [39] consists of complete hemispherical corticectomy, conserving the diencephalon and basal ganglia. The most feared complication is superficial hemosiderosis, late in onset, which has been considered the result of repeated subdural bleeds into the cavity left by the hemispherectomy. The clinical picture consists of hydrocephalus, cerebral deterioration, intracranial hypertension, and death. Dural plication designed to diminish the empty space, subdural drains, or the performance of a "functional hemispherectomy" have been proposed as prophylactic or alternative procedures. The latter procedure was described by Rasmussen in 1983 [40]. It consists of separating functionally the involved hemisphere from the contralateral hemisphere and from the brainstem...without interrupting the vascular supply. In 1995 Comair reported that the results are quite satisfactory both in controlling the seizures and in avoiding the complications [41].

Disconnection Surgery

Callosotomy

Partial or total longitudinal sectioning of the corpus callosum may be performed in single or multiple sittings [42], either surgically or with stereotaxic radiofrequency. The purpose is to interrupt the propagation of the discharges from one hemisphere to the other, which finds its clinical expression in avoidance of the devastating effects of synchronous bihemispherical seizures. And, in light of the fact that this procedure only rarely results in total disappearance of the seizures (a totally unexpected result!), it is indicated as a palliative in patients with pharmacoresistant seizures not expected to respond to other surgical procedures. The most common goal is to avoid the sudden and dramatic falls due to bilateral synchronous spreading of the seizure.

Table 9.7. Callosotomy results

	No. patients	Total disappearance of the seizures	Significant improvement > 50%	Insignificant improvement
Engel [44]	197	5%	71%	24%
Wyler [45]	66	11%	68%	18%

Recently [43], a relationship between the preoperative slow-wave EEG activity and positive clinical results from such a procedure have been reported. Regarding extent of section of the corpus callosum, today most epilepsy surgeons prefer to limit it to the anterior two-thirds at the first sitting. If necessary, they suggest returning at another time to bring the sectioning posteriorly across the hippocampal commissure.

Results

Notwithstanding the understandable criticisms (based mainly upon the newness and as yet not understood physiopathology) of this procedure, the overall results seem encouraging, as Table 9.7 illustrates. Great controversy rages over the value of a second sitting, to extend the sectioning posteriorly. Some [45] sustain that this is useless, others support it [46]. A third group [47] see no relationship between extending the resection and the clinical results.

Complications

Pilcher and Ojemann [48] divide the complications into surgical and functional: meningitis, hydrocephalus, hemiplegia, edema, frontal infarction, disconnection syndrome (90%), split brain syndrome (30%), and realization of a latent neurological deficit which had been masked or compensated by interhemispherical connections (15%).

Alternative Surgery

Multiple Subpial Transections

Multiple subpial transections, introduced by Morrell, consists of performing multiple sections across the epileptogenic cortex, thus interrupting the horizontal interneuronal connections. This impairs recruitment within the epileptic discharge without damaging functioning of the cortical area, and is thus indicated when the epileptogenic zone is located in an eloquent area. It may be used as an isolated procedure or in conjunction with corticectomy of an adjacent area.

In 1995 Whisler [49] reported a series of 45 patients, with acceptable results. The limitations of this procedure are that one may not cut through into the sulcal area. His results, presented in accordance with Engel's scale, are:

Class I	55%
Class II	11%
Class III	16%
Class IV	18%

Consequently multiple subpial sectioning is to be considered a supplemental form of surgical treatment.

Chronic Intermittent Vagal Stimulation

This procedure was introduced during the late 1980s. The vagal nerve is mixed, somatic, and visceral. Its visceral fibers have their trophic center in the ganglion nodosum, which holds most of the visceral sensory fibers, which have retransmission nuclei localized in the fasciculus solitarius and to a lesser extent the dorsal motor nucleus, bulbar reticular formation, and Burdock's nucleus. The nucleus of the fasciculus solitarius projects upon the nucleus ambiguous, the dorsal motor nucleus, amygdala, hypothalamus, thalamus, and the insular cortex. Such extensive involvement of so many structures by the vagal afferents is the anatomical basis of vagal stimulation: these effects depend upon stimulation frequency and intensity. The most interesting for us are the effects regarding sleep, EEG synchronization, and desynchronization, especially the "C" fibres [50]. It is precisely in the desynchronization of the central nervous system that chronic intermittent vagal stimulation is effective, antagonistic to the neuronal synchronization which is typical of seizure discharges.

The surgical procedure consists of implanting a vagal stimulator with bipolar electrodes at the supraclavicular level. At times, such collateral effects as hiccups, hoarseness, and throat scratchiness may occur. However, early results have been encouraging [51, 52], but not uniformly so [53].

Clinical Evaluation

Because of the irregular irregularity of epilepsy surgery results, it is best to follow a single set of criteria. We suggest those proposed by Engel [44] in 1987:

Class I Seizure-free
- A. Completely seizure-free since surgery
- B. Aura only since surgery
- C. Some seizures after surgery, but seizure-free for at least 2 years
- D. Atypical generalized convulsions with antiepileptic drug withdrawal only

Class II Rare seizures ("almost seizure-free")
- A. Initially seizure-free but has rare seizures now
- B. Rare seizures since surgery
- C. More than rare seizures after surgery, but rare seizures for at least 2 years
- D. Nocturnal seizures only, which cause no disability

Class III: Worthwhile improvement
- A. Worthwhile seizure reduction
- B. Prolonged seizure-free intervals amounting to greater than half the follow-up period, but not less than 2 years

Class IV No worthwhile improvement
- A. Significant seizure reduction
- B. No appreciable change
- C. Seizures

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10 Tumors

"Hands, hold you poison or grapes?"
DYLAN THOMAS, Ears in the Turrets Hear

Introduction [1]

Brain tumors account for 18.6% of all cancers diagnosed in children, second only to leukemia [2]. It has been reported that the incidence of brain tumors in children is approximately 20 per million patients who are less than 16 years old [2, 3].

There are several significant differences in the management of brain tumors in children from that in adults suffering from the same disease entities: (a) there is a higher incidence of infratentorial tumors than in adults, and hence complicated triventricular hydrocephalus is more common in children; (b) malignant cerebral edema associated with the glioblastoma (the most common glioma in adults) does not occur in children; (c) the relatively more central location of the basal cisterns and greater volume of the sylvian and central fissures allow for more ample exposure of the parasellar and hemispherical areas, even in the absence of hydrocephalus; and (d) convexity and basal tumors (such as meningioma and schwannoma) are rare, whereas intraventricular and parenchymal tumors are common. Cerebral edema, independent of hydrocephalus, is an unusual occurrence. The rather remarkable differences in histological characteristics between tumors of children and adults are also noteworthy, with the meningioma and glioblastoma dominating in adult life and the medulloblastoma and infratentorial astrocytoma (benign) dominating in the pediatric age groups. Pituitary gland tumors are almost nonexistent in children under the age of 12 years, and there is a somewhat greater incidence of craniopharyngioma in children than in adults. The midline medulloblastoma is more common in children, whereas the cerebellar hemisphere (desmoplastic) medulloblastoma is more common in the older age groups.

The present work describes personal experiences by the author (AJR) with a continuous series of pediatric brain tumors over a 35-year period of time. The medulloblastoma was the most commonly encountered malignant tumor. It is of interest that because of the high surgical mortality in the treatment of medulloblastoma prior to 1969 [4], and the *theoretic* risk of disseminating malignant tumor cells during the operative procedure, some authors had recommended aspiration biopsy through a small craniectomy or twist drill opening and then radiation therapy [5, 6]. Recent experiences in many centers have proved that craniectomy (or craniotomy) with radical removal of the tumor is followed by both a much lower surgical mortality-morbidity and a much great postoperative longevity [7, 8]. The use of precraniotomy ventriculoperitoneal or III ventriculostomy *shunting* in the treatment of triventricular hydrocephalus, which results from tumor occluding the fourth ventricle, compensates immediately and completely the increase in intracranial pressure, minimizes the need for pre- and intraoperative steroid treatment, eliminates cerebral and peritumoral edema, reduces postcraniotomy morbidity, and provides the operating surgeon with a relaxed operative field for removal of the medulloblastoma, or posterior fossa astrocytoma [9–11].

Maximum resection of the *medulloblastoma* debulks maximally the tumor mass, and increases considerably the effectiveness of roentgen therapy, which is universally used for the treatment of the tumor site and prophylatic tumor "sterilization" of the supratentorial compartment and spinal axis. *Our experiences with the use of radiation therapy suggest that it is responsible for diminished intellectual function, diminished levels of growth hormone and thyroid-stimulating hormone (TSH) in many patients, and severe retardation of body growth* [12]. We have, accordingly, recommended that attention be given to the following: (a) diminishing significantly, or eliminating, prophylactic supratentorial radiation treatment, (b) recalculating dosage and timing of radiation therapy (using *radiosurgery* techni-

ques) to the tumor site so as to have the option of reirradiating the tumor site for local recurrences (since this has been noted to be the most difficult complication to treat), and (c) considering irradiation of the spinal cord with spot treatments when drop metastases become evident, rather than "prophylactic" treatment when there is no cytologic, clinical, or radiologic evidence of seeding or tumor growth. It is not clear to us why anyone would recommend irradiating such radiosensitive organs as the spinal cord and brain of children when there is no evidence of tumor. In the era before cerebrospinal fluid (CSF) cytology, magnetic resonance imaging (MRI), and high-resolution computed tomography (CT) scanners, this was theoretically understandable. The present state of science, and art, of medicine suggests reevaluation of the soundness of prophylatic whole brain and spinal roentgen therapy for medulloblastoma and ependymoma [13]. Roentgen therapists have different experiences and opinions concerning these matters [14]. Chemotherapy [15, 16] has not been shown to be effective in the management of medulloblastoma, and it has also been shown to be associated with a relatively higher morbidity (A. E. Evans, personal communication). This is no longer true! Our oncology colleagues are now equipped with pharmacologic substances which are very much more effective, less toxic, and indicated even before surgical treatment. Bone marrow harvesting and reimplantation techniques have been required.

The majority of *cerebellar astrocytomas* are benign; the incidence of malignant astrocytomas is reported to range from 4% to 15% [17, 18]. Winston et al. [19] classified cerebellar astrocytomas into glioma A and glioma B, according to their histological features, with group B identified as the malignant variant, occurring in approximately 20% of their series. Previous articles have reported that grossly cystic cerebellar astrocytomas range from 60% to 80% [17, 18, 20] of the total incidence of astrocytic tumors of the cerebellar hemispheres [18, 20] and vermis, that the tumor invariably recurs after simple evacuation of the cyst, and that extension of cystic compartments into the pons is unpredictable. In our own series, the cystic cerebellar astrocytomas have been much less common than the solid tumors. Not a single cystic tumor has recurred (because we resect the entire tumor wall at the time of surgery). On the other hand, those cystic tumors referred to us after initial drainage or simple removal of the mural nodule have often recurred, despite our efforts to resect secondarily the tumor wall. Cushing [21] reported that it was sufficient to remove the tumor nodule in cystic astrocytomas, whereas Matson [18] stated that it is necessary to remove the cyst wall as well as the tumor nodule, although he also stated that it is not necessary to remove *all* the cystic wall. Gol [17], with whom we agree, reported that 45% of cystic astrocytomas consist of cystic fluid completely surrounded by cyst wall composed of neoplastic tissue. Unfortunately, if the cyst wall extends into the brainstem (pons or medulla oblongata), it is not possible to resect the tumor capsule completely, even though there is almost invariably a clear plane of demarcation between tumor capsule and surrounding normal brain tissue.

We have not recommended X-ray therapy for grades I and II astrocytomas, observing very good long-term results in these patients after complete resection. A note of caution is expressed here, however, since some authors have reported recurrence of cerebellar astrocytomas as long as 20 years after complete resection [22], although histological study of these recurrent tumors revealed that they remained benign, and reoperation was followed by excellent results [23]. We have observed malignant transformation of a tumor found to be initially benign on histological examination, especially among the *pilocytic* and *fibrocytic* tumors.

Brainstem tumors are considered technically operable, as gliomas elsewhere in the brain, but histologically curable only in accordance with the histological nature of the individual tumor, as elsewhere in the brain! Indeed, some of these gliomas, as gliomas everywhere else, may have, at their center, cysts of varying size and form. We do not recommend biopsy of brainstem tumors, nor drainage of the microcystic cavities they may contain. Panitch and Berg in 1970 [24] described that there was no apparent relationship between any surgical procedure and survival time. It should be noted that Lassiter et al. in 1971 [25] and Reigel et al. in 1979 [26] undertook to biopsy these tumors and that their results do not indicate an increased morbidity. This has not been our observation.

Although rare, *glioblastoma multiforme* is a tumor which does occur in childhood and which occurs with approximately equal frequency in the cerebral hemispheres and the brainstem. Those tumors treated with both chemotherapy and roentgen therapy have an average survival of only 1.4 months longer than those treated with roentgen therapy alone [26].

Most authors, consequently, agree that surgery and roentgen therapy for the supratentorial glioblastomas [27] and roentgen therapy alone for the brainstem tumor [28] (as determined by imaging studies) provide maximum relief of symptoms, with the average 5-year survival for brainstem tumors ranging from 16% to 38% [29, 30].

Many authors have written imprecisely of "*pinealomas*" but it is far preferable to speak of tumors in the pineal region as "*pineal tumors*" and then to subdivide them histologically, since glioblastoma, teratoma, pinealoma, medulloblastoma, primitive neuroectodermal tumors, and others all grow in this area and are indistinguishable from each other preoperatively. The location, form, density, and vascularity of pineal tumors on

imaging studies do not permit precise histological diagnosis.

There was initially a great deal of trepidation in approaching surgically pineal tumors, but more recently an audacious group of neurosurgeons have advocated immediate operation on all tumors with an attempt to remove radically, or at least subtotally, neoplasms from within the area of the pineal gland. A more rational approach is to treat first the hydrocephalus that is invariably present in patients with pineal tumors, taking CSF samples from the ventricular system and the cisterna magna, so as to study the CSF for α-fetoprotein, β-HCG, and neoplastic cells [3, 4]. If the studies are positive, X-ray therapy [31] should be undertaken without [32] direct surgical attack on the tumor, and then the patient should be followed with imaging. Recurrence of tumor or growth of tumor during X-ray therapy could represent indications for surgical exploration and biopsy or removal. Negative CSF studies for α-fetoprotein and neoplastic cells represent adequate justification for direct attack on the pineal region, frozen section to learn whether the tumor is benign or malignant, and an attempt at complete removal if the tumor is benign. In the past, the surgical mortality for the treatment of pineal [33] tumors had been reported to range from 25% to 70% [34–37]. Presently, surgical mortality for removal of tumors from the pineal region ranges from 0% to 10% [38–42]. More than 70%–80% [43, 44] of these tumors have been reported to be highly radiosensitive. Many authors have reported long-term survivals from simply shunting and X-ray therapy, either directly to the tumor site or a "T" port [43] because of ventricuar dissemination.

Both *diencephalic tumors* and *brainstem gliomas* have long been considered neurosurgical lesions and, consequently, malignant because of anatomical location, irrespective of the histological characteristics of the tumor. Some authors have, however, proposed craniotomy and biopsy of diencephalic tumors for the purpose of obtaining histological diagnosis, since the malignant tumors are treated with radiation therapy and some of the benign tumors have been compatible with long-term survival without radiation therapy. Although "decompression" has been considered a justification for surgery by some authors [45–47], most do not subscribe to this [48–51]. Stereotaxic needle biopsy is presently being performed in some centers [52–54].

Gliomas of the optic pathway represent an extraordinarily interesting type of tumor in that they may be limited to one optic nerve, the optic chiasm, or extend from an optic nerve into the chiasm and either posterolaterally into the optic tracts or, very rarely, up the opposite optic nerve. This benign and slowly growing tumor is associated in approximately 33% of the cases [55], and extends into the hypothalamus in more than half of the children in whom the optic chiasm is involved [56]. Most authors have advocated surgical exploration and biopsy prior to X-ray therapy [18, 57, 58]. Some authors have reported that there is an equal incidence of long-term survivals and good quality of survival in those patients not irradiated as in those receiving roentgen therapy [55, 59]. Most authors, however, agree that when the tumor is limited to an optic nerve, located entirely within the retrobulbar portion, it is impossible to distinguish between an optic nerve glioma and meningioma, especially if the patient has von Recklinghausen's disease. Even Hoyt and Braghdassarian [55], who doubt the value of surgery, agree that exploration and resection of an optic nerve glioma is justified for the relief of severe proptosis in a blind eye.

Our own recommendations are that optic nerve tumors invariably be explored and that the sheath surrounding the optic nerve be opened to inspect for the presence of a meningioma, or remove the optic nerve if the eye is already blind. We recommend roentgen therapy for optic chiasm tumors invading the hypothalamus, since we have often observed significant improvement in visual acuity in some patients with this type of lesion.

Craniopharyngioma, the most common parasellar tumor in childhood, is limited entirely to the intrasellar area in only 10% of the patients, extending to the suprasellar and intraventricular areas in the remaining children [56]. The middle fossae, anterior fossae, and sylvian fissures, as well as the posterior fossa, may be invaded by lobular extensions of this epithelial tumor or its areas of cyst transformation and calcification. Consequently, there is no uniform clinical picture. Visual disturbances, pituitary insufficiency, hypothalamic inadequacy, hydrocephalus, and long tract signs all occur with varying frequency and severity. Although surgical resection (complete) had once been considered impossible, some authors now consider it possible in almost every instance and, indeed, recommend that every effort be made to attain a complete resection at the first operation. Careful study of papers reporting successful "complete" [18, 60] resections on a longitudinal basis have revealed steady increases in recurrences of those "totally resected" tumors [61, 62]. The surgical mortality for craniopharyngioma has decreased steadly over the past 20 years, with most authors now reporting no operative mortality. There is almost unanimity that the factors limiting total resection are adhesions of the tumor capsule to the hypothalamus with extension of epithelial plugs into the hypothalamus itself, as well as dense adhesions and calcifications aherent to the tumor on one hand and either the optic chiasm or major arteries (carotid, basilar) on the other. Size and location of extension of tumor into the third ventricle, interpeduncular cistern, or posterior fossa, as well as fungation of the major mass of tumor into the retrochiasmatic area, seriously limit the possibility of complete resec-

tion. Those tumors, however, which are primarily cystic and which have a capsule that does not breach the arachoroidal barrier and penetrate the hypothalamus are totally resectable.

It has been our observation that tumors may *not* be totally removed (although this is a surgical judgment and not an anatomical fact) when the vein of Rosenthal at the anterior perforated substance indicates extension of tumor into the hypothalamus and basal ganglia, precluding complete anatomical removal. *We wish to stress that complete surgical removal and complete anatomical removal are two separate factors,* and that complete anatomical removal cannot be evaluated reliably at the time of surgery. We even express reservations concerning the reliability of present-day MRI studies as definitive indicators of "total" resection.

The rate of recurrence of craniopharyngioma treated with simple aspiration and biopsy is quite high in comparison with those treated with subtotal or total resection [60, 62], irrespective [22] of follow-up treatment with roentgen therapy. We do not accept the view that these tumors are best treated with simple biopsy, aspiration of cystic fluid, and X-ray therapy. It has been our observation, based upon our own material and a careful review of the literature [62, 63], that every effort should be made to remove as much of the tumor as possible and that postoperative roentgen therapy should be reserved for those patients who have massive tumors that cannot be totally (surgically) or subtotally removed. Radiosurgery is another matter! It is, in fact, being reported to provide excellent results.

Although most authors have reported benign tumors of the astrocytic series to be more common in the supratentorial compartment, Matson's [18] experience expresses an approximately equal incidence of benign and malignant astrocytomas of the cerebral hemisphere, and Zulch [64] noted that ependymomas were actually the most common tumor of the cerebral hemisphere. The surgical management of these tumors has, in the past, been predicated upon a concept of complete resection, with various authors suggesting that as radical a resection as possible be undertaken. Matson [18] states, "in children with a low-grade astrocytoma or mature ependymomas, we may elect to re-explore in 7–10 days after initial operation in order to examine the tumor bed with meticulous care under magnification vision for evidence of residual tumor." Today, reexploration is certainly unacceptable. It has not been our experience that complete resection of benign or malignant gliomas is either possible or advisable; the younger the child, the truer this has been. Indeed, the very young (less than 1 year of age) child with a glioma, benign or malignant, suffers extension of the tumor into the internal capsule, brainstem. Very young children have small frontal lobes and even smaller temporal lobes, so that the supratentorial gliomas tend to extend into or emanate from the parietal lobes and, consequently, grow inferomedially through the centrum semiovale into the internal capsule.

We do not recommend roentgen therapy for low-grade astrocytomas (I or II), although some authors have taken a position in favor of roentgen therapy, making value judgments on the management of the individual tumors which appear to be based more on empirical grounds [20, 65]. There is unanimity that the malignant tumors should be treated with roentgen therapy.

The *choroid plexus papillomas* in childhood are located primarily within the lateral ventricles, although some occur within the third ventricle and, quite rarely, in the fourth ventricle. They are associated with hydrocephalus, which in approximately 50% of children persists even after the choroid plexus papilloma has been totally resected [66, 67]. This form of hydrocephalus is due to hypersecretion of cerebrospinal fluid, and if the tumor is located in either the third or fourth ventricle it may also be responsible for causing obstructive hydrocephalus. We have always attempted to resect totally the choroid plexus papillomas [67]. We have not had any experiences with preoperative roentgen therapy, as suggested by Carrea and Polak [68], in order to shrink the tumor and its vascular stroma.

Concerning general data and *statistics*, in a very busy pediatric neurosurgery clinic, one may expect that approximately (48.8%) of children with brain tumors suffer malignant tumors and (43.1%) benign tumors.

By far, the benign astrocytoma is the most commonly occurring tumor, with the medulloblastoma the second most common, and the craniopharyngioma third (Table 10.1). The malignant astrocytoma, the fourth most common in incidence, occurs in only 7.5% of children, and the ependymoma, the fifth most common in incidence, in only 6.3% of children. One commonly assumes that the malignant astrocytoma is a rare tumor in childhood and that the ependymoma is common. This is incorrect. We found seven cases of glioblastoma multiforme (2.1% of our entire series), whereas the malignant tumors of the astrocytoma series made up 9.6% of the total. Adding the four (1.2%) ependymoblastoma tumors to the other tumors of the ependymal series only brings this total to 7.5% of the total, with astrocytic and ependymal tumors occurring with about even frequency.

We classify the tumors as *infratentorial*, *pineal-quadrigeminal*, and *supratentorial* (Table 10.2). The infratentorial tumors are subclassified into those involving the cerebellum and IV ventricle, those involving the brainstem, and those occupying the cerebellopontine angle but not including exophytic extensions of either cerebellar of brainstem tumors. The tumors in the pineal-quadrigeminal area are subclassified on the basis of anatomical location, disregarding entirely the histological classification of atypical teratoma, astrocytoma, etc.

Table 10.1. Relative percentages of pediatric brain tumors in our experience

Histological type	(%)
Benign astrocyoma	(29.2)
Medulloblastoma	(16.9)
Craniopharyngioma	(8.4)
Malignant astrocytoma	(7.5)
Ependymoma	(6.3)
Choroid plexus papilloma	(3.3)
Dysgerminoma	(2.4)
Glioblastoma multiforme	(2.1)
Olgiodendroglioma	(1.8)
Primitive neuroectodermal tumor (PNET)	(1.8)
Meningeal sarcoma	(1.5)
Metastasis	(1.2)
Ependymoblastoma	(1.2)
Undifferentiated	(1.2)
Meningioma	(0.9)
Hamartoma	(0.9)
Giant cell astrocytoma	(0.9)
Medulloepithelioma	(0.6)
Dermoid tumor	(0.6)
Teratoma	(0.6)
Chorioepithelioma	(0.3)
Pineoblastoma	(0.3)
Lymphoma	(0.3)
Colloid cyst	(0.3)
Chordoma	(0.3)
Ewing sarcoma of occipital bone	(0.3)
Neurinoma	(0.3)
Embryonal carcinoma	(0.3)
Unknown	(8.1)

Table 10.2. Location of brain tumors in infancy and childhood

Infratentorial	47.0%
Cerebellum-fourth ventricle	75%
Brainstem	22%
Cerebellopontine angle	3%
Pineal-quadrigeminal	6.0%
Supratentorial	47.0%
Cerebral hemisphere	40%
Suprasellar-anterior third ventricle	37%
Diencephalon	20%
Lateral ventricle (choroid plexus papilloma)	3%

Table 10.3. Tumors of the diencephalon

Type	Total %	Location	
		Medial %	Lateral %
Benign astrocytoma	40	41	49
Malignant astrocytoma	13	25	75
Glioblastoma multiforme	16	50	50
Primitive neuroectodermal tumor	6	50	50
Choroid plexus papilloma	6	100	0
Ependymoma	3	100	0
Colloid cyst	3	100	0
Oligodendroglioma	3	0	100
Unknown	23	24	75

Consequently, in this category of tumor we encountered atypical teratoma, pineoblastoma, astrocytoma, medulloepithelioma, and so on. The supratentorial tumors are subclassified into those occupying the cerebral hemisphere, the suprasellar-anterior third ventricle (parasellar), the diencephalon, and the lateral ventricle. The specifics regarding lateral ventricle tumors are elaborated upon in that section in this chapter.

Among the cerebral hemisphere tumors we treated 23% were benign astrocytoma, with the distribution of the tumor complying directly to the volume of cerebral parenchyma: frontal most common and occipital least common. Meningeal sarcomas grew directly from the dura mater and not from the meningeal portion of either the tela choroidea of the roof of the third ventricle or tissue of the choroidal fissure. There were only two metastatic tumors to the brain, one benign meningioma, and one each of dermoid and teratomatous tumors. We chose to identify diencephalic tumors as a discrete entity from both clinical and therapeutic points of view (Table 10.3). In turn, these tumors were subdivided according to whether they were located in the medial or lateral portions of the diencephalon, since the former presented with endocrine disturbances.

The suprasellar and anterior third ventricle tumors were separated from the diencephalic tumors already described (Table 10.4). In the suprasellar anterior third ventricle category we treated, 50% suffered craniopharyngioma, 26% optic pathway gliomas, and 7.0% germinomas. All of our patients with dysgeminoma were girls, all but one presenting with precocious puberty and/or diabetes insipidus, with follow-up extending from 1 to 6 years.

These comments concerning the dysgerminoma are of particular interest when considered in light of our patients with pineal-quadrigeminal area tumors. We encountered the same percentage of dysgerminomas in the pineal quadrigeminal area, as well as pineoblastoma, choroid carcinoma, and embryonal carcinoma. Whereas all of the children with parasellar dysgerminomas were girls and all but one presented with diabetes insipidus and/or precocious puberty, *none* of the children with pineoquadrigeminal dysgerminoma or pineoblastoma-type tumors were girls, and *none* presented

Table 10.4. Suprasellar-anterior third ventricle tumors

Type	Location				
	Suprasellar (%)	Extrinsic (%)	Optic chiasm (%)	Hypothalamus (%)	Other (%)
Craniopharyngioma	50	100	0	0	0
Benign astrocytoma	30	0	88	12	0
Dysgerminoma	7	0	0	100	0
Malignant astrocytoma	3.0	0	50	50	
Ependymoma	2.3	0	0	0	100
Lymphoma	2.3	0	0	0	100
Pituitary adenoma	2.3	0	0	0	100
Unknown	3.0	0	50	50	0

Table 10.5. Most common posterior fossa tumors

Type	(%)	Location		
		Cerebellum and fourth ventricle (%)	Brainstem[a] (%)	Cerebellopontine angle (%)
Medulloblastoma	34	100	(11)	0
Benign astrocytoma	31	77	22(4)	0
Malignant astrocytoma	6	40	60(10)	0
Ependymoma	7	100	(80)	0
Ependymoblastoma	2	100	0	0

[a] Numbers in parentheses indicate percentage tumors invading the brainstem.

with precocious puberty. Indeed, we have never seen a tumor of the pineal area with associated *precocious puberty!* Of our children with tumors in the pineal area, hydrocephalus was present, severe, and the primary cause for consulting the physician. None of the children with parasellar dysgerminoma suffered hydrocephalus. The types of pineal tumors encountered, in descending order of occurrence, were as follows:

Dysgerminoma
Medulloblastoma
Medulloepithelioma
Benign astrocytoma
Glioblastoma multiforme
Ependymoblastoma
Pineoblastoma
Choriocarcinoma
Embryonal carcinoma
Malignant astrocytoma
Unknown

Posterior fossa tumors were found in 47% of our children. The most commonly encountered posterior fossa tumor was the medulloblastoma and the second most common was the astrocytoma (Table 10.5). Because of the fact that extension of the cerebellar-fourth ventricle tumor into the brainstem affects directly both resectability and survival (chronologic and quality), we looked carefully to see whether the tumor extends from the cerebellum into the brainstem: this we found in 11% of the medulloblastomas, in 5% of the benign astrocytomas, in 25% of the malignant astrocytomas, and in 72% of the ependymomas.

Although it is generally possible to suspect, either on the basis of clinical or imaging observations, extension of cerebellar-fourth ventricle tumors into the brainstem, we are not able to do this reliably. Indeed, we are not able to diagnose with certainty the ependymoma preoperatively, or to distinguish the cerebellar hemisphere medulloblastoma from the benign solid astrocytoma.

The most common complicating clinical entity resulting from a brain tumor in childhood is hydrocephalus: found in 78.2% of infratentorial tumors, 100% of pineal quadrigeminal tumors, and 39% of supratentorial tumors. One is not surprised to see that 94% of the cerebellar-fourth ventricle tumors are complicated by hydrocephalus, but our observation that 32.3% of the brainstem tumors are complicated by hydrocephalus is different from what others have reported in the literature [18]. Consequently, the presence of hydrocephalus in a child with a history or findings suggestive of a

Table 10.6. Hydrocephalus associated with brain tumor

Location of tumor	Incidence of hydrocephalus (%)
Infratentorial	
Cerebellum-IVth ventricle	94.0
Brainstem	32.3
Cerebellopontine angle	20.0
Subtotal	78.2
Pineal-quadrigeminal	100.0
Supratentorial	
Cerebral hemisphere	3.3
Suprasellar-anterior III ventricle	50.0
Diencephalon	73.3
Lateral ventricle (choroid plexus papilloma)	100.0
Subtotal	39.0
Total % of children with hydrocephalus	61.1

brainstem tumor does not exclude the presence of a brainstem tumor. Conversely, in the light of our observations, if a child is suspected of having a pineal-quadrigeminal tumor either because of signs or imaging observations and if the child does not have hydrocephalus, one may seriously doubt the existence of a pathogenic tumor mass within this area.

We were very much surprised to observe that fully 33% of our children with cerebral hemisphere tumors have hydrocephalus as a complicating clinical entity (Table 10.6). This is quite different from what one observes in older adolescents and adults. The 50% incidence of suprasellar-anterior third ventricle tumors and 73% incidence of diencephalic tumors is predictable, since these tumors do not invariably occlude the CSF pathways either within the parasellar cisterns or the third ventricle.

Often enough, their growth is extraordinarily slow, allowing either the basal cisterns or third ventricle to mold themselves so as to permit CSF circulation. The 100% incidence of hydrocephalus in lateral ventricle tumors in and of itself is of particular interest, but of more interest still is the fact that *70% of these children remain hydrocephalic even when the tumor is totally removed.* It has long been known that the lateral ventricle choroid plexus papilloma produces enormous amounts of CSF, and it has been suspected that this has been the primary cause for the hydrocephalus.

However, there must be some other cause since removal of the tumor (and, consequently, removal of the causative factor for CSF hypersecretion) does not cure the hydrocephalus in the majority of the children. Whether the high protein associated with CSF hypersecretion by choroid plexus papillomas causes changes in the anatomical and physiological characteristics of the absorptive surfaces is impossible to state with certainty.

Since 1960, we have advocated the insertion of a precraniotomy shunt (before operating) on children who have a brain tumor complicated by the presence of hydrocephalus, with papilledema and other significant signs of increased intracranial pressure (ICP). The craniotomy is performed when the papilledema and/or other signs of increased ICP disappear. This has resulted in compensating the hydrocephalus and eliminating the increase in ICP resulting from hypertensive hydrocephalus and the cerebral edema associated with it before direct attack on the tumor and, consequently, in a smoother intraoperative and postoperative course.

Recently, III ventriculotomy has become an alternative used by some, but there is still no (statistically significant) publication reporting indications, results, complications, etc. *We must await these to form an objective evaluation.*

The incidence, indications and results of precraniotomy shunting are published elsewhere [10, 11, 69] and are discussed below. We do think, however, that one may best appreciate the beneficent effects of precraniotomy shunting procedures for treatment of the hydrocephalus if the complications are also known, so we present in tabular form the complications of this procedure (Table 10.7). As "complications" we included malfunctioning, overproduction of CSF, upward herniation, transfalcical herniation, subdural hematoma, intratumoral hemorrhage, and infection.

Eighty-four percent of our posterior fossa tumor patients with complicating hydrocephalus were shunted, 100% of our pineal region tumor patients, and 71% of our supratentorial tumor patients ... 82% overall, precraniotomy ventriculoperitoneal shunts performed.

In 75% of the craniopharyngiomas, we were able to obtain a radical (visually complete) resection at the first operation, but reoperations for recurrences were necessary in 20% of these children, with an additional 10% undergoing *reoperation twice* and one patient three times, in each instance with the postoperative diagnosis of "complete resection" (Table 10.8). Hence, we consider resection to be "surgically radical," reserving the

Table 10.7. Nature and incidence of complications of precraniotomy shunt

Total	(%)
Malfunction	3.0
Overproduction of CSF	3.0
Upward herniation	2.0
Transfalcical herniation	2.0
Subdural hematoma	2.0
Intratumoral hemorrhage	0.6
Infection	0.6
Prolonged coma in 2.0%, children with upward herniation	

Table 10.8. Extent of resection

	Cases	Radical	Subtotal	Partial	Biopsy	Exploratory	Nonoperative
Infratentorial							
Cerebellum-fourth ventricle	53	15	23	10	2	0	3
Medulloblastoma	53	15	23	10	2	0	3
Cerebellar astrocytoma	42	28	14	0	0	0	0
Ependymoma	14	4	10	0	0	0	0
Other	8	4	4	0	0	0	0
Brainstem	34	0	6	3	9	5	11
Cerebellopontine	5	2	2	1	0	0	0
Pineal	21	3	9	2	1	1	5
Supratentorial							
Cerebral hemisphere	61	12	39	6	3	0	1
Suprasellar-anterior third ventricular	28	21	5	2	0	0	0
Craniopharyngioma, other	28	2	6	5	10	3	2
Diencephalon	30	6	10	5	10	3	2
Lateral ventricle (choroid plexus)	8	6	2	0	0	0	0

Table 10.9. Surgical mortality after primary craniotomy

	Craniotomy	Death
Infratentorial		
Cerebellum-fourth ventricle	116	6
Brainstem	23	2
Cerebellopontine angel	5	0
Pineal-quadrigeminal	16	1
Supratentorial		
Cerebral hemisphere	61	0
Suprasellar-anterior third ventricle	54	0
Diencephalon	25	1
Lateral ventricle (choroid plexus papilloma)	8	0
Total	308	10[a] (3.2%)

[a] Death of *any cause* within 1 month of surgery. Of these ten deaths, six were in infants under 12 months of age, four with medulloblastoma, one with metastatic sarcoma, one with diffuse astrocytoma.

"anatomicalally radical" designation for those cases where recurrence never occurs.

In evaluating postoperative mortality, we consider a postoperative death as one resulting from any cause within 1 month of surgery or at anytime after surgery if related directly or indirectly to the surgical procedure (Table 10.9). The longest time a child remained in the hospital after surgery was 3 months; the average time was 2 weeks and the minimum (for posterior fossa surgery) 5 days. It is of particular interest that of our perioperative deaths (3.2%), 60% were in infants (all but one had highly malignant tumors). There were three intraoperative deaths. Six percent of the operated children suffered transient coma or stupor, 5% brainstem signs, and 4% hemiparesis. There was considerable overlap in the morbidity.

Considerable attention has been given recently to dissemination of neoplastic cells through ventriculoperitoneal or ventriculojugular shunts, especially since the reports of Hoffman et al. [60]. Accordingly, we analyzed our clinical material in order to learn how many of our shunted children suffered systemic metastases of their brain tumors through the shunt. There was a total of seven children in our series who had suffered systemic metastases; four of them had no shunts inserted and three did. None of our posterior fossa tumors suffered lung or bone metastases. Two patients with supratentorial tumors suffered intraperitoneal metastases through their ventriculoperitoneal shunts; one suffered lung and bone metastases, which we cannot consider related to the ventriculoperitoneal shunt. Two patients with supratentorial tumors suffered intraperitoneal metastases through their shunts. Of the four patients who suffered systemic metastases but did not have a shunting procedure, the metastases were to the retroperitoneal space, lymph nodes, lung, pleura, and skin. *We conclude that there is no significant difference in incidence of metastases in children with or without implanted shunts: the*

Table 10.10. Five-year survival and reoperation for recurrence

	Five-year survival (%)	Reoperation for recurrence (%)
Cerebellar astrocytoma	90.0	12.5
Craniopharyngioma	76.5	78
Optic glioma	71.6	13
Benign cerebral tumors	57.6	17
Diencephalon tumor	50	4
Medulloblastoma	38.5	14
Malignant cerebral tumor	34.6	4
Pineal region tumor	33.3	19
Posterior fossa hematoma	14.3	15
Brainstem tumor	6	0

total incidence of metastases through a shunt is 1.8%, whereas 2.4% of children without shunts suffer metastases.

The observations to be stressed are those concerning the complicating effects of radiation therapy on mental development, body growth, and endocrine function [12]. Of the 19 children we reported in that study who survived 3 years or more, 3 were not tested for intellectual function because the parents refused in 2 cases and 1 child suffered high cervical myelopathy radiation damage that rendered him quadriplegic and ventilator dependent. Of the 16 we were able to test, only 3 had normal intelligence quotients (none of these had an IQ greater than 100), and 13 were retarded! When we tested a similar group of children with cerebellar astrocytoma we found that the IQ was normal in all 11 children we were able to test. The one child we were not able to test suffered cerebral damage from a cardiac arrest during surgery.

As one would predict, the 5-year survival is greatest in children with cerebellar astrocytoma and second best in children with craniopharyngioma (Table 10.10). Interestingly enough, 71% of the children with optic pathway glioma survived for 5 years and 50% of the children with diencephalic tumors survived for 5 years. Brainstem tumor and posterior fossa ependymoma were the tumors that caused the highest incidence of mortality and the lowest incidence of 5-year survival (6.0% and 14.3%, respectively).

Surgical Approach and Removal

Probably the best place to start when undertaking a description of the surgery of brain tumors in children is with a clear distinction between brain and skull on one hand, and between *neurocranium* and *splanchnocranium* on the other (Fig. 10.1). Skull base surgery has come into its own ("with a bang" one might add) since the first edition of this book. However, there is no intent at this time to extend the text across the basal cistern barrier. Rather, the description remains classical, including orbit, sella, clivus, foramen magnum and some aspects of the subtemporal area in broad terms.

The separation of neurocranium from splanchnocranium is not intuitively possible: one wonders why a large portion of the orbit is splanchnocranium and why the pterygoid plates are neurocranium, the zygomatic arch is neurocranium yet auditory canal and styloid process splanchnocranium. Therefore, Fig. 10.1 (redrawn from Pernkopf) allows the reader to identify at a glance which is which...and to realize that all of the brain (eyes included) drain through the dural sinuses of the neurocranium.

Bone Tumors

Excluding tumors of the scalp, such as the lipoma, neurofibroma, dermoid, and epidermoid, conventional classification of neoplasm involving the skull proceeds anatomically from the outer table through the middle table (diploë) and to the inner table. Regarding outer table tumors, one most prominently observes osteoblastic or mixed tumors, much less often ostoplastic lesions; regarding the middle table, the hemangioma is rather typical, but the epidermoid and dermoid are far more common in children; regarding the inner table, those neoplasms considered to originate from this structure are, in essence, more directly related to the meninges, such as the chondroma or osteoma.

The younger the child the more true the maxim "skull tumors may not be classified anatomically as outer table, diploic, or inner table, since the thickness of the skull is such that all three tables may become involved." The pediatric neurosurgeon never sees patients with skull metastases; the meningioma is not a tumor of the newborn, infant, or toddler, though it may occur in a juvenile with von Recklinghausen's disease; dural chondroma has not been reported in childhood; the ostoma rarely grows from the inner table of the skull into the intracranial compartment, but almost invariably expands from the outer table of the skull as a relatively smooth, broad-based, bony-hard excrescence and is a clinical oddity in childhood.

Dermoids (the most common skull tumors seen in childhood), epidermoids, neurofibroma, and lipoma are associated with a central radiolucency on the skull X-rays, and an intact layer of ossified tissue on either the inner table or the outer table, or both. Dermoid, epidermoid, teratoma, and hemangioma may all be located entirely within the subgaleal space, may destroy one or more layers of the skull, or may have varying degrees of primary or secondary calcification associated with them. The eosinophilic granuloma and aneurysmal bone cyst, such as fibrous dysplasia, involve all three layers of the skull and may be tender to touch.

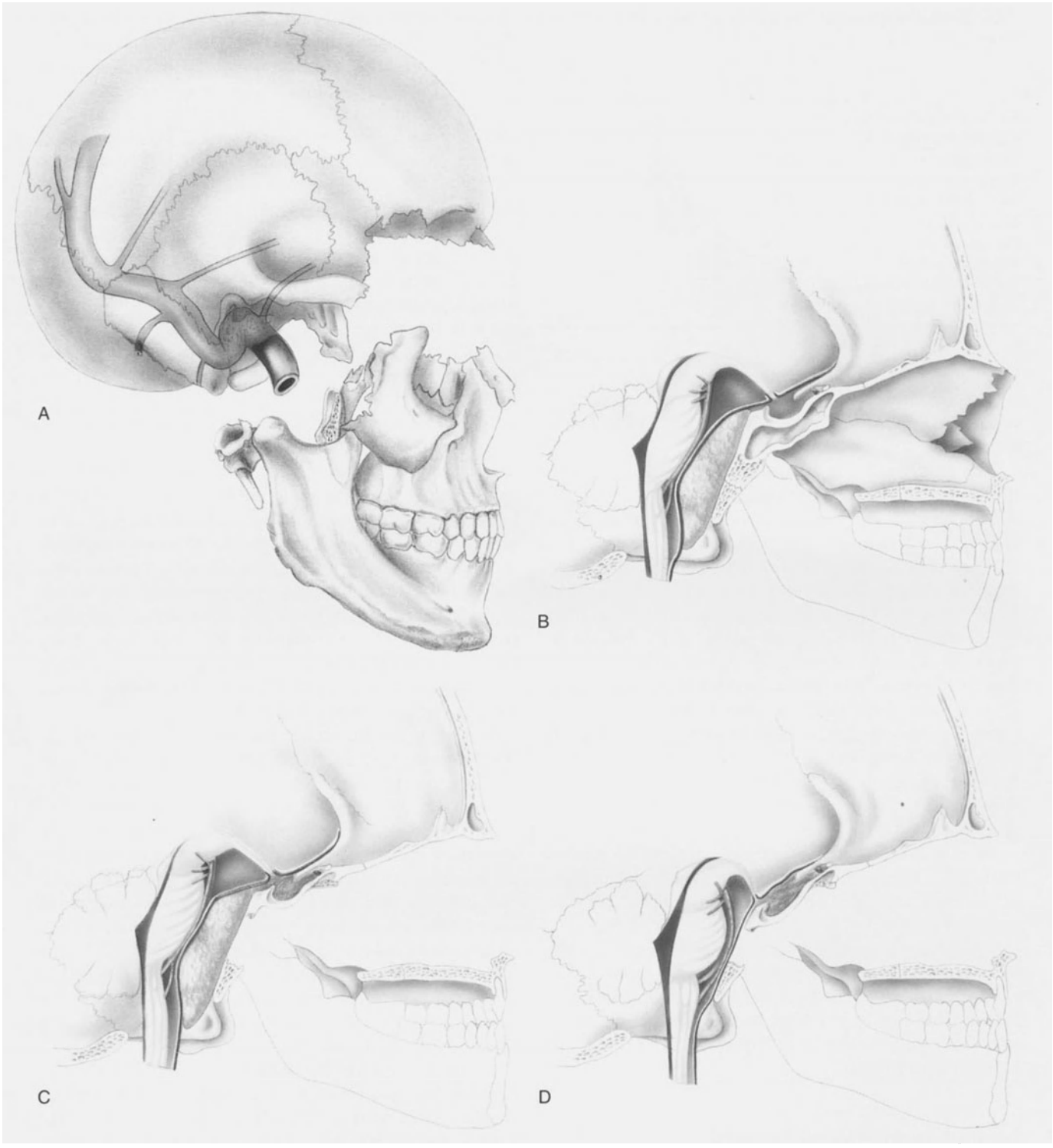


Figure 10.1. (A) Drawing of the neurocranium separated from the splanchnocranium. (B) This is a clival chordoma in a child, unusual indeed: of approximately 650 personally managed brain tumors in children, I have only seen four chordomas and operated on – without positive effects – only two. The purpose of this illustration is to show that the chordoma initially develops around the spheno-occipital synchondrosis along the clivus, that its early expansion is extradural, and that access to the tumor with present-day techniques may be achieved by the transphenoid route. These drawings are a conceptualization of such a case operated on at the University of Rome by P. Cantore and G. Iannetti. They show (B) the tumor in a retroclival/extradural location, (C) the transphenoidal route, (D) postoperative reapproximation of the dura.

The histological diagnosis of bone tumors is always a long time coming because of the difficulties inherent in processing the tissue, and the preoperative diagnosis is only conjectural since it is based purely on radiologic criteria. Therefore, once a bony "tumor" is diagnosed, one makes a decision to operate or to observe. The following description pertains to operative findings and management.

Dermoid and Epidermoid Tumors (Figs. 10.2, 10.3)

The dermoid and epidermoid tumors are removed by incising the periosteum along the limbus of the tumor, where the skull flutes out at its junction with the neoplastic expansion. This frees the intraosseous mass from the overlying periosteum and permits one to dissect the inner surface of the mass from the inner table of the skull or the dura, if the inner table of the skull is

Figure 10.2. This dermoid tumor, growing within the region of the pterion, consisted of fibrocollagenous tissue which was semisolid. When the extruding material herein shown was removed, a cartilaginous, intradiploic, portion was excised in toto.

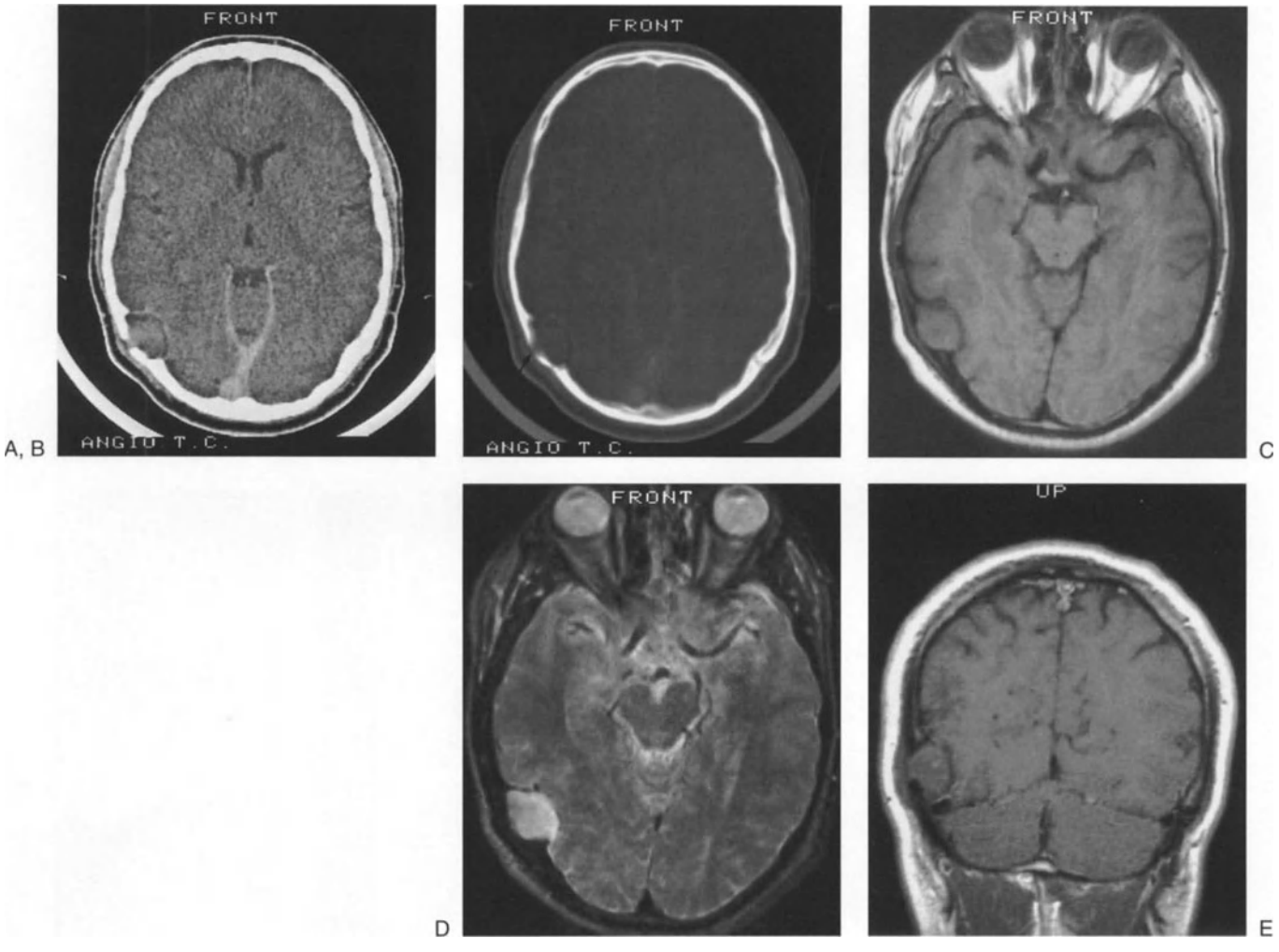


Figure 10.3. The CT and MRI scans are extraordinarily revealing for this type of pathology. In (A) one sees discontinuity of the calvarium (window level for brain parenchyma) in the right parieto-occipital area, immediately anterior to the suture. In (B), a window level for bone, the *arrow* indicates the break in bone continuity. (C), (D), and (E) show axial and coronal slices revealing, respectively, a low signal mass on T1, a high signal on T2, and nonenhancing characteristics on the post-gadolinium scan.

eroded. Classically, one removes bony tumors with rongeurs, Leksell or Kerrison. However, we have found it preferable to use high-speed drills. When these tumors are located along the midline, caution should be taken lest the superior sagittal sinus be damaged. When they are located along the lateral surface of the skull, they may be limited to the skull and sutures or they may communicate with the meninges.

Eosinophilic Granuloma

The eosinophilic granuloma is removed with either a rongeur or a high-speed drill armed with a bur tip. Once the thin, osseous, outer table of the skull has been removed, the granulomatous tissue may be taken away either with a curette or dissected out with an Oldberg periosteal elevator.

Aneurysmal Bone Cyst

Aneurysmal bone cysts bleed inordinately! If one encounters very bloody tissue after opening into the tumor, it is best to stop the bleeding with bone wax and to desist. X-ray therapy is curative; surgical removal is unnecessary.

Fibrous Dysplasia and Juvenile Aggressive Fibromatosis (Fig. 10.4)

There is no curative surgical treatment for fibrous dysplasia when it is polyostotic. The treatment for juvenile aggressive fibromatosis is cytoreductive. The unsightly excrescences of fibrous dysplasia which often extend across suture lines should be removed with the use of a high-speed bur, brushing the protuberances away until a smooth contour results.

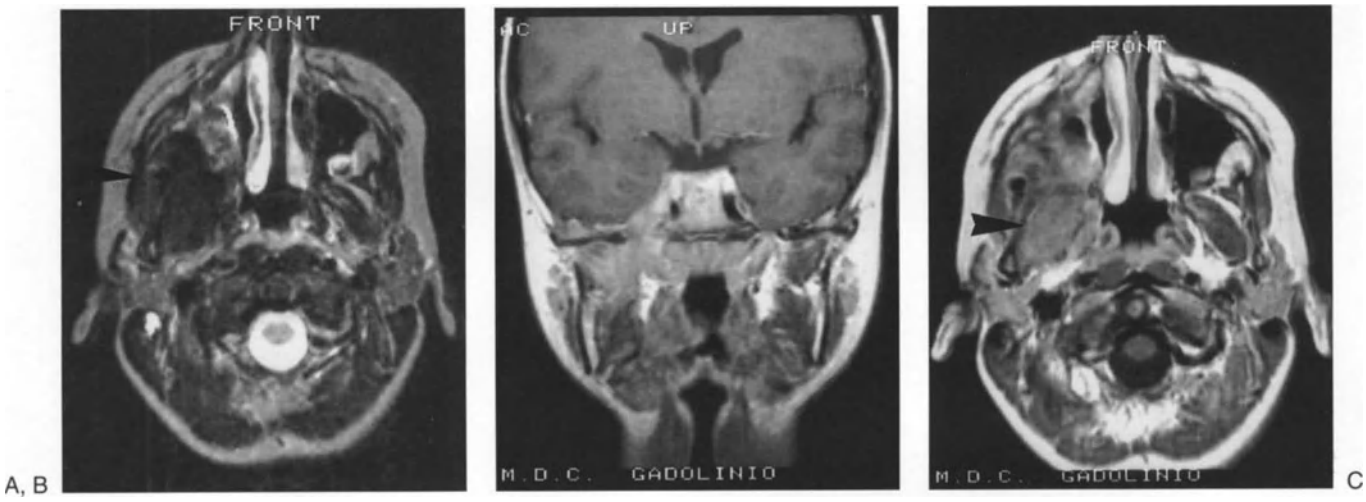


Figure 10.4. These three MRI studies are of a child with an invasive tumor of the splanchnocranium and neurocranium, occupying the right infratemporal fossa and extending through the foramen ovale, with involvement of the right cavernous sinus. These images, without (A) and with (B) and (C) gadolinium, show that the aggressive *juvenile fibromatosis*, hypointense on the T2 weighted images, is an extensively invasive mass occupying the right infratemporal fossa. On the coronal T1 weighted image (D) the extension of the mass through the temporal floor, via the foramen ovale and very probably foramen spinosum, is as well seen as is the massive involvement of the cavernous sinus (constricting the internal carotid artery). Enhancement of the lesion (*arrow*) is shown in this axial T1 weighted image.

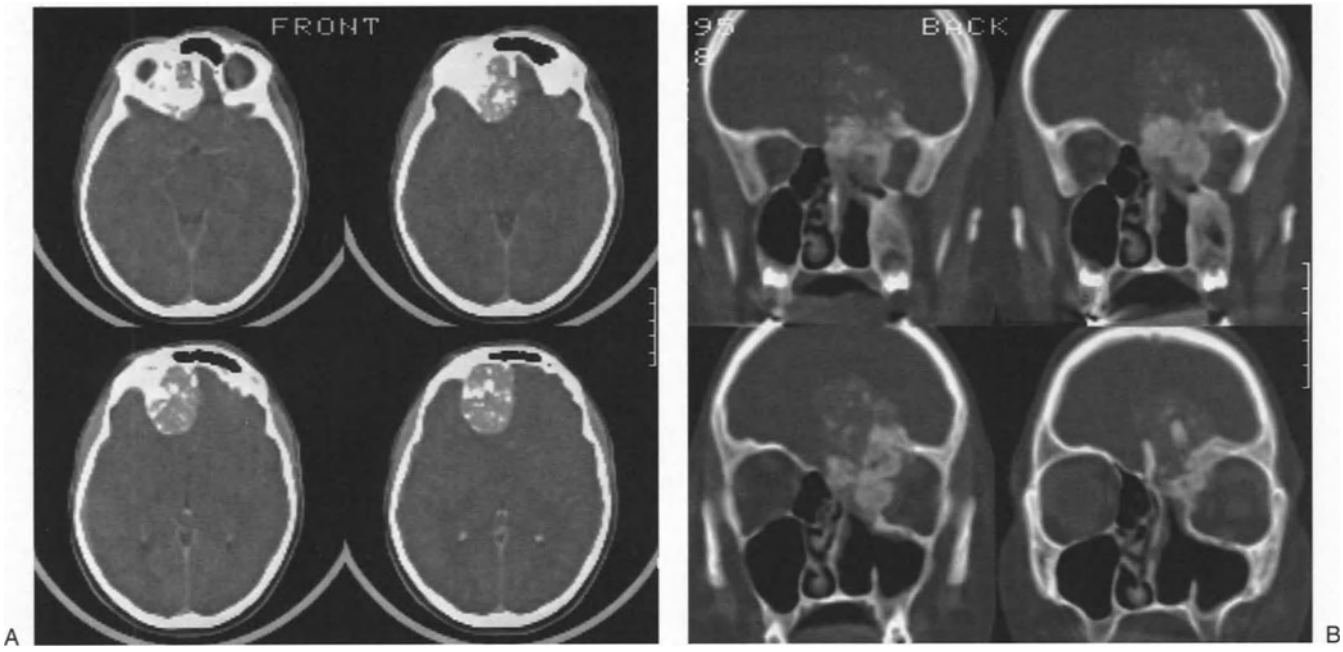


Figure 10.5. Intra- extracranial recurrence of frontal ethmoidal *osteosarcoma.* In (A), the axial views are shown and in (B) the coronal views. There is a right frontal/ethmoidal mass of bone density in addition to a soft tissue mass extending into the neurocranium, the orbit, and the left ethmoid sinus.

As the outer table is breached, one notes that the diploë have a cavernous appearance and that bleeding is moderate. One may chisel the surface and the diploë without fear of weakening the skull.

Monostotic fibrous dysplasia should be treated when it involves the sphenoid bone and constricts the optic nerve or occludes the superior orbital fissure, threatening vision, exophthalmos, or cranial nerve palsy. Treatment consists of using a bur to remove the hyperostotic bone from the neural structures it is compressing. Once these have been freed, nothing may be gained by continuing to resect the bony overgrowth, unless this is done for cosmetic purposes!

A particular problem may be encountered when the bone is very thick, and there is overgrowth from the inner table into either the intracranial or intraorbital compartments. If one wishes to use a craniotome, it is necessary first to reduce the skull thickness by "brushing" away the outer surface (exostosis) with a high-speed drill, then using the craniotome to reflect the flap.

The plastic surgery literature holds reports of cases of fibrous dysplasia which have, over the years, undergone malignant changes, so that some plastic surgeons recommend their radical removal. One wonders whether the reported cases were not osteogenic sarcomas.

Osteoma

The excresence of the osteoma is simply shaved away with a bur, until the normal contour of the skull has been restored. It is not necessary to perform a craniectomy, though doing so offers no particular disadvantages: no bone needs to be grafted if the defect is small, and rib is excellent as donor tissue if the defect is large.

Dural- and Osteo-Sarcoma (Figs. 10.5, 10.6)

Though the meningioma does not occur in the newborn or infant, and is extremely rare in the toddler and juvenile, except in the occasional child suffering neurofibromatosis, the dural sarcoma is, in the infant and toddler, relatively common, occurring as often as the primitive neuroectoermal tumor (PNET). On CT scan this tumor presents as a parenchymal mass, occupying one or more lobes, generally the frontotemporal area extending deeply into the anterior and middle fossae. The diagnosis of dural sarcoma, consequently, is seldom expected. Angiographically these tumors appear to be highly vascularized lesions, centered at the pterion or lesser wing of the sphenoid, taking arterial supply from the external and internal carotid systems. At surgery, the tumors give the surgeon the impression that he is dealing with a glial sarcoma, an invasive, highly malignant, tumor of white matter, cortex, dura, and (in the infant) bone.

Despite the malignant histological appearance, every effort should be made to resect these tumors totally,

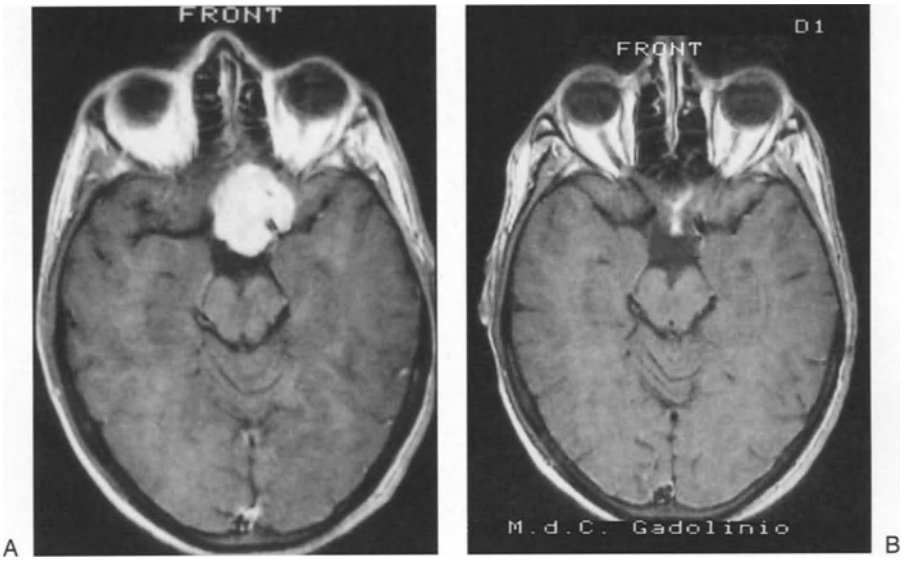


Figure 10.6. Though in children we generally do not consider metastasis, except of course in leukemic patients, to be very frequent, they do occur; and quite often may be both surprising and very responsive to chemotherapy, radiosurgery, and surgical excision. *Ewing's sarcoma* is a tumor which often metastasizes to the dura mater and, in fact, is quite difficult to separate histologically from the *primitive neuroectodermal tumor.* In (A) an axial post-gadolinium MRI scan, a very large parasellar mass is growing from the dura at the tuberculum sellae and the cavernous sinus. The internal carotid artery is partially engulfed by the tumor. (B) This postchemotherapy study shows the remarkable diminution in volume of the tumor, with only a linear enhancement along the plane of the dorsum sellae.

since their biological activity is not reflected by the histological appearance. Therefore, the surgical approach should consist of resection of (noneloquent) parenchyma and dura.

If one attempts to remove the tumor with standard neurosurgical technique, all of the feeding vessels should be isolated before entering into the main mass of the neoplasm. This should be done circumferentially, working at the periphery of the mass, separating it from the surrounding brain. This presents no difficulties as long as one is working along the frontal or temporal lobes, but subsequent descent along the lesser wing of the sphenoid brings the surgeon into a highly vascularized area, prolonging the operative procedure and rendering complete resection increasingly difficult. Avoid the temptation to move into the center of the tumor with cautery and suction, because of the significant blood loss which may occur.

Since resection of the tumor entails resection of parenchyma and dura, one finds oneself exposing bone. This complicates the closure and increases the risk of cerebrospinal fluid fistulae, since it is difficult to attain a dural closure. In addition to this, the bone flap would be approximated only over a portion of the remaining cerebral tissue, so that the only barrier between subarachnoid spaces and the environment is the skin. If the dura cannot be closed, grafts of periosteum or fascia are used. If periosteal grafts are ineffective, *fascia lata* grafts may be used if extensive defects remain. If the ventricle has not been used, such dural substitues as Goretex may be used. *Avitene* strips, lain along the surface of the skull and then packed onto it by using soaked fluffy cotton, are most useful in sealing possible leakage, diminishing the risks of cerebrospinal fluid rhinorrhea or otorrhea. Postoperative roentgen therapy should not be begun until there is complete healing of the skin and no indication of cerebrospinal fluid leak. This tumor is very responsive to chemo- and radiation therapy, so no effort should be made to follow it into eloquent areas.

Orbital Tumors

Periorbital Tumors: General Comments (Fig. 10.7)

Some mention should be given to nonorbital tumors, which both impinge upon and change the size of the orbit, which occur after the bony sinuses have developed, such as the *mucocele.* The interesting aspect of this tumor is that it results from accumulation of fluid secondary to the obstruction of the sinus ostium.

As the mucocele expands, it thins the bony walls of the skull around it, pushing into the orbit, causing proptosis and resulting in diminished orbital volume. Other lesions which may, as the mucocele, result in diminished orbital volume are fibrous dysplasia, edentulous maxillary cyst, adamantinoma, and papillary tumors of the paranasal region.

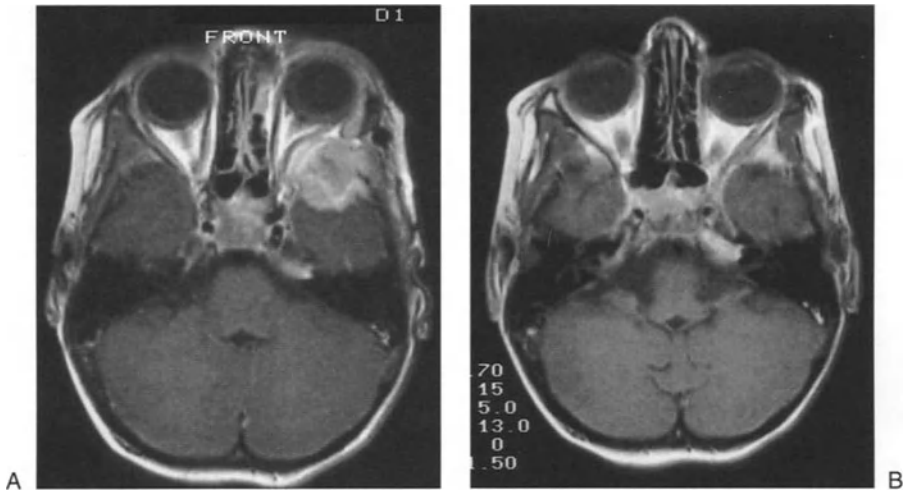


Figure 10.7. *Rhabdomyosarcoma* is the most malignant tumor of the orbital adnexa. Seldom are these tumors operable, except when a total orbital exenteration is possible, but they do respond favorably to chemotherapy. (A) An axial MRI scan after gadolinium injection reveals an in situ recurrence of a lateral rectus rhabdomyosarcoma which was surgically excised. In (B), also a post-gadolinium study, a very significant reduction in tumor volume occurred after chemotherapy.

Intraorbital Tumors

The intraorbital tumors which a neurosurgeon may encounter are *neurofibroma* (especially the plexiform type), *retinoblastoma, rhabdomyosarcoma, lacrymal gland tumors,* the *hemangioma* (which, incidentally, is the second most common cause for unilateral exophthalmos), *lymphangioma, and dermoid* and *epidermoid tumors.*

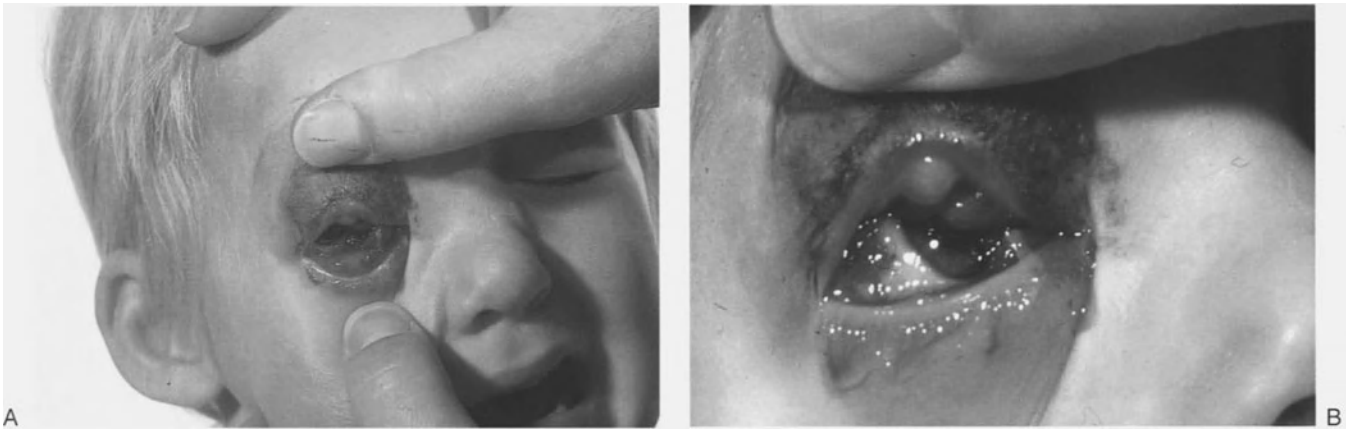
The most superficial tumors, and the most common, are the *dermoid* and *epidermoid tumors* (and the *lacrymal gland tumors*, which they closely resemble in appearance and location). They are located in the superolateral portions of the orbits, at times deepening the lacrymal gland fossa. The dermoid and epidermoid tumors of the orbital wall are removed with the same technique as those of the skull.

It would be misleading to suggest that there are different techniques for histologically different intraorbital tumors, as indeed there are for such intracranial tumors as medulloblastoma, solid astrocytoma, papilloma, craniopharyngioma, and so on. In fact, very likely the great attention given to histological classification of human brain tumors resulted from the distinctly different techniques used in resecting the individual histological tumor types, with the exception of the gliomas.

Whereas the surgeon attempts to remove as completely as possible such malignant intracranial tumors as medulloblastoma, ependymoma, and primitive neuroectodermal tumors, no such attempt is made for intraorbital tumors: it is impossible to resect them completely without irreparably damaging the globe, optic nerve, orbital vasculature, or extraocular muscles, any of which results in severe visual disturbances.

By and large, the general neurosurgical principle for intraorbital tumors is that malignant tumors (rhabdomyosarcoma, retinoblastoma, neuroblastoma) need neither biopsy nor resection. Some intraorbital benign tumors (hemangioma) should be operated on and resected completely. Diffuse, invasive, deforming tumors such as the plexiform neurofibroma are tragedies which should not be compounded by surgical intervention. Periorbital tumors (fibrous dysplasia, osteoma, mucocele, some dermoid and epidermoid tumors) should be removed immediately they are diagnosed, and the resection accompanied by decompressing the orbital rim, orbital roof, greater wing of the sphenoid, superior orbital fissure, or optic foramen. These tumors damage by compressing, so an adequate decompressive procedure is indicated. A radical resection of all neoplastic tissue, irrespective of whether it is compressing structures related to vision, is not indicated.

Hemangioma (Fig. 10.8) and lymphangioma (Fig. 10.9), the most common primary intraorbital tumors, are very prominent in younger children, and very often disappear spontaneously. Their tendency is to invade the adnexa of the orbit and the lids, and commonly causing orbital enlargement and proptosis. These tumors originate primarily within the muscular cone, diffusely spread throughout its contents, the extraocular muscles, and the periocular fat. Consequently, they have a rather invasive nature, though they are seldom malignant. They also tend to calcify, as do all hemangiomas or arteriovenous malformations. Surgical removal is difficult to accomplish without producing unacceptable deficits, and is to be discouraged unless the tumor progresses in volume (as the child ages) and threatens vi-



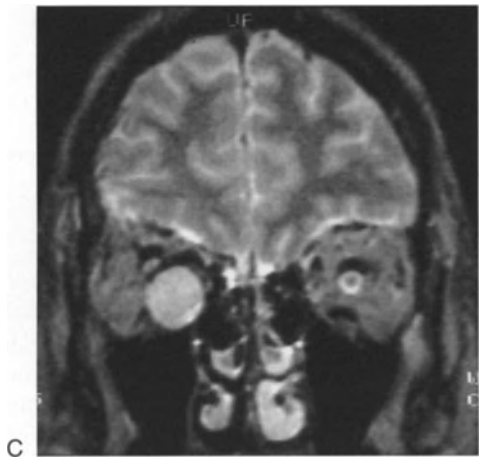


Figure 10.8. (A) Hemangioma has caused an hemorrhagic conjunctivitis, so that the eye is nonfunctional. (B) This is a closeup shot of the conjunctiva and sclera, illustrating the weeping hemangioma. (C) This T2 post-gadolinium, coronal MRI scan is of a child with a cavernous hemangioma located in the superomedial portion of the left orbit. The child in whom this imaging study was performed presented clinically in exactly the same manner with the same observations as the child illustrated in (A) and (B).
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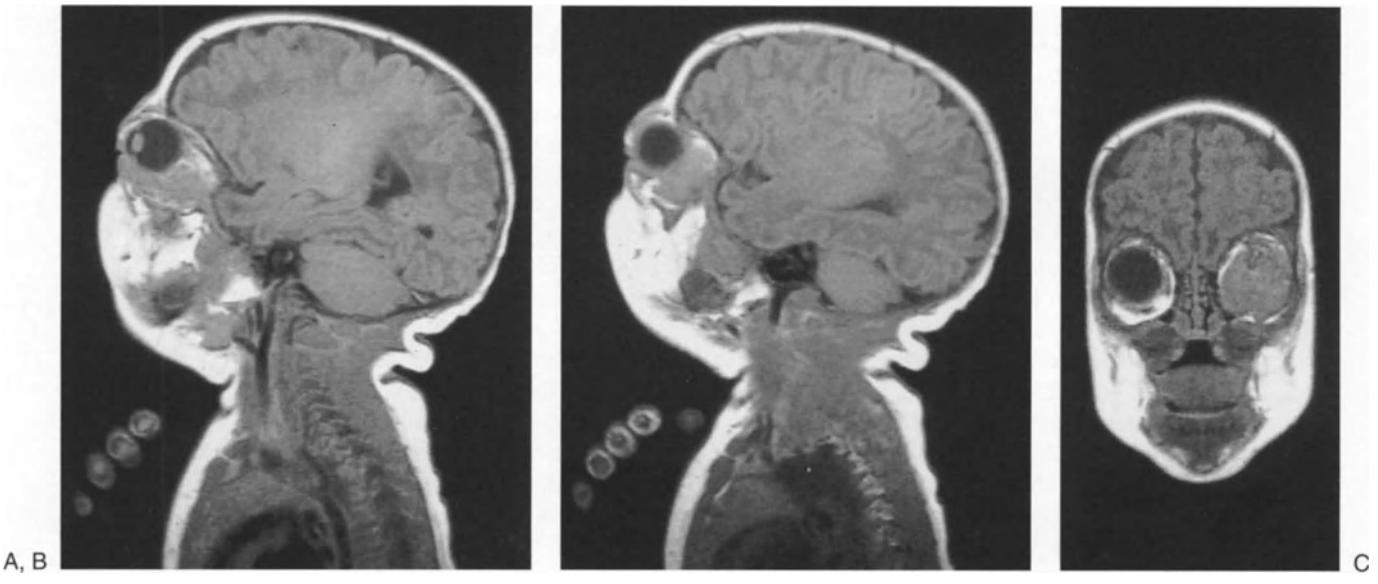


Figure 10.9. This figure is of particular value in that it provides adequate information to permit the surgeon, wishing to perform a biopsy, to plan the orbital approach. The child, 1 month old, has a *lymphangioma* of the left orbit which progressed steadily in volume, as determined by the exophthalmus. In (A) the tumor mass is revealed to occupy the inferior splanchnocranial portion of the orbit, displacing the globe superiorly and anteriorly. In (B), a more laterally placed MRI slice, one notes that the tumor mass extends laterally and, even more inferiorly. This is an excellent surgical indication for the Krönlein incision, much easier and less involved for a biopsy than the transfrontal/transorbital approach. (C) The lymphangiomatous mass may be seen to occupy the anteroinferior portion of the orbit, permitting the surgeon access along the line of the inferior orbital rim.

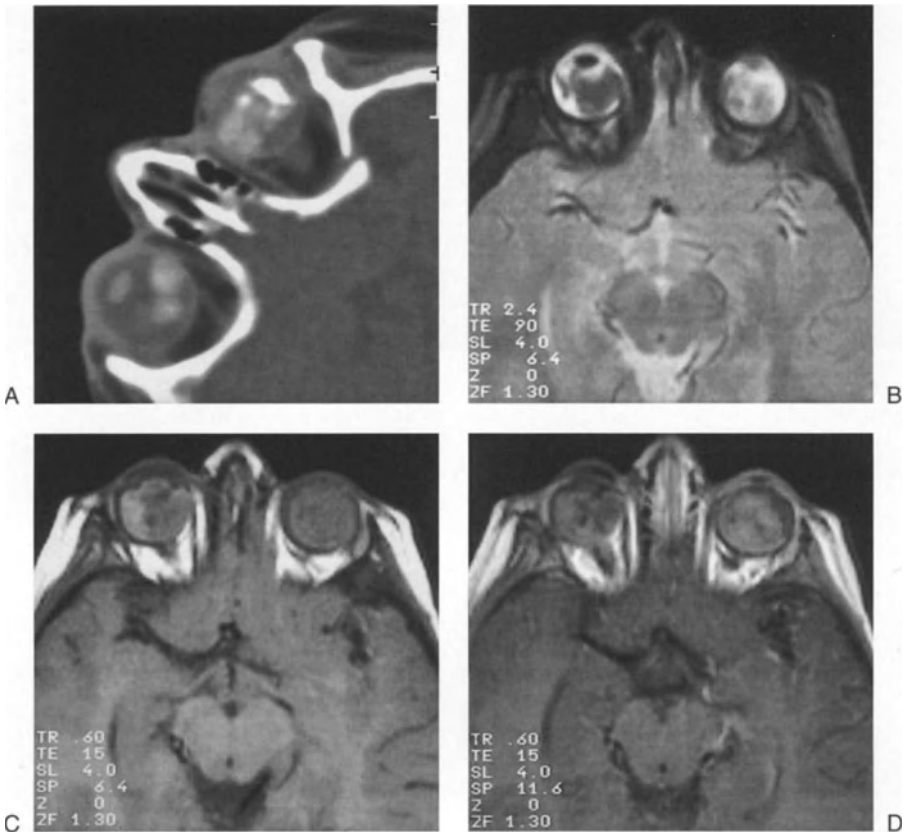


Figure 10.10. Bilateral *retinoblastoma*. In (A), an axial CT scan without contrast enhancement, one sees multiple bilateral calcifications in the vitreous body, in (B) an axial MRI T2 weighted sequence, the hypointensity lesions punctuate the vitreous body as they do in (C), a noncontrast enhancement study. (D) The post-gadolinium study reveals diffuse lesion enhancement bilaterally, with the nonenhancing portions representing flecks of calcification.

sion. If it produces unsightly vascularization of the sclera and lids along with such severe impairment of vision as to render it useless, and the eye cosmetically unacceptable, radical resection and implantation of a prosthesis is a positive alternative. More often, the tumor bleeds, causing an intraorbital hematoma or calcified mass. These should be removed.

The *plexiform neurofibromas* are invasive, slowly growing, and destructive. They, also, most unfortunately, may result in unsightly deformities of the orbital and periorbital region. They are not resectable, and are difficult tumors to deal with even if one wishes to attain only an agreeable cosmetic result. Here, restraint is advised since the tumor is highly vascularized, diffusely invasive, and without definable confines.

Retinoblastoma (Figs. 10.10, 10.11) and *rhabdomyosarcoma* are both highly malignant tumors, with the former tending to grow along the optic nerves into the intracranial compartment and, at times, to expand within the optic chiasm. The rhabdomyosarcoma, on the other hand, is an invasive tumor, extending into the orbital adnexa, eroding and destroying bone. Neither of these tumors is operable.

Neuroblastoma may present as a primary orbital tumor, when, in fact, the orbital lesion is a metastasis.

Surgical Considerations

Intracranial Approach to Orbital Tumors (Fig. 10.12)

The intracranial approach to orbital tumors may be more correctly termed transcranial, since the technique entails the reflection of a frontal flap, an extradural approach to the roof of the orbit and its rim. The dura is dissected from the roof of the orbit, which is exposed from the pterion, laterally; the lesser wing of the sphenoid, posteriorly; and the cribriform plate, medially. One should expose the base of the anterior clinoid and the most medial portion of the planum sphenoidale.

The anterior medial dissection of the dura from the orbital roof to the ethmoid bone allows for complete visualization of the intraorbital contents, once the orbit is unroofed, without risk of stripping rootlets of the olfactory nerve from the perforations of the cribriform plate. Retraction of the frontal lobe beneath the dura mater is carried out slowly and gently, since no cerebrospinal fluid is being aspirated.

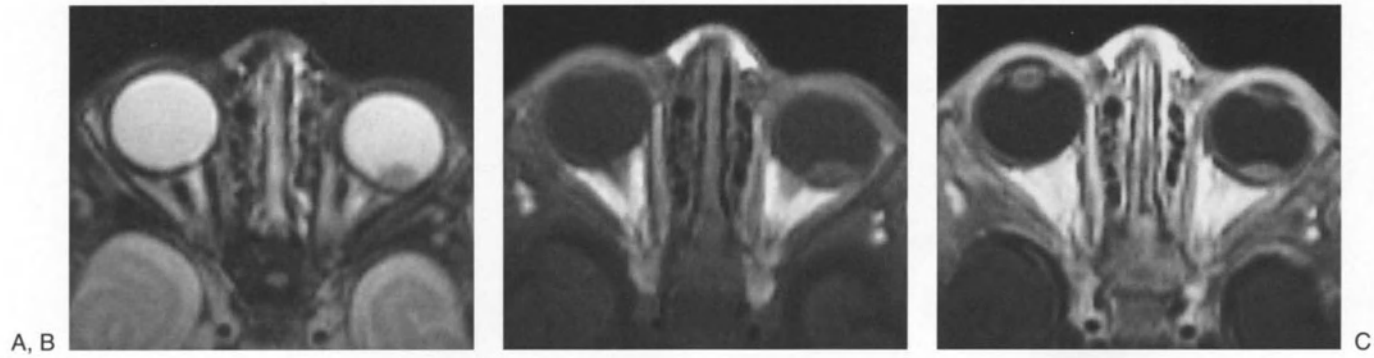


Figure 10.11. Retinoblastoma of the left globe. Indifferently, retinoblastoma may be limited to one globe, bilateral, or a diffuse optic pathway tumor extending from the globes to the geniculate bodies. In this figure, a unilateral tumor (limited to the retina of the left eye) is illustrated. (A) and (B) are images which show a mass in the left globe, in T2 and T1 weighted sequences with low signal intensity. The post-gadolinium study (C) reveals enhancement.

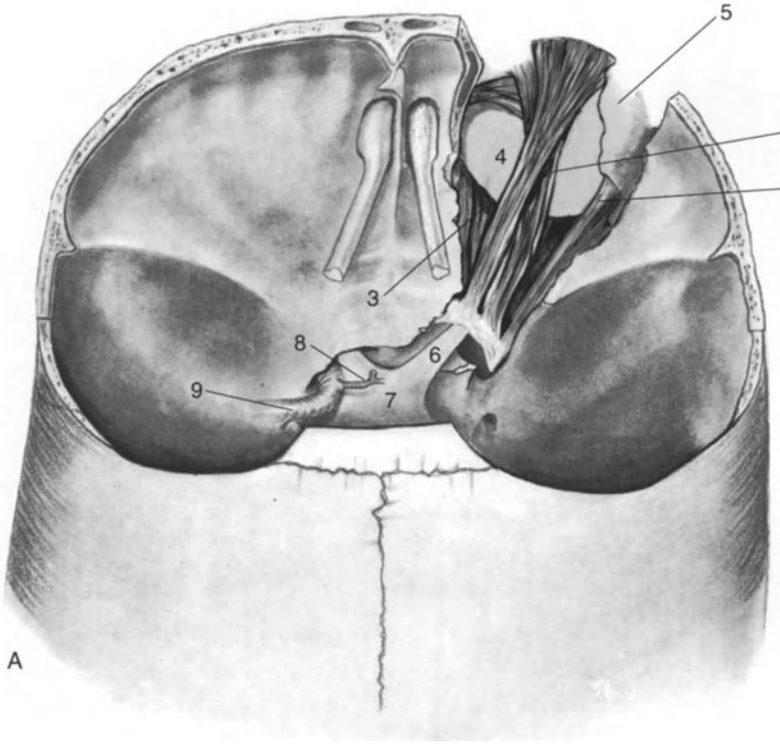


Figure 10.12. (A) The orbital roof and supraorbital rim have been removed, exposing the superior (*1*), lateral (*2*), and medial (*3*) rectus muscles, as well as the globe (*4*) and lacrimal gland (*5*). The optic nerve (*6*) and chiasm (*7*), as well as the anterior (*8*) and middle (*9*) cerebral arteries are included for anatomical orientation. (B, C) see p. 199.

The orbital roof and supraorbital rim are removed. The periorbita is opened and the frontal nerve identified on the superior aspect of the superior rectus. The levator palpebrae and superior rectus muscles are dissected along the lateral and medial surfaces of their bellies. They are then transected and reflected out of the operative field, exposing the orbital cone from the globe to the optic foramen.

Dissection of the contents of the cone is difficult because of the abundant fatty tissue, the volume of the globe, the rapidly tapering bony confines of the orbit, and the unpredictability of displacement of intraorbital arteries and veins. The use, consequently, of self-retaining retractors is mandatory if one desires a precise and complete dissection of the tumor, which may, in its broadest terms, be located freely within the cone, between optic nerve and its sheath, or entirely within the optic nerve.

The instrumentation for resection of an intraorbital tumor has classically consisted of suction, microdissecting instruments, and bipolar cautery. The technique is directed toward isolating the tumor from the surrounding tissue and then removing it, either *en bloc* or in fragments. This requires maintaining a clean plane of

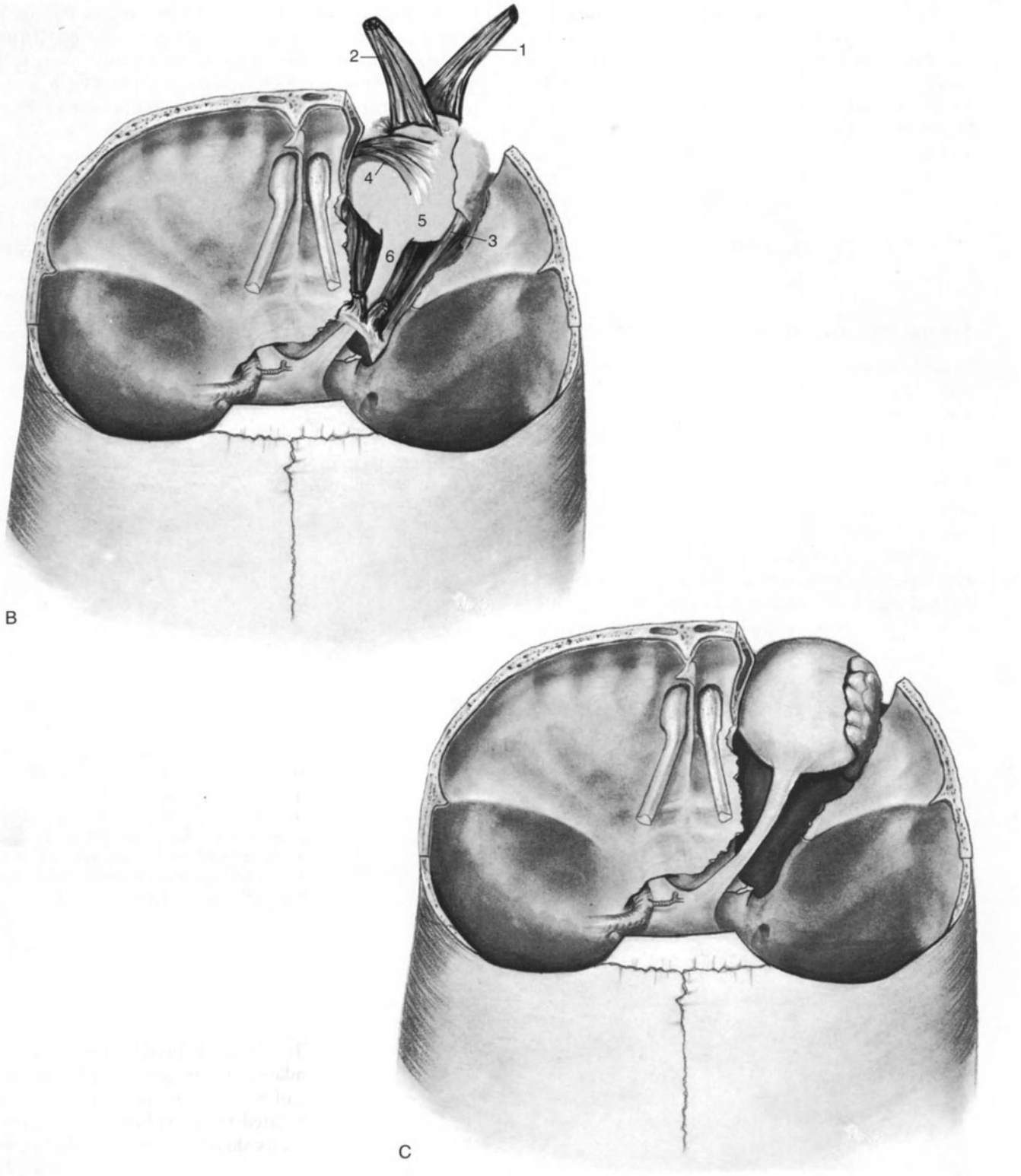


Figure 10.12. (B) Transecting the levator pallpebrae (*1*) and superior rectus (*2*) exposes lateral rectus (*3*), superior oblique (*4*), globe (*5*), and optic nerve (*6*). (C) All muscles have been removed to illustrate the course of the optic nerve in the unroofed orbit. It follows a posteromedial course from the globe to the optic foramen, gradually approaching the orbital roof.

cleavage between the tumor surface and neural or adnexal elements. Modern technique allows for the use of the laser to vaporize or the Cavitron to aspirate ultrasonically the tumor in situ, avoiding completely the need for teasing tumor away from healthy neural or muscular tissue. The optic nerve, ophthalmic artery, lachrymal gland, and globe are covered with Telfa strips before the laser Cavitron microdissection techniques are used, with a setting of 4–6 W and intermittent beam for the laser. The Cavitron is very effective, but is bulky and not safe where tissue and vital vasculature are intermixed.

Anterior Cone Tumors (Fig. 10.13)

Those tumors which occupy the anterior cone, and which are not excrescences from the globe or optic nerve, nestle between the four rectus muscles externally, the optic nerve centrally, and the posterior surface of the globe anteriorly. The lachrymal gland is well out of the tumor area. Fibrous bands, very similar in appearance to arachnoidal tissue, bridge the space between tumor and globe. They may be dissected free by using a cotton fluffy or, preferably, the tumor may be shrunken with bipolar cautery, aspirated with the Cavitron, or vaporized with the laser. Either of these two latter techniques are preferable to attempts at *en bloc* removal because of the minimization, or elimination, of traction. Both techniques provide progressively greater exposure of the area as the tumor diminshes. One may follow the globe directly into the optic nerve, maintaining a clean plane of separation between tumor and the neural tissue as the former is taken from the latter.

If the tumor is located on the ventral surface of the optic nerve, it is necessary to take it from the globe and lateral surface of the nerve first. One may then move on to the globe and medial surface of the nerve. Resection of tumor from the inferior surface of the nerve is to be reserved for the very end of the procedure, since this avoids the need to displace tumor and nerve from side to side, and to work constantly on the full bulk of the tumor while manipulating the optic nerve.

Though every care is taken to avoid compression, or excessive displacement, of the optic nerve, one must not think that gentle and firm retraction of the nerve is to be avoided at all costs. The optic nerve is a robust structure, one with a high degree of mobility. Cover it with tiny strips of Telfa and, if necessary, retract it.

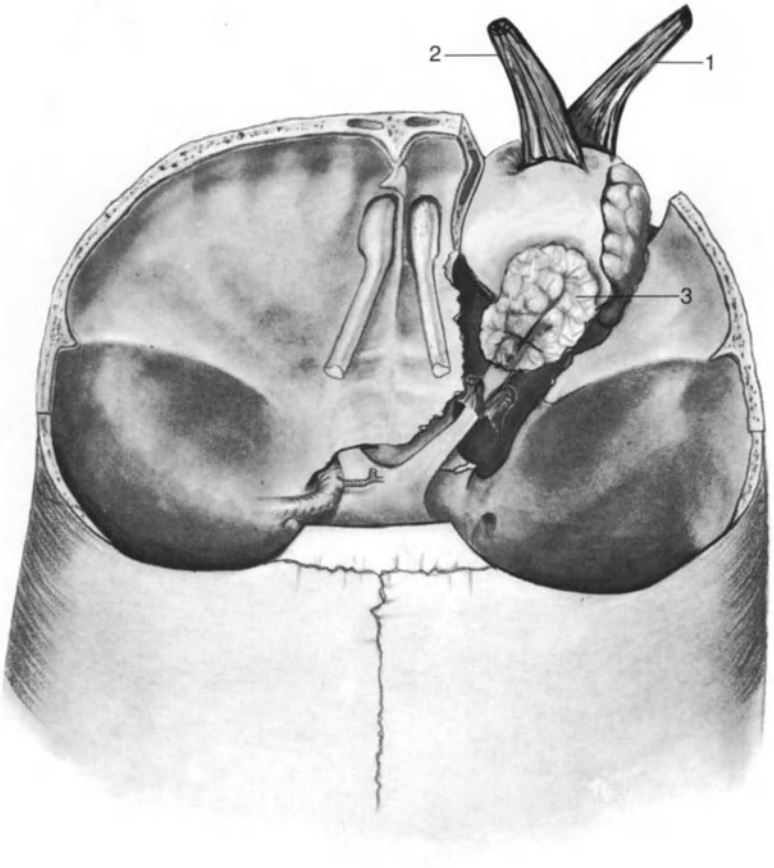
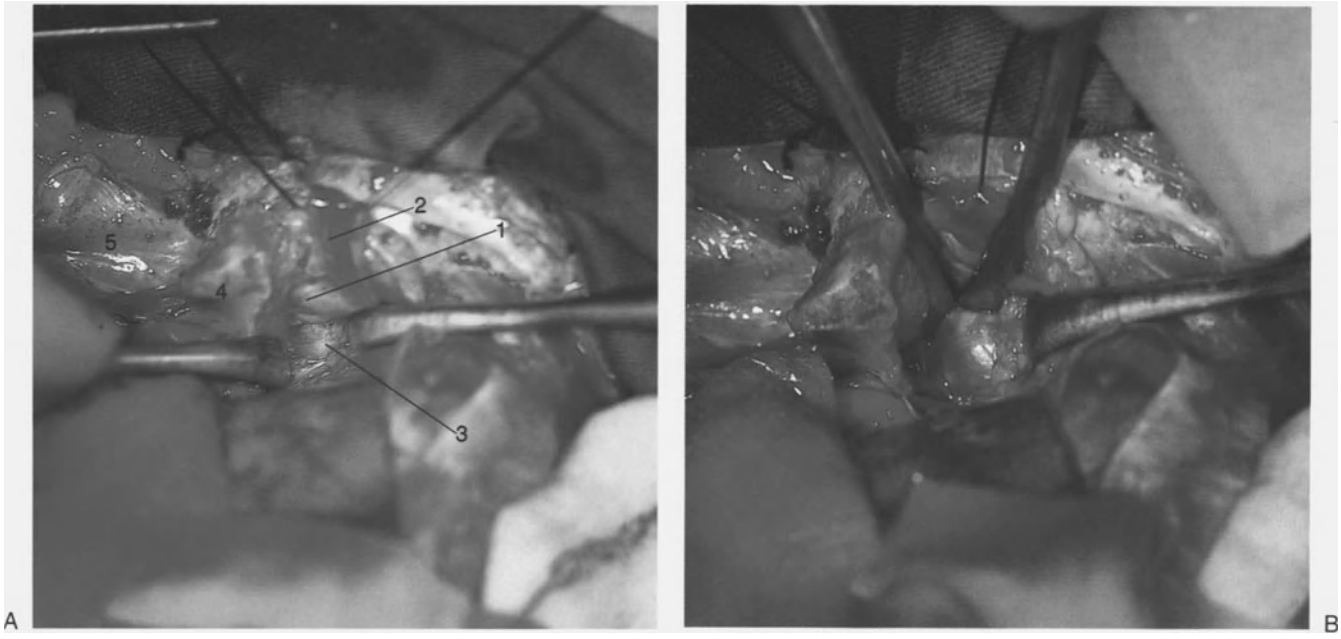


Figure 10.13. This anterior cone tumor nestles within the base of the cone, between the globe and the optic nerve. Reflection of the levator (*1*) and superior rectus (*2*) muscles exposes completely the tumor (*3*) within the anterior portion of the cone. The intraorbital fatty tissue has been removed.



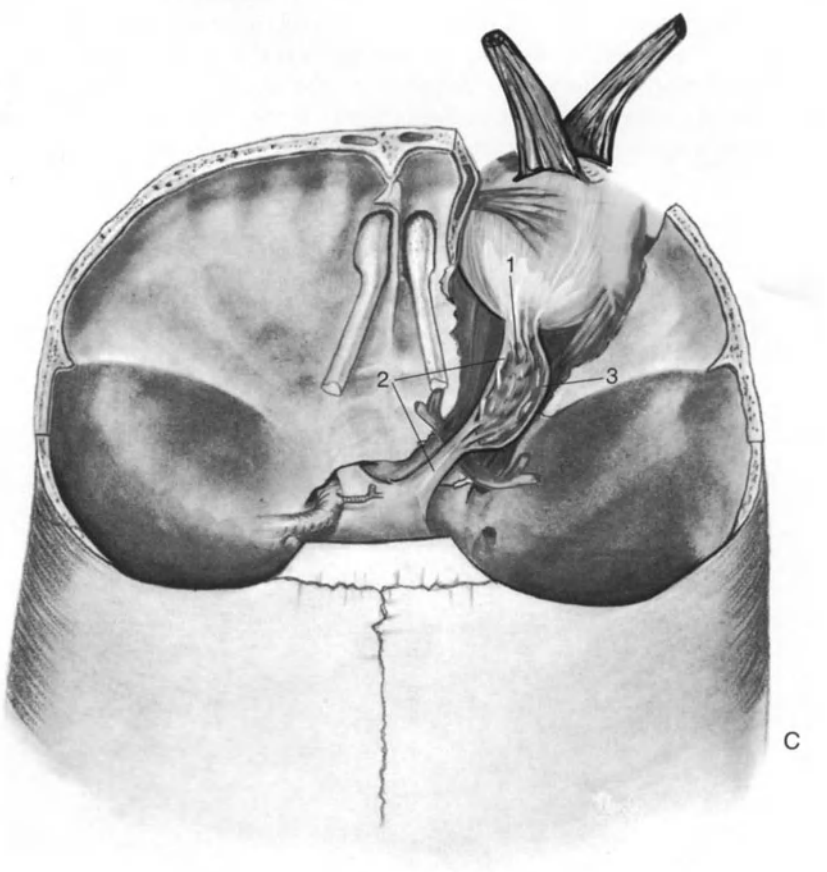


Figure 10.14. (A) This child had a meningioma growing from the optic nerve sheath. The globe (*1*) has been exposed after the levator palpebrae and the superior rectus muscles (*2*) were transected and reflected anteriorly. The widened optic nerve (*3*) is exposed between the two Penfield dissectors. It is not possible at this time, before opening the nerve sheath, to be certain as to whether this tumor is an optic nerve glioma or a nerve sheath tumor. The zygomatic process of the temporal bone (*4*) and retracted temporalis muscle (*5*) are labeled. (B) Once the sheath has been opened, however, the mesenchymal nature of the tumor becomes clear and one is able to dissect it from the underlying optic nerve. (C) This is a drawing of a mesenchymal tumor (*1*) located between the optic nerve (*2*) and its sheath (*3*).

Nerve Sheath Tumors (Fig. 10.14)

One may separate space-occupying lesions that cause an enlargement of the optic nerve sheath into those located between the sheath and the nerve, and those located within the optic nerve. The deformity of the nerve sheath appears the same in both instances, so that the surgeon may not be certain of what he is dealing with until he has incised the perineurium. Even then, on occasion, the optic nerve glioma may be an excrescence from the optic nerve, fungating out at a given point on the nerve only to be reflected back into it by the nerve sheath.

Incision of this sheath with a #11 blade gives egress to tumor tissue, allowing the surgeon to inspect it be-

fore proceeding with a resection. The sheath should be opened from the globe to the optic foramen before the dissection is undertaken. If the tumor is independent of the optic nerve, it may be dissected from its adhesions to the nerve by extending the dissection from anterior to posterior, lifting the tumor from the nerve as one proceeds. If a laser is used, extreme care must be taken to protect the optic nerve by continuously placing Telfa or fluffy cotton between the tumor and the nerve as the former is vaporized. Equal attention must be given to these structures when using the Cavitron.

Optic Nerve Tumor (Figs. 10.15, 10.16)

If the tumor is within the optic nerve, it may be confined to the orbit, but it may also extend through the optic canal and into the subarachnoid portion of the optic nerve and chiasm. Removal of these tumors is incompatible with vision, so the only indication for excision is an unsightly exophthalmus in a blind child who has an optic nerve tumor which is entirely intraorbital. If the proptosis is so severe as to result in corneal damage, the tumor should be removed irrespective of whether it is confined to the orbit, unless one chooses orbital exenteration and implantation of a prosthesis. If these criteria are met, then the optic nerve is sectioned at the globe and at the optic foramen, isolating the neoplastic nerve so that it may be lifted from the field.

There is no indication for extending the dissection intradurally. An enlarged optic foramen on CT or MRI will already have informed the surgeon whether there is extension of tumor into the subarachnoid portion of the optic nerve.

Age and Brain Tumors

It is of value to discuss characteristics particular to specific (newborn, toddler) age categories now, so as to permit the reader to continue to identify the very real differences between these age groups and those of the juvenile and adolescent...just as we see in craniocerebral trauma, hydrocephalus, vascular pathology, etc.

The incidence of brain tumors has been reported to range between 1.3% [70] and 5% [71] of the childhood population. Unfortunately, the literature holds little information concerning incidence in the newborn and infant. With the increase in attention given to the care of the newborn, the specialization of pediatric neurosurgery, and the use of CT, we may expect both increased numbers and more detailed reporting of brain tumors in specific (anatomophysiological rather than chrono-

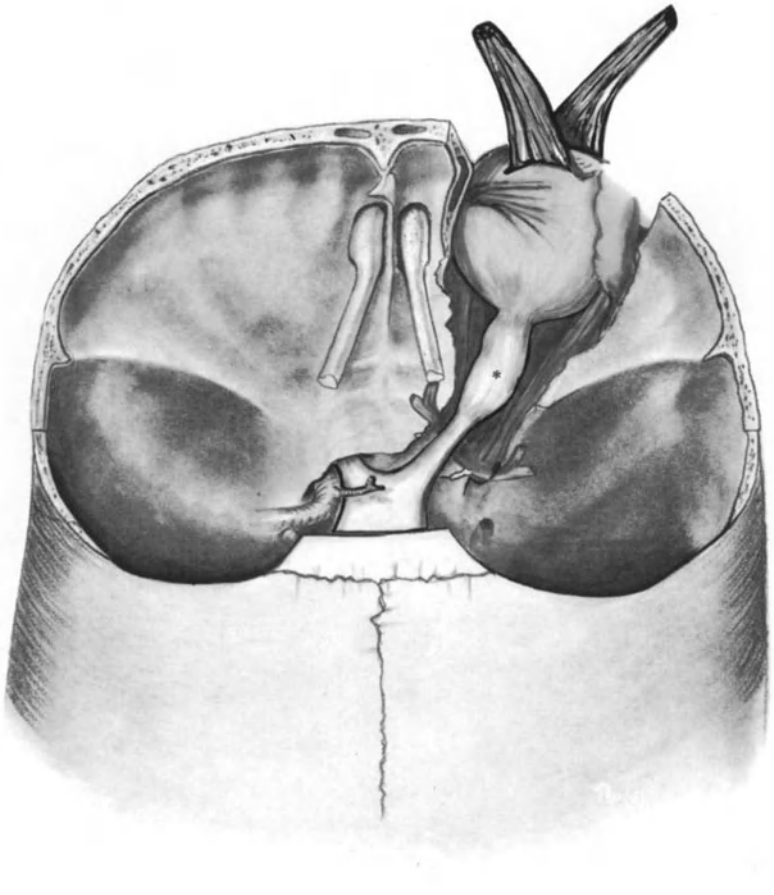


Figure 10.15. An optic nerve glioma (*) growing entirely within the intraorbital portion of the optic nerve does not extend through the optic foramen.

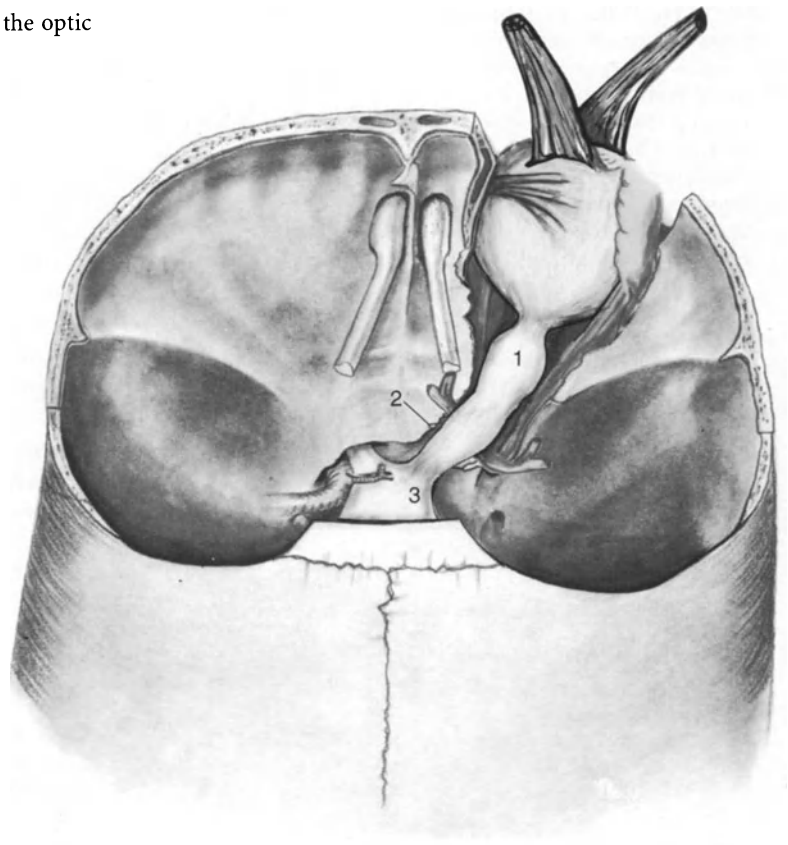


Figure 10.16. An optic glioma (*1*) extending through the optic foramen (*2*) and up the optic chiasm (*3*).

logic) age categories: newborn, infant, toddler, juvenile, and adolescent.

It has been described in the literature that brain tumors of infancy occur much more frequently in the supratentorial compartment (almost twice as common) [20, 72], that they are not unusually large in size at the time of detection, that there is a high incidence of lateral ventricle choroid plexus papilloma and ependymoma, and that postoperative survival is extremely poor in this age group [73, 74]. Our experiences gainsay these statements. Because of the expansile cranium, open sutures, fontanelles, as yet undeveloped central nervous system, widened extracellular spaces [75], and basal cisterns which allow external and internal decompression, most infants suffer surprisingly mild symptoms...in spite of the magnitude of the tumor.

It is of value to review our experiences (at Northwestern Unversity–Children's Memorial Hospital, Chicago) with 39 intracranial tumors detected during the 1st year of life, specifically, with regard to (1) location and histological pattern, (2) symptomatology, (3) treatment and results, (4) criteria for predicting prognosis, and (5) "quality of survival."

Materials and Methods

Of the 510 children with intracranial tumors treated, 55 of these were diagnosed (histologically proven) during the 1st year of life (weeks to 12 months). Of these, 29 were supratentorial, 19 were infratentorial, 7 involved both brain compartments, and 31 were histologically malignant tumors.

Sexuality proved significant in this age group: all choroid plexus papillomas and three-fourths of the medulloblastomas occurred in males; and two-thirds of the benign astrocytomas in females. The ratio of supratentorial to infratentorial tumors was 1.8 to 1.

Twenty infants were diagnosed before 6 months of age, 9 of whom were under 2 months of age. Thirty-five patients were symptomatic before 6 months, 15 during the neonatal period. Though the onset of symptoms and signs diagnostic of brain tumors may appear during the 1st year of life, histological confirmation may not be possible until later. In contrast to others [73–77], we have chosen to consider, like Sunder-Plasman [78] and Sunder-Plasman and Jellinger [72], only those diagnosed histologically during the 1st year of life. We did this because symptomatology, management, and prognosis are different for children on whom histological verification is made during the 1st year of life.

In a review of all our childhood brain tumors for age of onset, we note that 12, of 78, medulloblastomas

Table 10.11. Order of frequency of signs of brain tumor, reasons for consultation, and location of brain tumors

Signs of brain tumor	
Bulging fontanelle	Asymmetrical head
Increased head size	Ataxia
Papilledema	Hemianopia
Poor head control	Spasticity
Hemiparesis	Stiff neck
Abducens palsy	Vocal cord palsy
Facial weakness	Torticollis
Optic atrophy	
Reasons for consultation	
Increased head size	Failure to thrive
Vomiting	Stridor
Delayed milestones/developmental arrest	Blindness
Lethargy/irritability	Sudden coma
Seizure	
Location of brain tumors	
Supratentorial	
Cerebral hemisphere	
Intraventricular	
Parasellar	
Infratentorial	
Cerebellar-fourth ventricle	
Brainstem-diencephalon	

(15.5%), 11 of 159 benign astrocytomas (7.0%), and 6 of 33 malignant astrocytomas (18%) occurred during the 1st year of life. It is important to note that 8 of 13 choroid plexus tumors occurred at this age. The most common tumor was medulloblastoma. Benign astrocytomas were the second most common tumor, followed by choroid plexus papillomas.

Half of the patients with abducens palsy and most of the infants with hemiparesis had supratentorial tumors. Optic atrophy or hemianopia was found predominantly in the cases of supratentorial tumor. All infants with deformities of the head had either cerebral hemsiphere or lateral ventricle tumors, and the bossing was ipsilateral to the tumor [74]. One-fifth of the children with normal head size were found to have hydrocephalus. The most common signs were a full, bulging fontanelle and rapid increase in head circumference, expressive of a flexible skull and open sutures (Table 10.11) [72, 73, 79, 80]. Despite this increase in ICP, the incidence of papilledema was low. Other signs of increased ICP were vomiting, abducens palsy, lethargy, irritability, developmental arrest or delayed milestones.

Infantile brain tumors are frequently associated with hydrocephalus, either obstructive or CSF overproduction, expressed as the midline syndrome [70]. Cerebral or intraventricular tumors, which were invariably huge, also presented only subtle lateralizing signs, attributable to the immature development of the cerebrum and wider extracellular spaces characteristic of this age group. Stiff neck, vocal cord palsy, or torticollis indicate the presence of a mass in the posterior fossa. Optic atrophy was usually associated with a suprasellar mass.

Eighty percent have hydrocephalus complicating the brain tumor. If a posterior fossa or pineal region tumor is present, *III ventriculotomy is now the shunting procedure* (CSF is shunted from the III ventricle into the interpeduncular cistern) of choice before attacking the tumor, though chemotherapy is now a very important alternative or addition. Shunting (or chemotherapy) prior to the craniotomy results in a decrease of the intracranial pressure and marked improvement in the physiological condition of the infants [69].

The postoperative mortality is high, approaching 20%, and the causes are gastrointestinal bleeding, intraoperative bleeding, and inappropriate (ISNDH) ADH syndrome.

Brain tumors are the cause of 5% of neonatal deaths; yet they have seldom been diagnosed antemortem [81]. Despite the common misconception that childhood tumors usually occupy the infratentorial region, brain tumors in neonates and infants, in fact, occur more frequently in the supratentorial region. This observation of ours has already been reported, with the ratio being 2.8:1 in one series [20] and 3.6:1 in another [72].

Fessard [74] reported the supratentorial/infratentorial ratio increased during the first 6 months of life, becoming nearly equal during the second 6 months, and reversing after the 1st year. We, however, observed the ratio to be equal – after correcting for pineal area tumors.

Histological patterns of brain tumors in this age group are quite interesting. Sunder-Plasman and Jellinger [72] describe spongioblastoma as the most common brain tumor in stillborns. It has been reported that choroid plexus papilloma, ependymoma [82] and medulloblastoma [73, 83] are the most common tumors. Our experiences are that medulloblastoma and benign astrocytoma are the most common, with choroid plexus papilloma and meningeal sarcoma ranking third and fourth. Takaku et al. [80] report teratoma as the most common tumor in the neonatal period.

Brain tumors have been considered "congenital" by many authors. However, there are no reliable data available regarding the growth rate of tumors to justify classifying them as "congenital." Some authors have defined "the congenital origin" to be restricted to tumors presenting during the neonatal period [84–86]. Jellinger and Sunder-Plasman [87] defined brain tumors found before 2 weeks of age as definitely "connatal." Bell and McCormick [88] consider "congenital" tumors, such as teratoma, chordoma, nasal glioma, or cholesteatoma, to originate from embryonal rests present in the brain at birth. All of these are rare, indeed, as occurrences in the neonate and the infant. Russell and Rubinstein [89] stated: "Although the existence of congenital tumors is

seldom established at the time of birth, it is permissible to calculate on clinical grounds that certain others which are verified in the neonatal period have been congenital. Most of the congenital expamples are primitive neuroectodermal tumors such as retinoblastoma, medulloblastoma, neuroblastoma of the adrenal medulla, and medulloephithelioma.

It has been our observation that most tumors, benign or malignant, which present in early life are huge at the time of diagnosis. Therefore, considering the size of the tumor and the short duration of postuterine life, we feel most of these tumors are "congenital" to the extent that they are present at birth or during intrauterine life. It is our thought, and suggestion, that this be distinguished from "developmental," a term which suggests persistence of "primitive" cells within mature tissue and organ systems. We cannot accept the "developmental" theory.

Hydrocephalus is frequently associated with supratentorial tumors, "midline" tumors, or choroid plexus papilloma. Precraniotomy shunt has been proposed to relieve the increased intracranial pressure due to hydrocephalus [10, 84, 90]. This is especially significant in infants who are physically ill because of persistent vomiting and failure to thrive. The precraniotomy shunt allows them time to improve their physical and nutritional status while waiting for the definitive surgery, and facilitates reexpansion of brain parenchyma. This is of particular importance when performing posterior fossa surgery where the aqueduct of Sylvius is unplugged by resection of the tumor, thus allowing CSF to escape suddenly and potentiating the risks of hypotension syndrome or subdural hematoma.

Postoperative radiation therapy has been recommended for malignant tumors. We express caution and reservations concerning the tumor dosage and indiscriminate, routine "prophylatic" radiation to the entire brain and spinal cord in children [12] who have discreet tumors either of the vermis, pineal region, or parasellar area irrespective of the tissue type and age. We cannot accept the unfounded concept that irradiating a tissue before it becomes prey to tumor invasion is of value. We consider it dangerous! Present CT techniques allow one to monitor the brain regularly, thus enabling physicians to treat metastases precisely if, and only when, they occur. Because of the immature brain cells, the effects of radiation are more serious than in older children and adults [91]. When indicated, radiation therapy should be carefully instituted in the form of radiosurgery in consideration of the mental development of infants [12].

According to *Fessard* [74], only 5 of 62 infants showed completely normal findings during the period of survival; *all 5 had posterior fossa tumors.* Farwell et al. [73] reported that 8 were retarded or handicapped, 3 were normal and 5 were unknown among 17 survivors of more than a year. Raimondi and Tomita [7] called attention to the superior results obtained when "total resection" of the tumor was achieved.

Previous reports were not encouraging in terms of prognosis. In 10 cases reported by *Sato* et al. [77] there were 5 deaths less than a year after surgery and only 19 of 66 infants and children under 2 years of age survived more than a year [74]. Adding up the higher incidence of failed or delayed diagnosis and intraoperative death, even the benign choroid plexus papilloma in the newborn or infant is associated with poor overall prognosis [67].

One is advised to separate, however, neonatal brain tumors from those diagnosed in later infancy, since the former have a poorer prognosis: only 4 out of 14 neonates reported by Takaku et al. [80] survived more than 1 year after surgery. The opposite is true for tumors occurring during the second 6 months of life. As a prognostic indicator, posterior fossa tumors in which all but one were malignant in the present series showed brief survival. As we have already reported, medulloblastoma is associated with a poorer prognosis in the very young [7]. Infants with meningeal sarcoma, despite microscopic evidence of malignancy, enjoy a longer survival after radical resection and postoperative radiation therapy. On the contrary, choroid plexus papilloma has the best overall prognosis [67, 76, 79], though 80% of our patients remained shunt-dependent. Astrocytomas, benign or malignant, of this age group, usually invade diffusely the cerebrum, diencephalon, or brainstem, and are associated with a poor prognosis. We take note of the fact that some authors have reported series of optic pathway glioma associated with a good prognosis, but that the reports included cases ranging very widely in age from the very young through adolescence [55, 58]. Our experiences are that there is a poor prognosis for children under 1 year of age, especially *when a diencephalic syndrome is present* in a child with a hypothalamic-optic pathway glioma (75% dying within 2 years). The prognosis is relatively better if a diencephalic syndrome is not present at the time of diagnosis.

The fact that 10% of our children have survived 5 years or longer, without evidence of recurrence and without neurological or mental deficits, provides supporting evidence for the treatment of brain tumors during the 1st year of life, but obliges one to be measured in the prognosis.

Hemispherical Tumors

Solid Hemispherical Tumors

The solid hemispherical tumors in childhood may be highly vascular or avascular. They range from the vascular ependymoma and optic pathway glioma (which

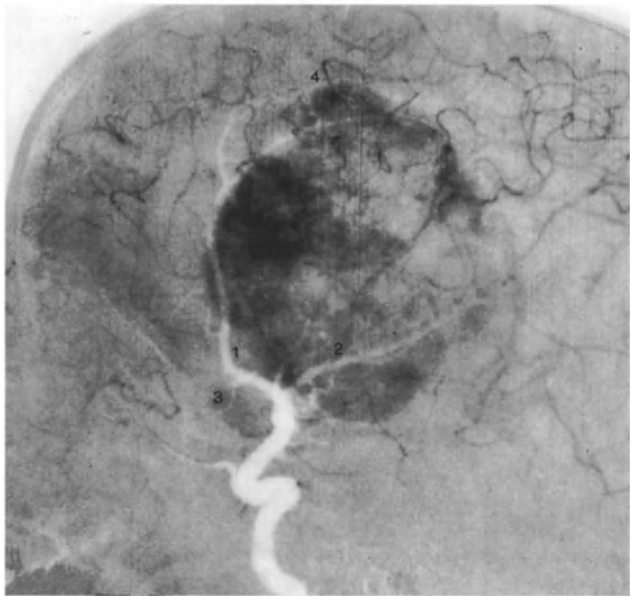


Figure 10.17. Solid vascular hemispherical tumor. This study reveals the superimposition of the anterior (*1*), middle (*2*), frontobasal (*3*), and sulcomarginal (*4*) arteries upon the tumor stain, allowing one to appreciate extension of the tumor within the rostrum, genu, and body of the corpus callosum as well as into the septum pellucidum and III ventricle.

extends into the temporal lobe from the optic tracts), to the totally avascular hamartoma and astrocytoma diffusum.

Exactly as in the adult, CT characteristics permit no insight into the vascularity of the tumor nor does enhancement on CT studies offer incontrovertible proof that the lesion is neoplastic.The cerebral hemisphere glioma has a very poor prognosis: only 20% survive more than 5 years. Radical resection does not alter the course of this tumor, irrespective of its histological appearance! Consequently, one is not justified in attempting to remove all tumor tissue. Unfortunately, radiation therapy is as ineffective in curing these tumors as are surgery or chemotherapy, but a combination of the three offers palliation.

Highly Vascular Hemispherical Tumors

(Figs. 10.17, 10.18)

The operative procedure and, indeed, decision whether to operate or to proceed directly with roentgen therapy is to a great extent made on the basis of the vascularity of the tumor: highly vascular invasive intraparenchymal lesions in children, in general, but infants, in specific, are malignant, moderately sensitive to roentgen therapy, and impossible to resect completely.

Another characteristic of highly vascular intraparenchymal tumors is their tendency to take blood supply directly from the circle of Willis or the main trunk of

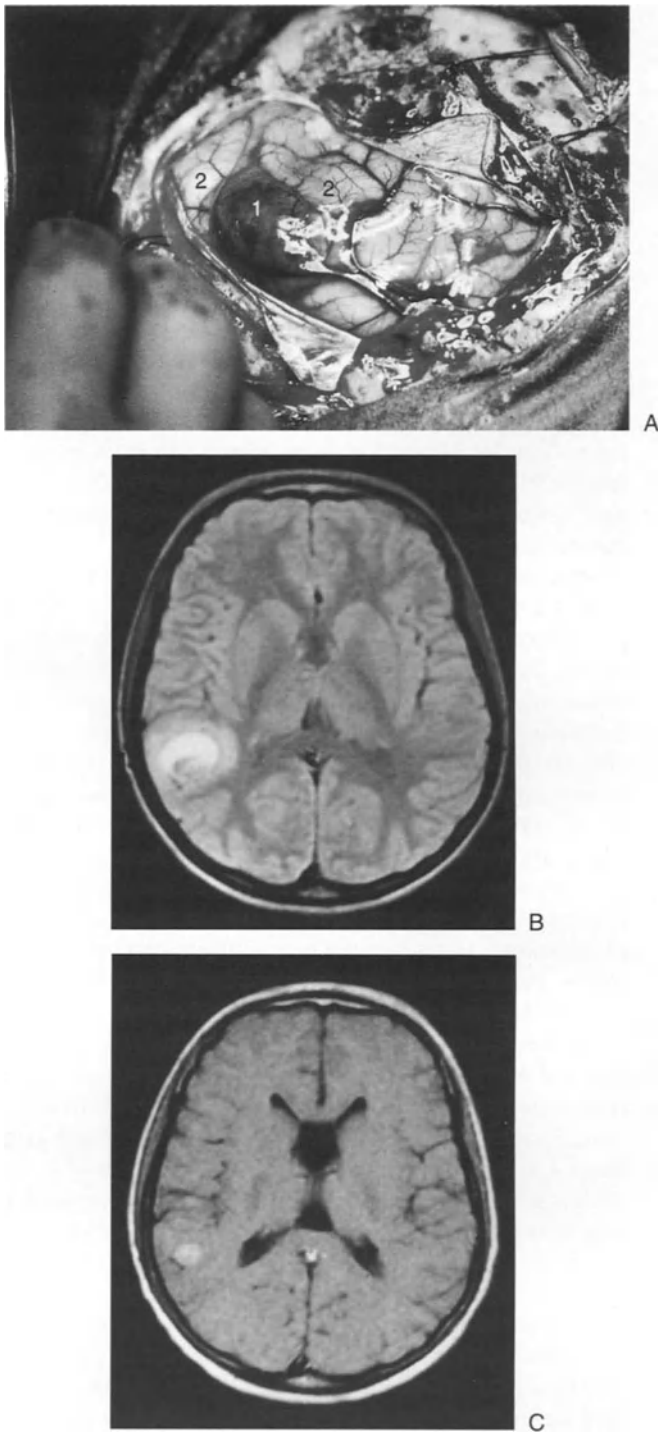


Figure 10.18. (A) This highly vascularized, well-circumscribed mass was resected by working around the periphery of the tumor where it borders normal cerebral tissue, after having occluded the main feeding vessels. The bridging vascular structures between the tumor and surrounding tissue were coagulated and transected as the dissection proceeded freeing the tumor (*1*) completely from the surrounding cortex and white matter (*2*). The two axial MRI scans (B) and (C) are respectively T2 and T1 post-enhancement images of the cavernoma (considered for the purposes of this text to be a highly vascularized tumor).

the posterior cerebral artery. This renders resection all the more difficult, because it necessitates isolation of the main trunks of the feeding vessels to the tumor prior to the initial cerebrotomy for lobectomy. To proceed directly with incision of highly vascularized intracerebral tumors before isolating and occluding the feeding vessels is ill advised.

The craniotomy and dural opening selected for a highly vascular hemispherical tumor should be such as to provide access to all main-trunk arterial and venous system structures without the need to approach them in a transcerebral manner. A frontal or occipital lobe vascular tumor would necessitate, respectively, a medial frontal, or a temporoparieto-occipital flap, so as to isolate immediately the anterior cerebral or posterior cerebral arteries and the cortical bridging veins. These tumors are approached from the far end, that is to say from the surface more remote to the convexity of the hemisphere, to minimize blood loss. The efferent vessels to the tumor are dealt with one at a time, coagulating, or (clipping and transecting) them until the tumor is completely devascularized of arterial blood.

One does not take the venous channels prior to perfoming the cerebrotomy: the neoplasm also draws blood from the surrounding healthy brain tissue. Shunting of oxygenated blood into venous channels is common. If this were to be done the neoplastic lobe would swell up immediately, forcing an impromptu resection. Rather, it is preferable to proceed to the cerebrotomy, working at the periphery of the neoplasm, by coagulating arachnoid, pia, and gray matter, along a linear plane, tediously stopping each minute bleeder as it is encountered. Though these vessels, especially within the white matter, appear thin-walled, they necessitate transection. Simple coagulation results in their being torn along their course through healthy white matter, with parenchymal outpouring of blood in a spongelike fashion. Extension of the cerebrotomy from the gray into the white matter proceeds only after the overlying arachnoid and pia have been coagulated and transected. Once the cerebrotomy has been completed and the involved lobe isolated, the bridging veins are coagulated and transected.

It is not unusual for the tumor to receive vascularity from the dura mater, falx, and tentorium. It is common for abnormal venous channels to drain into the dura at the orbit, the ethmoid plate, the sphenoparietal sinus, and the tentorium.

Avascular Hemispherical Tumors (Figs. 10.19–10.26)

Avascular intraparenchymal tumors are removed either with the cerebrotomy or lobectomy techniques, depending upon the size and extent of the mass. It is unusual for a glioma in childhood to occupy only a portion of a lobe. The rule is for these tumors to be quite large, to extend into one or more additional lobes: frontoparietal and frontotemporoparietal. Also, they extend into the internal capsule and basal ganglia. The lobectomy technique affords maximum tumor removal; it is not directed toward internal decompression. The cerebrotomy technique is reserved for the unusual, well-circumscribed, small tumors. Parietal lobe, basal ganglia, and thalamic tumors are not resectable because of the very serious functional consequences. Radiosurgery and/or chemotherapy is the treatment of choice for tumors in these strucutres since it is associated with a minimal morbidity, resulting in a far better quality of survival.

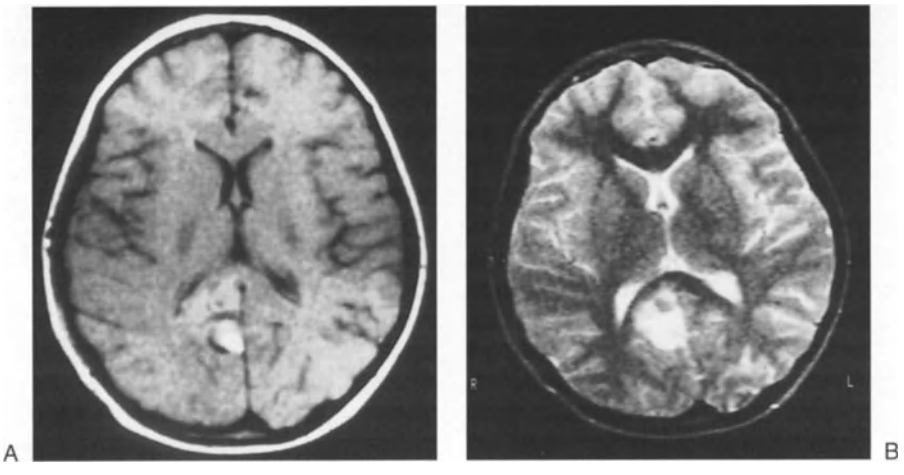


Figure 10.19. This avascular (oligodendroglioma) tumor of the right medial parietal lobe is shown in axial T1 (A) and T2 (B) weighted images. The only difficulty a tumor such as this presents is one of access, which needs to be parasagittal, along the falx.

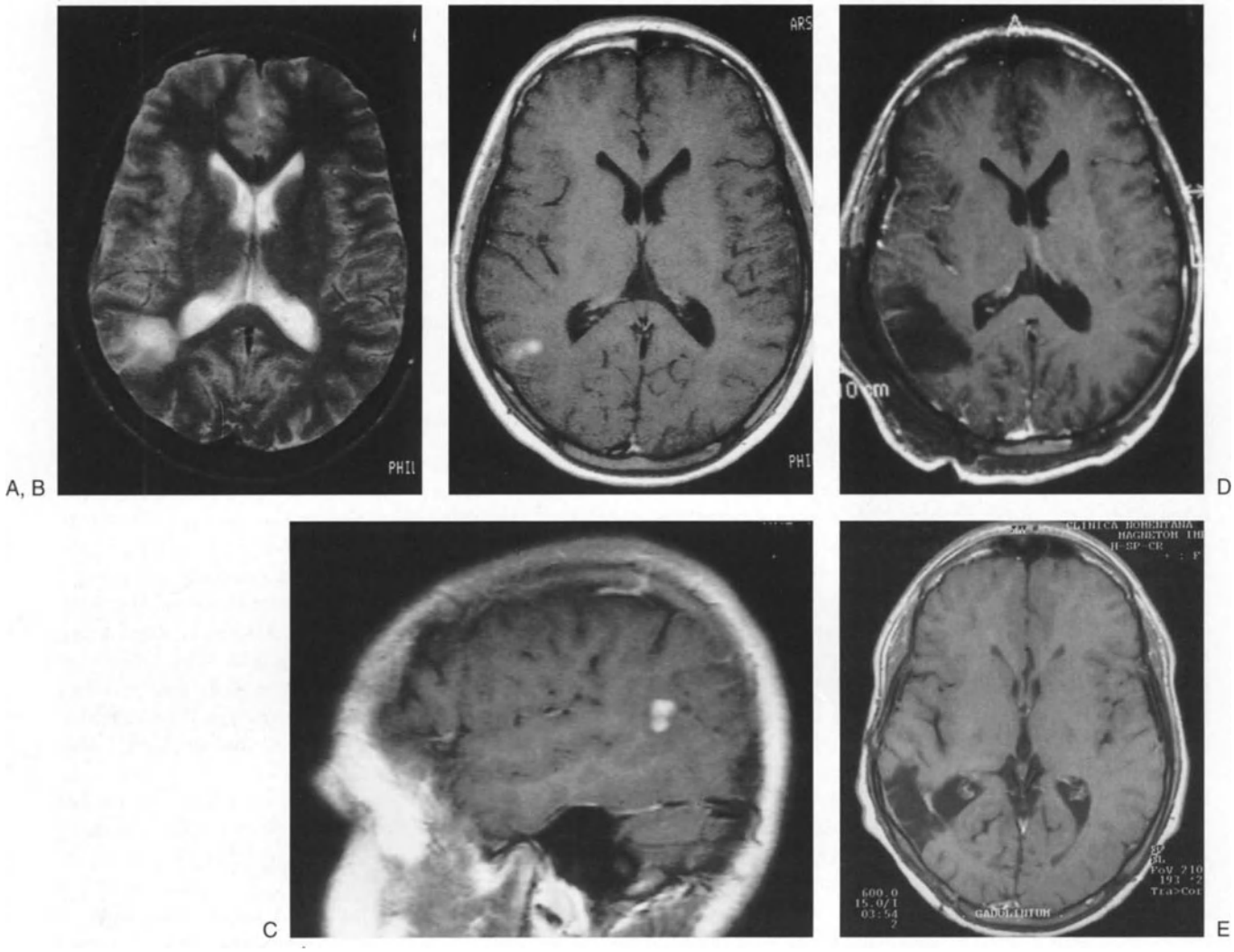


Figure 10.20. A very interesting tumor of the cerebral hemispheres, one composed of neural cells, most often resembles a malformation rather than a neoplasm in that it is most probably malformative in origin. Though the tumor has a certain neural cell component, the frequency and distribution of these elements is variable, with interspersing of glial cells. Thus, what is in adult neurosurgery a very rare tumor, in pediatric neurosurgery is relatively common, and the sites most frequently harboring these tumors are the temporal lobe, the tuber cinerium, and the para (III) ventricular area. In this figure, a right subcortical temporo-occipital lesion is illustrated. In (A), there is a homogeneous area of high signal intensity; in (B), the post-gadolinium study, one is able to identify the solid enhancing portion of the neoplasm; in (C) the post-gadolinium image in the sagittal projection reveals the mass located in the area of the supramarginal gyrus. (D) and (E) are the two postoperative gadolinium studies, respectively, 3 days and 7 months after surgery, revealing the surgical cavum with no enhancing residual mass and subsequently filling the cavum with cerebrospinal fluid but no residual tumor. It is the natural history of this tumor to be susceptible to complete cure when a total resection is effected.

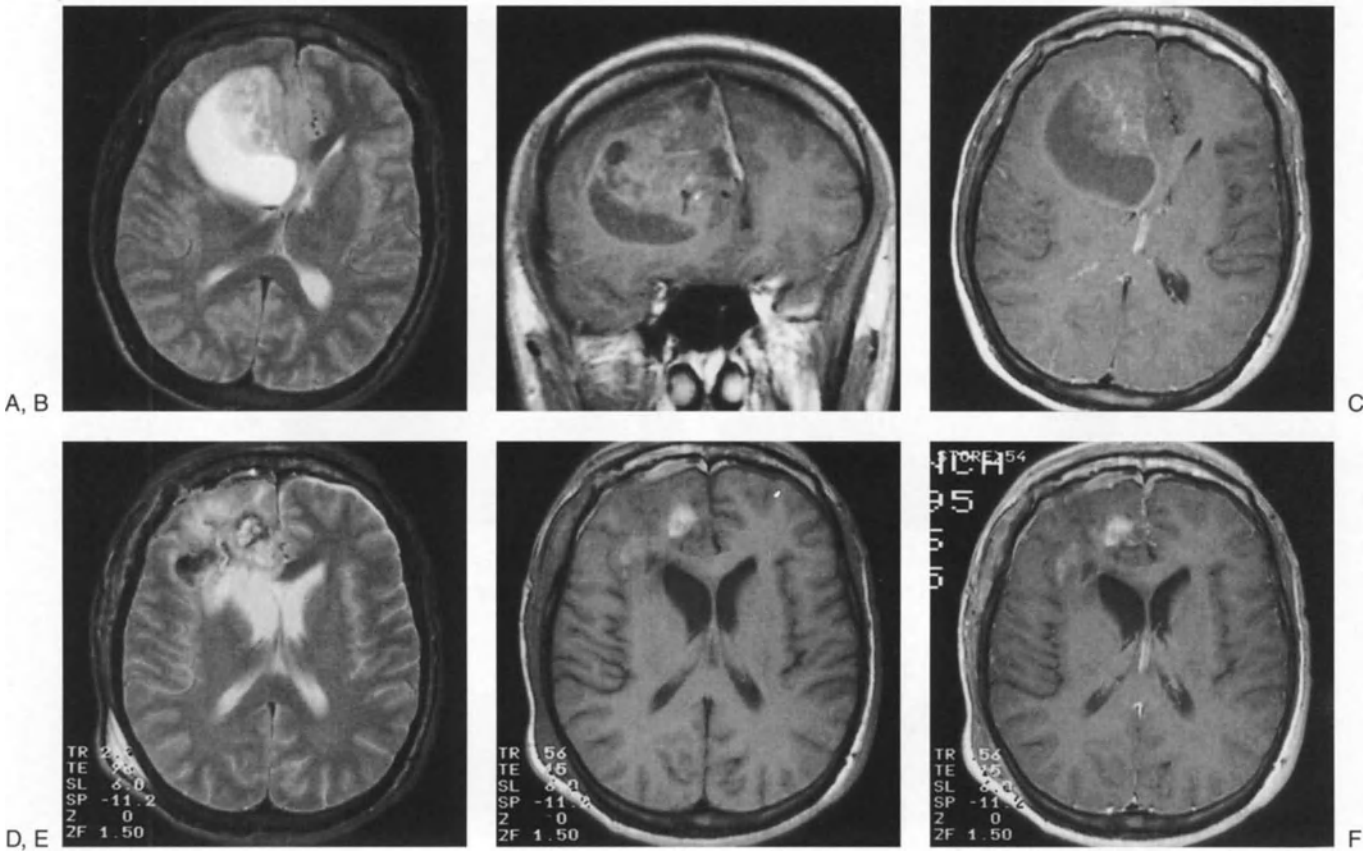


Figure 10.21. The *primitive neuroectodermal* tumor (PNET) represents a purely paradigmatic problem since the time of its introduction by Hart and Earl in 1973 [186]. The World Health Organization has proposed a classification of PNET which includes *neuroblastoma, pineoblastoma, medulloblastoma, and ependymoblastoma.* Whatever the histological orientation of the reader, it suffices to say that this is a highly malignant tumor with varying responses to the combination of treatments surgery/irradiation/chemotherapy. In this figure a right frontal lobe mass is illustrated. In (A) the lesion shows a hyperintense T2 weighted semilunar fluid portion with a low signal nonenhancing mass visible in the sagittal (B) and axial (C) sections. The mass shows an isosignal on both T2 and T1 weighted images in (A), (B), and (C), with foci of high signal on T2, representing necrosis. The post-gadolinium studies (B) and (C) reveal the lesion to have few, diffuse areas of enhancement. In the postoperative studies, less than 4 days after surgery in the (D) T2 weighted image, (E) the T1 weighted image, before gadolinium injection, and (F) the post-gadolinium T1 weighted image, one sees an area of high signal in (E) represented by blood in the surgical site. The cystic portion of the mass is no longer evident (D), and no residual enhancing tumor is evident. This child died 8 months after surgery. He received neither chemo- nor radiation therapy.

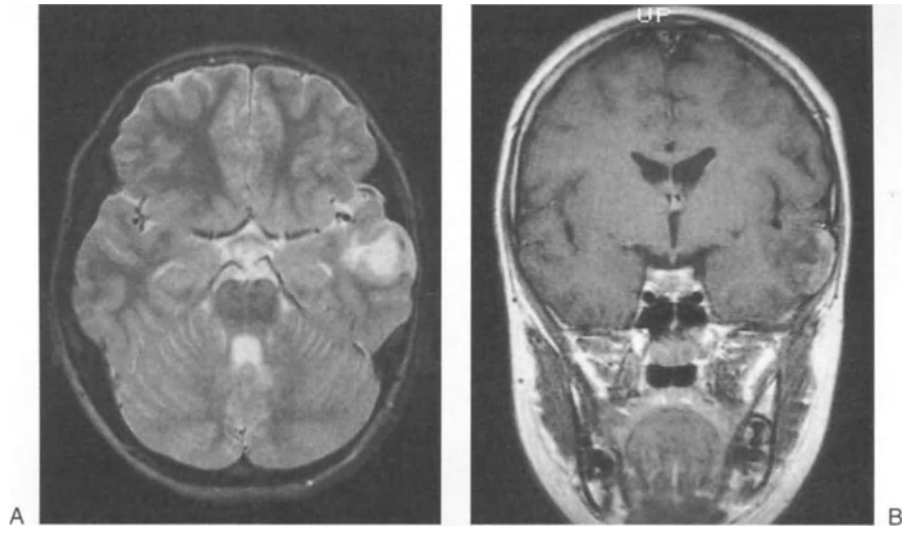
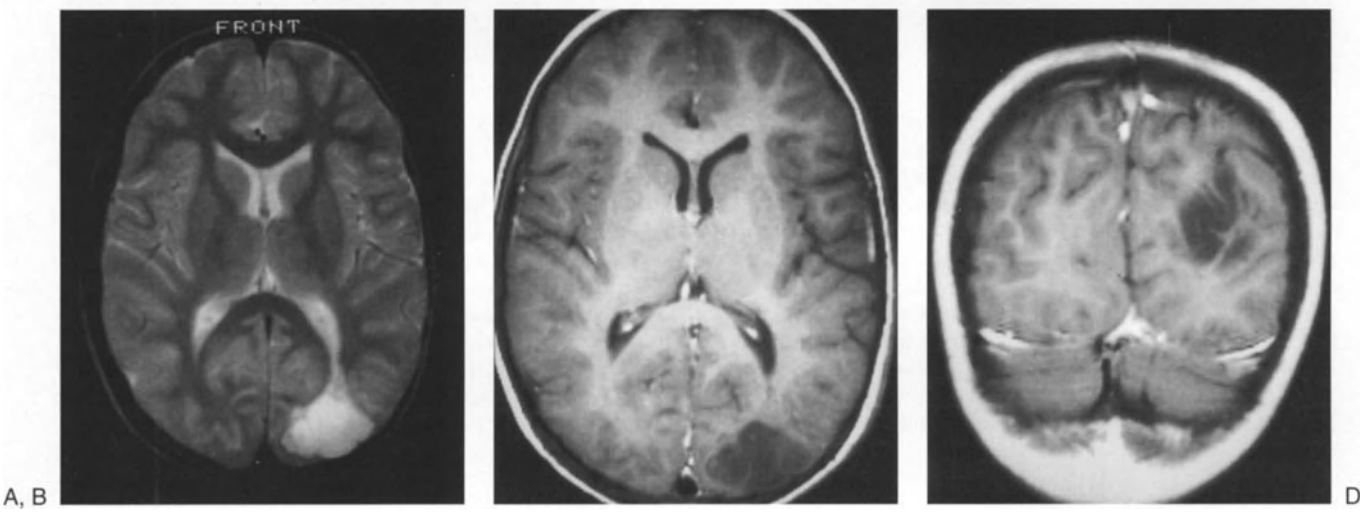


Figure 10.22. For the present, disappointingly true, we are not able invariably to classify precisely from a histological point of view *cerebral hemisphere tumors.* This grade 2 *astrocytoma* is presented as such and no implication is made concerning a relationship between magnetic resonance imaging and histological nature. What, however, we are able to do more consistently, more effectively, and more precisely, is to identify the anatomical parameters of a lesion, thereby rendering surgery more precise. (A), an axial projection, reveals a high signal intensity in the cortico/subcortical strata of the left mid-temporal lobe. In (B), the post-gadolinium image, one notes perifocal enhancement of the mass which causes remodeling of the temporal cortex in the superior and mid-temporal gyri at the temporal operculum of the insula.



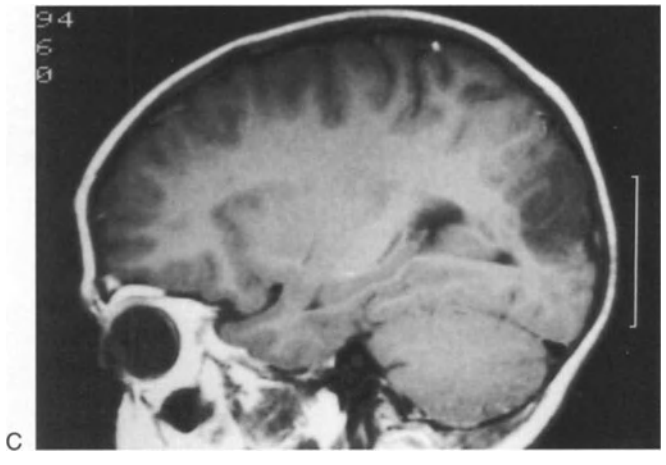


Figure 10.23. *Dysembryoplastic neuroepithelial tumor.* These tumors, which may be considered either neuronal and/or mixed neuroglial in histological characteristics, are located in the cortical surface, composed of a series of nodules of astroglial elements with focal dysplasia, and have very specific glioneuronal microstructural formations. They are generally located on the cerebral convexity, produce drug-resistant seizures, and may be identified on the basis of T1 weighted imaging hypointensity and T2 weighted imaging hyperintensity. Resecting the tumor cures the child's epilepsy. In (A), a T2 weighted image, one sees a homogeneous high signal intensity lesion involving the cortical and subcortical left parieto-occipital area. (B), (C) and (D), axial, sagittal, and coronal sections of this gadolinium enhanced study, respectively, reveal the nonenhancing nature of the tumor and its specific location.

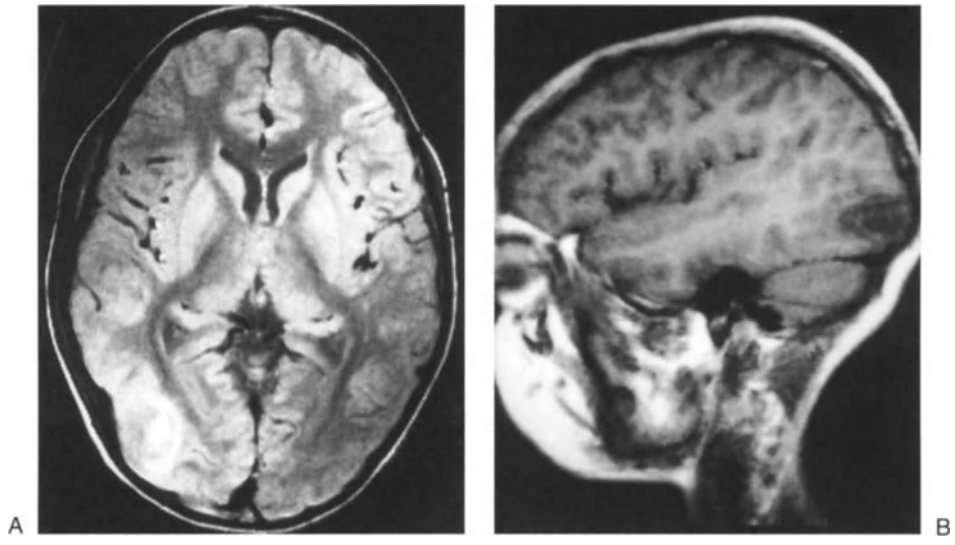
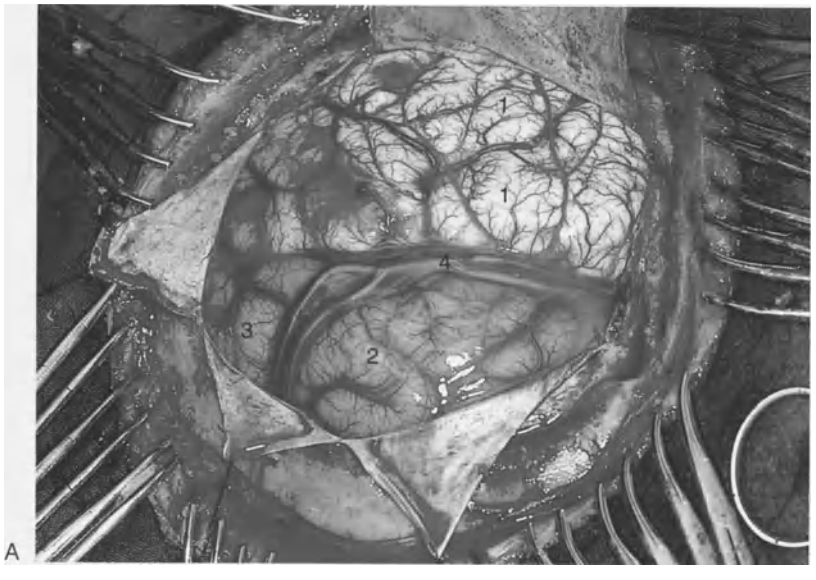


Figure 10.24. This is another child with a *dysembryoplastic neuroepithelial tumor* which is located in the right occipital lobe. The axial proton density image (A) reveals an incompletely homogeneous, hyperintense lesion in the cortical-subcortical strata of the right occipital lobe. The sagittal section (B) of a post-gadolinium injection study reveals the nonenhancing hypointense magnetic resonance characteristics of this very typical tumor.



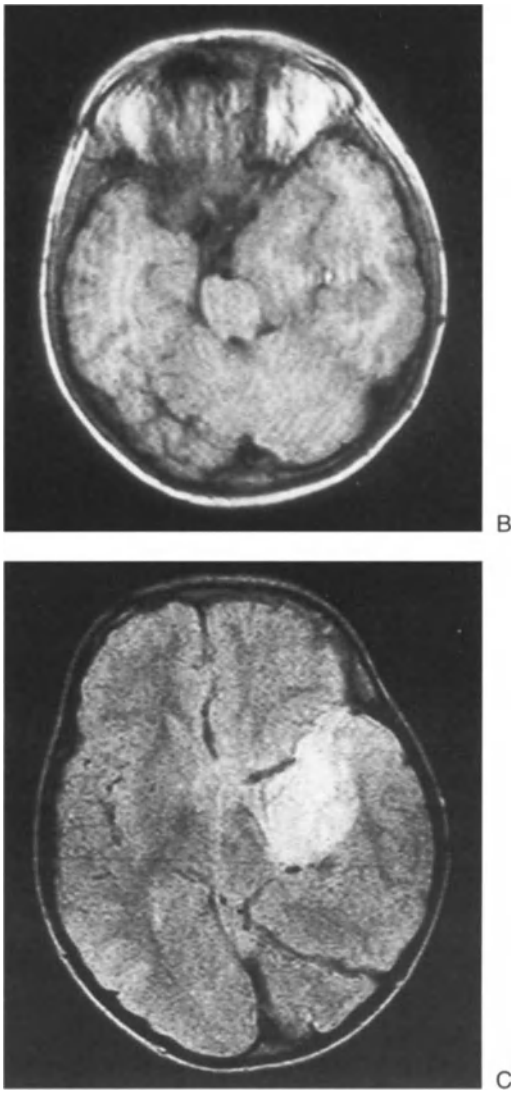


Figure 10.25. (A) This is an interesting and unusual case: hamartoma (*1*) of the temporal lobe. In this, the surgeon's view of the hamartoma of the left temporal lobe, one notes the frontal operculum (*2*), the parietal operculum (*3*), and the exit of the vein of Trolard (*4*) from the insular region. A complete temporal lobectomy was perfomed. (B) On magnetic resonance, axial section, one sees the remarkable hypertrophy of the hamartomatous temporal lobe; and, in (C), that in the post-enhancement study the uncus and hippocampal formation light up much more markedly than the rest of the hamartomatous temporal lobe (C). The temporal lobe epilepsy was not cured by lobectomy.

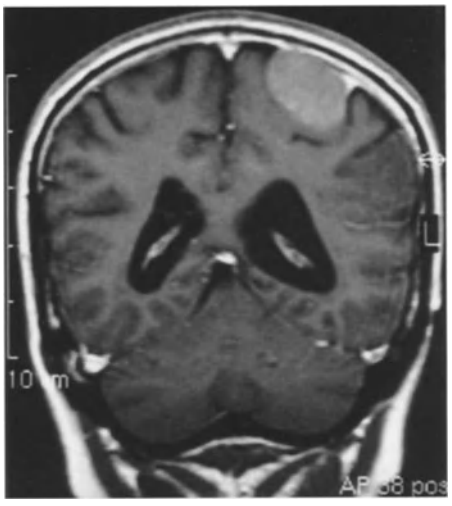


Figure 10.26. It is necessary to recognize objectively the value of radiation therapy, and to see this treatment modality in its evolution as dispassionately as we do with the surgical treatment of brain tumors. Of course, as with the surgical treatment of brain tumors, radiation therapy has its complicatioins. Here is one: an avascular meningioma.

Cystic Hemispherical Tumors (Fig. 10.27)

Cystic tumors of the hemispheres may be approached along the convexities, since vasculature is not a problem and one need not concern oneself with isolating the main trunks of the carotid or terminal basilar systems. These tumors also assume enormous proportions but, different from the solid tumor, more often cause remarkable transfalcical herniation. Cerebral decompensation, when it occurs, is precipitous. Cystic tumors have identifiable limits, displace other structures rather than invade them, and their resection is followed by clinical improvement (the compressive damage of surrounding structures is alleviated).

Apart from adequate exposure of the cortical surface for removal of the cystic neoplasm, three primary characteristics of these tumors warrant consideration: the capsule, the cyst fluid, and the nodule.

Capsule (Figs. 10.28, 10.29)

After the cerebrotomy has been performed and the surgeon comes upon the capsule, dissection is followed along its surface but only enough to provide adequate area to enter the cavity and drain the fluid.

En bloc resection is discouraged. Most glioma, and all congenital (dermoid, epidermoid), capsules are thick enough to permit surface exposure if one is gentle. Fluffy cottons, which have been kept moist, facilitate exposure of the dome of the capsule, then its separation from the surrounding cerebral parenchyma, allowing the surgeon to open his working space between capsule and gliotic tissue.

Once the capsule has been exposed, one may use first a Penfield #1 dissector, then a #2, resting the tips upon and parallel to the capsule, to separate gliotic tissue from capsule. Slight compression upon the cystic mass by the tip of the forceps minimizes entrance into parenchyma. One should not attempt to isolate the entire cyst in this manner until after the contents have been drained, since it unavoidably compresses cyst against brain (liquid is noncompressible).

After the dome of the cyst wall has been delivered into the field, Telfa is placed over the surrounding hemisphere in a circumferential manner, laying the individual strips over the capsule by brushing them along the cyst, thus isolating the tumor. The Telfa should not be forced along the brain surface because this results in opening a plane within the substance of the white matter, not in separating cleanly the cyst capsule from the white matter, The spatulae are brought along the dome of the cyst between it and the Telfa strips. As the spatulae are advanced, new Telfa strips may be positioned by sliding them over the spatulae, then withdrawing the spatulae, once more repositioning the spatulae beween Telfa and tumor, and so on.

Cystic Fluid

The cystic fluid is never crystal clear. It ranges in color from yellow through amber to chocolate brown. If crystal clear fluid is found, the diagnosis is ependymal or arachnoid cyst. Cysts associated with developmental tumors (teratoma) may contain emerald green fluid, or material which looks like liquefied fat. Dermoid and epidermoid cysts may contain ectodermal derivatives as well as, or in place of, fluid. The liquid content, except in cysts associated with dermoid sinus tracts, is sterile. It may, however, produce a chemical meningitis or irritate the cortex. Consequently, care should be taken in removing it, preferably performing this by inserting a needle into the cyst cavity and aspirating some fluid before entering the cavity. After the cyst cavity has been drained of fluid, the puncture site is extended into an incision large enough to permit complete visualization of the cavity wall and nodule. Telfa should be placed between gliotic tissue and the cyst wall before entering the cavity. Once the cavity has been emptied, one notes relaxation of the surrounding brain and collapse of parenchyma into the void left by the evacuation of cyst contents. At this time, bridging cortical veins may be stretched or torn, so one should support, with self-retaining retractors, the surrounding brain, holding the cyst cavity open for the completion of the surgery and diminishing the risks of tearing vascular structures. Telfa should be placed between the retractor and the brain.

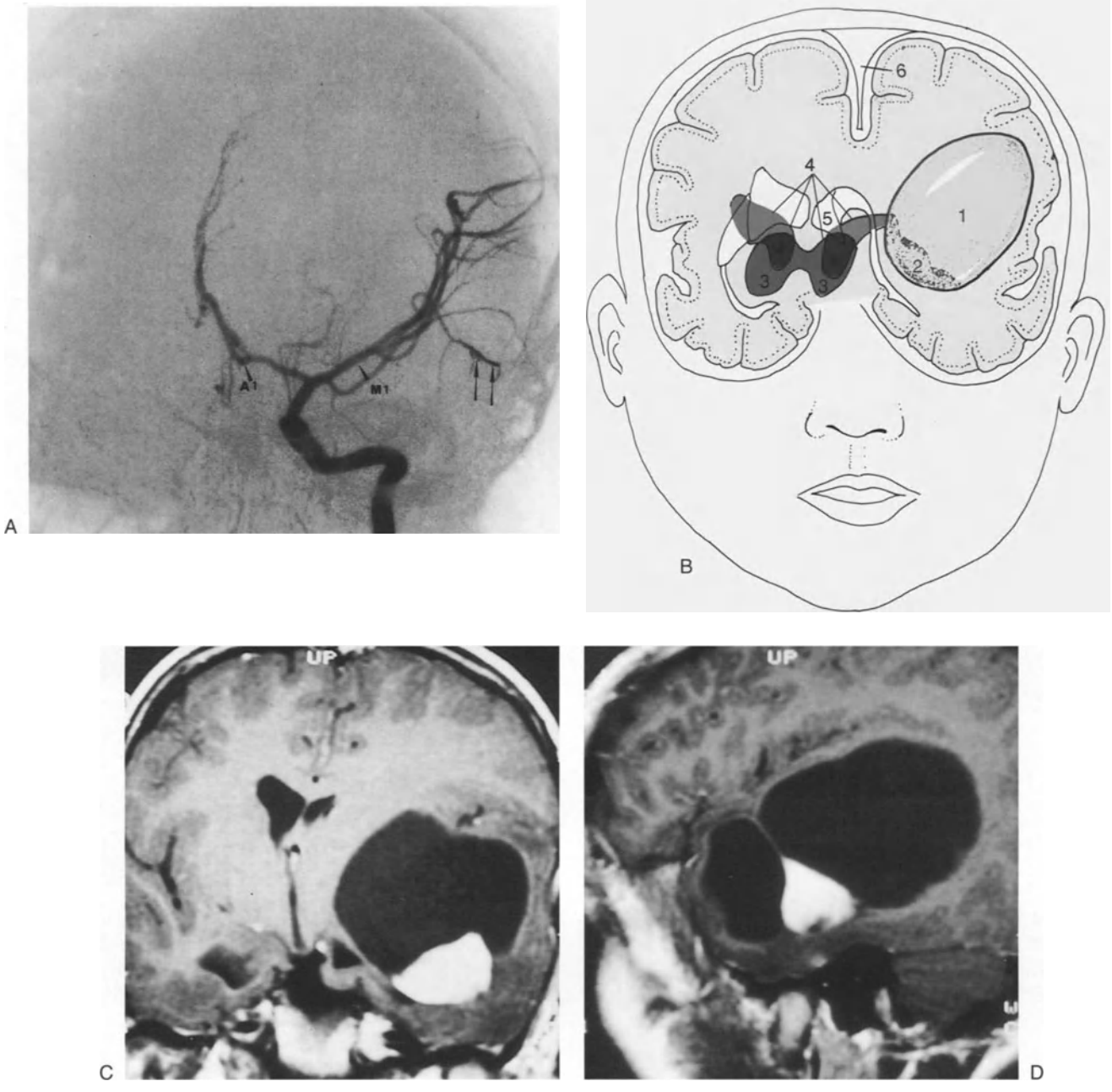


Figure 10.27. (A) This cystic astrocytoma is located in the frontal lobe, anterior to the central (Rolandic) sulcus and superior to the Sylvian sulcus. The A-1 portion of the anterior cerebral artery is displaced posteriorly, as is the M-1 portion of the middle cerebral artery. The tumor nodule is displacing Sylvian branches (*arrows*). There is remarkable transfalcical herniation of the A-2 portion of the anterior cerebral artery. (B) The centrencephalic location of a cerebral hemisphere cystic tumor (*1*) and, in this particular child, the location of the tumor nodule (*2*). The drawing puts into relief the inferomedial displacement of the thalamus (*3*), the medial displacement of the head and body of the caudate nucleus (*4*), and the remarkable transfalcical herniation expressed by displacement of the lateral ventricles (*5*) to the contralateral side, beneath the falx cerebri (*6*). (C) This coronal, post-enhancement, MRI study reveals the enormity of the cyst, enhancement of the nodule, and apparent isodensity of cyst fluid and cerebrospinal fluid. (D) The parasagittal section through the temporal lobe puts into relief septum formation, a not uncommon observation in supra- and infratentorial cystic gliomas.

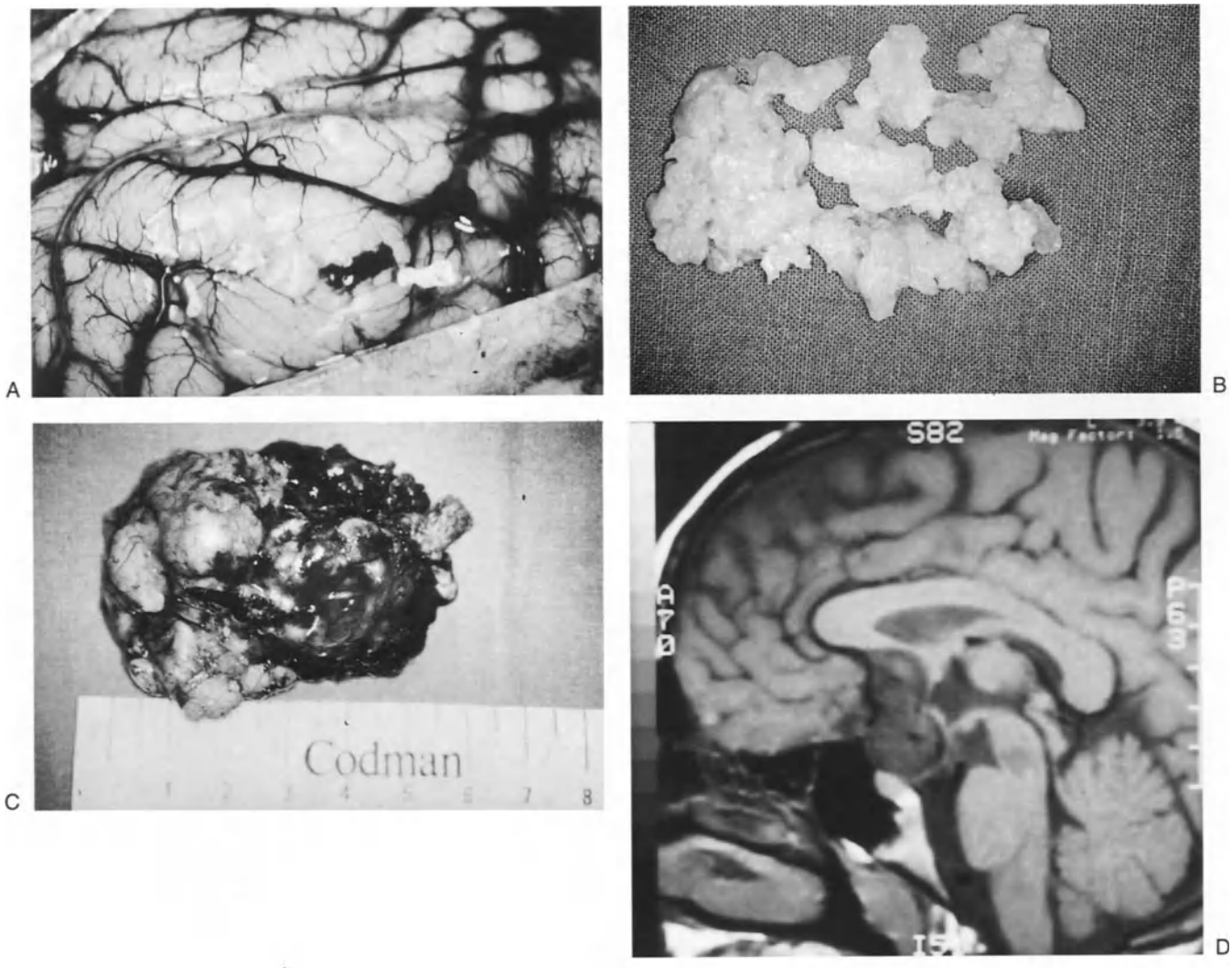


Figure 10.28. Surgery, tumor, and cystic fluid from three children with intraparenchymal epidermoid tumors. One notes the surface of the brain (A), and (pearly appearing) epidermoid tumor after removal (B). After the cystic fluid was removed, the tumor bulk was diminished considerably so that removal of the pearly tissue became quite simple. (C) The solid component of another epidermoid, from a third child. (D) This sagittal MRI study reveals the epidermoid tissue, as illustrated in (B), in a patient with a parasellar epidermoid.

Nodules

Nodular masses within cystic tumors are not common. When present, they may be avascular (the majority) or vascular. The nodules are best removed by coagulation ultrasonic aspiration, or, preferably, vaporization with laser, whether they are vascular or avascular.

Either technique destroys the nodule without subjecting the surrounding brain to stretching damage, which is unavoidable when the nodule is pulled away with a pituitary forceps or a sucker. If laser or ultrasonic aspirator is not available, Takahashi (biopsy) forceps may be inserted into the cavity to grasp the mural nodule. The nodule is then withdrawn. As this occurs one notes that the walls of the capsule strip away with the nodule, though portions become frayed. One may use a Penfield #1 dissector to separate the fraying portions of the cyst capsule from the surrounding tissue, though this is seldom necessary if the surgeon has already dissected the cyst capsule from the surrounding white matter with Telfa and spatula.

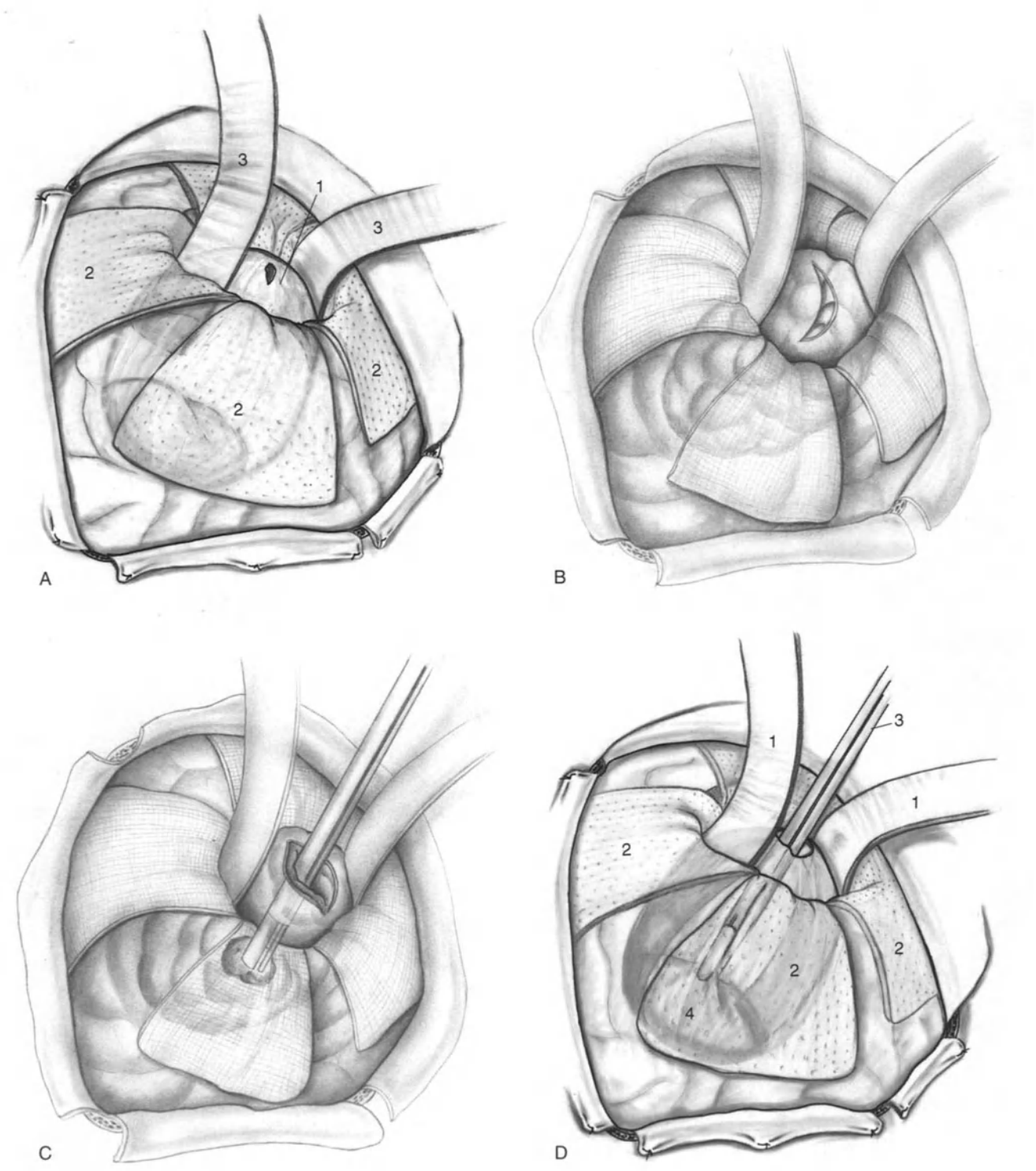


Figure 10.29. (A) The cerebrotomy has been performed and the surface of the tumor exposed, so that its dome (*1*) delivers itself through the opening. Telfa strips (*2*) are laid over the capsule, separating it from the surrounding brain. Spatulae (3) may be insinuated along the tumor surface after the cyst has been opened and the fluid drained. (B) The incision in the tumor capsule is extended. (C) The tumor foreps are inserted into the cystic cavity and the nodule grasped. (D) The spatulae (*1*) have been positioned over the tumor capsule, with the Telfa (*2*) protecting the cerebral surface. Micropituitary forceps (*3*) are used to remove the tumor nodule (*4*), taking care to work within the cyst cavity. (E, F) see p. 216.

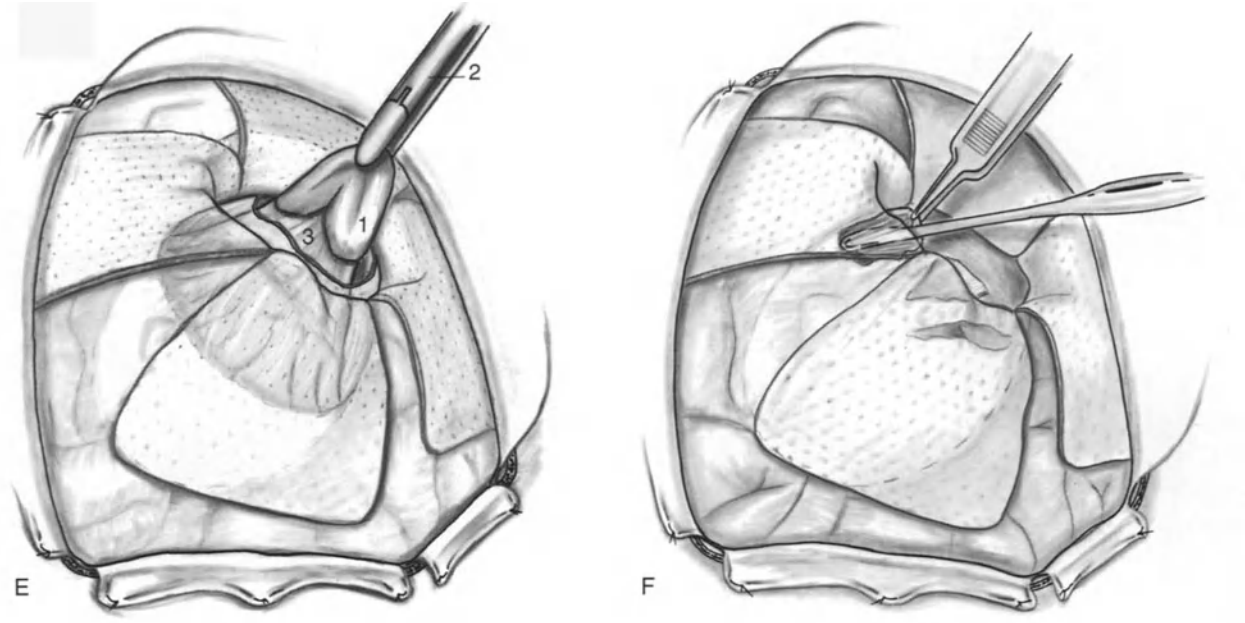


Figure 10.29. (E) Gentle retraction of the tumor nodule (*1*) by the forceps (*2*) generally strips much of the capsule (*3*) from the surrounding brain. This "classical"technique is acceptable, but the use of the Cavitron to aspirate the tumor nodule and capsule is much preferable! (F) A Penfield #4 dissector may be used to separate the frayed portions of the cyst capsule from the surrounding tissue.

Ventricular Tumors (Figs. 10.30, 10.31)

Lateral ventricle tumors, in the... true... sense of the word are only the choroid plexus papilloma, the extremely rare meningioma, and the ependymoma. However, subependymal gliomas, the tubers of Bourneville's disease (giant cell subependymal astrocytoma), and those gliomas which fungate into these two CSF chambers, practically speaking, are to be considered intraventricular tumors.

Apart from the choroid plexus papilloma – and the rare meningioma – intraventricular tumors are either *ependymal* or *glial* in origin, with the ependymal tumors taking their origin from the ependymal lining of the ventricles and the glial tumors arising from the subependymal astrocytes. Tubers are enormous masses of giant astrocytes which may collect into tubercle-appearing structures which line the ventricular surface, or giant tumors extending from the central white matter into the ventricle. Though the ependymomas may occur in either the lateral or IV ventricles, the subependymal gliomas are found only in the lateral and III ventricles. The choroid plexus papilloma occurs more frequently in the lateral ventricles, less frequently in the III ventricle, and is extremely rare (in childhood) in the IV ventricle. This latter observation is quite the opposite of what one notes in the adult, where the choroid plexus papilloma is most common in the IV ventricle and least common in the lateral ventricles. Intraventricular meningiomas have been reported in children, but are exceptional occurrences, even when one considers the increased incidence of meningioma in children with von Recklinghausen's disease.

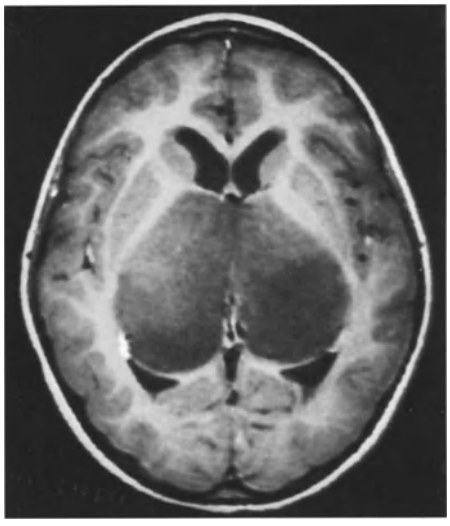


Figure 10.30. This is a bithalamic glioma, low signal intensity and nonenhancing, which collapsed the III ventricle and caused biventricular hydrocephalus.

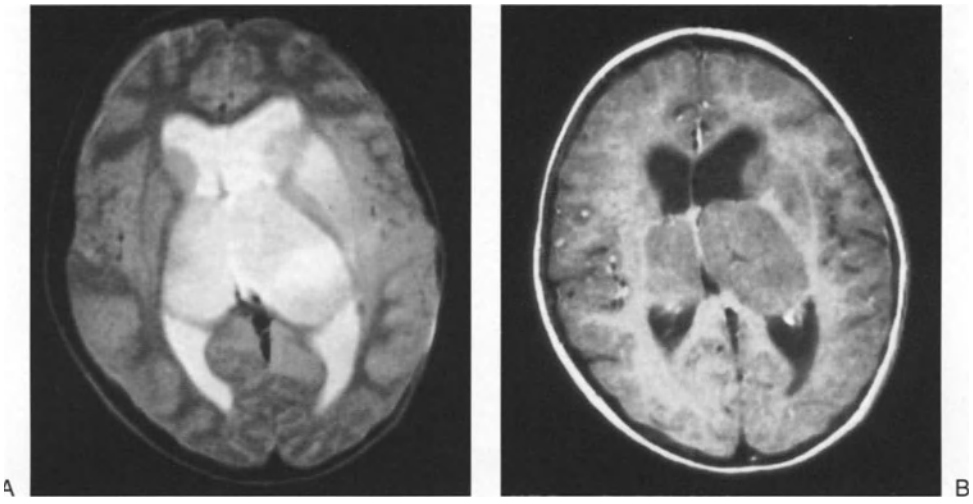


Figure 10.31. A bithalamic glioma involving also the basal ganglia. T1 (A) and T2 (B) images.

The primary surgical considerations in approaching intraventricular tumors have to do with the presence and type of hydrocephalus (mono-, bi-, triventricular), the extent and vascularization of the pedicle, the entrapment of either a ventricular horn or an entire ventricle, extension of the tumor into another ventricle, and neoplastic invasion of such vital structures as the hypothalamic or pontomedullary nuclei. Extension of the tumor into the basal ganglia is rare, whereas thalamic tumors often fungate into a ventricle.

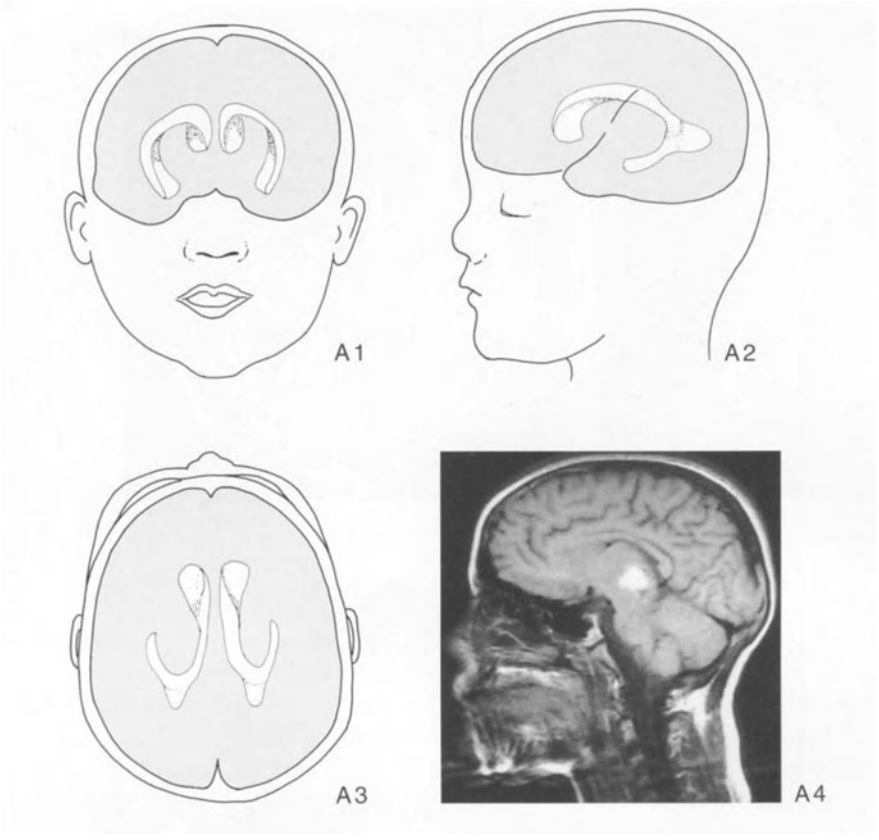
As the intraventricular tumor expands, the ventricles dilate, either from complicating hydrocephalus or tumor expansion in and of itself. Ependymomas, for example, may expand within the lateral ventricular system, taking on the form of the ventricles (plastic ependymomas) as they grow, gradually replacing the cerebrospinal fluid. The glioma, teratoma (typical or atypical), and papilloma, on the other hand, tend to expand circumferentially, displacing neural structures and obstructing cerebrospinal fluid flow. These characteristics influence the surgical approach and operative technique for removal, since a lesion which expands to fill uniformly and snugly a single lateral ventricle is quite different from one which expands into the III and/or the opposite lateral ventricle.

Similarly, a plastic ependymoma entails a different surgical and microsurgical technique for total removal than a papilloma expanding at the trigone and obstructing the temporal and occipital horns. Teratomas of the pineal region, extending into the III and lateral (at the trigones) ventricles, necessitate a much more extensive and versatile craniotomy than a pineal tumor occupying only the III ventricle, or simply extending along the quadrigeminal plate and pushing into the posterior fossa. Consequently, preoperative evaluation of intraventricular tumors is demanding: it obliges the surgeon to conceptualize the anatomical confines of the ventricular system and the extent to which the tumor either expands to occupy them or breaches their walls and invades the cerebral parenchyma.

Lateral Ventricle Tumors: General
(Figs. 10.32–10.40)

Though complete resection of these tumors, cystic or solid, benign or malignant, is enticing, the reality is that they tend to have occupied the entire ventricle (plastic types) by the time the diagnosis is made. For the malignant, partially resected, or cystic extraventricular ependymoma, radiosurgery is indicated, but not prophylactic spinal irradiation.

Conceptualization of lateral ventricular tumors is facilitated greatly if one first envisions the craniofacial region in the three basic anatomical projections (coronal, sagittal, axial), and then the basal ganglia and thalami within the center of the intracranial compartment. The thalami border both the lateral surfaces of the III ventricle and the posteroinferomedial surfaces of the lateral ventricles. The caudate nuclei make up the anteromedial and posterolateral surfaces of the frontal horns and bodies of the lateral ventricles, and the entire medial surfaces of the temporal horns. This anatomical configuration is particularly fortuitous because it allows the surgeon access to the ventricular chambers, without the risk of entering the nuclei of the basal ganglia and thalamus. Thus, one may, in approaching the superior surface of the lateral ventricle, penetrate the ventricular wall through either the frontal, the anterior temporal, or the occipital horns without any risk of causing neurological deficit.



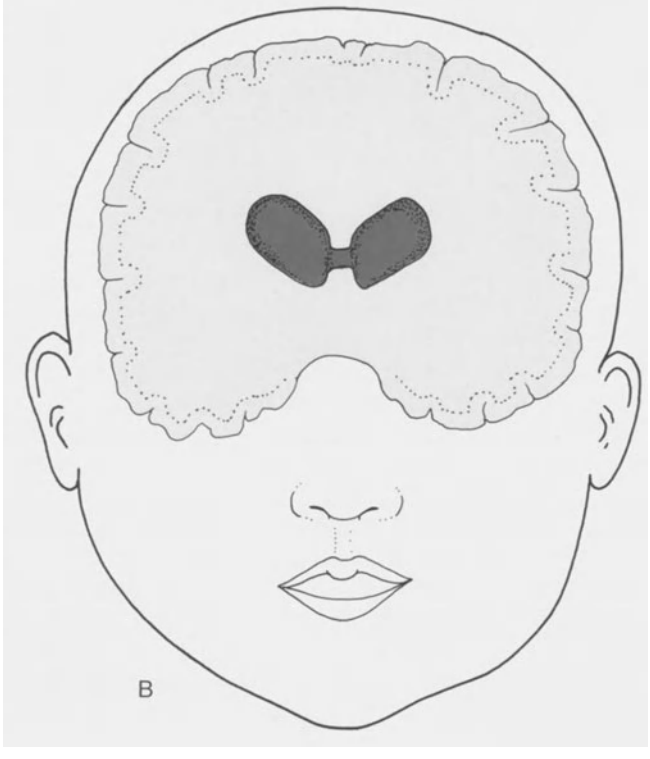


Figure 10.32. In evaluating the location, extent and complicating ventricular dilation in a child with an intraventricular tumor, it is best to conceptualize the craniocerebrum in anterior-posterior, axial, and lateral projections (A1,2,3,). The lateral ventricles are drawn in three basic projections, allowing the reader the opportunity to study the general size and spatial orientation of each ventricle in each of these projections. (A4) This postenhancement, parasagittal section, MRI study reveals the enhancing nodule anterior and superior to the pulvinar, and diffuse tumefaction of the body of the thalamus involved by a glioma. It puts into relief alterations in the body of the lateral ventricle. (B) The thalami have been drawn in, to allow appreciation of their location at the most inferomedial portions of the cerebral hemisphere. (C) The head, body, and tail of the caudate nucleus have been drawn on the thalami, permitting one to appreciate the anatomical and spatial relationships between these two structures. (D) The lateral and III ventricles, thalami, and caudate nuclei permit one to appreciate the details of how the tail of the caudate nucleus (*1*) extends into the temporal horn (*2*), but the head of the caudate nucleus (*3*) is located along the medial surface of portion 2 of the lateral ventricle, and its body (*4*) courses over the top of the thalamus. (C, D) see p. 219.

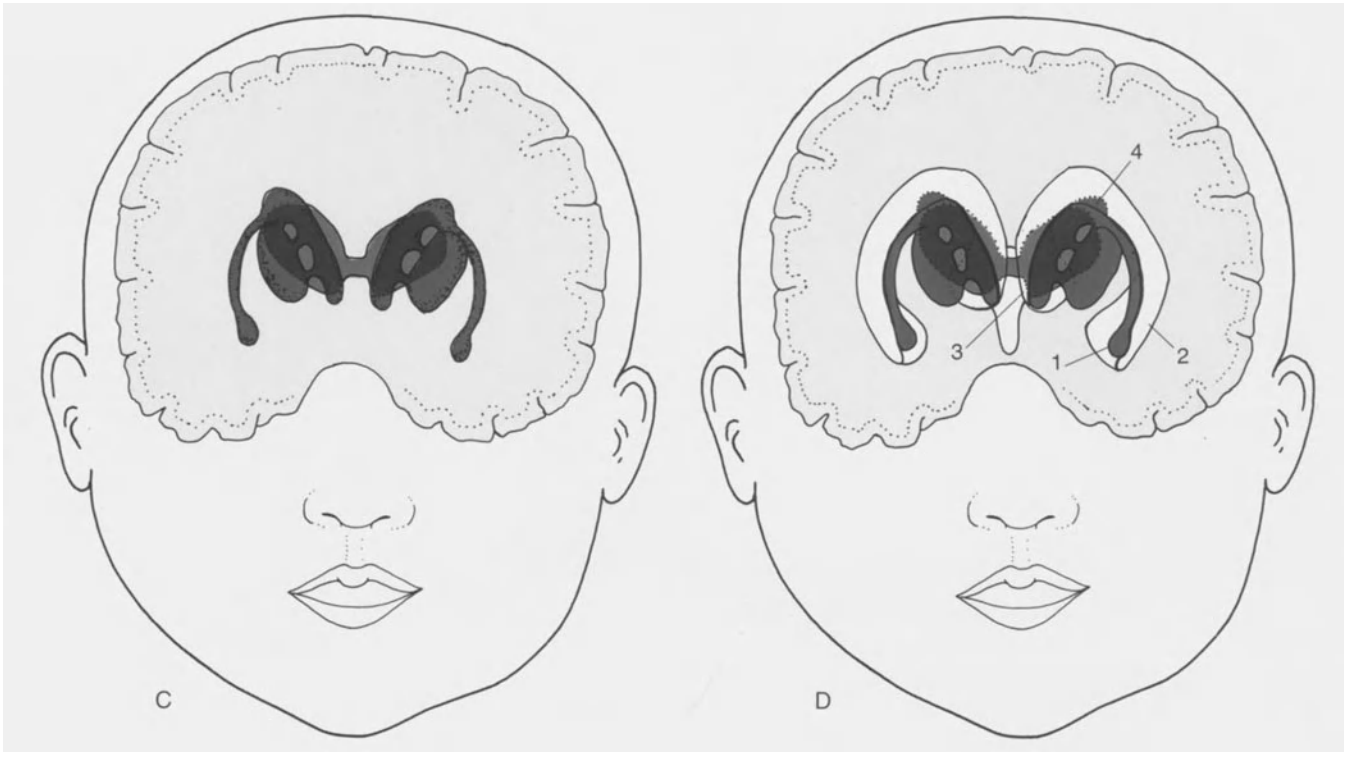


Figure 10.32. Legend see p. 218.

Entrance into the body of the lateral ventricle may be gained anywhere along the corpus callosum, from the genu to the splenium, simply by dissecting laterally from the midline (along the indusium grisium and within the pericallosal sulcus). This latter route is ideal if one wishes to expose the entire lateral ventricle, or any portion of the lateral ventricle, along with the III ventricle. It must be remembered, however, that we still do not have definitive evidence concerning neurologic deficit or right-left disorientation, which result from sectioning the corpus callosum. It is, consequently, advisable to breach only a limited portion of this commissure, never to split all of it from the genu to the splenium.

When a lateral ventricle tumor obstructs the foramen of Monro, there ensues remarkable dilation of the homolateral ventricle, with shift to the opposite side, resulting from the combined pressure vectors of the tumor and the asymmetrical hydrocephalus. Though this combination of forces aggravates drastically, and acutely, the clinical situation, it provides the surgeon with an enlarged ventricular chamber through which he may work in resecting the tumor.

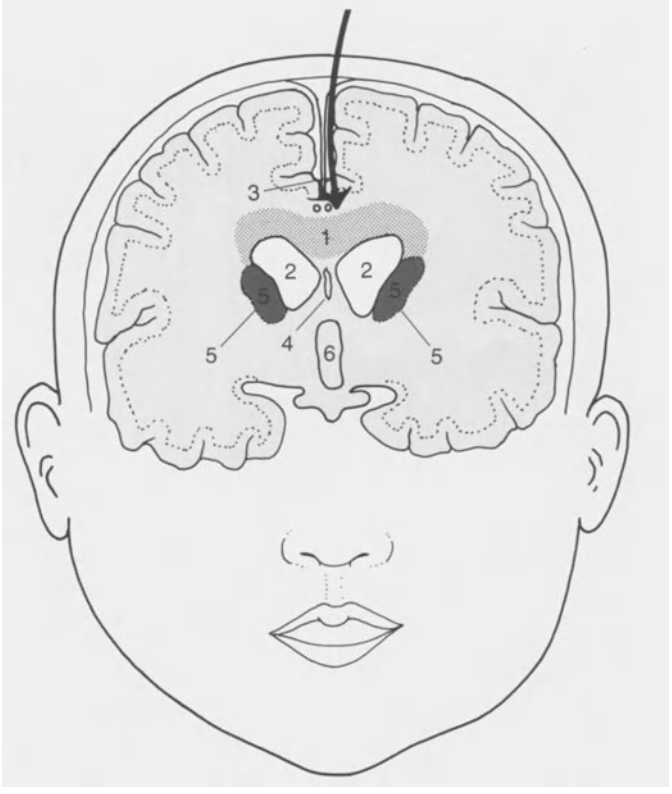


Figure 10.33. How dissection along the parasagittal surface of the corpus callosum (*1*) allows one to enter the body, portion 3, of the lateral ventricle (*2*). For orientation purposes, the falx cerebri (*3*), septum pellucidum (*4*), head of the caudate nucleus (*5*), and III ventricle (*6*) are also labeled. The slightly curved arrow indicates the parasagittal approach to the corpus callosum and ventricular system.

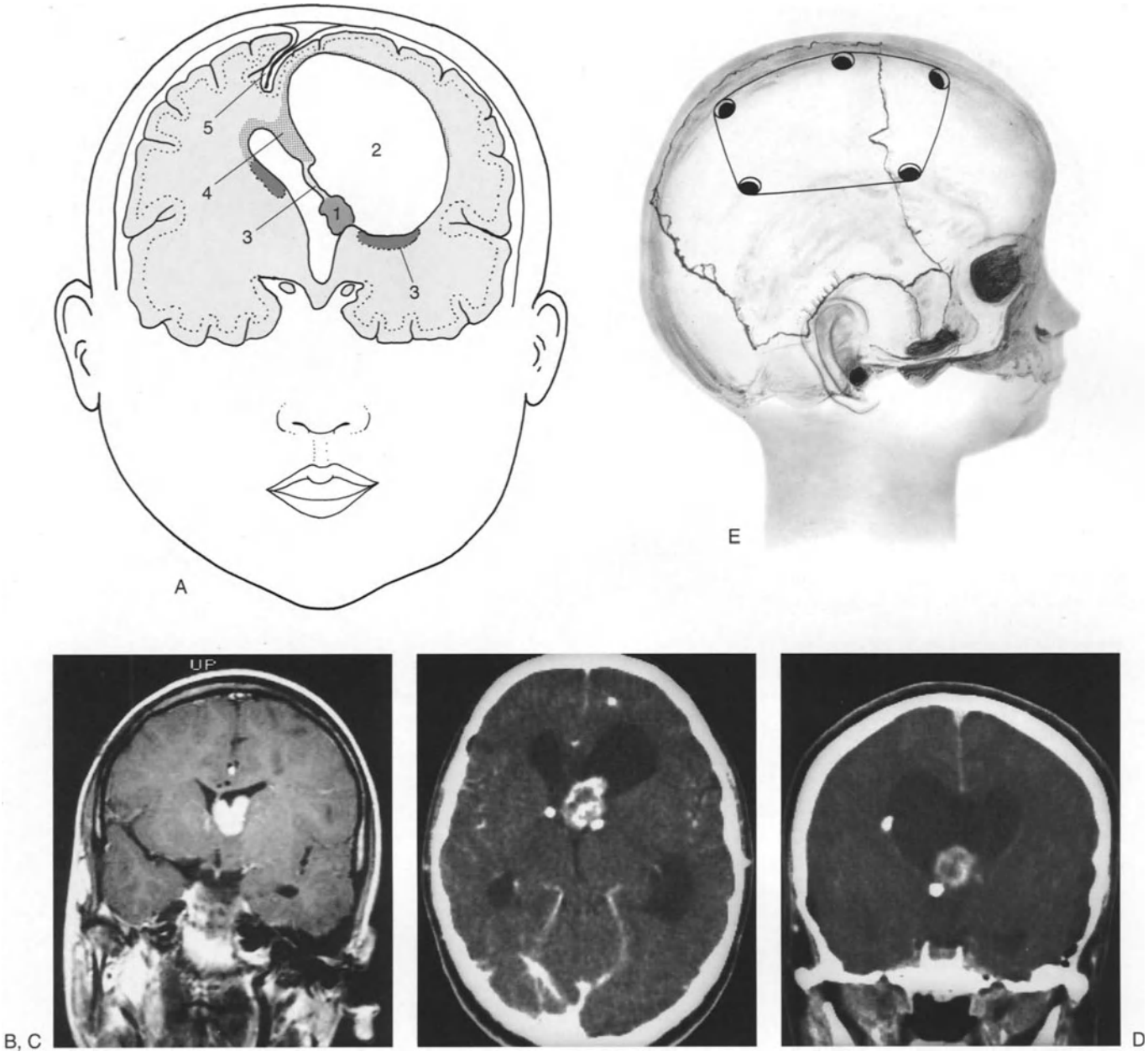


Figure 10.34. Tuberous sclerosis is one of the phacomatoses (a dysgenetic syndrome which most commonly causes obstructive asymmetric hydrocephalus resulting from tuberous nodules at one or both foramina of Monro) is a familial disease, autosomal dominant, of variable inheritance penetrance, tuberous sclerosis (*Bourneville's disease*), with the gene located on the long arm of chromosome 9q of the tumor suppressor gene 16 T. Histologically, this tumor is a *giant cell subependymal astrocytoma*, and it is very often associated with rather typical cerebral convolution, cytoarchitectually deranged, groupings of bizarre neurons and giant astrocytes known as "tubers." These same "tubers" are found along the ventricular surface, the subependymal area, occasionally within the centrum semiovale: observations which permit one to deduce that abnormal migration is the effect of the gene abnormalities. In (A) the obstructing lesion (*1*), at the foramen of Monro, dams the cerebrospinal fluid in the lateral ventricle, causing it to dilate (*2*) and resulting in a shift of the septum pellucidum (*3*) and corpus callosum (*4*) to the opposite side. In children, the interhemispherical fissure (*5*) is often shifted because the falx tends to be hypoplastic. (B) A coronal MRI post-gadolinium study shows homogeneous enhancement of a mass at the left foramen of Monro: a typical aspect of giant cell subependymal astrocytomas. (C), (D), coronal and sagittal sections, respectively, of a CT study, reveal a giant cell subependymal astrocytoma at the foramina of Monro expanding from the left thalamic surface, asymmetrical hydrocephalus similar to that represented diagramatically in (A), and numerous tubers along the subcortical areas of the frontal and insular gyri (B) and involving the ependyma (C) and (D). (*Continued* on p. 221).

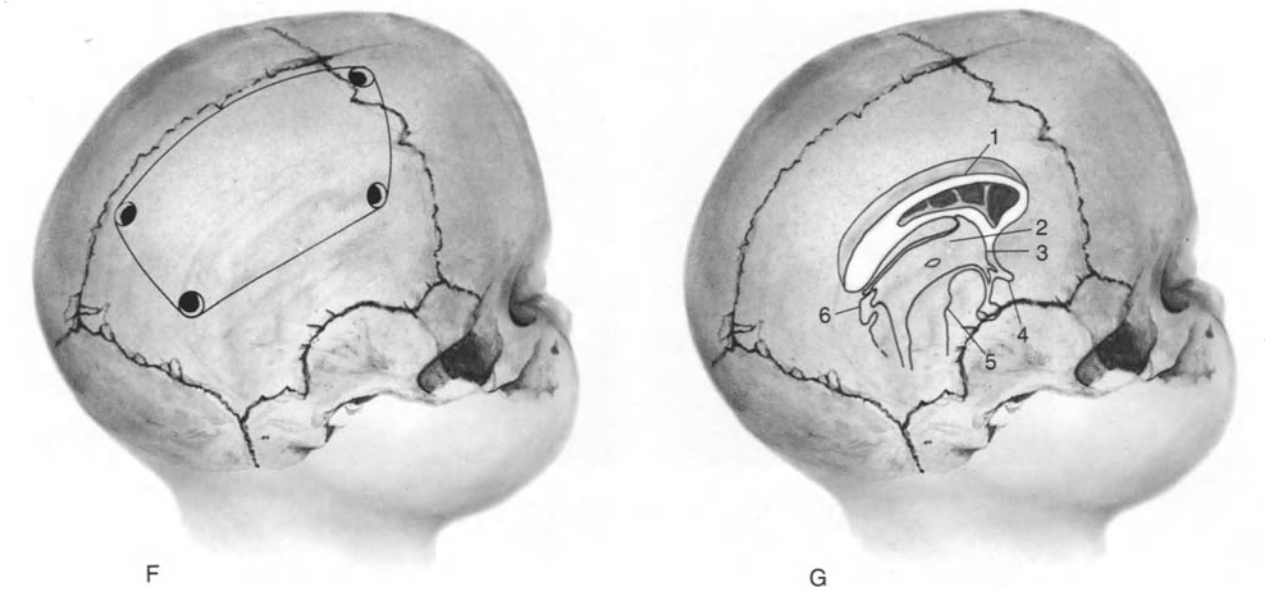


Figure 10.34 (*continued*). The schematic drawings (same as Figs. 3.13A–3.15A) of frontoparietal and parietal bone flaps, and of the midline structures and ventricular system, permit the "visualization" of transfrontal or parasagittal approaches to the III ventricle for resection of (E) a colloid cyst of the III ventricle, (F) a subependymal glioma occluding the aqueduct, or (G) an intraventricular (astrocytoma) tumor [corpus callosum (*1*), III ventricle (*2*), septum pellucidum (*3*), optic chiasm (*4*), midbrain (*5*), pineal gland (*6*)]. The choroid plexus papilloma of the III drains directly into the internal cerebral veins in the roof of the III ventricle, providing clear indications for a transcallosal approach

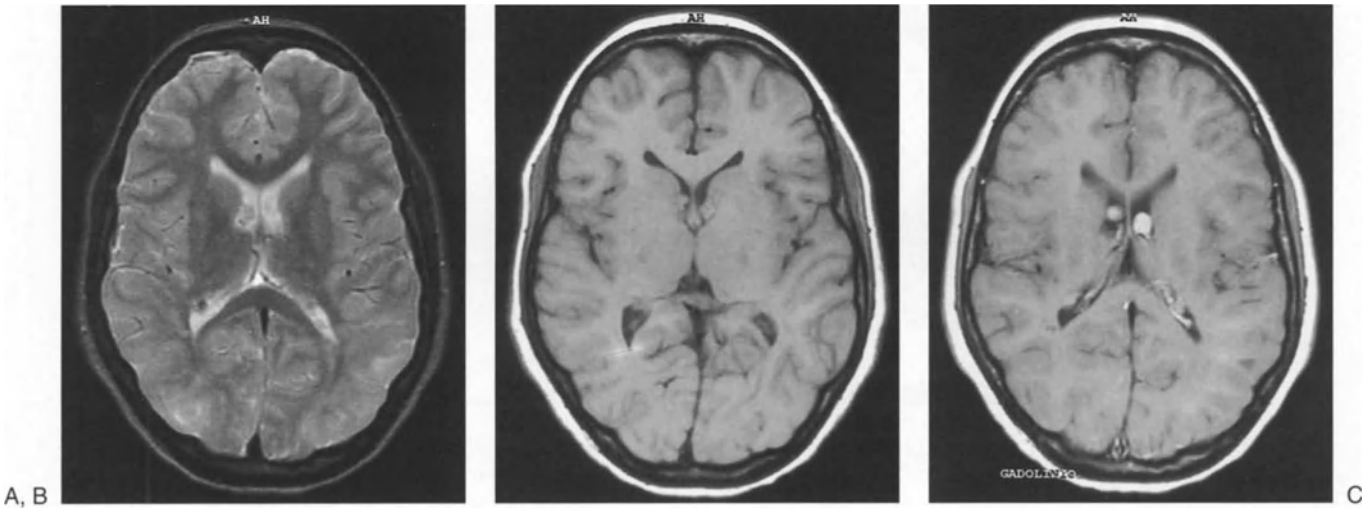


Figure 10.35. This is another child with Bourneville's disease, from whose MRI scan these axial sections are herein reproduced. A T2 weighted image (A) reveals multiple subependymal nodules with low signal. In (B), T1, the nodules are shown to be isosignal and in (C), the post-gadolinium study, to enhance markedly. If one now returns to the T2 weighted image reproduced in (A), the parieto-occipital cortical tubers may be identified bilaterally.

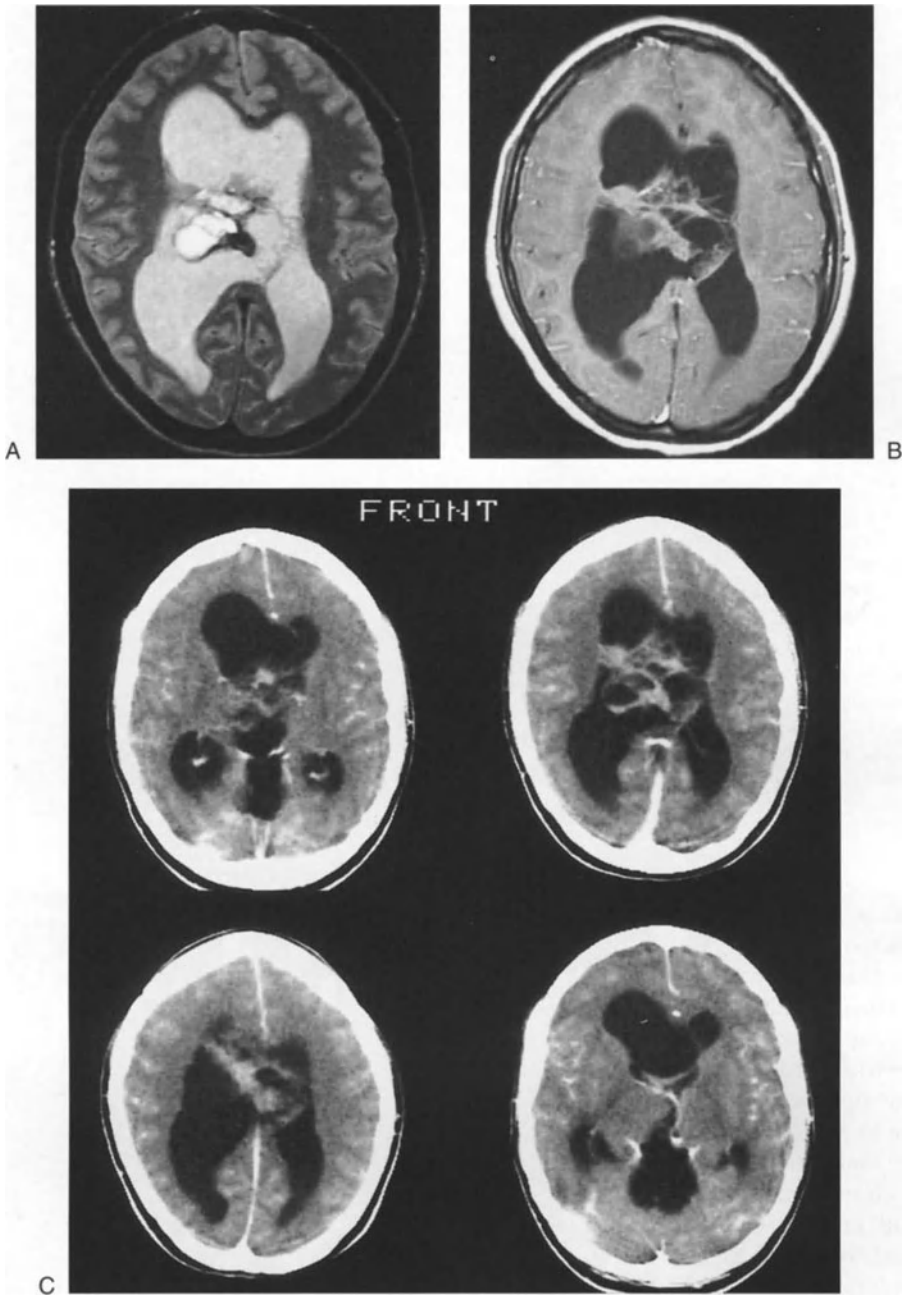


Figure 10.36. Earlier classifications of intra- and paraventricular tumors identify lesions we now call *central neurocytoma* as either *ependymomas* of the region of the foramina of Monro or *oligodendroglioma*, because of the fact that the cellular elements are derived from the subependymal plate groupings which developmentally transform into neurons, and from the nuclear gray matter of the septum pellucidum and the fornix. Hence, *central neurocytoma*. In (A) is such a tumor composed of high and low dishomogeneous intensity signals involving the region of the septum pellucidum and expanding irregularly into both ventricles, the left more than the right, causing asymmetrical hydrocephalus. In (B), the nonenhancing characteristics of the tumor are evident. (C) reveals the asymmetrical hydrocephalus, high-density, and dishomogeneous nature of the tumor.

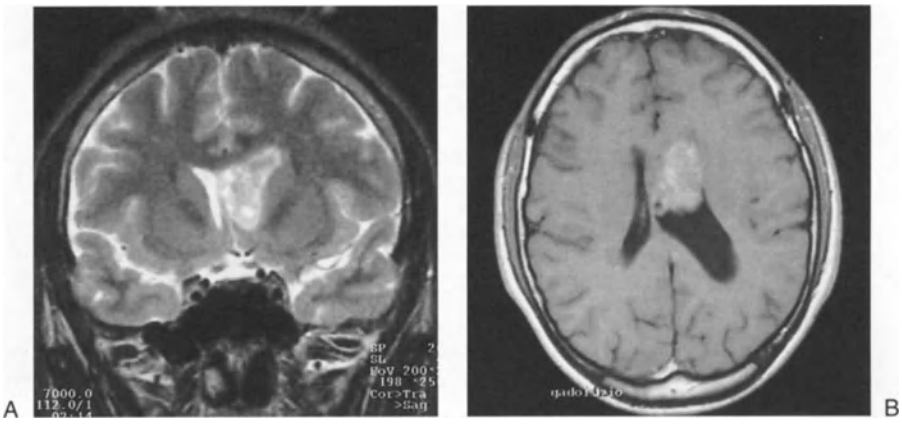


Figure 10.37. This is another neurocytoma which, in the (A) T2 sequence, reveals a large hypointense left ventricular mass with scattered high signal areas within it; and (B), the T1 sequence gadolinium study, which reveals the monoventricular hydrocephalus, minimal enhancement of the intraventricular mass. In the very early classifications of human brain tumors, the *spongioblastoma* was considered to be a highly malignant neoplasm. Subsequently, the tumor which had previously been classified as spongioblastoma was given a new name: *glioblastoma*. The term "spongioblastoma" was then used to describe tumors of the astrocytic series which had a piloid appearance, and later still astrocytic tumors whose cytological characteristics were such as to recall the primitive spongioblasts and whose biological behavior was malignant. Certainly, today no one would call a tumor with the histological characteristics of a glioblastoma, a spongioblastoma. Therefore, this histological terminology, spongioblastoma, still has value, especially in light of the fact that these tumors grow from the intra-III ventricular surface of the optic chiasm, the hypothalamus, the optic nerves, or at the lamina terminalis/septum pellucidum area. Perhaps, the history of the nomenclature of the spongioblastoma assists us to understand why some tumors growing, for example, from the optic nerve or optic chiasm or hypothalamus into the III ventricle are benign (optic pathway gliomas) and others are highly malignant (spongioblastoma).

It is intuitive that shunting from the side opposite the tumor (and dilated ventricle) would only aggravate the shift, and that shunting from the side in which the tumor is located could result in puncturing the tumor and, thereby, causing hematoma to develop within the very ventricle one wishes to decompress. For these reasons, direct attack upon intra-(lateral) ventricular tumors is preferable to attempting to shunt for the (complicating) hydrocephalus first. This is particularly true if the tumor is a choroid plexus papilloma, because of its vascularity and growth within the choroid plexus from the choroidal fissure in the temporal horn, past the trigone, to the foramen of Monro.

Lateral Ventricle Ependymal Tumors

Lateral ventricle ependymal tumors are soft, gelatinous in consistency, and highly vascular. Those portions of the tumor which extend into the brain substance change in consistency, but not in vascularity. One of the most important surgical considerations in this tumor is the matter of identifying precisely the area of attachment, since the ependymoma tends to have a rather sessile base from which it extends into the cerebral parenchyma, sulci, and cisterns. Therefore, one is confronted with a tumor which has all the characteristics of a glioma within the cerebral substance, and which lobulates with great redundancy upon itself within the intraventricular chamber.

Since this tumor may expand freely within the ventricle, it generally grows without causing focal neurologic deficit and presents clinically with an increase in intracranial pressure: the midline syndrome. The gradual increase in intracranial pressure is accepted well by the child until the *midline syndrome* [69] becomes evident. Consequently, the tumor may grow from one lateral ventricle to its foramen of Monro and into the III ventricle. On occasion, it may actually extend through to the contralateral ventricle, resulting in a tumor mass which assumes the form of the ventricular system: *plastic ependymoma*. No effort should be made to follow this tumor through the foramen of Monro and into the III ventricle, or the contralateral ventricle. The natural history of ependymoma is not affected favorably by radical or "total" resection!

As the ventricle is entered and the surface of the tumor exposed, one may extend the opening of the ventricular wall, from anterior to posterior, exposing the entire tumor. Telfa is then placed along both the lateral and medial ependymal surfaces of the floor of the lateral ventricle, taking care to bring its edges to the pedicle of the tumor prior to beginning the resection. No normal ependyma should be left unprotected.

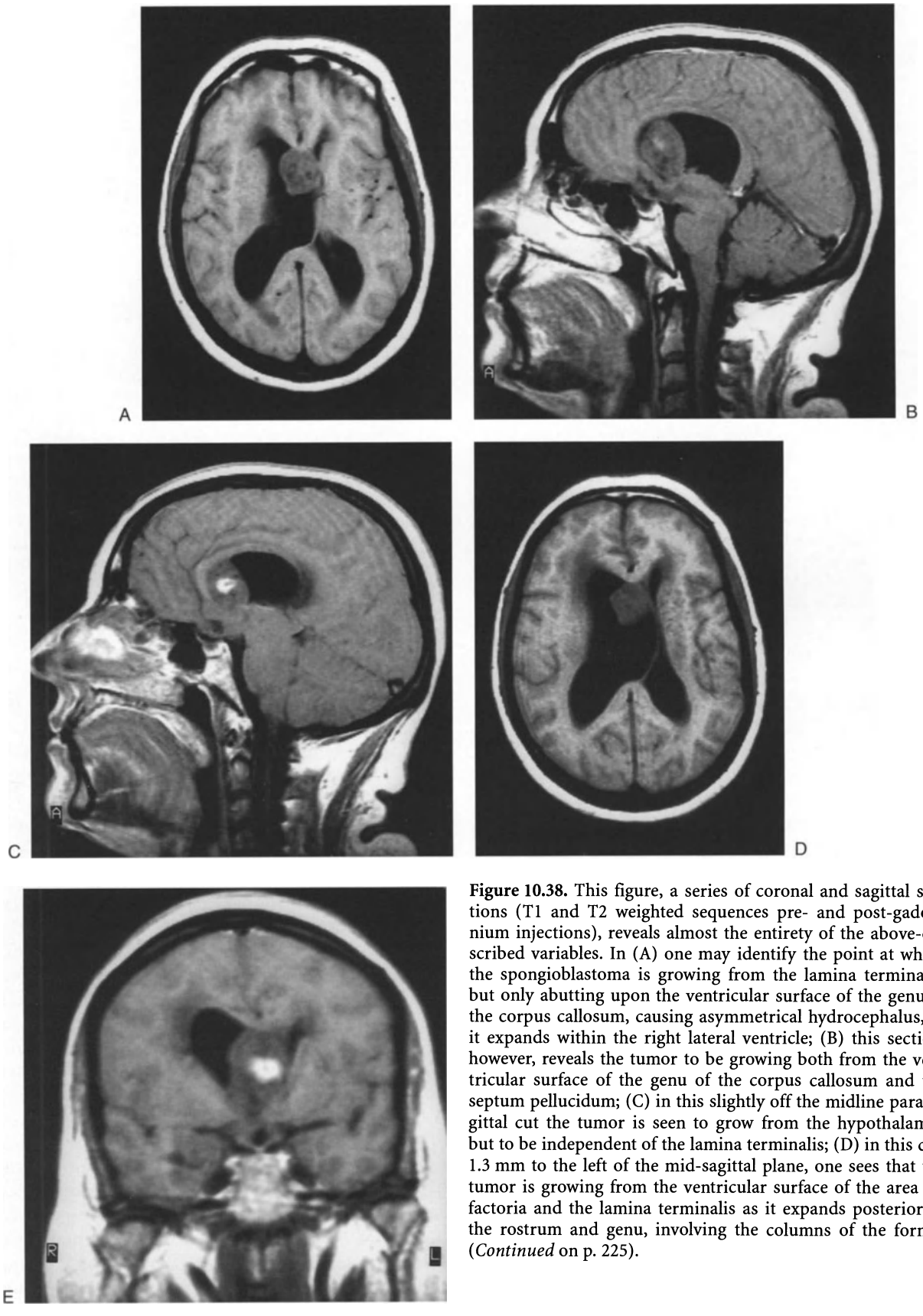
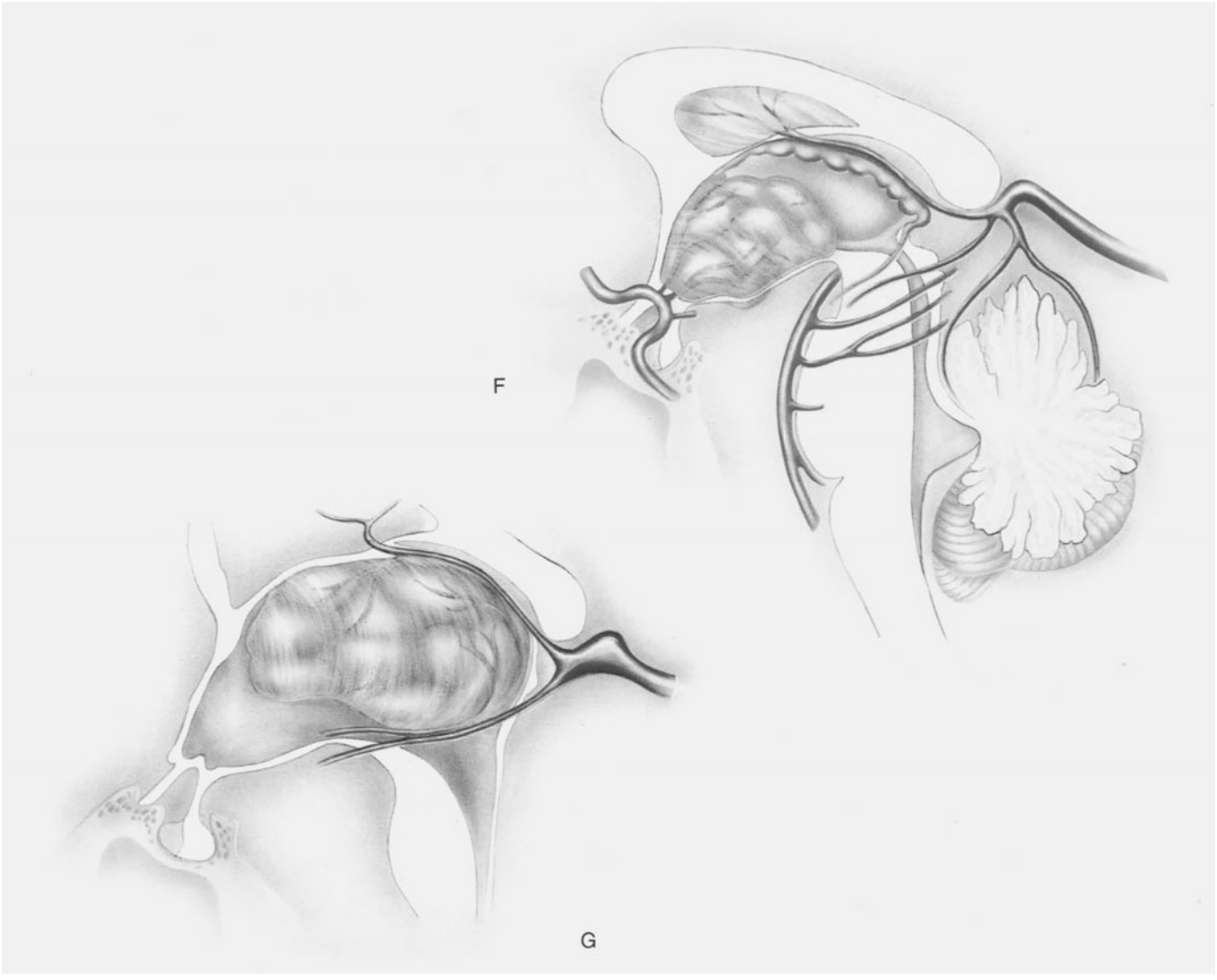


Figure 10.38. This figure, a series of coronal and sagittal sections (T1 and T2 weighted sequences pre- and post-gadolinium injections), reveals almost the entirety of the above-described variables. In (A) one may identify the point at which the spongioblastoma is growing from the lamina terminalis, but only abutting upon the ventricular surface of the genu of the corpus callosum, causing asymmetrical hydrocephalus, as it expands within the right lateral ventricle; (B) this section, however, reveals the tumor to be growing both from the ventricular surface of the genu of the corpus callosum and the septum pellucidum; (C) in this slightly off the midline parasagittal cut the tumor is seen to grow from the hypothalamus but to be independent of the lamina terminalis; (D) in this cut, 1.3 mm to the left of the mid-sagittal plane, one sees that the tumor is growing from the ventricular surface of the area olfactoria and the lamina terminalis as it expands posterior to the rostrum and genu, involving the columns of the fornix. (*Continued* on p. 225).



◀ **Figure 10.38.** (E) This coronal section reveals the tumor growth along the septum pellucidum, inferiorly, at the region of the mammilothalamic tract and the mammillary bodies, and superiorly at the inferior surface of the corpus callosum. In sum, tumors involving the septum pellucidum and lamina terminalis, by definition, may involve the columns of the fornix, the optic nerve, the optic chiasm, the intraventricular portion of the anterior perforated substance, the hypothalamus, the mammillary bodies, etc. In (F) such a tumor is schematically represented in the sagittal section, to allow the reader to identify the above possible contiguous relationships of such a neoplasm and the vascular supply from perforating branches of the anterior circle and the choroid plexus of the III ventricle. In (G), on the other hand, the III ventricular tumor is illustrated to extend inferiorly from the junction of the septum pellucidum and the bodies of the fornix, in a manner similar to that which occurs in III ventricle choroid plexus papillomas.

Preferably, the tumor resection is performed with the Cavitron. Before beginning, fluffy cottons are rolled from one edge of the neoplasm to the other, continuously wetting them, changing them, and sucking them dry so as to draw the vascular portion of the pedicle up into the fluffy. Attempts to identify individual bleeding vessels and coagulate them are futile. This results only in extending the area of dissection into the brain substance, generally the basal ganglia or thalami, and efforts to stop the bleeding are in vain. As the entire insertion of the pedicle is separated from the underlying brain and the fluffy cottons, gradually extended around the tumor's edges, along the pedicle, the mass is lifted somewhat. Fluffies and Telfa embrace the pedicle and protect the surrounding ependyma.

One begins to aspirate the tumor at its periphery. The aspiration should extend, in broad sweeps, over the entire surface of the tumor, taking care not to work in one limited area, producing gutters or craters. Bleeders should be coagulated immediately.

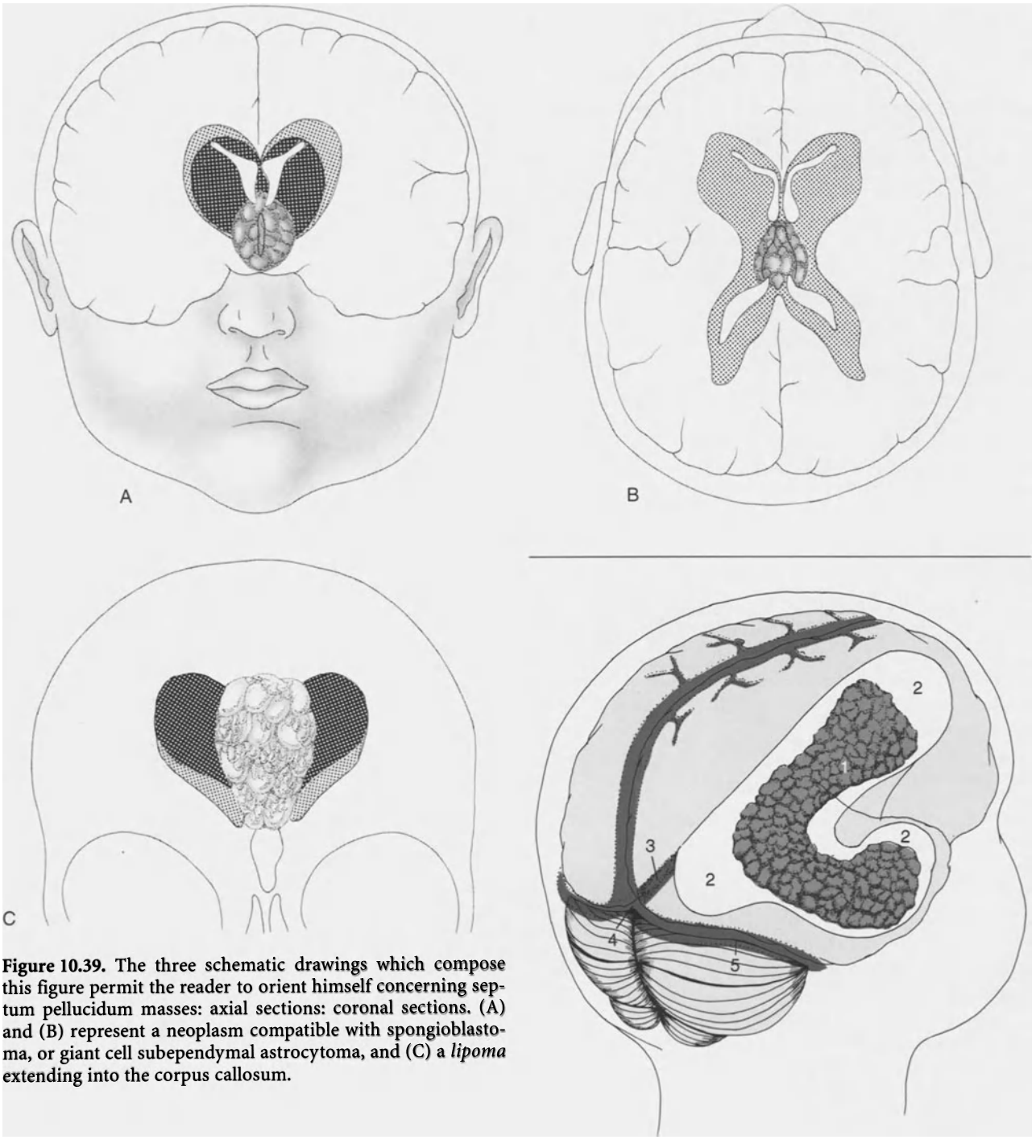


Figure 10.39. The three schematic drawings which compose this figure permit the reader to orient himself concerning septum pellucidum masses: axial sections: coronal sections. (A) and (B) represent a neoplasm compatible with spongioblastoma, or giant cell subependymal astrocytoma, and (C) a *lipoma* extending into the corpus callosum.

Figure 10.40. An intraventricular tumor, in this case a choroid plexus papilloma (*1*), causing dilation of the entire lateral ventricle (*2*). For orientation purposes, the straight sinus (*3*), torcular Herophili (*4*), and tranverse sinuse (*5*) are illustrated.

Because of the biologically malignant nature of (intralateral ventricle) ependymoma, irrespective of its histological appearance, excessive efforts to attain a complete resection are to be avoided. The surgeon is best to limit himself to as complete a resection as possible, given the limitations of controlling the bleeding and following the tumor into the brain substance.

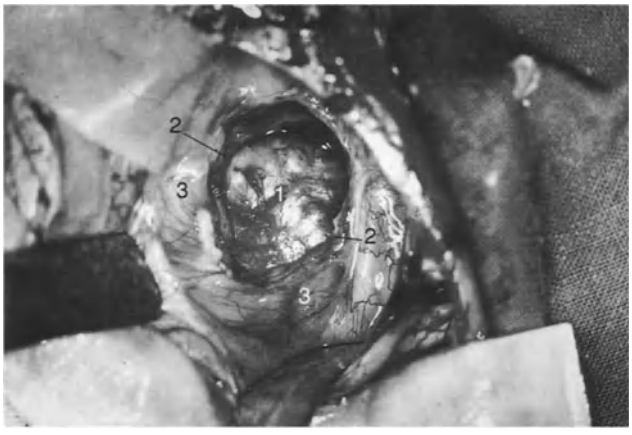


Figure 10.41. Giant subependymal astrocytoma, which occupied the entirety of the left lateral ventricle. The tumor (*1*) is seen through the cerebrotomy opening (*2*), allowing one to appreciate both its mass and density, which were most likely responsible for the gliotic appearance of the overlying cerebral hemisphere (*3*).

Glial Tumors (Fig. 10.41)

Though glial tumors of the hemisphere, or the corpus callosum, may bulge into a lateral ventricle, compressing it completely, as the neoplastic hemisphere shifts beneath the falx, clinically significant excrescences into a ventricle are not such as to justify classifying these as intraventricular gliomas.

Subependymal Gliomas

The subependymal glioma expands almost exclusively within the ventricle from either its superolateral wall or the septum, indistinguishable from ependymoma or choroid plexus papilloma in that they present clinically with the *midline syndrome* and neuroradiologically as enhancing, intraventricular masses, which may cause symmetrical or asymmetrical biventricular hydrocephalus.

Although those gliomas arising from the inferior and lateral surfaces of the ventricle expand only within the body of the lateral ventricle, those arising from the septum pellucidum may expand into one or both lateral ventricles, as well as the III ventricle inferiorly or the pineal recess posteriorly. The intraventricular glioma is glistening in appearance, grayish yellow, and has an irregularly lobulated contour. One finds that subependymal gliomas are quite dense. They do not obliterate the normal subependymal veins. Therefore, the surgeon may obtain the incorrect impression that the tumor is, in essence, a deformed tumefaction of the ventricular wall.

These are beingn tumors, they seldom invade the basal ganglia or thalamus, and they are best treated by removing only the intraventricular portion of the tumor and, at that, just enough to guarantee opening of the cerebrospinal fluid pathways. The clinical symptomatology these children suffer is a result of the complicating hydrocephalus. This does not mean to imply that a simple shunting procedure would be preferable: shunting from the homolateral ventricle results in repeated shunt obstructions because of compressive occlusion of the proximal end of the shunting system by the intraventricular tumor.

Since the Cavitron represents the ideal instrument for removal of intraventricular glioma, techniques using sharp dissection, suction, or cautery loops are acceptable only when one is unavailable. Sharp dissection is awkward and imprecise within the ventricles, suction technique is incomplete because of the density of the tumor, and electrocautery loops suffer the definite disadvantages of potential sectioning of deep-seated vessels within the tumor mass. The Cavitron permits the surgeon precise ultrasound aspiration of the tumor without the need of retraction, and the ability to shave away the neoplastic cells, layer by layer.

Those intraventricular gliomas that take origin from the inferolateral surface of the ventricle are best approached through a cerebrotomy performed within a silent cortical area. This brings the surgeon into the lateral ventricle along a plane that gives him direct access to the tumor pedicle. Once the ventricle is entered and opened, so as to expose completely the tumor pedicle from anterior to posterior, wet Telfa is slid over the intraventricular tumor excresence, separating it from normal ependyma. The surgeon proceeds from the superior surface of the tumor to its medial aspect, taking care to identify the terminal and anterior septal veins before retracting the tumor from the surface of the foramen of Monro. *This permits the surgeon to "dome" the lesion: to work around its surface to its base.* At times, neoplastic adhesions may bridge across the ventricle from tumor surface to the foramen of Monro. Fluffy cotton may be used to dissect the medial extension of tumor from the foramen of Monro, moving cautiously along the terminal and anterior septal veins. In the event there are adhesions, or it appears one of these veins may be occluded or at risk of rupture, one may coagulate either without concern.

With the superior and medial surfaces of the tumor freed, the surgeon may pass a Telfa along the inferior intraventricular portion of the tumor, from anterior to posterior, so as to separate it from the superomedial and inferomedial ventricular surfaces. This frees the superior and medial surfaces, allowing the surgeon to turn the tumor on the stretch, but augments the risk of tearing subependymal veins.

With the pedicle of the tumor visualized along its superior surface, and walled off from the underlying inferomedial wall of the ventricle along its inferior surface, the surgeon is prepared to cut along an imaginary

line separating tumor from the inferolateral surface of the ventricle. This permits *en bloc* removal of the glioma. Occasional bleeders may be encountered along the surface of the tumor bed, but this is rare and the bleeders are small and easy to occlude. No effort is made to remove glioma located within the cerebral parenchyma because of the risk of extending the dissection into the internal capsule, or of causing edema of this and surrounding structures.

Gliomas of the Septum Pellucidum

Though the subependymal tumors are very often multiple, masses of the septum pellucidum (and lamina terminalis, which must be considered with them because there is so very often contiguity of growth) are solitary. They are *spongioblastomas*.

Gliomas of the septum pellucidum very often occlude both foramina of Monro, and extend (to greater or lesser degrees) into both lateral ventricles. Consequently, one may choose to remove these tumors, as illustrated, through a cerebrotomy in a silent area along the convexity of the right hemisphere, or via a parasagittal transcallosal approach (see Fig. 10.34). This approach affords excellent exposure of both lateral ventricles, permitting maximum opportunity to appraise the degree of tumor invasion of the fornices, and to protect one or both during the tumor resection. The approach through a cerebrotomy positions the surgeon so that he is looking at the lateral extension of the dome of the tumor, and places him on the blind side of the fornices and the extension of tumor into the contralateral ventricle.

After an S-shaped skin incision has been made and a medial frontoparietal craniotomy reflected, the dura is opened and the right parietal lobe elevated from the superior sagittal sinus and the falx. Since these tumors are rare in newborn and infants, the surgeon will encounter a full complement of cortical bridging veins, which may be so arranged as to oblige him to sacrifice one in order to expose, appropriately and safely, the corpus callosum. As the hemisphere is elevated and held in place with a self-retaining retractor, care must be taken in separating the superomedial surface of the hemisphere from the superior sagittal sinus, and in assuring that there is not undue stretch placed on the anterior and posterior bridging cortical veins.

The incision in the corpus callosum should be just long enough to permit exposure of the tumor and visualization of those portions of the bodies of the lateral ventricles into which the tumor is bulging. Complete section of the corpus callosum may result in either a disconnection syndrome or memory deficits. Limited incision of the genu, body, or the splenium is safe. The body of the corpus callosum, overlying the tumor, is generally flattened and thin, so one may split it cleanly and bloodlessly by using a Penfield #4 dissector in long, cutting sweeps. The neoplastic septum pellucidum is encountered. Cerebrospinal fluid flows into the field from each lateral ventricle as it is opened. Telfas are brought into each lateral ventricle, over the lateral extension of tumor from the septum pellucidum, so as to protect the ventricular surfaces. Both anterior septal veins and both foramina of Monro are identified. One then proceeds to identify the terminal veins bilaterally, so as to guarantee protection of the internal cerebral veins at the foramina of Monro, where they receive their two major tributaries (the anterior septal and terminal veins). At this point. the columns of the fornix are identified as they curve gently into their direct extensions, the bodies of the fornices. Though section of one fornix results in only a transient memory loss, section of both fornices is to be avoided at all costs because of the permanent and totally disabling intellectual consequences!!

Removal of these tumors is carried out by using the Cavitron. Traction or suction may damage permanently the fornices without the surgeon being aware of what is happening. Here, again, as in resection of inferolateral intraventricular glioma, the purpose of the operation is to open the cerebrospinal fluid pathways. Once the foramina of Monro have been cleared of tumor, and the septum pellucidum opened so as to permit flow of cerebrospinal fluid between the two lateral ventricles, the operation may be considered completed. Pursuing the dissection along the septum pellucidum puts the bodies of the fornices at considerable risk.

Papillomas

The surgical implications and considerations of lateral ventricle papillomas include the asymmetrical communicating hydrocephalus and expansion of the tumor mass at the trigone; occlusion and subsequent cystic transformation of either the temporal or occipital horns; extension of the tumor mass along the choroid plexus through the foramen of Monro and into the III ventricle, with resultant obstruction and progressive dilation of the lateral ventricle; and extension of the tumor mass through the choroidal fissure and into the quadrigeminal cistern, from whence it may enter either the opposite lateral ventricle or the posterior fossa.

Asymmetrical Hydrocephalus

Asymmetrical hydrocephalus and elevated CSF protein (with complicating adhesive arachnoiditis) (Fig. 10.42) is treated with external ventricular drainage, since the volume of cerebrospinal fluid secreted and the extremely high protein levels preclude the peritoneal cavity as an absorptive surface and a valvular system as a conveyor. The drainage should be continued until the ventricles are decompressed and the child is ready for surgery, no longer. Almost all of these children will need a permanent shunt.

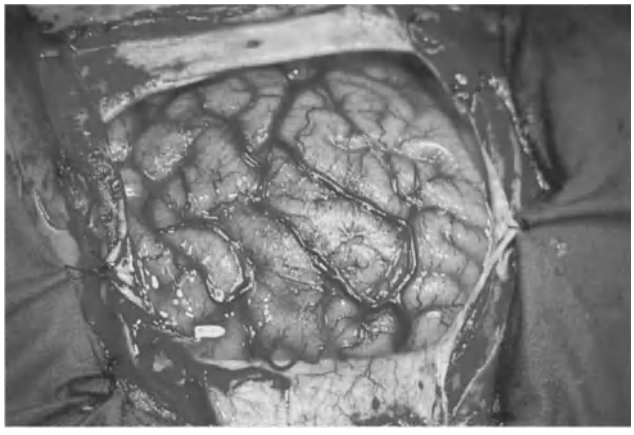


Figure 10.42. Cortical adhesive arachnoiditis (secondary to extraordinarily high CSF protein) in a child with a choroid plexus papilloma of the lateral ventricle. The granular appearance of the arachnoid over the right hemisphere is quite typical of this entity.

Choroid Plexus Papilloma of the Glomus

The choroid plexus papilloma (Figs. 10.43, 10.44) may expand from the glomus to occupy the entire lateral ventricle, in which instance drainage of the cerebrospinal fluid from the temporal horn is *obstructed*. Accumulation of xanthochromic cerebrospinal fluid within the now *encysted temporal horn* results. It is not at all unusual for the ependymal lining of the trigone, or fibrocollaginous material, to coat the surface of the tumor, taking on the appearance of a capsule. This causes a cyst to form over the surface of the papilloma: neuroradiologically this looks like porencephaly or tumor.

Trigone papillomas are best resected through a temporoparietal flap, which is large enough to give access to the temporal pole anteroinferiorly and the angular gyrus posteroinferiorly. Upon opening the dura, one notes flattening of the gyri. Puncture of the encysted temporal horn, before opening the dura, with drainage of its densely xanthochromic fluid, affords immediate decompression (from 10 to 60 ml CS). A linear cerebrotomy, 3–4 cm, within a sulcus located behind the angular gyrus, provides generous access to the trigone. It is not necessary to resect a cone of cerebral tissue, since a long linear incision provides adequate exposure of the ventricular system immediately over the trigone tumor mass. The cerebrotomy is best made in the most flattened pathologic portion of the cortex, which is overlying the tumor. The cerebral mantle is already maximally compressed from the underlying tumor and cyst. Bipolar coagulation and then sectioning of the arachnoid with microscissors, followed by bipolar coagulation of the cerebral mantle, assure hemostasis of this highly vascular area at the time of cortical incision.

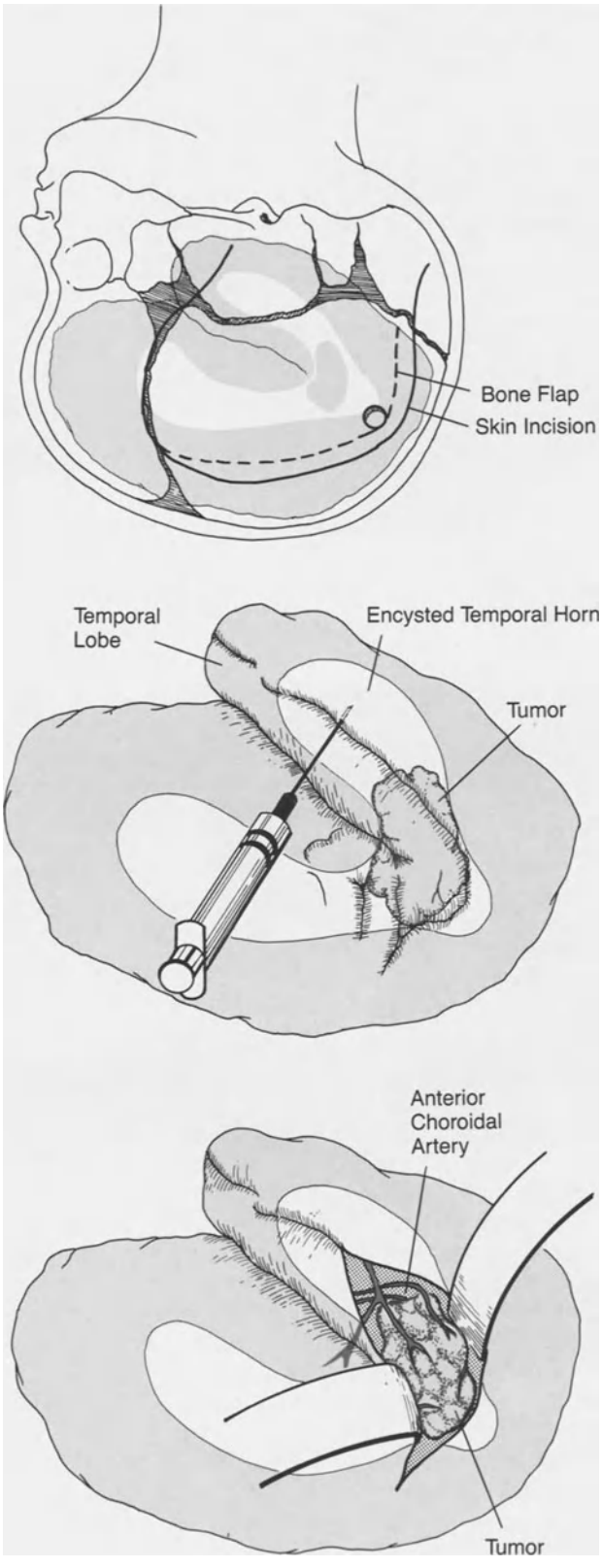
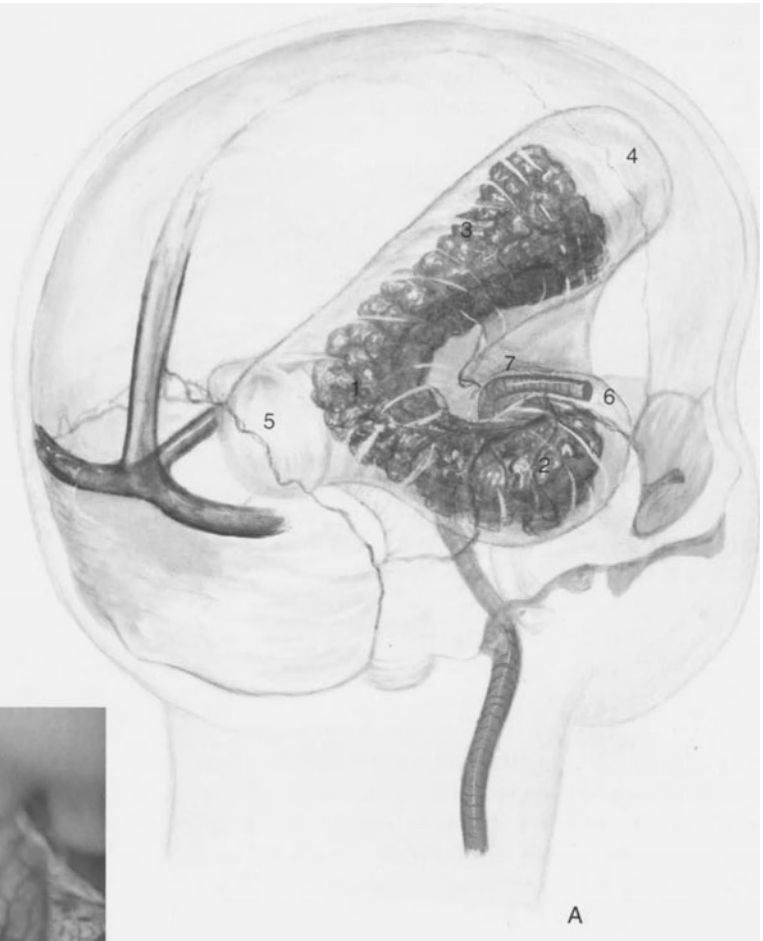


Figure 10.43. Head position, recommended skin incision, drainage of encysted temporal horn, and resection of papilloma at the trigone.

Figure 10.44. (A) This laterale ventricle papilloma occupies the trigone (*1*), temporal horn (*2*), and body (*3*) of the lateral ventricle. The frontal (*4*), occipital (*5*), and temporal (*6*) horns are encysted. The middle cerebral system (*7*) is flattened between expanding frontal and temporal horns. This drawing illustrates the importance of access to the temporal horn to control the anterior choroidal artery, to the trigone for the glomus, and to the foramen of Monro for the internal cerebral and arterior septal veins.



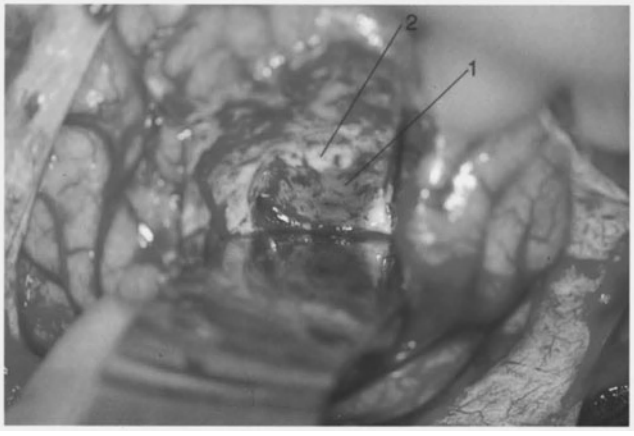


Figure 10.44. (B) After the cerebrotomy has been made and the dissection carried through the white matter, the spatulae are positioned at either extremity of the cerebrotomy, exposing the ependyma (*1*) and a thin rim of white matter surrounding it (*2*).

The dissection through the white matter is performed with two spatulae, thereby minimizing the area of cerebrotomy and maximizing the depth of opening. Once the ependyma over the surface of the tumor is identified, it may be coagulated and sectioned, thereby unsheathing the tumor proper. Layers of fibrocollagenous tissue may be stratified between the xanthochromic ependyma and the glistening or opaque lobular surface of the papilloma.

Cotton fluffies are pulled into the desired size and form, and then freshly soaked in normal saline at body temperature, prior to being used to dissect the surface of the tumor from the surrounding ependyma and parenchyma, with which the tumor may have established continuity or adhesions. Telfas are placed between the tumor and the ependyma of the ventricular wall, and then snuggled into position around the pedicle of the papilloma. This isolates completely the tumor from the wall of the ventricle, and permits one to define clearly the size and extent of the pedicle of the tumor. Thereafter, fingerlike pieces of cotton fluffies are brought circumferentially over the lobular mass until they surround the pedicle or the tumor at its inferomedial surface, the line of entry of choroidal arteries, and exit of draining veins.

It cannot be overstressed that these tumors must not be removed piecemeal either with monopolar loops or pituitary forceps, because of their high degree of vascularity! Rather, they must either be removed *en bloc* after the pedicle has been coagulated and sectioned (an archaic technique to be used only when *the most limited* facilities are available), or gradually shrunk with bipolar forceps. Irrespective of the size of the tumor, it is almost always possible to identify the vascular pedicle

and then to deal with it hemostatically under direct vision. Identification of the pedicle by digital palpation and "blind" application of hemostatic clips to the pedicle *was* an acceptable technique at one time, but is no longer acceptable.

Linear cerebrotomy through the encysted temporal horn permits ample access to the tumor pedicle, and to the feeding branches of the anterior choroidal arteries, into the tumor at its posteroinferomedial surface, if the tumor is moderate in size. If, however, it is large, one may not obtain a direct line of vision to the vascular pedicle, something which may not be known preoperatively. Consequently, the surface of the tumor should be coagultaed, using bipolar forceps to shrink individual lobules, applying the blades to the surface of a lobule and activating the current without closing the blades. This coagulates the papilloma with only minimal risk of tearing small vessels. The tumor may be diminished considerably in size with this technique, permitting the surgeon access to the pedicle.

The vascular pedicle may extend anteroinferiorly, from the trigone, along the choroidal fissure, toward the hippocampus, or superomedially along the choroid plexus toward the terminal sulcus and foramen of Monro, penetrating the choroidal fissure to enter the quadrigeminal and galenic systems. It may also extend downward to enter the supraculminate system. In either event, it is best dealt with within the lateral ventricle where it may be coagulated and transected. Most of the small feeding arteries may also be coagulated and transected, but the larger ones should be clipped prior to sectioning. Following removal of the tumor mass, wait for approximately 10 min after filling the ventricle with normal saline so as to be certain that there is no active bleeding, or oozing. Inspect the entire lateral ventricle, especially the foramen to look for a clot which may be plugging the outflow of cerebrospinal fluid.

The above technique predated perfection of bipolar thermocautery or the laser for neurosurgery, and is described both for historical pruposes and for the benefit of those confronted with the need to resect a choroid plexus papilloma but without thermocautery or laser facilities and instrumentation. The CO_2 laser should be set at 8 W, continuous mode, and the beam maximally defocused, so as to provide a vaporizing beam.

A cutting beam is not used in such a vascular tumor as this! If 8 W is not enough, 12–15 W, but not beyond this because of the friability and vascularity of the papilloma, are such as to render working at higher voltage unsafe.

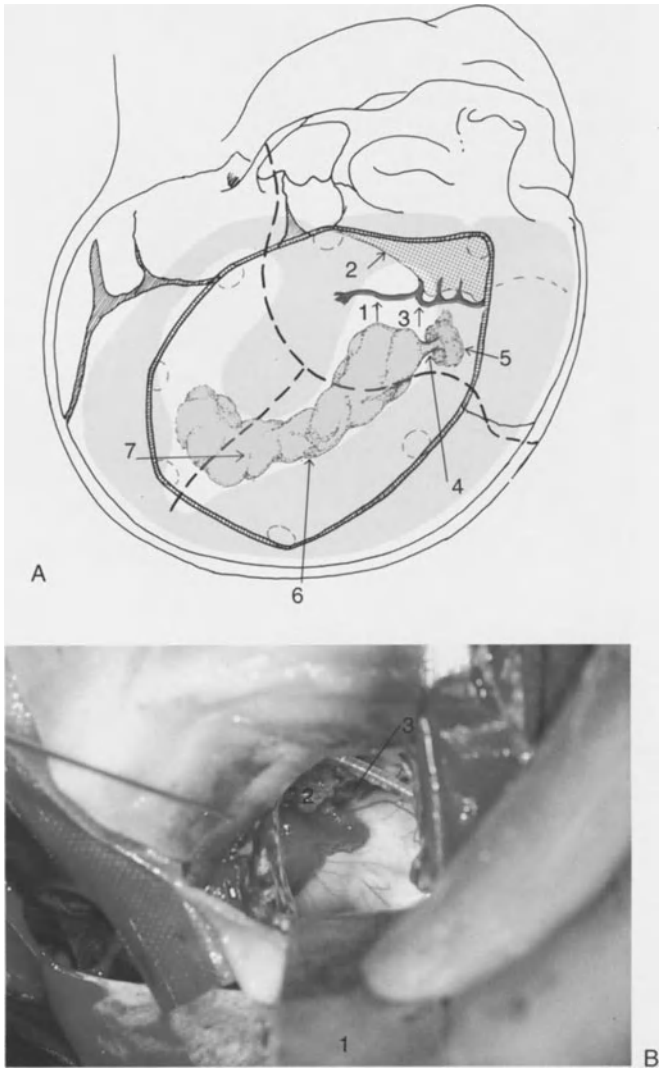


Figure 10.45. (A) Recommended position, bone flap, lines of dissection, and extent of tumor in papillomas entering the III ventricle through the foramen of Monro. The middle cerebral artery (*1*), lesser wing of the sphenoid (*2*), anterior cerebral artery (*3*), tumor extending through the foramen of Monro (*4*) and into the III ventricle (*5*), tumor in the lateral ventricle (*6*) and at the trigone (*7*), and skin incision (*broken line*) are illustrated. (B) The spatula (*1*) is elevating the trigone and body of the right lateral ventricle, exposing papilloma (*2*) over the thalamus and plugging the foramen of Monro (*3*). The proximal tubing for drainage of cerebrospinal fluid from the right lateral ventricle crosses the papilloma just anterior to the foramen of Monro.

Lateral Ventricle Choroid Plexus Papilloma

The lateral ventricle (Figs. 10.45–10.47) choroid plexus papilloma may extend along the choroid plexus into the *terminal sulcus*, and anteriorly as far as the foramen of Monro, entering the *III ventricle*. When this occurs and the foramen of Monro is occluded, one may encounter encysted occipital and frontal horns as the mass occludes the trigone and enters the III ventricle.

A temporofrontoparietal flap (allowing access to the angular gyrus posteriorly, the middle frontal convolution anteriorly, the rostrum and genu of the corpus cal-

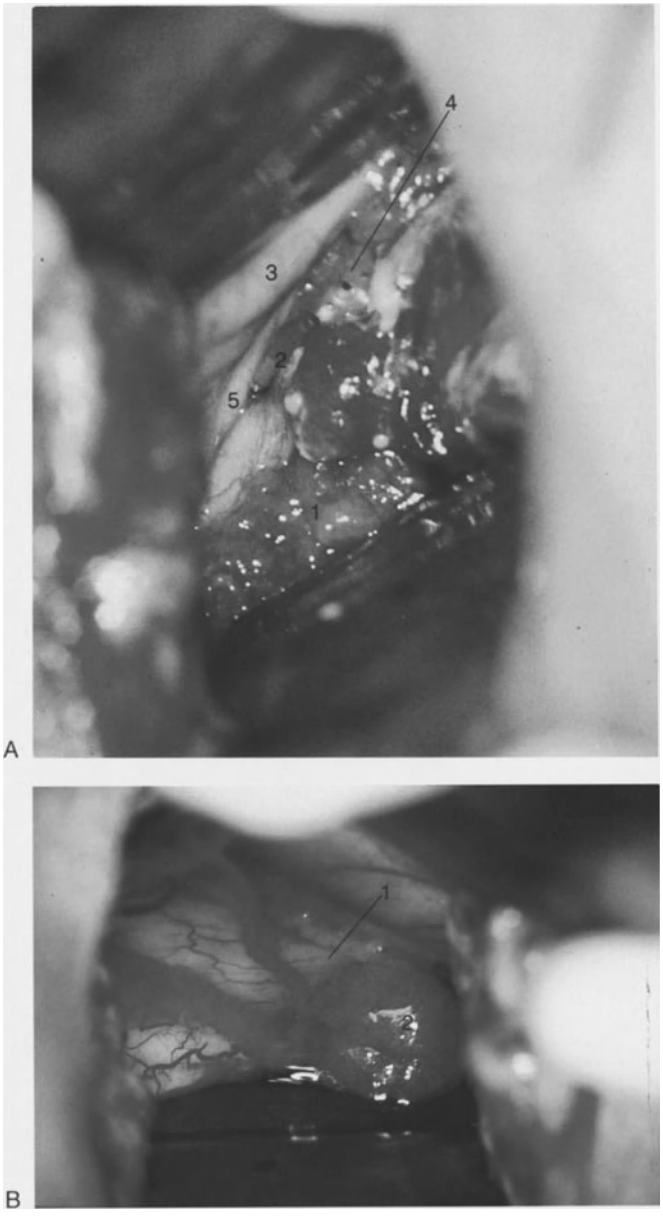


Figure 10.46. Technique and exposure for opening the septointerthalamic portal to the III ventricle: specifically, the attachment of the *lamina affixa* to the thalamus at the terminal sulcus, the line along which the choroid plexus also runs. Brushing the choroid plexus from the surface of the thalamus, along with the *lamina affixa*, establishes a direct communication between the lateral ventricle and the III ventricle, displacing medially the roof of the III ventricle, inferior to the septum pellucidum. (A) The choroid plexus papilloma (*1*) and the *lamina affixa* (*2*) have been brushed from the thalamus (*3*), exposing the membranous roof of the III ventricle (*4*). The body of the fornix (*5*) has been displaced with the papilloma and *lamina affixa*. (B) This photograph was taken through the operating microscope. The *lamina affixa* has been freed from the terminal sulcus (*1*), allowing the papilloma within the III ventriccle (*2*) to well up into the operative field, since there is now direct communication between the lateral ventricle and the III ventricle.

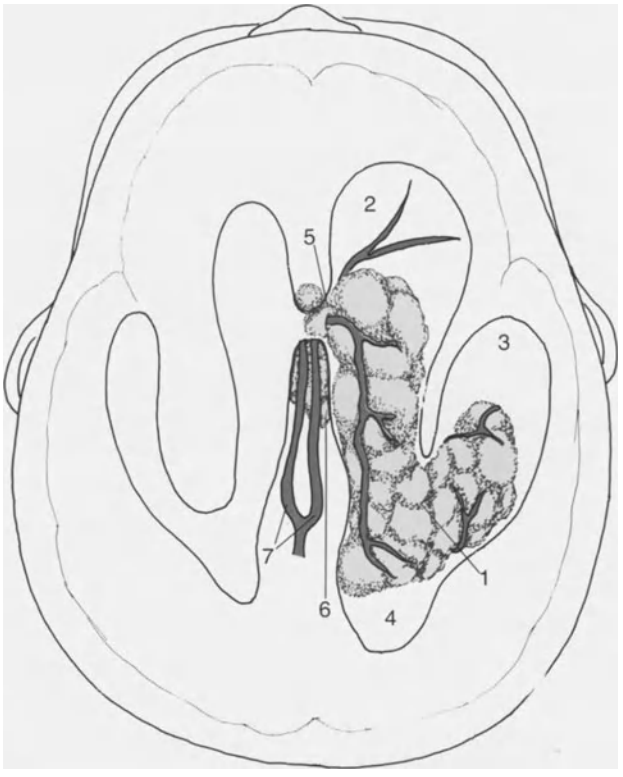


Figure 10.47. Diagrammatic representation of a full axial view of tumor occupying the trigone (*1*), encysting the frontal (*2*), temporal (*3*), and occipital (*4*) horns, and extending through the foramen of Monro (*5*) into the III ventricle (*6*). The veins draining the tumor enter the internal cerebral vein at the foramen of Monro. Tumor in the III ventricle elevates the tela choroidea and deforms both internal cerebral veins (*7*).

losum anteromedially, and the temporal pole anteroinferiorly) is necessary for such a procedure to be effective. Before opening the dura, the encysted ventricular chambers (frontal, temporal, occipital horns) are punctured, allowing egress of the encysted ventricular fluid.

A cerebrotomy of 3–4 cm in length is made behind the angular gyrus. Vaporization or bipolar coagulation of the tumor facilitates its isolation and idenfitication of the pedicle. This should be clipped and sectioned at the point where the feeding branches from the anterior choroidal artery at the base of the trigone enter the tumor from its bed along the terminal sulcus and choroidal fissure. Then, advancing toward the foramen of Monro and coagulating minute perforating feeders along the bed, one may either vaporize the tumor or lift it from its bed within the terminal sulcus. This anteromedial dissection is not carried beyond the point at which the body of the fornix continues into the crus fornices. At this point a cerebrotomy is made in the middle frontal gyrus so as to gain access to the encysted frontal horn.

The foramen of Monro is identified, and obliteration of the perforating feeders to the tumor carried out by proceeding posteriorly from the foramen of Monro to approximately the point at which the body of the fornix ends. This frees the tumor from its attachment to the floor of the trigone and the body of the lateral ventricle.

Extension of tumor from the lateral into the III ventricle, through the foramen of Monro, is variable in amount, vascularity, and adhesions (to the foramen of Monro, the roof of the III ventricle, the anterior surface of the III ventricle). However, there is always a constricted area at the foramen of Monro. If the surgeon chooses the transforaminal entry into the III ventricle, this must be enlarged, after coagulating and clipping the neoplastic extension through the foramen.

An alternative technique for entering the III ventricle, one which is equally complete in exposing the entire III ventricle, and which has the advantage of avoiding dissection along the columns of the fornix, entails stripping the tela choroidea of the roof from its insertion into the superior surface of the thalamus along the terminal sulcus. At this point the tela choroidea of the roof of the III ventricle is continuous with the choroid plexus of the lateral ventricle. The dissection is carried out with fluffy cotton, rolling it backward over the surface of the thalamus from lateral to medial (from the terminal sulcus just medial to the terminal vein). This strips the choroid plexus of the roof of the III ventricle from the thalamus, laying open the entirety of the III ventricle, establishing a continuity between it and the lateral ventricle. In essence, the compartmentalization of lateral and III ventricles is taken down, the fornix is rolled back over the superior aspect of the thalamus where the horizontal surface of the thalamus (floor of the body of the lateral ventricle) turns inferiorly to become the vertical surface of the thalamus (lateral wall of the III ventricle). When this is done any lesion within the III ventricle is visualized immediately. The III ventricle in this way is exposed from the lamina terminalis to the pineal gland.

The advantage of this opening of the III ventricle is that it allows the surgeon to inspect the posterior surface of a tumor for the presence of veins draining into the posterior portion of the internal cerebral veins and the great vein of Galen.

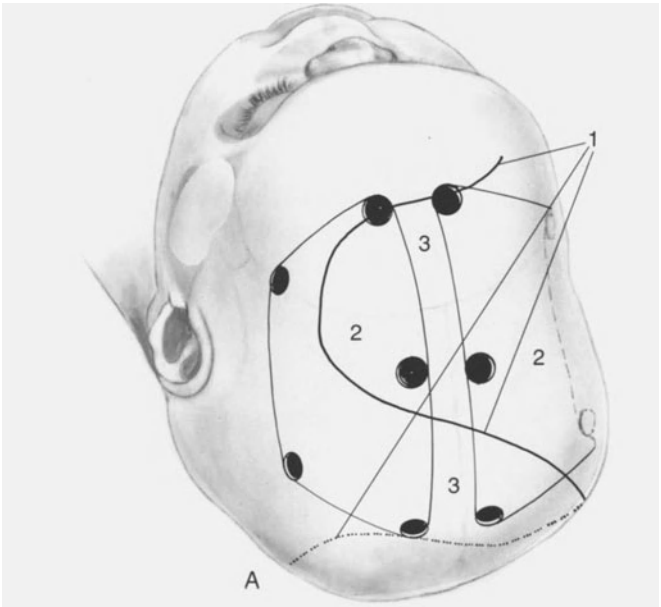
If one chooses to use the septothalamic approach, one must first assure oneself that tumor is not growing through this potential space directly into the III ventricle. If not, then the tela choroidea of the roof of the III ventricle is stripped from its attachment to the superior surface of the thalamus medial to the terminal sulcus, along the floor of the lateral ventricle, taking great care not to damage the body of the fornix! A wide and free communication between lateral and III ventricles is thereby established. This permits separating the main tumor mass (which preferably should be shrunken with bipolar coagulation) from its extension into the III ventricle, and subsequent removal of the tumor in two to attempt to dissect it from the III and lateral (lateral and III ventricular) completely shrunken portions.

After the tumor has been lifted from the lateral ventricle, the surgeon may inspect the tumor within the III ventricle, taking care to assure himself that the anterior septal, terminal, and internal cerebral veins are freed completely both from the tumor and adhesions to it. Appropriately fashioned Telfa strips are then inserted along the superior surface of the tumor, separating it from the roof of the III ventricle and the internal cerebral vein within this latter structure. Similarly, Telfa strips are placed along the lateral surfaces of the tumor and then along its anterior surface as far inferiorly as the floor of the III ventricle. When this is done, gently, and with soaking wet Telfas, one avoids damaging the vascular or nuclear structures bordering upon the III ventricle and one is assured of freeing the intraventricular mass.

At this time a large fluffy is placed in the lateral ventricle, at the foramen of Monro, and the line of dissection is moved from transventricular to parasagittal by retracting the frontal lobe laterally. This gives access first to the genu and rostrum of the corpus callosum and then to the lamina terminalis (anterior border of the III ventricle), as far inferiorly as the chiasm. If the tumor is adherent to the walls of the III ventricle, one may choose to enter this chamber through its anterior margin. This requires performing a cerebrotomy from the rostrum of the corpus callosum inferomedially along the fornix, and then through the lamina terminalis to the supraoptic recess. Complete visualization of the anterior and superior portions of the III ventricle, of the anterior septal vein, and of the foramen of Monro is achieved. Since all vascular supply and drainage to the III ventricular portion of these tumors is along the tela choroidea, one need not be concerned that there may be vessels within the area of the floor of the III ventricle or the lamina terminals.

Bilateral Lateral Ventricle Papilloma

Lateral ventricle papillomas (Fig. 10.48) may extend through the *choroidal fissure* into the *quadrigeminal cistern* and, at times, even into the *contralateral ventricle, becoming bilateral lateral ventricle papillomas.* Those papillomas extending directly medialward through the choroid fissure, and into the quadrigeminal cistern, may either expand "dumbbell fashion" on either side of the choroid fissure or into a nodule within the quadrigeminal cistern. They may then penetrate the contralateral ventricle through its choroid fissure, thereby becoming indistinguishably intermingled with its glomus. In either event, the tumor compresses the quadrigeminal plate, becomes adherent to the commis-



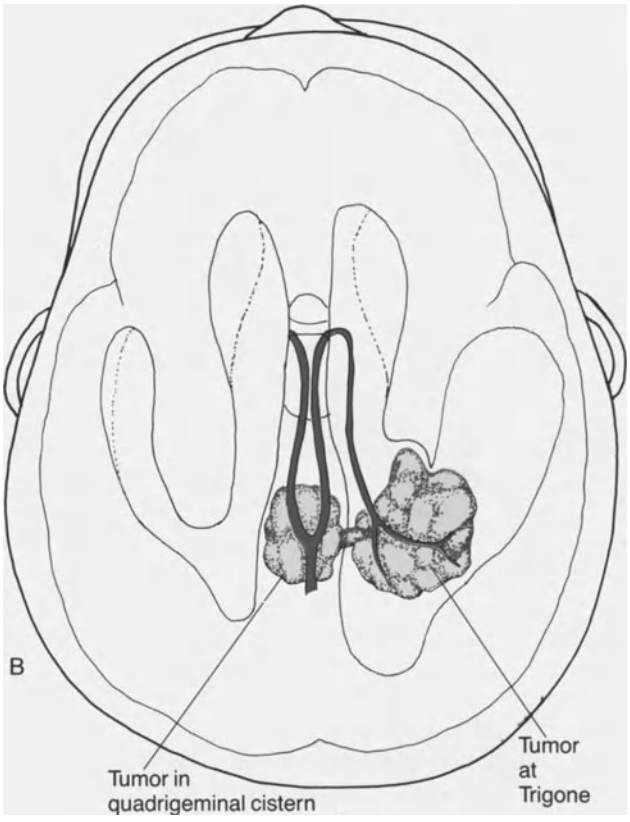


Figure 10.48. (A) Recommended S-shaped skin incision (*1*), biparietal bone flaps (*2*), and strip of bone over the superior sagittal sinus (*3*) for combined approaches to tumor at trigone, in the quadrigeminal cistern and at the foramen of Monro. (B) Extension of a choroid of a choroid plexus papilloma from the trigone of one lateral ventricle, through the choroidal fissure and into the quadrigeminal cistern. It illustrates not only the extension of the mass along the choroid plexus and through the choroidal fissure, but also the advantages of an S-shaped skin incision and biparietal bone flap for access to such a tumor. The specific detail illustrated here is that the tumor is extending posterior to the III ventricle and inferior to the suprapineal recess. Consequently, one must have access to the trigone of the lateral ventricle and to the splenium of the corpus callosum in order to remove the extension from the quadrigeminal cistern. (C) A papilloma (*1*) growing from the glomus, at the trigone (*2*), directly medially toward the choroidal fissure (*3*) and through it.

sure of the fornix, and elevates the splenium of the corpus callosum. There is obstruction to the passage of cerebrospinal fluid from the ambient cistern into the quadrigeminal cistern, forward displacement of the pineal gland and the suprapineal recess of the III ventricle, and to some degree extension of tumor along the anterior surfaces of the culmen monticuli and lobulus centralis of the cerebellar vermis.

The tumor is fed through the medial and lateral branches of the posterior choroidal arteries on both sides: quadrigeminal and inferior-retrosplenial arteries. It drains into the internal cerebral veins (lesser galenic system), the veins of Rosenthal, the greater galenic system, the supraculinate system, and the anteromedial occipital veins.

A biparietal craniotomy through an S-shaped skin flap permits access to the angular gyri on both sides and to the region of the quadrigeminal cistern. After puncture of the temporal horn (bilaterally when the tumor is expanding in both trigones), the dura is opened and the tumor is dissected free from its ventricular adhesions down to its pedicle at the choroidal fissure, from the pes hippocampus anteriorly to the collateral eminence posteriorly, through a cerebrotomy posterior to the angular gyrus.

The line of dissection is changed from transventricular to parasagittal only after the intraventricular (on

both sides when the tumor extends into the contralateral ventricle) portion of the tumor has been removed.

At this time, fluffy cottons are in the lateral ventricle along the choroidal fissure. The dissection of the tumor within the quadrigeminal cistern is performed from the parasagittal approach, coming down bilaterally on either side of the falx cerebri. The anterior and posterior branches of the lateral posterior choroidal artery, going to the tumor with the quadrigeminal and inferior retrosplenial arteries, are identified as a group, occluded, and transected. This dissection is carried out from side to side and from behind forward. It is necessary to identify the anterior choroidal arteries, coming posterolaterally through and over the tela choroidea of the III ventricle. Venous drainage into the lesser and greater galenic systems, and their tributaries, diminshes considerably once these last feeding vessels are transected.

However, removal of the tumor without identifying, occluding, and transecting the bridging veins results in the opening of these major, normal venous structures.

The tumor, consequently, may be removed in two or three pieces which have been maximally shrunken by bipolar coagulation, depending upon whether it has extended, dumbbell fashion, only into the quadrigeminal cistern or has entered the contralateral ventricle.

Again, it is advisable to wait approximately 10 min in order to assure oneself that there is no oozing before proceeding with filling of the ventricular system with normal saline and closure. As in all choroid plexus papillomas, the resection is best accomplished with laser or by shrinking the mass with bipolar forceps.

Midline Tumors

Tumors of the midline may be intraventricular (III, aqueduct, IV), intracisternal (parasellar, pontine, medullary, quadrigeminal, magna), or intraparenchymal (mid-brain, pons, medulla, vermis). In this text, we discuss them as *midline ventricular tumors, brainstem tumors, vermian tumors, cisterna magna tumors, pineal region tumors, and parasellar tumors.*

Midline brain tumors in childhood are particularly interesting because together they make up a statistically significant percentage of neoplasm afflicting the child's nervous system. Furthermore, individual locations are the site of specific tumor types: sella turcica – craniopharyngioma; hypothalamus – astrocytoma; IV ventricle floor – ependymoma; pons – biologically malignant glioma. Each anatomopathological entity tends to have its own clinical symptom/sign complex: III ventricle floor germionoma – precocious puberty; midbrain glioma – imperceptibly slow onset and progression of hydrocephalus; medulloblastoma – explosive triventricular hydrocephalus. All midline tumors of childhood have in common the growth triad of intraventricular intraparenchymal intracisternal expansion, with the first and last occurring either as sessile or pedunculated (exophytic) excrescences of neoplastic tissue, which may be resected with relative ease and to the child's benefit.

The intra-axial (intraparenchymatous) growths have recently piqued the neurosurgeon, his aroused curiosity bringing him to focus upon utilizing new technology for surgical eradication of these old enemies. The results have been far from dramatically successful, so chemotherapy is being explored as an adjuvant method. The negative effects of radiation therapy seem to have been understood by all, but full-dose whole head/tumor/spinal cord therapy is still used regularly, and the neurosurgeons reporting their experiences and results continue to consider the complication of hydrocephalus a real enough problem to insert regularly precraniotomy shunts. They all comment on the nonexistence of seeding of neoplastic cells through the shunting system as a clinical reality.

Midline Ventricular Tumors

Third ventricle tumors, especially those growing within the posterior portion of the III ventricle, may extend posteriorly into the quadrigeminal cistern or inferiorly into the superior cerebellar cistern; superior III ventricular tumors may grow inferiorly into the aqueduct, both plugging it and occupying its entirety; IV ventricular tumors, such as medulloblastoma or ependymoma, all – too – often fungate into the posterior portion of the III ventricle. Therefore, one may justify considering an anatomical description of the midline ventricular system as it relates to extension of tumors from one ventricular chamber into the region of another and, more directly, planning of surgical flaps for access to these tumors.

Because of the enormity of the basal cisterns and the absence of the sphenoid air sinus, the III ventricle resting superior to the IV ventricle, the brainstem appears to extend directly from the center of the hemispheres, in a vertical course which is perpendicular to the horizon. Consequently, although the III ventricle is directly superior to the IV ventricle, it is not in the same coronal plane, being located slightly anterior to it.

As will be discussed subsequently, *tumors of the III ventricle* are divided (from a surgical point of view) into anterior, superior, and posterior III ventricle tumors. This has a rational basis, since it assists the surgeon in approaching the III ventricle through the foramen of Monro or the lamina terminalis (anterior III ventricular tumors); the transcallosal/roof of the III ventricle route, or the transventricular/transeptal route (tumors within the roof of the III ventricle); and the occipital/transtentorial route (tumors of the posterior III ventricle).

Tumors of the pineal region may:

1. Expand superior to the roof of the III ventricle, in which case they are approached as for roof of III ventricle tumors.
2. Remain within the pineal and quadrigeminal cisterns, in which case they are approached as for posterior III ventricle tumors.
3. Extend into the posterior fossa by growing primarily within the superior cerebellar cistern and displacing the lobulus centralis of the vermis posteriorly, in which case they are approached through a suboccipital supracerebellar craniotomy.

The incidence of expansion of pineal tumors into the III ventricle, between the III ventricle and the body of the corpus callosum, or into the posterior fossa is equal. The decision concerning which of these approaches is best for pineal tumors will be discussed subsequently. It is predicated entirely upon the direction of tumor displacement of the deep venous structures: lesser and greater galenic systems.

Fourth ventricle tumors are approached through a midline suboccipital craniotomy, as for tumors of the inferior cerebellar triangle, without removal of the arch of C1. If the IV ventricle tumor is very large, one may expect to find that it fungates into the aqueduct of Sylvius and III ventricle superiorly; the lateral recesses and, at times, the pontocerebellar cisterns laterally; and the vallecula and cisterna magna, medially. Access to the vallecula, IV ventricle, and aqueduct is adequate through a simple midline craniotomy, but special consideration must be given to exposure of the lateral recess. In fact, this does not entail simply extending the width of the craniotomy laterally on both sides, since the lateral recesses are paramedian structures, located at the point where the pontocerebellar junction nestles into the petrous apex. Access to this area is achieved by elevating and displacing laterally the cerebellar tonsils. This permits one to expose the lateral recess from within the IV ventricle, and then to elevate the cerebellar hemisphere from the squamous occipital bone, so as to view the lateral recess over the IX, X, and XI cranial nerves at the pontocerebellar junction. This is all possible if the rim of the foramen magnum is reflected with the inferior triangle craniotomy.

The suboccipital craniotomy affords ample bone opening for complete exposure of IV ventricle tumors and their extensions, facilitating access to portions of the III and IV ventricles.

Third Ventricle Tumors

General Discussion (Fig. 10.49)

The III ventricle is a diverse anatomical structure delicately situated at the inferomedial aspect of each hemisphere, wedged between the thalami and mesencephalon, and capped superiorly by the internal cerebral veins. From a functional point of view the lamina terminalis and tela choroidea anteriorly and superiorly, respectively, represent silent areas, whereas the inferior and posterior surfaces, the hypothalamus and midbrain respectively, are eloquent. When viewed from the lateral perspective, one may note that the internal cerebral vein and choroid plexus of the roof of the III ventricle outline the superior surface of the III ventricle, whereas the basal vein of Rosenthal describes a concave arch around its inferior surface. If this were to be projected from the axial perspective, it would be seen that the internal cerebral veins delimit the medial aspect of the III ventricle, and the basal veins of Rosenthal its lateral surfaces. Its posterior borders are the pineal gland and the iter to the aqueduct of Sylvius, its anterior borders the lamina terminalis, the supraoptic and infundibular recesses.

The infundibulum and tuber cinerum are the anatomically identifiable structures within the membranous portion of the floor, whereas the surface of the midbrain constitutes the anatomical structures for the solid portion of the floor. The columns of the fornices, running from the mammillary bodies to the foramen of Monro, and the crus fornices, running from the foramen of Monro posteriorly to the level of the pineal gland, represent the eloquent anatomical structures along the lateral surfaces of the III. One may, therefore, understand immediately that the safest, but by no means the most convenient, partial entry into the III ventricle is through the lamina terminalis; the second safest, and most dangerous from the vascular point of view, is through the roof; and the most awkward and difficult access is through the posterior surface where the pineal gland is lodged. If one wishes complete access to the III, with assurance that neither the fornices nor the vascular strucutres within the roof may be damaged, then consideration should be given to entering it through the lateral ventricle: separating the choroid plexus from the lamina affixa of the terminal sulcus as described for papillomas extending into the III from the lateral ventricle (see previous section). This establishes an open communication between lateral and III ventricles.

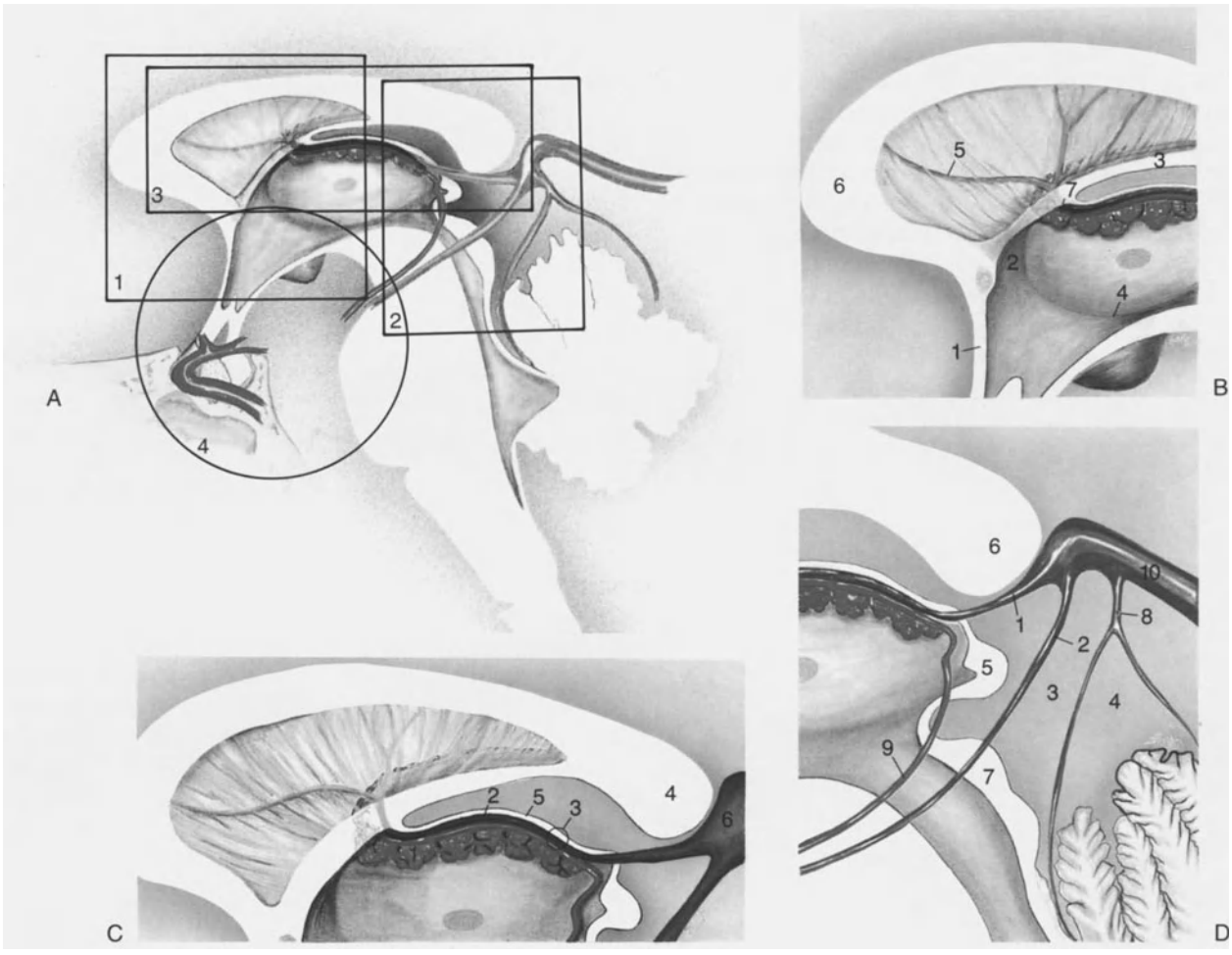


Figure 10.49. (A) One may reasonably divide the III ventricle into anterior (*1*), posterior (*2*), and superior (*3*) portions. This distinction is rational since the differences in these three sections of the III ventricle are both anatomical and functional, despite the fact that there is overlap between the superior and anterior portions anterosuperiorly, and the superior and posterior posterosuperiorly. The fourth area, a portion of which is occupied by the III ventricle, is the parasellar area (*4*). Therefore, one may speak of anterior, superior, and posterior III ventricular tumors in addition to parasellar tumors. The distinction is predicated upon primary origin of the tumor, e.g., posterior III tumors originate from the pineal gland, superior III tumors from the choroid plexus, anterior III tumors at the foramen of Monro and lamina terminalis, and parasellar tumors from the pituitary gland and optic chiasm. (B) A coned-down view of the *anterior III ventricle*, for considering removal of intraventricular tumors located within this area, reveals the lamina terminalis (*1*), foramen of Monro (*2*), body of the fornix (*3*), and hypothalamic sulcus (*4*). The course of the anterior septal vein (*5*), from the genu of the corpus callosum (*6*) to the point at which the terminal vein curves through the foramen of Monro to enter the internal cerebral vein (*7*), allows one to appreciate the significance of considering these venous structures when working within the region of the foramen of Monro or anterior portion of the III ventricle. (C) The superior *III ventricle* contains only vascular structures medially, and is in direct continuity with the body of the fornix slightly lateral to the midsagittal plane. The choroid plexus of the roof of the III ventricle (*1*) fishes freely within this chamber, whereas the internal cerebral veins (*2*) are located in the tela choroidea (*3*). The splenium of the corpus callosum (*4*) overhangs the posterior portion of the roof (*5*) and the great vein of Galen (*6*). (D) Within the area of the posterior III ventricle are the most posterior portions of the internal cerebral vein (*1*) and the basal vein of Rosenthal (*2*), the quadrigeminal (*3*) and supracerebellar cisterns (*4*), the pineal gland (*5*); superiorly, the splenium of the corpus callosum (*6*); inferiorly, the quadrigeminal plate (*7*) and, posteriorly, the supraculminate veins (*8*). The medial posterior choroidal artery (*9*) and the great vein of Galen (*10*) complete the major structures bordering the posterior III.

General Comments Concerning Access to III Ventricle Tumors

Access to the anterior III ventricle may be obtained through a subfrontal approach, sectioning the lamina terminalis. This is adequate if one wishes to perform a III ventriculostomy, but does not provide the surgeon proper exposure for a tumor within the III ventricle. All of the vascular structures come either from the superior or posterior aspects of the III ventricle and, consequently, would be located behind the tumor mass. Colloid cyst of the III ventricle, the most common anterior III ventricle tumor in the adult but a true rarity in the child, may be removed by the subfrontal, translamina terminalis approach. The risk of anosmia does not justify this approach for such a benign lesion, one which may be resected as effectively through a transventricular approach.

The craniopharyngioma often grows superiorly, at times displacing the floor of the III ventricle as it bulges more or less completely into this chamber and at others fungating freely into and then within it...in either instance very often occluding one or both foramina of Monro. Consequently, it may be necessary to enter the III to remove tumor, in which case, already working subfrontally, the lamina terminals is the portal of preference. However, one must weigh the advantages – direct visualization of tumor – against the disadvantages – postoperative panhypopituitarism. Translamina terminalis resection of craniopharyngioma – a tumor which originates around the infundibulum immediately beneath the III ventricle – invariably results in irreversible damage to the inferomedial hypothalamic nuclei and the pituitary stalk.

Anterior III Ventricle Tumors

Anterior III ventricular tumors are best approached through a lateral frontal craniotomy and cerebrotomy with transventricular access to the foramen of Monro. Since this is a much simpler craniotomy, it is associated with no neurological deficit and allows for identification of the anterior septal, terminal, and internal cerebral veins while removing the tumor.

Superior III Ventricle Tumors

Superior III ventricular tumors are best approached along the *parasagittal/transcallosal* route when the tumor is located between the roof of the III ventricle and the corpus callosum (as indicated angiographically by downward displacement of the internal cerebral veins), but by the *occipital transtentorial* approach when the intra-III ventricular tumor is displacing the internal cerebral veins upward.

Posterior III Ventricle Tumors

Posterior III ventricular tumors are approached through either a medial occipital or a parietal parasagittal bone flap, followed respectively by either a transtentorial or transcallosal approach to the pineal region. The exception is those which expand primarily within the superior cerebellar cistern, in which case the *suboccipital/supracerebellar* approach is preferable. These criteria are described in detail in the section on pineal tumors.

Specific Comments Concerning Access to Tumors of the Anterior III Ventricle

Lamina Terminalis Approach (Fig. 10.50)

The *subependymal glioma* may grow from the lamina terminalis. This is a rare tumor, one which is impossible to distinguish from hypothalamic or optic pathway gliomas. The only relatively common tumor of the anterior III ventricle is the *colloid cyst.* The colloid cyst is also referred to as a paraphyseal cyst, since it is thought to take origin from the paraphysis and to contain a substance which is colloid in consistency. This tumor is well encapsulated, nonvascularized, and loosely attached to the anterior surface of the lamina terminalis at the foramina of Monro. It may be easily mobilized and readily separated from the ependyma of the walls of the III ventricle. Currently endoscopic surgery permits one to resect the colloid cyst through a bur hole opening.

Lateral frontal craniotomy and cerebrotomy afford access to the junction of the frontal horn and the body of the lateral ventricle, and then visualization of the enlarged foramen of Monro with the capsule of the colloid cyst bulging into the lateral ventricle. Care should be taken to identify the anterior margin of the foramen of Monro so as to orient oneself concerning the precise location of the columns and body of the fornix, the anterior septal vein, and the terminal vein.

These latter structures, tributaries to the internal cerebral vein, should be separated from the superior surface of the capsule of the tumor by using tiny fluffy cottons and a microdissector, running this latter instrument within the space between the rim of the foramen of Monro and tumor capsule. It should be directed from anterior to posterior and from superior to inferior, with the point of departure being the junction of the anterior septal and terminal veins. In some patients there is a false venous angle: the terminal vein enters the internal cerebral vein well behind the foramen of Monro – rendering identification of the terminal vein difficult. One is forewarned by studying the preoperative venogram.

It is often possible to free the colloid cyst from the surrounding foramen of Monro with dissection, and to deliver the cyst directly through the unopened foramen.

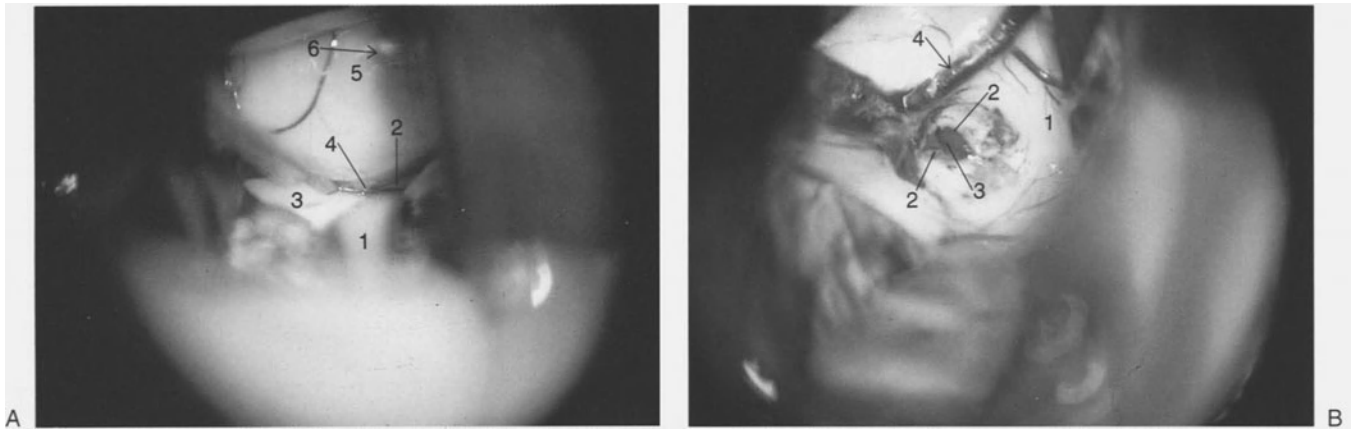


Figure 10.50. Transventricular approach to III ventricle colloid cyst. It does not matter whether one enters the lateral ventricles through the middle frontal convolution or along the parasagittal route and through the corpus callosum. In fact, the parasagittal route is quite easy in the child, though it is actually very difficult in an adult because of the significant size of the bridging cortical veins and the multitude of pacchionian granules. Once the lateral ventricle is entered, one identifies the septum pellucidum and the foramen of Monro. (A) A Penfield #4 dissector (*1*) has been placed along the anterior inferior limb of the foramen of Monro (*2*) and a small cottonoid (*3*) is being inserted between the posterior rim of the foramen of Monro and the sliverlike visible portion of the colloid cyst (*4*). The septum pellucidum (*5*) is being opened with the CO_2 laser. Note the defocused beam (*6*). The CO_2 laser is particularly effective in situations such as this, since its vaporizing activity is stopped immediately when it encounters water. (B) The septum pellucidum (*1*) has been opened (*2*) with the CO_2 laser. The colloid cyst (*3*) is clearly visible through the opening in the septum pellucidum, as are the anterior septal (*4*) and subependymal veins.

However, the size of the colloid cyst, or its adhesions to the surrounding ependyma, may be such as to discourage "delivery." In these circumstances, one should open the foramen of Monro by cutting inferiorly or directly posteriorly, avoiding either an anterior or a superior cut since these put the column and the body of the fornix, respectively, at risk. Such a cut may be performed either with microscissors or the laser. When this is done, the cyst may be seen to consist of a pedunculated structure hanging posteriorly and inferiorly from the anterior surface of the foramen of Monro. The peduncle may be coagulated with bipolar forceps and then cut with microscissors, allowing the surgeon to lift it through the foramen of Monro and out of the III ventricle. There is nothing gained by removing the colloid cyst *en bloc*. One may drain the contents and then work one's way around the collapsed capsule, freeing it from the surrounding ependyma as one goes.

Tumors of the Roof of the III Ventricle (Fig. 10.51)

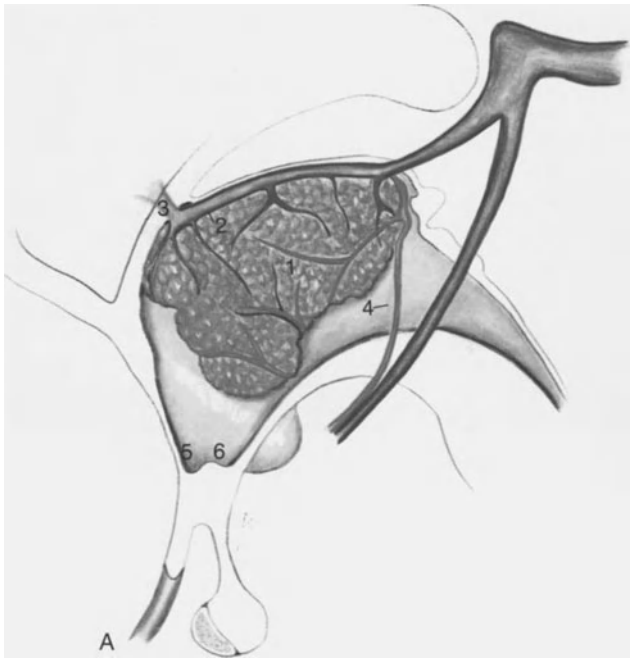
The only tumor which may be considered to grow from the roof of the III ventricle in childhood is the *choroid plexus papilloma*. Although meningiomas, theoretically, may occur here, they do not occur in childhood and, for all intents and purposes, there is no clinical or surgical difference between menigioma and choroid plexus papilloma.

Since the choroid plexus papilloma of the roof of the III ventricle attains enormous size, is clinically silent until it produces symmetrical, biventricular hydrocephalus, and routinely causes hypersecretion of cerebrospinal fluid if it is impossible to operate on immediately, one has no choice other than to put the child on bilateral external ventricular drainage, and to maintain the drainage functional until the child's condition permits surgery, or to shunt the child.

The choroid plexus papilloma of the III ventricle is approached by using the septo-interthalamic route already described for papillomas extending into the III through the foramen of Monro. It may, however, be approached through either the transcallosal or transforaminal routes, with the former being more familiar (and dangerous) and the latter technically simpler and safer.

Transcallosal Approach to Superior III Ventricle Tumors

The transcallosal approach, using a medial parietal bone flap and parasagittal access to the corpus callosum, entails splitting of the body of the corpus callosum from the genu to the splenium, *an undesirable option*. This also subjects the child's intellectual function to greater risk. Once the splenium of the corpus callosum has been split, the strawberry-red multilobulated tumor and the flattened internal cerebral veins immediately



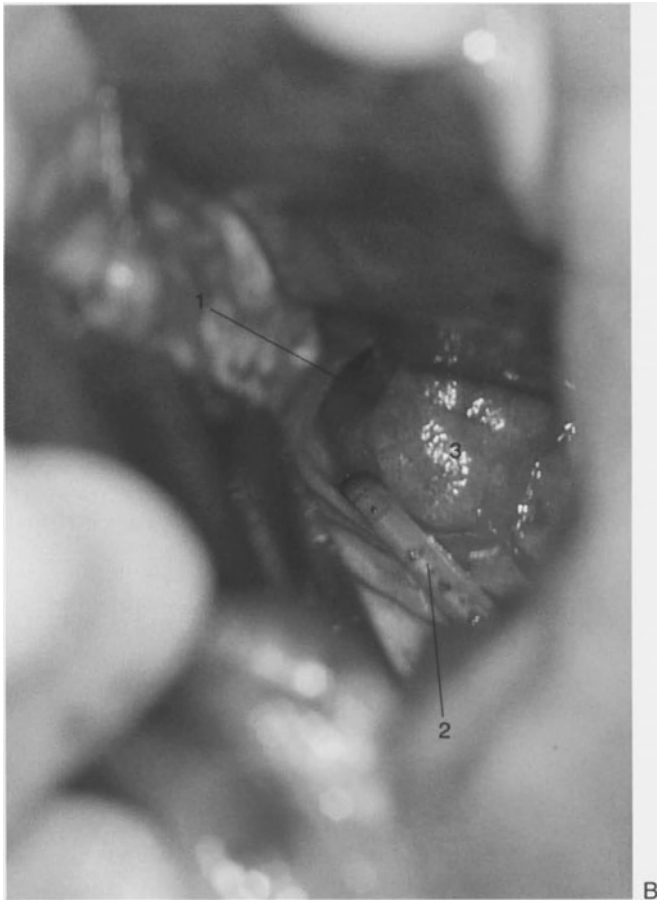


Figure 10.51. (A) The papilloma (*1*) elevates the internal cerebral vein (*2*) from the foramen of Monro to the suprapineal recess. The tumor always extends into the foramen of Monro (*3*). The major feeding vessel is the medial posterior choroidal artery (*4*). Posteroinferiorly, the mass may plug the aqueduct. It generally dilates the inferior III ventricle, deforming the supraoptic (*5*) and infundibular (*6*) recesses. (B) The split corpus callosum has been retracted and the roof of the III ventricle (*1*) opened; the proximal end of the shunt tubing (*2*) is seen between the tela choroidea and a lobule of the III ventricle papilloma (*3*).

come into view. The tela choroidea should be cut along its attachment to the thalamus lateral to the homolateral internal cerebral vein after the former has been coagulated. As this is done the tumor oozes into the opening, filling it entirely as the tela choroidea is unfolded. The tumor may be rolled over, onto the roof of the III ventricle and the internal cerebral veins, by inserting Telfa strips into the III ventricle, bringing them around its anteroinferior and posterior surfaces. The Telfa may be used to hold the multilobulated tumor after it has been freed from the strands of fibrocollagenous tissue which attach loosely to the ependyma of the III ventricle. The surgeon may begin to remove it from the ventricular surface of the roof by coagulating it with bipolar at the junction between roof and tumor. If this is difficult because of inability to identify completely the pedicle, one may choose to shrink the tumor with bipolar cautery, a relatively simple and very safe procedure.

If a laser is available it may be used to vaporize the tumor, but care must be taken when using it in and around the III ventricle: the beam may open the lesser or greater galenic systems, or it may damage the nuclei along the ventral and lateral hypothalamic surfaces. It is safer to use bipolar cautery to shrink tumors in this location.

Once the mass of tumor has been reduced, either by bipolar shrinkage or laser vaporization, to such an extent that the surface of attachment may be identified, one proceeds to coagulate and section along this line until the entirety of the tumor has been freed from its attachment to the roof of the III ventricle. The anterior landmark is the foramen of Monro, the posterior the suprapineal recess or pineal gland. Only when the mass is floating *freely* within the III ventricle should it be lifted from the field. Doing so before this may be complicated by tearing an anterior septal or internal cerebral vein, the consequences of which are catastrophic because of the sudden and uncontrollable bleeding – lest one is fortunate enough to find and coagulate the internal cerebral vein.

After the tumor has been lifted from the III ventricle, its bed should be identified and complete hemostasis assured.

Septo-interthalamic Approach to Superior III Ventricle Tumors

If the transforaminal (transparenchymal, transventricular) approach is selected, a frontoparietal bone flap and cerebrotomy are used to gain access to the lateral ventricle. This exposes the insertion of the tela choroidea of the III ventricle at the terminal sulcus, and the choroid plexus of the lateral ventricle running over this line. Fluffy cotton is used to strip the tela choroidea and choroid plexus from the line of the terminal sulcus, and over the superior surface of the thalamus, until the lateral and III ventricles are in free communication with one another. At this time, the choroid plexus papilloma will float into the interval between the thalamus inferiorly and the body of the fornix superiorly. Resection is then performed in the manner already described.

Posterior III Ventricle Tumors

The posterior III ventricular tumors are, in the broad sense of the term, *pineal tumors*. Specifically, pineal tumor is an anatomical, a surgical-anatomical, term which indicates any tumor occupying the pineal region, whether it be glioma, teratoma, pineocytoma, germinoma, or other. This avoids the conflicting and confusing terminology predicated upon histological characteristics.

For the purposes of surgical indications and considerations, we prefer that the general, anatomical term "pineal tumors" be used for all tumors in the region of the pineal gland, and that such developmental, histological, or congenital terms as "dysgerminoma," "pinealoma," and "teratoma" be reserved exclusively for neuropathological descriptive purposes.

Of our 72 children – age range from 7 months to 16 years – treated for pineal tumors during the 35-year period 1962–1997 inclusive, all presented with hydrocephalus and most necessitated ventriculoperitoneal shunts. This incidence of pineal tumors represents 13.0% of the total number of brain tumors in childhood managed during this period of time. Sixty percent of the children were operated on for direct attack of the tumor, with the postoperative mortality being 1 (as defined by death preceding discharge from hospital): a 7-month-old child with a medulloepithelioma which rapidly, within 3 weeks, invaded the thalamus, corpus callosum, brainstem, and cerebellum, causing death.

It is recommended that cerebrospinal fluid (CSF) be taken from the ventricular system at the time of insertion of the ventriculoperitoneal shunt and that this fluid be analyzed for α-fetoprotein (AFP), human choronic gonadotrophins (HCG), and carcinoembryonic antigen (CEA), and that cytology be performed to search for neoplastic cells. If the marker studies are positive, or malignant neoplastic cells are identified at cytology, then it is recommended that radiosurgery be given to the patient and that no direct surgical attack of the tumor be attempted. On the other hand, if the marker studies and cytology are negative, direct surgical attack is recommended after the ventriculoperitoneal shunt, when all signs of an increase in intracranial pressure disappear. Third ventriculotomy is now a desirable alternative to V-P shunting. None of our patients had steroid management for the increase in intracranial pressure. The shunt sufficed. We suggest that the surgical approach (parasagittal, occipital-transtentorial, suboccipital-supracerebellar) be planned on the basis of the direction of displacement of the galenic (internal cerebral vein, and great vein of Galen) and supraculminate (precentral and superior cerebellar) venous systems.

Following Krabbe's [92] introduction of the term "pinealoma" in 1916, most authors reporting on tumors of this area have chosen to use this term indiscriminately, describing or defining all varieties of neoplasms ranging from dysgerminoma through typical teratoma to glioma of the brainstem. This has been unfortunate.

A great deal of confusion has resulted from the term "pinealoma," disseminated by Cushing [93] in 1927 and Bailey [94] in 1948. Krabbe's definition of pinealoma was histological. The majority of tumors within the pineal region are not tumors of the pineal gland. In this text, consequently, posterior III ventricular-pineal tumors will be described from a purely anatomical point of view and more specifically as regards surgical technique.

Clinical Criteria

Our experience with pineal tumors represents 13% of all childhood brain tumors we treated. This is the highest percentage of childhood pineal tumors in the Western literature. When compared directly to the Eastern literature, it even surpasses the 6.7% reported by Araki and Matsumoto [95], and the 2.7% reported by Sano and Matsutani [96] for all age groups. We do not, however, consider this incidence to be in any way expressive of the epidemiology of pineal tumors, since ours has been a referral practice which would not receive naturally occurring numbers of individual tumor groupings.

We, as others, have noted that the commonest indicence is from the latter period of the 1st decade of life to the early 3rd decade, though we have treated infants with chorioepithelioma, embryonal carcinoma, and medulloepithelioma of the pineal region. The predominance in male patients has been only for germ cell and pineal cell tumors, with sex distribution being even for glial tumors, mesenchymal-type, and cysts. The male-female ratio was 18:8 and the age range was 7 months to 16 years.

The parasellar germinomas which we have observed are all excluded from this series, never occured as pineal

region seeds, never occurred in association with pineal region germinomas, and all responded immediately and in a dramatically positive manner to focal roentgen therapy until 1985, and radiosurgery thereafter. The clinical, endocrinologic, and neuroradiologic characteristics of the parasellar dysgerminoma are such as to permit one to make a diagnosis without performing a craniotomy for either inspection or biopsy of the involved area.

In general, we may categorize the symptoms and signs present in a child with a pineal tumor as those expressive of hydrocephalus (which has invariably been present in our cases), of compressive invasive damage of the surrounding structures, or of endocrine disorders. The hydrocephalus causes headaches, lethargy, either optic atrophy from compression of the optic chiasm by a dilated III ventricle or papilledema from diffuse cerebral edema, and ataxia from either compression of the superior cerebellar peduncles or distention of the lateral ventricles. After shunting, these signs generally disappear, though optic atrophy and ataxia (from direct tumor compression of the superior cerebellar peduncles) persist.

Disruption of fibers coming from areas 18 and 19 and going to the superior colliculi is likely to be responsible first for the paralysis of upward gaze and (terminally in its evolution) then for the forced downward gaze. The periaqueductal syndrome – conversion nystagmus, retraction nystagmus – is expressive of tegmental involvement.

Because of the antigonadotrophin effect exerted by the functioning pineal gland, mediated by melanin and thus light sensitive, it has been postulated that the precocious puberty once thought to be typical of pineal tumors results from glandular destruction and unimpeded gonadotrophic activity. Blinded animals, or those confined to darkness, have been reported to show either gonadal retardation or hypotrophy, most suggestive of being indicative of adaptive species characteristics of birth in the spring to express the suppressive activity [97]. Arita et al. [98] reported five boys with precocious puberty who had high β-(glycoprotein)HCG titers: two with choriocarcinoma, two with germinoma, one with an unverified lesion. Two of these boys had pineal tumors, two parasellar tumors, and one a combined tumor. We did not observe precocious puberty in any of our cases. This was not true in our children with parasellar dysgerminoma, who very often show signs of precocious puberty. Consequently, we wonder whether two of Arita's five patients with precocious puberty may not have had an extension of a hypothalamic glioma into the pineal region, especially in light of the fact that infundibular gliomas are often quite small and the infundibular area is difficult to study neuroradiologically and almost impossible to see surgically.

Calcification was invariably present in our pineal tumor children. Because of the fact that nonpathogenetically associated calcification of the pineal gland does not occur in black children, we considered calcification in the pineal region as being diagnostic of a pineal tumor when identified in a black child.

The value of magnetic resonance imaging (MRI) and computed tomography (CT) scanning here, as in diagnostic medicine in general, is so well known as to need no comment. MRI-angiography is extremely important, cannot be overemphasized, and must not be considered a diagnostic study of the past. It diagnoses the lesion, provides essential information concerning its vascularity, and permits the surgeon to plan with precision and confidence the most convenient approach – parasagittal, occipital, or suboccipital.

The indications for surgery should be clearly identified, since the nature and location of pineal tumors is so variable as to oblige the surgeon to consider that treatment alternatives to direct operative intervention are equally variable. Germinomas, pineoblastomas, and malignant germ cell tumors cause CSF seeding in a very high percentage of cases, though this may not necessarily be diagnosed with cytologic examination in all cases. In fact, only 33% of our cases with CSF seeding showed positive cytology.

Regarding markers, choriocarcinoma produces β-HCG; endodermal sinus tumors produce AFP; and embryonal carcinoma produces HCG and AFP. Unfortunately, teratomas produce no marker substances. Different from gonadal germinoma, the intracranial germinoma may produce β-HCG; despite the absence of synsytiotrophoblast, the commonly accepted site of HCG production. Though the presence of a given marker is indicative of a germ cell tumor, there is no specific correlation between a given marker and a specific histological type of germ cell tumor. The same applies to positive CSF cytologic studies for malignant tumor.

If the marker studies are positive, then one knows one is treating a primitive germ cell, nonresectable, highly radiosensitive tumor. Positive CSF cytology may be indicative of a pineal cell tumor or malignant glioma, carcinoma, sarcoma, etc. Consequently, positive CSF marker studies and/or cytology render surgery for either resection or biopsy of a pineal tumor both unnecessary and unnecessarily dangerous. If, on the other hand, these studies are negative, one is advised to operate and biopsy only if the frozen section is positive for malignant tumor, and to do a total resection if a nonmalignant tumor is found.

With or without surgery, and with laboratory evidence of a malignant tumor, radiosurgery is highly recommended, *though full head and spinal cord treatment should be reserved for those children with positive cytology*! Otherwise, only the tumor site should be radiated, since high dosage "prophylactic" radiation of the child's

brain and spinal cord has severe negative effects on intelligence, growth, etc. [97, 99].

One should, preferably, begin a discussion of the surgical management of "pineal tumors" with the description of the antomical confines of the area in question, and then go on to discuss incidence, pathophysiology, and surgical management. Accordingly, one would begin by considering the pineal region anatomically as being that bordered by the splenium *dorsally*, the colliculi *ventrally*, the posterior surface of the III ventricle rostrally, and the culmen monticuli of the cerebellar vermis caudally. This perspective has been put forth by many authors in discussions of neuroradiologic diagnosis, and the natural history of treated and untreated tumors in this area.

The same degree of sophistication of thought has recently been applied by Rubinstein [100], who, in 1972, deleted the term "pinealoma" from his histological classification of tumors in this region:

1. Germ cell tumors: teratoma, germinoma, choriocarcinoma, embryonal carcinoma, endotermal yolk-sac tumors
2. Pineal cell tumors: pineocytoma, pineoblastoma
3. Glial tumors: astrocytoma, glioblastoma, oligodendroglioma, ependymoma
4. Other tumors: mesenchymal origin tumors such as meiningioma, rhadbomyosarcoma, angioma, hemangiocytoma
5. Cysts: epidermoid, arachnoid, glial

De Girolomi and Schreidek added chemodectoma and glioma to Rubinstein's category 2 (pineal cell tumors).

Gliomatous and gangliogliomatous differentiation often occurs within parenchymal tissue surrounding pineal cell tumors. Pineal tumors vary in histological evolution, typical teratomas changing into tumors with rapid regrowth rates and invasive characteristics from one portion of the tumor to another. Similarly, teratoma and epidermoid tumors have variable cell differentiation in different regions of the tumor, necessitating multicentric examination of the tumor mass before being able to identify with precision the histological nature. *These are some of the factors which render such procedures as stereotaxic biopsy unreliable or misleading.*

It appears that with improving diagnostic techniques pineal tumors are becoming easier to diagnose and, therefore, *apparently* more commonly encountered clinical entities. In addition to this, one notes rather remarkable effectiveness of treating the complicating bi- or triventricular hydrocephalus, successful results of radiation therapy, and reduction of operative and postoperative mortality rates to those identical to other major pediatric intracranial procedures. Consequently, one may now deal effectively with tumors in the pineal region, providing the child with immediate relief from the signs and symptoms of hydrocephalus and with excellent prospects for palliation or cure of the primary lesion.

The problems confronting neurosurgeons today are no longer those of whether a pineal tumor may be operated on or removed completely, but whether direct surgical attack of the tumor is preferable to roentgen therapy and, if so, which (the parasagittal, occipital, or suboccipital) approach should be used. In the past, the decision to operate or irradiate varied from one clinical center to another, and surgical approach to the pineal region varied from one surgeon to another.

There are at the present time adequate clinical criteria to provide information upon which to base a decision concerning when it is in the best interests of the patient to irradiate the lesion and when to operate. The present state of the art concerning the use of such biological markers as HCG, carcinoembryonic antigen, and AFP as indicators of germ cell tumors is such that one may proceed directly with roentgen therapy if cerebrospinal fluid cytology studies for malignant cells do not permit the precise identification of the specific tumor type. They provide enough information to justify roentgen therapy without biopsy of the tumor. Unfortunately, negative cerebrospinal fluid cytology or biological marker studies, especially on ventricular cerebrospinal fluid, do not exclude a malignant germ cell, pineal cell, glial, or mesenchymal tumor.

Craniotomy and biopsy of pineal tumors when the cerebrospinal fluid studies are negative may be considered. However, it is preferable to prescribe a "trial dose" of radiosurgery to see whether the tumor responds. If not, surgery is indicated. The next decision is which route should be used to approach the pineal tumor for biopsy and, if the tumor is benign, complete removal. Each of the three classical approaches (parasagittal/transcallosal, occipital/transtentorial, suboccipital/supracerebellar) has distinct advantages and surgical proponents. It is probable that no single approach is ideal for all patients, and that this represents the reason for the relatively higher postoperative mortality rates reported by those authors who have used only one approach when compared to those who varied the approaches.

Since tri- or biventricular hydrocephalus, secondary to obstruction, respectively, either of the posterior half or the entirely of the III ventricle, has invariably been present in our patients, ventriculoperitoneal shunting is recommended as the *initial* procedure of choice. It provides the child immediate intraventricular decompression and relief from the signs and symptoms associated with increased intracranial pressure. If the hydrocephalus is triventricular, a unilateral shunt is inserted; if biventricular, a bilateral shunt. Steroids have not been used since the problems, hydrocephalus and the periventricular edema associated with it, are treated with

shunting. Hormonal treatment is indicated only in those children who also present with varying degrees and types of hypothalamic inadequacy.

Irrespective of histological characteristics, tumors of the pineal region (pineal tumors) may expand:

1. Anteriorly into the III ventricle
2. Superior to the III ventricle
3. Posteriorly, into the quadrigeminal and superior cerebellar cisterns
4. Inferiorly, along the collicular plate and invading or displacing the lobulus centralis and culmen monticuli of the cerebellar vermis

Intraparenchymal extension into the pulvinar, the tectum, or the hippocampus is not unusual. Similarly, multidirectional extension into, for example, the infrasplenial and trigone areas, of the ventricle and quadrigeminal cistern, are common observations. One may expect a pineal tumor to be intraventricular, intracisternal, intraparenchymal, or any combination of the three (Figs. 10.52, 10.53).

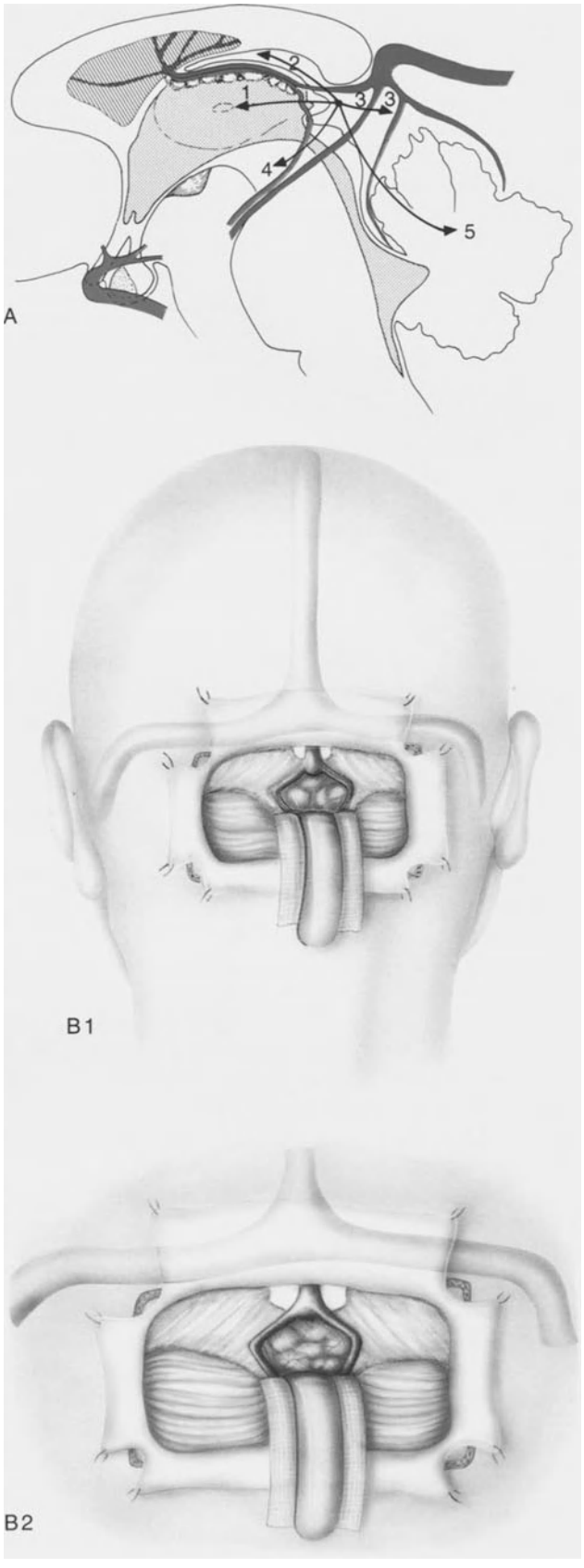


Figure 10.52. (A) Routes of pineal tumor expansion and invasion (*1*) into the III ventricle; (*2*) between the splenium of the corpus callosum and the roof of the III ventricle; (*3*) into the quadrigeminal and supracerebellar cisterns; (*4*) into the brainstem; and (*5*) along the anterior medullary velum and into the cerebellar vermis. (B1) When as in (*1*) in (A), the tumor extends into the III ventricle; the recommended approach is supracerebellar, infratentorial, through a rectangular quadrilateral superior suboccipital craniotomy, allowing the cerebelar hemispheres to fall away and using only a midline retractor over the vermis. It is necessary to section the bridging veins which pass from the superior surface of the cerebellum to the tentorium. This diagrammatic drawing illustrates the location of the tumor at the tentorial opening, embraced by the two veins of Rosenthal (which are splain apart by insinuation of tumor between them. The great vein of Galen is beneath the splenium of the corpus callosum. (B2) This drawing illustrates the spatula over the vermis, the two cerebellar hemispheres, the tentorial surface above them and running toward the midline, the great vein of Galen covering the medial portion of the splenium of the corpus callosum and the two veins of Rosenthal embracing the tumor. The surgeon may work freely between the two veins of Rosenthal and beneath the vein of Galen without fear of damaging a vascular structure, resecting the tumor until he enters the III ventricle. In (C1) and (C2) the parasagittal approach for access to pineal region tumors which grow between the internal cerebral vein and the body of the corpus callosum is illustrated. This approach is recommended because the internal cerebral veins are displaced downward by the tumor. It illustrates the head with its sagittal plane horizontal and the parietal lobe to be retracted superiorly: avoiding gravitation traction on the bridging cortical veins. (C, D) see p. 245.

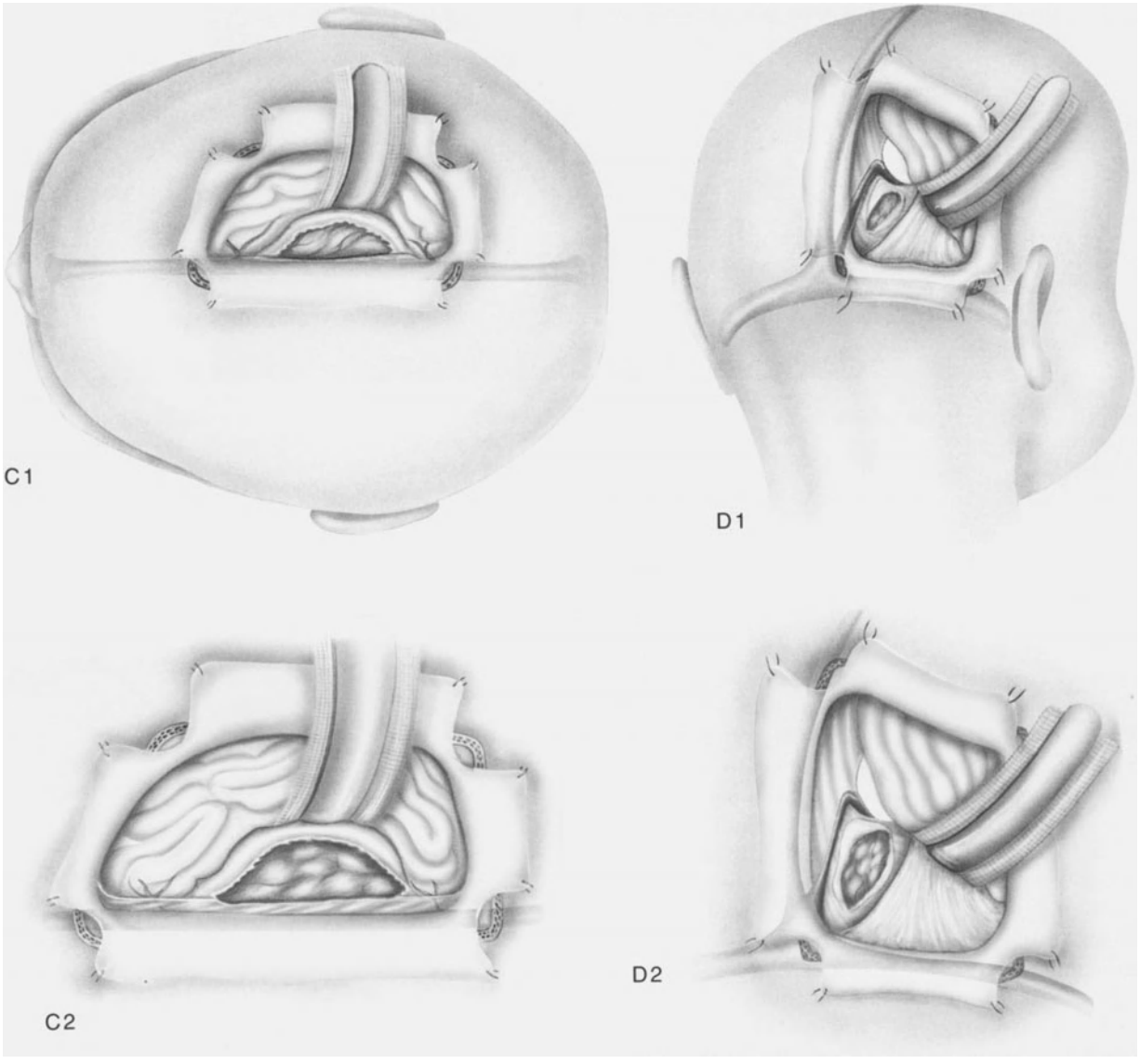


Figure 10.52. In (C2) the drawing has been magnified so as to permit clearer distinction of the extent of the corpus callostomy. (D1, D2) When the pineal tumor extends inferiorly, insinuating itself between the culmen monticuli superiorly and the lobulus centralis inferiorly...and the collicular plate anteriorly, the recommended approach is medial occipital bone flap/superolateral retraction of the occipital pole/transtentorial, with the child in the lounging position. In (D1), the reader notes that the child's head is rotated slightly to the right, the spatula is retracting superolaterally the occipital pole, permitting visualization of the splenium and great vein of Galen as well as the falx cerebri and tentorium cerebelli. In (D2) the tentorium has been incised and the tumor is visualized immediately. For venographic and sagittal schematic drawing representations, see ahead to Figs. 10.63, 10.65, 10.66, 10.68, and 10.69.

There has been considerable literature published concerning the three basic approaches to tumors of the pineal region: the parasagittal/transcallosal approach of Dandy [101], the occipital/transtentorial approach of Poppen [33] and Jamieson [102], and the suboccipital/supracerebellar approach of Krause [103]. It is our opinion that *no single approach should be used for all children with pineal tumors* but that, rather, the approach should be adapted to the specific location of the tumor with regard to its relationship vis-à-vis the deep venous structures. This allows the surgeon to place the tumor between the deep venous structures and himself, so that he is not obliged to work around the internal cerebral vein or the great veins of Galen in order to biopsy or resect a pineal tumor.

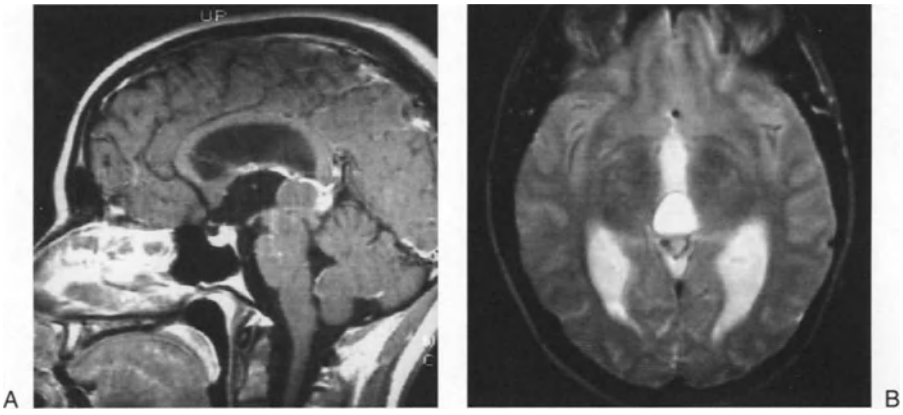


Figure 10.53. The previous illustration may be read as a basis for this hemorrhagic pineal cyst with hydrocephalus secondary to collicular plate compression and aqueductal occlusion. The (A) sagittal and (B) T2 weighted axial MRI studies show, respectively, pineal enhancement of the cystic lesion and hypointensity with intracystic posthemorrhagic fluid level.

Attempts to dissect completely the tumor from the great vein of Galen and the internal cerebral vein are to be avoided, since a multitude of venous drainers enter both of these vessels. *If the tumor is found to be malignant on frozen section, no effort should be made to debulk it, since there is no evidence to suggest that this offers greater survival and since such attempts may result in severe hemorrhage.* If the tumor is benign on frozen section examination, then its removal should be effected by shrinkage with bipolar cautery, until the great vein of Galen and internal cerebral veins are identified. At this time, it may be cut from its adhesions to these veins, with microscissors and under magnification.

If resection of a benign tumor proceeds smoothly, without major bleeding, one may go ahead and resect it totally. However, if difficulties are encountered, it is best to desist: the hydrocephalus will already have been compensated, so one may wait to reoperate if the tumor begins to cause neurological deficit independent of the complicating hydrocephalus.

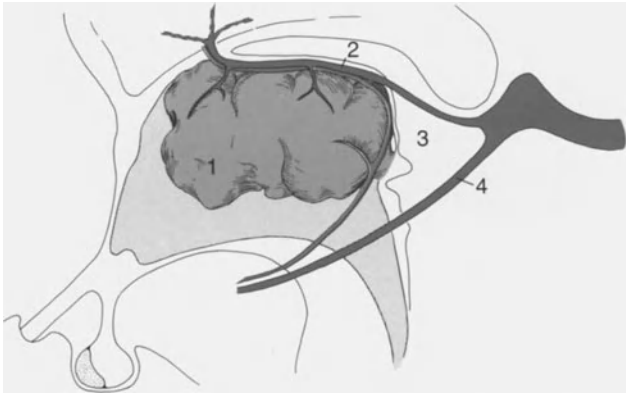


Figure 10.54. The intraventricular pineal tumor (*1*) elevates the internal cerebral vein (*2*), allowing one to conceptualize the advantage of working through the quadrigeminal cistern (*3*) and between the veins of Rosenthal (*4*).

Intra (III) ventricular Pineal Region Tumors: Suboccipital, Supracerebellar Approach
(Figs. 10.54, 10.55)

The specific considerations for the removal of pineal tumors in the III have to do with vascularity, since this tumor takes some of its blood supply from the medial posterior choroidal artery within the roof of the III ventricle. However, it is also supplied by the quadrigeminal and inferior retrosplenial branches of the posterior cerebral arteries, which course through the quadrigeminal cistern. This alerts the surgeon to the necessity of opening the posterior wall of the III ventricle even when it is not completely neoplastic, in order to occlude individual feeding vessels to the tumor mass.

When (MRI) angiography reveals elevation of the internal cerebral vein, this is indicative of primary extension of the tumor into the III ventricle. Extension of pineal tumors into the III ventricle occludes the aqueduct of Sylvius and is often so massive as to occlude both foramina of Monro. In either event, the internal cerebral vein is elevated. The illustration permits one to appreciate the justification for bilateral ventricular shunting procedures, since one may not predict with confidence whether both foramina of Monro are patent.

Elevation of the internal cerebral vein, as revealed by cerebral angiography, secondary to superior displacement by a mass within the III ventricle, suggests the desirability of avoiding a parasagittal/transcallosal approach to the tumor, since the surgeon would be obliged to work around the two internal cerebral veins in order to gain access to the inferiorly located tumor

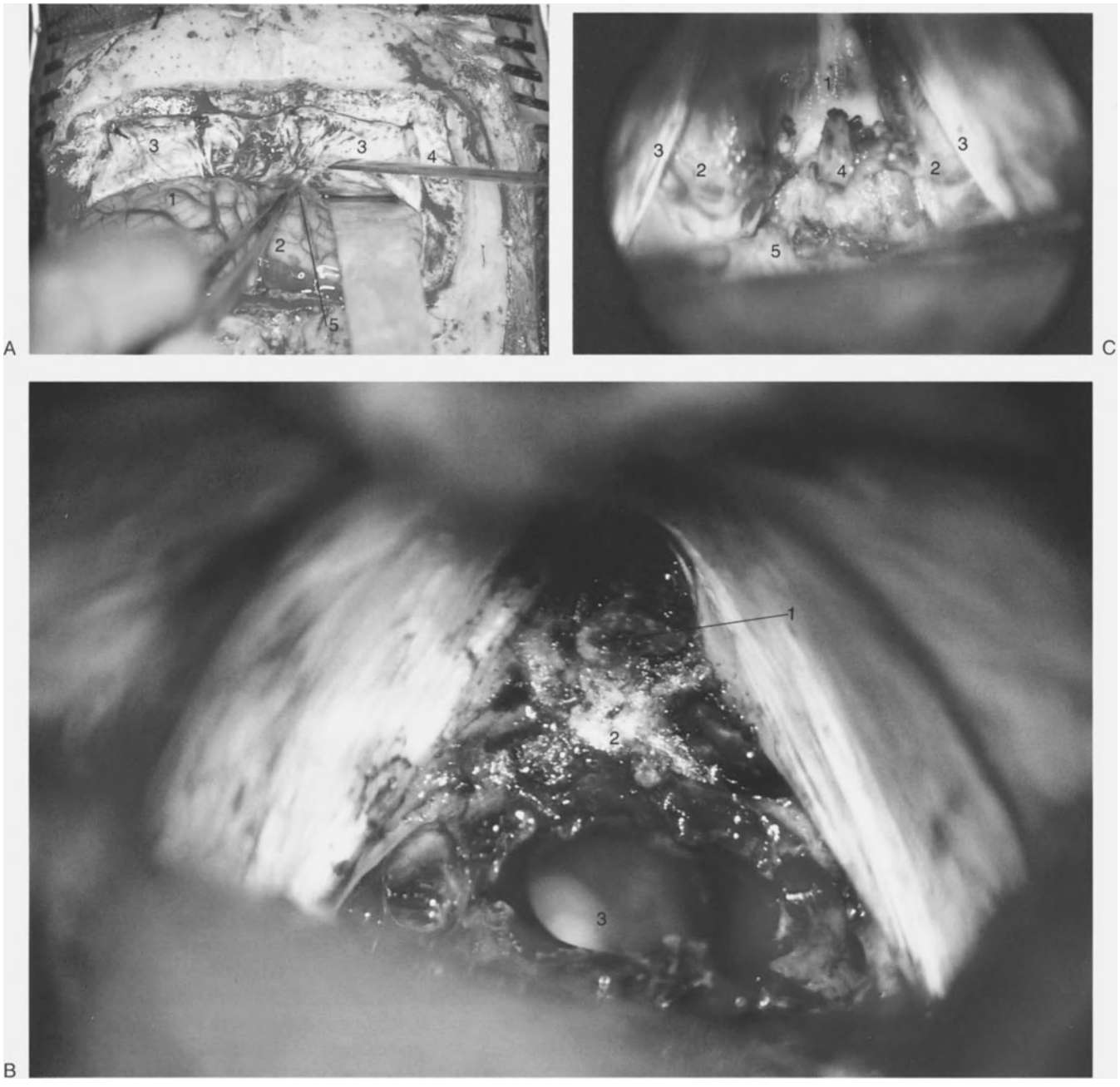


Figure 10.55. In reading this figure, refer to Figs. 10.52B1 and B2. (A) The cerebellar hemisphere (*1*) and vermis (*2*) have been exposed through this superior cerebellar triangle osteoplastic craniotomy, and the dural flap (*3*) sewn superiorly across the level of the transverse sinus (*4*). As a spatula holds the right cerebellar hemisphere, bipolar forceps are used to coagulate the bridging superior cerebellar veins (*5*) before transecting them. (B) Tumor adherent to the posterior wall of the III ventricle beneath both the tumorous pineal gland (*1*) and the most inferior portion of the great vein of Galen (*2*) has been removed, as has the tumor extension into the III ventricle. This results in opening the posterior inferior wall of the III ventricle so that one looks directly into it (*3*). The anatomical landmarks illustrated in the other illustrations of this figure cannot be seen in this photograph. (C) At the end of the procedure, after all of the tumor has been removed, the splenium (*1*) is allowed to fall back into position, blocking view of the III ventricle. The isthmus of the hippocampus (*2*) is seen bilaterally, just beyond the tentorial edge (*3*). The coagulated and transected supraculminate vein and a cluster of innominate vascular structures (*4*) are located between the splenium and the culmen monticuli (*5*).

mass. Subocccipital craniotomy permits an infratentorial/supracerebellar approach to the pineal region, and excellent exposure of both the tentorial opening and the entirety of the tumor mass.

Once the superior portion of the cerebellar hemispheres, the culmen declive, and the culmen monticuli, have been covered with Telfa, one should puncture the atlanto-occipital membrane with a 21- or 20-gauge spinal needle and give egress to cerebrospinal fluid from the cisterna magna. This causes the cerebellar hemispheres to nestle within the squamous portion of the occipital bone, and prevents the accumulation of cerebrospinal fluid in the cisterna magna. The IV ventricle also is collapsed by this procedure, as are the superior cerebellar and quadrigeminal cisterns. One has, in this way, excellent exposure of the tentorial opening and the pineal tumor without undue downward retraction of the cerebellar hemisphere. It is convenient to place a self-retaining retractor over the cerebellar hemispheres, for both protection and *minimal* retraction.

Dense arachnoidal tissue forming the parieties of the superior cerebellar cistern is the first structure encountered. It should be coagulated vertically, over its full extent, and then opened either with microforceps and microscissors, or the laser. The laser technique is very rapid and convenient, but wrought with danger. If used, care must be taken to identify the anatomy of the area precisely, lest the supraculminate vein be opened. After the arachnoidal membrane of the superior cerebellar cistern has been opened it may be reflected to either side, generally giving immediate access to the tumor mass. At this point, one takes note of the fact that the splenium of the corpus callosum may be draped around the (posterior III ventricular) pineal tumor so that the neoplasm is either more anterior or inferior than the surgeon would expect. The supraculminate vein and its tributaries are located in this area, so they should be identified, coagulated, and transected. This frees the posterior aspect of the neoplasm, permitting identification of the feeding vessels.

It is best to resect the tumor by working initially at (within) its core, the most posteroinferior portion, and then to proceed superiorly to either side of the midline. As the tumor is debulked, with whichever technique the surgeon uses (aspiration, cauterization and piecemeal removal, laser vaporization, etc.), one moves to either side along its border with the tentorial edges, gently freeing fibrinous adhesions, coagulating and transecting more dense or vascular bridges. The tentorial edges are free. This accomplished, the superior cerebellar, internal occipital, and posterior choroidal arteries are separated from the surface of the tumor. Each vascular structure along the ambient cistern should be identified, dissected from the tumor mass, and covered with Telfa. The basal vein of Rosenthal stands out prominently in this area.

Continued debulking of the center of the tumor should now permit one to note that the superior dome of the tumor comes into view. This is the most critical and dangerous portion of resection of pineal tumors using this approach: the two internal cerebral veins, the two basal veins of Rosenthal, and the great vein of Galen are often draped over the superior, posterior, and lateral aspects of the tumor's dome. Traction on tumor is to be avoided, lest one of these veins tears. Using the operating microscope, at magnifications of from 10 × to 20 ×, allows one to identify tumor and its capsule, and to separate these from the centrencephalic venous system, when this is possible. Complete resection, down to the last cell or cluster of cells, is most unusual and of doubtful value, so that portions of tumor tissue adherent to these vessels may be left

The splenium of the corpus callosum will steadily fall into the operative field as the dome of the tumor is removed, hanging over the posterior aspect of the III ventricle like a closing shade. A small spatula, or a small-bladed self-retaining retractor, may be used to hold the splenium upward, so that one may follow the dome of the tumor into the III ventricle, which it often invades for at least 2 cm.

Tumors in this location are continuous with the membranous superior and posterior thirds of the III ventricle, so the surgeon must identify the line of cleavage along which he chooses to separate the tumor from the surrounding tissue. Once this is done, the line of cleavage is coagulated with bipolar forceps. Microscissors are used to cut along the line of coagulation, but their use at such a depth suffers the risks of diminished depth perception. If the neoplasm is continuous with the brachium of the superior colliculi, the resection is stopped at least 2 mm superior to this structure.

Freeing the tumor from its attachment to the inferior portion of the posterior III ventricle necessitates bivalving it, so that one is not obliged to elevate the inferior pole in order to identify the collicular plate and brachia of the superior colliculi. Before the bivalving is accomplished, one places a strip of Telfa beneath the inferior pole of the tumor so as to suspend it, keeping it in view. This prevents the inferior pole from dropping out of sight. The cut edge of the superior pole of the tumor may then be held with a micropituitary forceps as the mass is cut from its attachment to the inferior plane of the posterior III ventricle. When this portion of the tumor is removed, the surgeon has a spectacular view of the entire III ventricle, being able to see the internal cerebral veins, the massa intermedia, and the foramina of Monro.

One may now direct oneself to removing the inferior pole of the tumor. This is generally not a difficult procedure, but may prove to be impossible if the tumor has invaded the culmen monticuli. Its resection is effected with the same technique as used for debulking the mass

and removing its superior pole, though one may exert gentle traction on the inferior pole in order to elevate it from the underlying superior cerebellar vermis and identify bridging vessels, which are coagulated and transected.

After the entire mass has been removed one may see the III ventricle, the centrencephalic venous system, the superior cerebellar and internal occipital arteries, the collicular plate, the inferior portion of the splenium, and the medial surfaces of the isthmus of the hippocampus.

Superior to III Ventricle Pineal Tumors: Parasagittal Approach (Figs. 10.56, 10.57)

Pineal tumors that expand superior to the roof of the III ventricle, beneath the body and splenium of the corpus callosum, depress the internal cerebral vein and displace the great vein of Galen posteriorly. The vasculature comes from the medial and lateral posterior choroidal arteries, as well as segments of the quadrigeminal branches of the posterior cerebral artery.

The most favorable surgical approach to a mass located in this area is along the parasagittal route and through the corpus callosum, since this affords the surgeon direct access to the tumor mass and does not oblige him to work around the internal cerebral veins. In this manner, the tumor may be removed from the interval between the body of the corpus callosum and the roof of the III ventricle without undue risk either to the lesser (internal cerebral) or the greater veins of Galen. A parietal flap, followed by dissection of the parietal lobe from the superior sagittal sinus prior to retracting it from the falx cerebri, permits exposure of the body of the corpus callosum. Splitting of the body (*not the entirety*) of the corpus callosum exposes the tumor mass

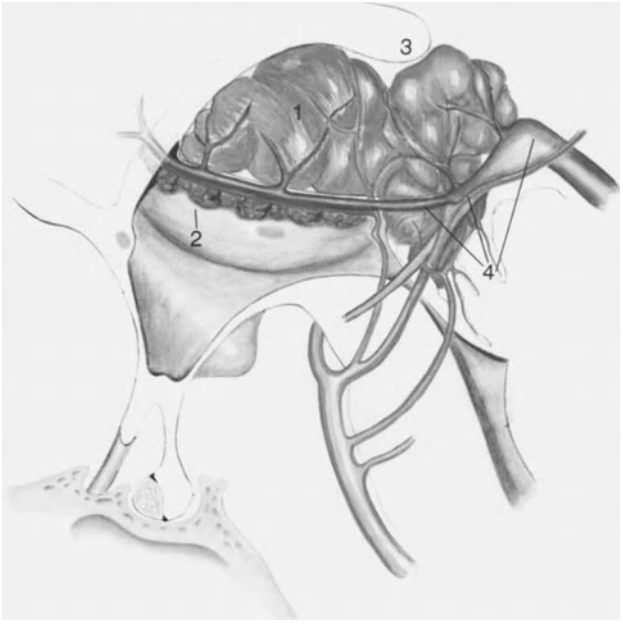


Figure 10.56. Tumor mass (*1*), located between the roof the III (*2*) and the inferior surface of the corpus callosum (*3*). This expresses the anatomical relationship between tumor and centrecephalic (internal cerebral) venous system (*4*).

and "delivers" it into the operative field. Separation of the tumor from the deep venous structures is best begun anteriorly, by taking it from the lesser vein of Galen and then working posteriorly, remembering that the major arterial feeders from the posterior choroidal and quadrigeminal cisterns enter the tumor posteriorly.

Bleeders encountered anteriorly are venous, but the posterior ones are both arterial and venous. When one reaches the area in which the lesser galenic system is tributary to the greater vein of Galen, one may encoun-

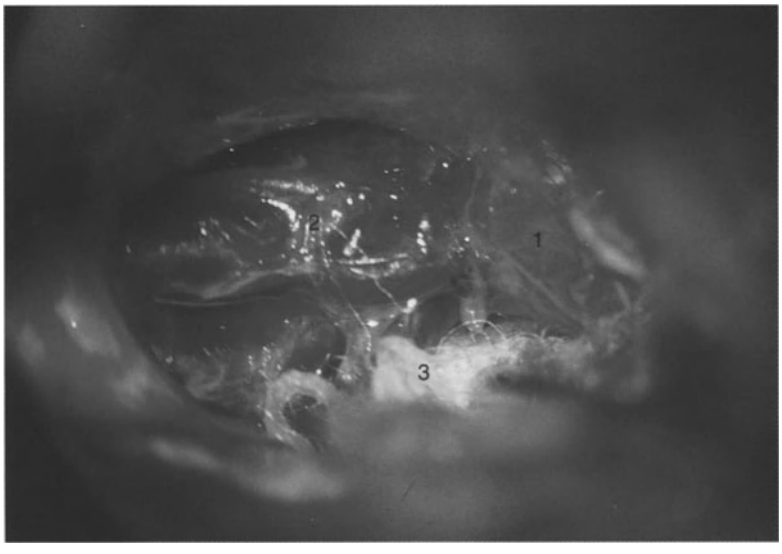


Figure 10.57. In reading this figure, review Figs. 10.52 C1 and C2. This photograph was taken through an operating microscope. The highly vascular pineal tumor has two distinctly different appearances, which were in fact expressive of a glial and teratomatous anterior component (*1*), and vascular cluster for a posterior component (*2*) (which histologically was an arteriovenous malformation). A fluffy cotton (*3*) is being used to tamponade bleeding vessels.

ter lobules of tumor extending superiorly, over the great vein of Galen, and inferiorly, wedged between the internal cerebral and basal veins. The use of cautery and microscissors must be applied in a plane parallel to the direction of the lesser (internal cerebral) and greater veins of Galen in order to avoid nicking these vessels! Similarly, laser must not be used. If one of these veins is opened, the bleeding generally signals the end of the operative procedure, because further manipulation only restarts the bleeding.

Pineal tumors which expand within the interval between the III ventricle and the body, or splenium, of the corpus callosum are characterized by extensive adhesions to surrounding structures: they are entirely within what is, anatomically, only a potential space. Therefore, one may not expect to find any free tumor surface, as when a tumor is partially or totally within either a cistern or a ventricle. This renders complete removal of pineal tumors in this location very difficult.

The tumor's confines are difficult to identify, and removing them entails entering *normal* structures repeatedly: the body of the fornix, the isthmus of the hippocampus, and the corpus callosum. If the tumor separates easily, with gentle traction, from these structures, it should be resected. If not, one does best to debulk the center, removing tiny fragments one at a time, until the impression is gained that all the tumor is removed which may be safely taken. It is not possible to have definite information concerning relative volumes of tumor resected and remaining, as when dealing with an intra III ventricular pineal tumor, until the postoperative MR scan is performed.

III Ventricle Pineal Tumors: Posterior and Inferior Occipital/Transtentorial Approach (Figs. 10.58, 10.59)

When the pineal tumor expands primarily into the region of the posterior fossa, superior to the collicular plate and anterior to the lobulus centralis and culmen monticuli, the supraculminate system is deformed as a result of posterior displacement and compression of the superior cerebellar vermis. This extension of tumor posterior to the quadrigeminal plate and anterior to superior cerebellar vermis also deforms the great vein of Galen.

An occipital flap, exposing both the superior sagittal and the transverse sinuses, provides the surgeon adequate space to perform a wide incision of the tentorium cerebelli and exposure of falx cerebri as well as the supra- and infratentorial areas. The tentorium cerebelli should be coagulated on a line which runs obliquely from the transverse sinus to the tentorial edge, using the microbipolar forceps, with the blades separated by a distance of 1 or 2 mm, and applying a drop of water so as to coagulate as completely as possible the surface of

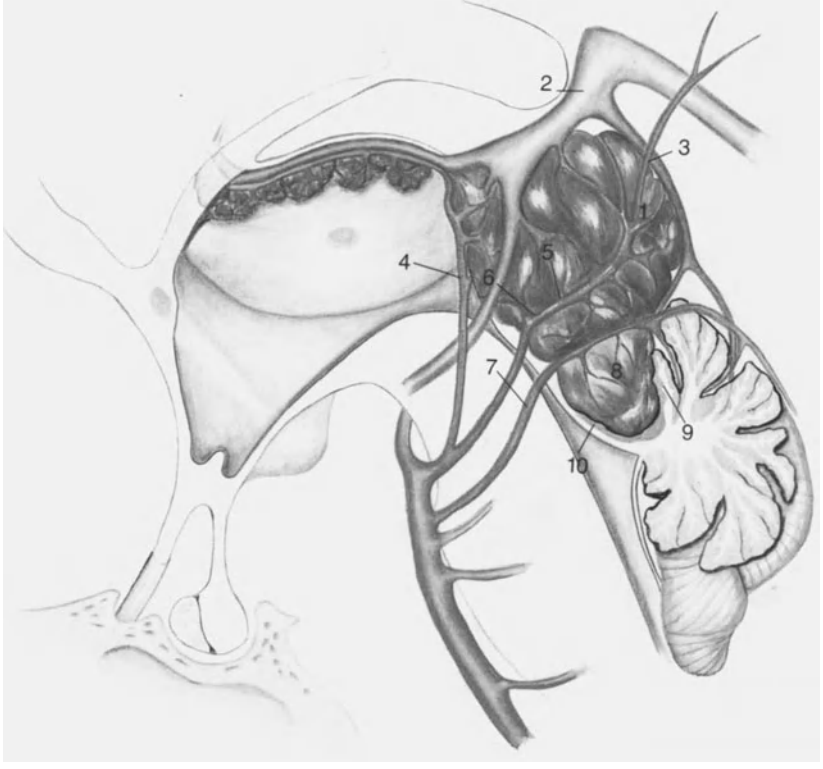
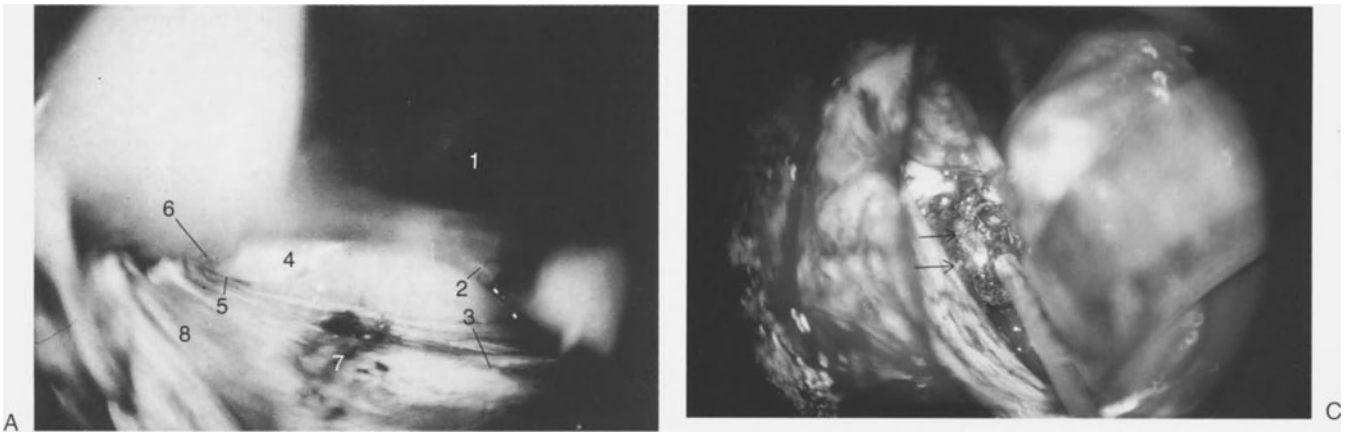


Figure 10.58. The pineal tumor (*1*) is expanding posteroinferiorly into both quadrigeminal and superior cerebellar cisterns, displacing the great vein of Galen (*2*) upward and the supraculminate vein (*3*) backward. The arterial supply comes from the medial (*4*) and lateral (*5*) posterior choroidal arteries, as well as small branches from the posterior cerebral (*6*) and superior cerebellar (*7*) arteries. The inferior pole of the tumor (*8*) often is insinuated between the lobulus centralis (*9*) and the brachium conjunctivum (*10*).



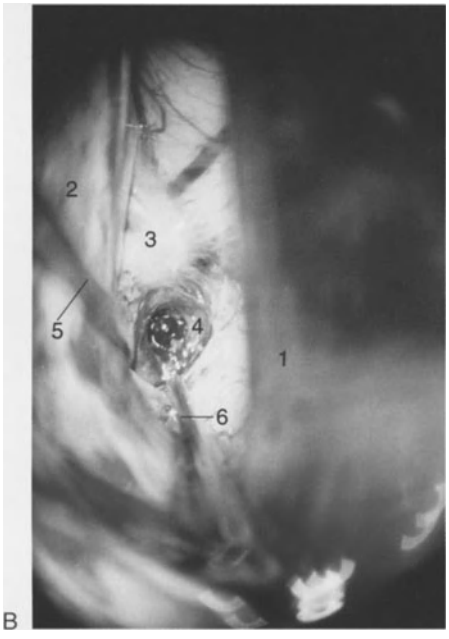


Figure 10.59. In reading this figure, review Figs. 10.52 D1 and D2. (A) The De Martel spatula (*1*) is being used to elevate the occipital lobe (*2*) from the tentorial surface (*3*), exposing the fibrosed arachnoid (*4*) at the junction of the ambient (*5*) and quadrigeminal (*6*) cisterns. Bipolar coagulation, using microforceps, was used to coagulate the surface of the tentorium along a line (*7*) approximately 4 mm from the straight sinus (*8*). (B) After the tentorium is opened, the retractor (*1*) is repositioned, so that its blade faces superolaterally, elevating the occipital lobe from the tentorium and retracting it from the falx cerebri (*2*). The fibrosed arachnoid (*3*) has been opened and the pineal tumor (*4*) within the quadrigeminal cistern exposed. A Penfield dissector (*5*) is pulling falx from the target area, and a sucker (*6*) is at the rim of the opened fibrosed arachnoid. (C) The laser is being used to vaporize the tumor in situ, minimizing retraction. One notes the neon laser (*arrow*) used as a visible tracer, since the CO_2 laser beam, well in the infrared wavelength, is invisible to the human eye. The carbonized tissue is lased blood. The Cavitron is actually superior to the laser for tumor resection.

the tentorium. The line of coagulation should not come closer than 5 mm to the straight sinus, which is centered at the midline. Remembering that the straight sinus may vary in width from 3 to 7 mm, depending upon the age and size of the child, care should be taken not to bring the line of coagulation closer than 8 mm to the midline.

If a laser is available, it should be used to cut the tentorium after coagulation, since it does this in an almost bloodless fashion, and permits one to shrink the dural tissues, avoiding the necessity to sew it away from the operative area. If a laser is not available, the tentorium should be cut in layers, using a #15 blade. The falx cerebri is opened in identically the same manner, extending from superior to inferior, from posterior to anterior, and taking care, here, not only to stay at a safe distance from the straight sinus, but also to avoid the great vein of Galen. If the laser is used, the great vein of Galen should be identified before the falx is opened so that it may be covered with Telfa or fluffy cotton. After the tentorium (and in those instances in which the surgeon chooses) the falx is (are) opened, one has complete exposure of the tumor inferior to the III ventricle and between the culmen monticuli and the quadrigeminal plate.

The splenium of the corpus callosum invariably overhangs the tumor, so that it must be elevated in order to separate neoplasm from eventual attachments to the posterior portion of the III ventricle. Debulking and medial manipulation of the lateral surfaces of the tumor, exposure of the dome, bivalving, and removal of the inferior pole, are all performed in the same order, and utilizing the same techniques, as for removal of pineal tumors within the posterior III ventricle. However, with this exposure, instead of working from below upward through the tentorial foramen, one works from above downward through an opened tentorium. The centrencephalic veins, the arterial structures, the sple-

nium, the collicular plate, the isthmus of the hippocampus must all be inspected and reinspected. At the end of the procedure, however, one does not have a complete view into the III ventricle: one's line of sight permits one to see only the posterior portion of the floor of the III ventricle and the iter to the aqueduct.

Hydrocephalus and Infratentorial Tumors

Although the posterior fossa tumor itself causes symptoms and signs, the complicating secondary hydrocephalus is often responsible for the increased intracranial pressure (ICP), thus superimposing the clinical picture of "midline syndrome" [69] (an increase in ICP without lateralizing signs) upon those signs resulting from the destructive and compressive effects of the tumor. Accordingly, the child with hydrocephalus complicating a primary brain tumor may be considered to have two distinctly different diseases which complicate one another and contribute to the complex clinical picture of increasing ICP: (1) a tumor and (2) hydrocephalus. Changes in cerebral blood flow that result from an increase in ICP and ventricular dilatation must be considered in the pathogenic cycle of events.

Neurosurgeons operating on posterior fossa tumors have long been aware of the severe complicating effects of hydrocephalus. Accordingly, many surgeons [65, 104], including Cushing [21], have recommended routine placement of a single occipital bur hole prior to opening the posterior fossa, so that the ventricular system could be punctured to decompress the enlarged ventricles. Ventricular needles were once taped to the bedside or to patient's charts during transportation, to be used for emergency ventricular drainage and intracranial decompression. The two obvious disadvantages of this procedure are uncertain decompression and increased risk of subdural bleeding secondary to rapid decompression. With the increase in diagnostic techniques that permit the preoperative diagnosis of the associated hydrocephalus, and the significant improvement in shunting procedures (III ventriculostomy by the open, Scarf, or sterotaxic, Raimondi, techniques; lateral ventricle/cisterna magna, Torkildsen, or canalization of the aqueduct, Lapras), neurosurgeons began also to suggest the insertion of a shunting device, external [105–107] or internal [10, 11, 69, 84, 108, 109], before proceeding with a craniotomy directed toward the definitive treatment of the tumor. All of these authors have reported excellent results, and many have commented on the remarkable improvement in the general condition of the child after the shunting procedure, along with a disappearance or improvement of symptoms, such as somnolence, headache, vomiting, loss of appetite, and double vision. All of these procedures have in common the shunting of CSF from one ventricle to another, from a ventricle to a basal cistern (magna or interpedundular), from the ventricle to another body chamber (blood stream, peritoneal cavity, subdural space), or externally.

Several reasons have been given in support of shunting before the definitive craniotomy: (1) false localizing signs, resulting from the associated hydrocephalus or subsequent herniation, diminish or disappear; (2) compensating the ICP by a shunt allows adequate time for stabilization of the intracranial contents, particularly cerebral blood flow, and diminution of the cerebral edema secondary to the hydrocephalus; (3) at surgery, the operative field is slack, thus easing the approach to the tumor and eliminating the need for hypertonic solutions, steroids, ventricular cannulation, or continuous spinal drainage; and (4) a smooth postoperative course is more likely.

The main theoretical disadvantage of a shunt in a child with an infratentorial tumor is that it may produce or aggravate "upward herniation." The likelihood of intracranial hypotension, which may lead to post-shunt subdural hematoma, the introduction of a foreign body as a focus for a possible infection and the possible dissemination of neoplastic cells via the shunt represent the theoretical reasons for removing a shunting device as soon as possible. Conversely, one may consider that the associated hydrocephalus may not always be secondary to simple mechanical obstruction of the CSF pathways, and that the child may indeed have permanent hydrocephalus not treatable by a ventriculostomy. The reported incidence of medulloblastoma tumor dissemination along the shunt track [108, 110, 111] has not been confirmed [7].

Of our patients with infratentorial tumors, 78.2% were found to have hydrocephalus. All of the patients with pineal-quadrigeminal tumors were hydrocephalic. The infratentorial tumors were subdivided into lesions in the cerebellar-fourth ventricle, brainstem, and clival-cerebellopontine angle regions.

Ninety-four percent of our patients with cerebellum/IV ventricle tumors suffered hydrocephalus by the time of diagnosis. Five clival cerebellopontine angle tumors were operated on. Only one patient, who had a meningioma, suffered hydrocephalus. The incidence of hydrocephalus is independent of the tumor type, benign or malignant. Rather, it is related to tumor location. The hydrocephalus was invariably symmetrical.

Regarding pathoanatomical factors complicating the surgery itself, the posterior fossa contents were reported to be "full" or tense in only 6% of the shunted children, but in 37% of an equal number not shunted.

Although the association of hydrocephalus with brain tumors is well known, very few papers have been published on its incidence, nature, natural history, and management. In 1959, Gol and McKissock [17] reported that cerebellar astrocytoma produces "clinical" hydrocephalus in 50% of tumors along the midline and in

22% of the cerebellar hemispheres. Brainstem glioma, the least common infratentorial tumor to produce hydrocephalus, has been reported in association with hydrocephalus in 20%–30% of the cases [112–114]. However, others [26, 28, 115] have reported a higher incidence of hydrocephalus associated with brainstem tumor. In 1968, Poppen and Marino [116] observed 36 cases of symmetrical hydrocephalus proven by ventriculography among 45 patients with pineal tumor. All of 19 cases with pineal tumors reported by Suzuki and Iwabuchi [42] were treated with preoperative external ventricular drainage. Cerebellar and fourth ventricular tumors have been described as invariably causing hydrocephalus [18, 20, 88].

The complicating hydrocephalus is responsible, in most instances, for the symptoms and signs that bring the child with a brain tumor to the neurosurgeon, and this increase in ICP so dominates the clinical picture that it "masks" the discrete neurological deficits resulting from tumor compression or destruction of neural centers, pathways, and nuclei. This becomes evident after a precraniotomy shunt compensates the hydrocephalus, at which time symptoms such as nystagmus, ataxia, and diplopia appear or "worsen." Persistence of headache, especially positional, Bruns' ataxia, vomiting, and pulse and respiratory changes are probably the result of mass effect, especially in midline posterior fossa tumors. It has been noted that the incidence of papilledema is proportional to the incidence of complicating hydrocephalus, and that it is present in 87% of infratentorial and 58% of supratentorial tumors in the pediatric age range [117].

It is this extraordinarily high incidence of papilledema and visual impairment, and many cases with pre- and postoperative (craniotomy) blindness or permanent visual disturbances, that puts the child at risk and justifies precraniotomy shunting to treat the hydrocephalus first. Cerebellar tumors cause papilledema in 90% of cases [17]. Cushing [21] stated, "It is the hydrocephalus that is responsible for the most serious symptoms of all from the standpoint of a useful life after a successful operation: namely, blindness." In his large series of brain tumors, among 61 patients with cerebellar medulloblastomas, impaired sight was noted at the time of admission in 26, 9 of whom were already blind. Forty of 76 patients with cerebellar astrocytoma had impaired vision at the time of admission, 22 of them having already become blind, or nearly so.

When a successful shunting procedure is performed, the improvement of papilledema is usually rapid and dramatic. Our experiences were documented by notes written by neurosurgical residents. Significant regression of papilledema was noted several days to 2 weeks after the insertion of the precraniotomy shunt. According to Duke-Elder [118], it may take as long as 2 months for papilledema to disappear (he was not reporting on postshunt children). Torkildsen [119] reported that the papilledema subsided in 5 weeks in 28% of cases with a third ventricle to cisterna magna shunt, although in some it took 3–4 months.

It is of interest to note that seizures related to posterior fossa tumors only occur in patients with hydrocephalus. Backus and Millichap [120] described a series of 165 children with infratentorial tumors, of whom 19 (12%) had seizures and hydrocephalus complicating a posterior fossa tumor. Seventy-nine percent of the 19 patients had an increase in ICP at the time of the first seizure. Wray [121] stated, "Seizures, not usually present in pineal region tumors, occurred in four of the cases with severe hydrocephalus, all of whom required a shunt procedure." Significant changes in cerebral blood flow, associated with acute obstructive hydrocephalus [122], as well as in the size and electrical characteristics of the extracellular space, changes secondary to the hydrocephalic process, likely indicate that compensating the hydrocephalus has a positive influence on control of the seizures.

"Upward herniation" is a well-known term but a poorly defined clinical picture, about which little has been written. Anatomically, this consists of anterosuperior displacement of the cerebellum and brainstem through the tentorial notch, compressing the dorsal surface of the mesencephalon, and deforming the posterior third ventricle. The veins of Rosenthal and Galen are compressed and distorted, raising the supratentorial pressure [123, 124]. Plum and Posner [125] described the clinical symptoms of "upward herniation" as progressive obtundation, hyperventilation, conjugate downward gaze, or loss of upward gaze. Rubinstein [100] theorized that the removal of a large supratentorial mass may cause upward cerebellar herniation. Emery [126] described two autopsy cases of long-standing shunts in hydrocephalus and meningomyelocele, in which there was marked upward movement of the brainstem and cerebellum, lifting of the hypothalamus, and stretching of the pituitary stalk. It has been postulated that in children with posterior fossa tumors this phenomenon is more accelerated because the posterior fossa contents tend already to prolapse through the tentorial notch. Hoffman et al. [108] described upward herniation in 6 of 96 patients (6%) who underwent precraniotomy shunting for posterior fossa tumor. They noted that symptoms usually occur 24–38 h after insertion of the precraniotomy shunt and that they are remedied by a prompt posterior fossa decompression. Their reported incidence of "upward herniation" is double that which we have seen. It is stressed that in our experiences over a 12-year period (1967–1979), with a minimum 2-year follow-up, published in 1981, 177 children with hydrocephalus complicating a posterior fossa or pineal region tumor, there were no permanent negative neurologic sequelae: the three patients with clini-

cally evident upward herniation recovered completely, the two shunt infections and four shunt malfunctions were without morbidity, the subdural hematoma was clinically silent, the onset of hemiparesis in four children and its worsening in one cleared in a matter of hours to days, and the vocal cord "paralysis" in a child with a vermian tumor was a paresis and temporary.

Endoscopic III ventriculostomy is now being used widely, so we may soon expect to read clinical reports documenting numbers of cases, results, complications. These reports are essential to evaluate before drawing comparative conclusions regarding complications encountered after the insertion of a pre-craniotomy shunt which causes hemorrhage into the tumor, although we doubt that there is a causal relationship between the two events. *Spontaneous hemorrhage into brain tumors occurs,* the majority of cases involving glioblastoma [127] or pituitary adenoma. Epstein and Mural [128] reported two cases of intratumoral hemorrhage (both posterior fossa astrocytoma) in 30 patients with precraniotomy shunts, and quite a few cases of intratumoral hemorrhage into medulloblastoma have been published [129].

Increase of cerebral blood flow and shift of brain structures after shunt insertion may play a role in intratumoral hemorrhage. There seems to be no real difference in supratentorial hydrodynamic changes resulting in subdural hematoma, whether the ventricles are decompressed by a shunt or by a posterior fossa craniotomy, with tumor removal. *The incidence of postoperative subdural hematoma was insignificantly different between these groups in our series.*

Cushing commented that, "Preliminary emptying of the cerebral ventricles is disadvantageous in one respect for it may so lower tensions in the posterior fossa that surface indications of a deep median wholly subcortical tumor are effectually concealed and may be overlooked [21]. This comment, since the advent of preoperative diagnostic procedures, must be considered only historic in value. The application of intraventricular cannulation has several problems: risk of intracranial bleeding secondary to acute decompression of ICP, danger of irreversible brain damage due to increased ICP at the time of induction of anesthesia, and unreliable [18, 21] decompression effect.

Following insertion of a shunt, the posterior fossa becomes quite slack, irrespective of whether the child is operated on in a sitting or prone position. This renders opening of the dura and separation of the tumor from the cerebellum easy and safe, and diminishes venous stasis and cerebellar prolapse.

We recommend removal of the shunt after the CSF pathway obstruction has been cleared. On occasion, we have removed the shunt directly, and at other times we have occluded it for varying periods of time before removal to be sure the child was not permanently hydrocephalic.

Despite radical resection of cerebellar tumors, with removal of the obstructing tumor from the fourth ventricle and/or the aqueduct, the patient may suffer permanent hydrocephalus from arachnoiditis secondary to subarachnoidal bleeding or local arachnoidal adhesions that were present before the operation [130–132].

Nine (20%) of 45 children with medulloblastoma required postoperative insertion of shunts in a series described by Mealy and Hall [133] and Stein et al. [134] reported that 30% of their patients with astrocytoma developed hydrocephalus in the immediate postoperative period, and 20% showed persistent postoperative hydrocephalus. None of their cerebellar astrocytoma patients had a precraniotomy shunt. In all brainstem glioma patients, once hydrocephalus develops, it is chronic, even after radiation therapy. Pineal-quadrigeminal tumors tend to produce chronic hydrocephalus. We have observed this with permanent stenosis of the aqueduct of Sylvius, meningeal adhesions, and cystic formation in the quadrigeminal cisterns, which are the main route of CSF circulation.

In summary, we conclude that precraniotomy shunting (ventriculocisternal or ventriculoperitoneal) for hydrocephalus complicating infratentorial tumors *provides the child a significant margin of safety from the negative effects of cerebral edema, papilledema, and rapid decompression of both the supra- and infratentorial compartments when the tumor is removed without previous shunting.* "Upward herniation" is a real, although very rare (3% in our series) complication. There is no statistical evidence to support the theoretical concept that "seeding" may occur along the shunting system or that this latter event increases the risk of systemic metastases from medulloblastoma or ependymoma.

Fourth Ventricular Tumors (Fig. 10.60)

The term "IV ventricular tumor" is intended to describe those tumors which occupy completely and primarily, but by no means exclusively, the IV ventricle. They generally present clinically with triventricular hydrocephalus and moderate signs of focal neurological deficit, expressive of involvement primarily of the brainstem and secondarily of the cerebellum. Consequently, preoperative insertion of a ventriculoperitoneal shunt is the treatment of choice, when the hydrocephalus is severe, since it compensates completely the hydrocephalus, relieving the child of the symptoms and signs of an increase in intracranial pressure, and allows time for the papilledema to disappear.

It is, in essence, the risk of loss of vision from the papilledema which presents the most feared complication of the hydrocephalus and cerebral edema resulting from triventricular obstructive hydrocephalus: blindness. *The dilated III ventricle compresses the optic chiasm and the edema involves optic nerves and optic pathways.*

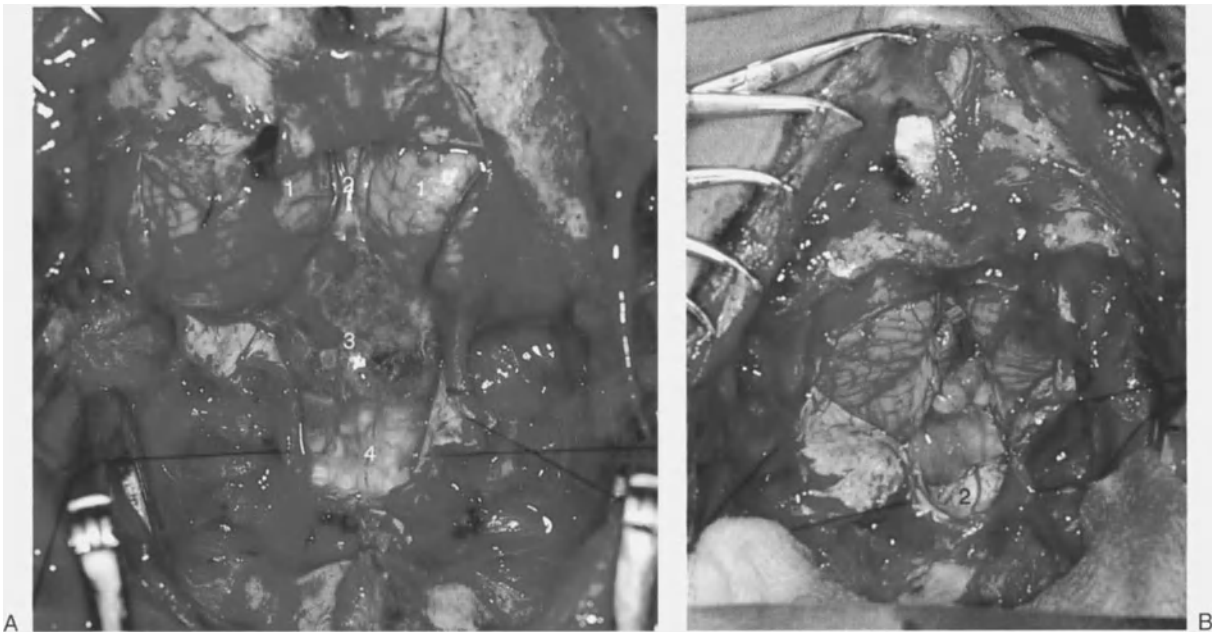


Figure 10.60. (A) A strawberry-colored, cauliflower-appearing tumor mass occupying the entire cisterna magna in a boy (age 15 years) with a choroid plexus papilloma of the IV ventricle, which invaded the entirety of the cisterna magna. The cerebellar hemispheres (*1*) are displaced laterally, and the inferior vermis (*2*) superiorly. The arachnoid of the cisterna magna is intact, permitting one to see the choroid plexus papilloma within it (*3*). The upper cervical cord, at the foramen magnum, is deformed (*4*). (B) This 11-year-old boy had persistent vomiting, but neither papilledema nor dilated lateral and III ventricles. Pneumoencephalography revealed filling of the III and lateral ventricles but not the IV ventricle. The basal and ambient cisterns were normal. Because of the unexplained persistent vomiting, the posterior fossa was explored (in 1973) and this polycystic (*1*) mass was found occupying the entirety of the IV ventricle, the vallecula, and a major portion of the cisterna magna. One notes the buckling of the superior portion of the medulla spinalis (*2*).

Acute decompression of the ventricles, which occurs when suboccipital craniotomy is performed as the primary operative procedure, may result in sudden and severe stretching on the optic nerves. This may damage permanently the already severely edematous optic pathways, with resultant blindness. And, this same sequence of events may occur if the ventricles are drained suddenly at the time of V–P shunting.

Medulloblastoma, midline glioma, ependymoma, and choroid plexus papilloma, in that order, represent the most common tumors obstructing completely the IV ventricle. The clinical history and signs, plus the neurodiagnostic procedures, allow one to distinguish brainstem glioma from the other three, so that if the neurosurgeon encounters a brainstem glioma upon opening the IV ventricle it is to be considered truly an exceptional event.

Brainstem gliomas are described in this section only to apprise the reader of their characteristics in the event they are encountered at surgery, not to recommend operating for biopsy, "debulking," or "cyst" drainage. There is no reason to conclude that imaging evidence of "isodense" areas within brainstem gliomas represent pooling of fluid within a significant cyst, one which may be cured by surgical drainage. Quite the opposite is true: *The fluid is within the intracellular compartment of the neoplastic cells, or, at most, within microcysts.* Egress of fractions of a cubic centimeter of fluid are no justification for surgery. Needling the intraparenchymal tumor, probing for fluid, with either open or stereotaxic techniques, is as likely to cause damage as to encounter a microcyst. It would be good if those who recommend surgery for brainstem gliomas were to publish results, in a controlled series, demonstrating that the operated patients do better than those treated with chemotherapy and radiation. Pre- and postoperative MRI scans are not enough.

The choroid plexus papilloma of the IV ventricle is so very rare in the age range dealt with in this book that it is included here only for completeness, since some reader may be called upon to treat one. In the adult, however, the IV ventricle choroid plexus papilloma is more common than the lateral ventricle papilloma.

One generally encounters, consequently, intraparenchymal tumors which fungate into and obstruct the IV ventricle: the medulloblastoma and the ependymoma. The ratio between the two is 3:1. It is not possible to diagnose the histological nature of IV ventricular tumors preoperatively, irrespective of the state of development of neurodiagnostic studies. Indeed, an effort to do

this by using clinical or neurodiagnostic criteria is ill-adivsed. One may, with some degree of certainty, determine whether the tumor is *primarily* in the vermis, the floor of the IV ventricle, or the cerebellar peduncles.

Fourth ventricular tumors should be removed through an inferior cerebellar triangle craniotomy. It is not necessary to remove the arch of the altas. After the dura has been opened one notes deformed cerebellar tonsils occupying the entirety of the cisterna magna in medulloblastoma and ependymoma, but the typical strawberry-colored, cauliflower appearance of tumor mass within the cisterna magna in choroid plexus papilloma.

The (exophytic) brainstem glioma fungating into the IV ventricle and cisterna magna is so very variable in growth and appearance that no general description is possible. In fact, it is not until the pedicle of the tumor has been identified, almost at the very end of the resection, that one becomes aware of the fact that one is removing a pedunculated, fungating glioma, extending out of the brainstem and into the foramen magnum.

After the arachnoid of the cisterna magna is opened and the tonsils separated, the pyramis of the vermis is elevated and the surgeon may inspect the region of the vallecula and IV ventricle. If the obex is flattened and elevated, and the floor of the IV ventricle deformed and tumefied, one may diagnose brainstem glioma and withdraw. Those gliomas characterized by a fungating tumor mass emanating from the floor of the IV ventricle, occupying its entirety and extending into the lateral recesses and the cisterna magna, take on the appearance of an astrocytoma of the cerebellar vermis or hemisphere. Occasionally, the tumor may degenerate in areas so as to give one the impression that it is a medulloblastoma.

Both the *ependymoma* and *medulloblastoma* have invasive characteristics. The former, emanating from the floor or walls of the IV ventricle, may extend into the inferior cerebellar vermis. The latter, generally but by no means invariably, emanating from the region of the posterior medullary velum at the uvula, may extend into the brainstem along the restiform bodies and/or brachia pontis. Consequently, gross inspection is unreliable in distinguishing precisely between medulloblastoma and ependymoma and, to a certain degree, the vermian glioma. Reliable frozen section is essential for precise diagnosis, though an intact floor of the IV ventricle speaks against ependymoma.

Medulloblastoma

In addition to as complete a surgical resection as possible, the surgeon undertakes:

1. To determine the benefit of chemotherapy as an adjuvant to radiation therapy and surgery
2. To evaluate the toxicity of chemotherapy
3. To evaluate the prognostic significance of the Chang staging system

If one looks at the total number of medulloblastomas diagnosed in the United States (data for 1973), taking note of the fact that 80% of them occur in children under 15 years of age, and then extrapolates from the childhood population, the median age at onset of symptoms is found to be 5 years. There are 250 medulloblastomas diagnosed annually in the United States.

The accepted form of treatment of a child with medulloblastoma (vermian tumor extending into the IV ventricle, occupying or occluding it, and invading to greater or lesser degrees the surrounding structures) is as complete a resection as possible followed by radiation therapy. This convention of following surgical treatment with radiation therapy is predicated upon the proposition that this tumor is radiosensitive and that the circulating CSF disseminates malignant cells throughout the cerebrospinal axis.

Still, with as radical a resection as possible, and radiation therapy to full biological tolerance, the tally of reported survivals has been: Chatty and Earle [134a], 12% 5-year survival among patients seen in their own institution; Bloom [134b], 32% 5-year survival for those referred to radiotherapy, 38% 5-year survival for those completing it (there had been a 25% operative mortality); Smith et al. [134c] obtained an overall 5-year survival rate of 25%, 32% if early deaths and incomplete removals were excluded; Jenkin [134d] compared patients treated between 1940 and 1952 to those between 1953 (when total CNS radiation was introduced) and 1965, reporting 5-year survival in the first group and 50% (8 of 15 completing radiation therapy) in the second if the entire CNS was irradiated.

Consequently, with results varying widely between 15% and 35% (depending upon whether all children diagnosed as having medulloblastoma were included or only those completing radiation therapy were considered), other treatment alternatives must be explored.

CCNU, vincristine (VCR), and prednisone may be used as chemotherapeutic treatment agents. The liposoluble CCNU is an attractive agent for brain tumors; VCR, although it is known not to cross the blood-brain barrier (BBB), may be used against tumors without a BBB; prednisone is added to the regimen to minimize the reactions to beginning or stopping drug treatment and to enhance the chemotherapeutic action.

Total resection of these tumors is the goal, but the reality is that this is far from always possible. Postoperative cure with radiation and/or chemotherapy is another goal, but the reality is that this is seldom possible, and the quality of survival when radiation therapy is given is discouraging and dismal, as first presented by the author in 1976, published in 1978, and con-

firmed repeatedly subsequently as late as 1994 [93]. These latter authors also report that the highest IQ scores obtained are by children who have been shunted!

Operative Technique: Medulloblastoma
(Figs. 10.61–10.64)

After the dura has been incised, its trapdoor sewn superiorly, and the inferior segment fashioned into a sling, the arachnoid of the cisterna magna is opened. Often, the surgeon will note the presence of small, seed-like deposits along the inner surface of the arachnoidal membrane. Though these may not be readily visible with the naked eye, magnification (either with surgical loops or the microscope) brings them into evidence. They represent seeding from the medulloblastoma. The cerebrospinal fluid within the cisterna magna is only of light or dense xanthochromic appearance.

The cerebellar tonsils are deformed, with a shift being present and the pyramis overlain by one tonsil or the other. Once the tonsils are separated and the pyramis split, after bipolar coagulation and transection of the vessels along the midline of the vermis, one comes upon the tumor, generally at a depth of no greater than 3 or 4 mm. The splitting of the vermis should extend superiorly to the folium, across the horizontal fissure, so as to permit adequate exposure of the superior pole of the medulloblastoma. It is safest to coagulate the inferior vermian veins on either side of the junction of tonsil, cerebellar hemisphere, and vermis because of the tendency of this tumor to swell and to bleed into its substance. This is followed by sectioning of the cerebellar vermis along the line of coagulation across the inferior vermian veins, horizontally, providing an inverted T incision in the cerebellum, with the long axis located within the vermis and the crossbar extending from one cerebellar hemisphere to the other.

The medulloblastoma most commonly grows from the nodulus, expanding within the IV ventricle. It flattens and depresses the floor, converting its surface into a concavity, and then lobulates into the aqueduct of Sylvius and through the foramen of Magendie. Occasionally, it extends through the foramina of Luschka. It seldom occupies the cisterna magna.

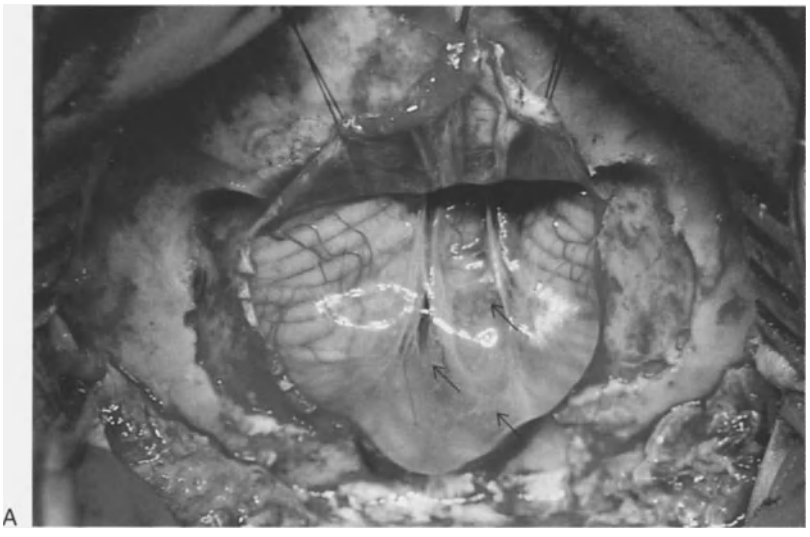
Before proceeding to dissect the dome of the tumor from the vermis, place a Telfa over the floor of the IV ventricle (under direct vision) by elevating the medulloblastoma from the floor as the Telfa is inserted with Cushing forceps. Two or three placements are generally necessary in order to bring the tip of the Telfa to the most superior portion of the IV ventricle, to the entrance of the aqueduct of Sylvius. After all of the floor of the IV ventricle, from the lateral recess on one side to the lateral recess on the other, and from the aqueduct of Sylvius superiorly to the obex inferiorly, is covered with Telfa, one may proceed with the resection of the medulloblastoma. To do so earlier exposes the floor to risk of damage.

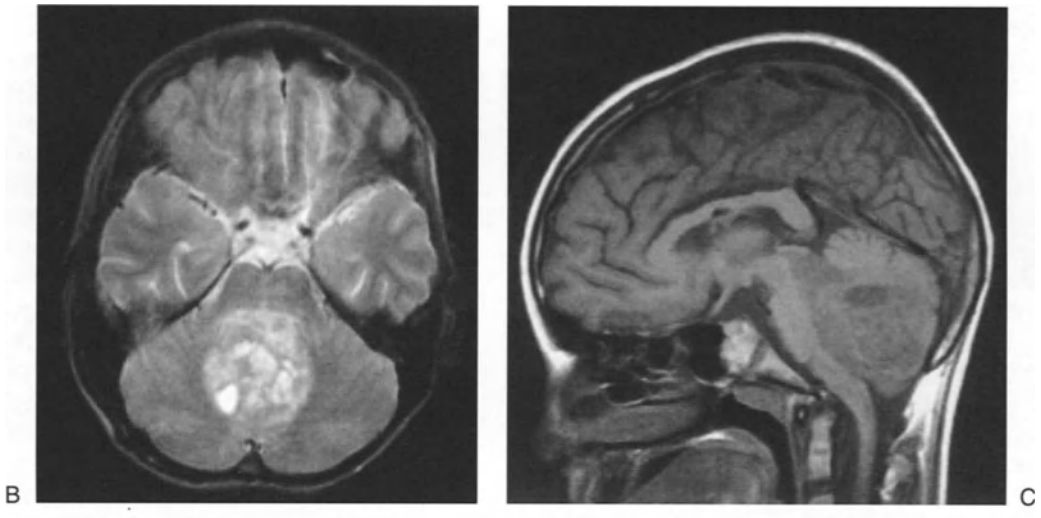
If a laser is available, the tumor may be removed by vaporization: this avoids traction. Similarly, the Cavitron permits delicate in situ tumor aspiration. A Telfa must be inserted between the inferior surface of the medulloblastoma and the floor of the IV ventricle before using the Cavitron or vaporizing the tumor. It may be necessary, because of the volume of the tumor, to insert the Telfa gradually, for distances of 3 or 4 mm, aspirating or vaporizing from the posterior surface of the tumor to the Telfa, as one proceeds. The dome or lateral surfaces of the tumor should not be removed until Telfa completely covers the floor of the IV ventricle.

The specific technique for *use of the laser to resect medulloblastoma* entails laser cerebellotomy, using a focused beam at 7–10 W, after the major surface arterial and venous structures have been coagulated with bipolar forceps.

Once medulloblastoma has been identified, the laser is used with a continuous wave, defocused beam, at approximately 12–15 W. A Telfa should be inserted along the border between cerebellum and tumor, over the posterior aspect of the medulla oblongata and the IV ventricle. A larger Telfa is placed over the cerebellar hemisphere and tonsils, with an opening cut to the size of the surface area of the exposed tumor. Vaporization of medulloblastoma proceeds quite rapidly, is almost bloodless, and results in progressive delivery of tumor into the operative field, as the surface of the tumor is vaporized. One must intermittently reposition the Telfa, taking care to look each time to identify neural and vascular structures. Particular attention should be given to identify the inferior vermian, retrotonsilar, supratonsilar, and retromedullary portions of the posterior inferior cerebellar artery (PICA), since these structures are greatly deformed by the expanding medulloblastoma. At times, tumor completely engulfs the PICA, closing this vessel within lobules. Vaporization should proceed within the center of the tumor, sweeping in broad layers rather than excavating gutters or craters.

Constant attention is given to identifying the lateral and superior surfaces of the tumor, so as to avoid perforating it and inadvertently vaporizing the surrounding neural structures. This technique permits bringing the periphery of the tumor ever closer to the center, the working area, gently taking it away from the surrounding neural structures upon which it borders or into which it extends. The tumor resection should be brought to Telfa covering the floor of the IV ventricle, with vaporization proceeding from dorsal to ventral, through the entire extent of the tumor, to this protective layer of Telfa. The laser sweeps should be continued until all visible tumor has been vaporized from the vermis, cerebellar hemisphere, tonsils, and cerebellar peduncles.





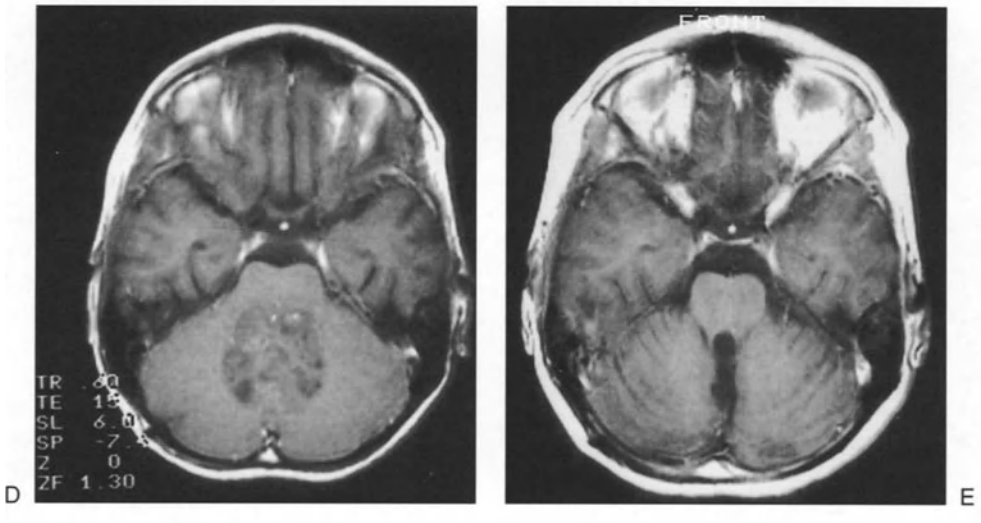
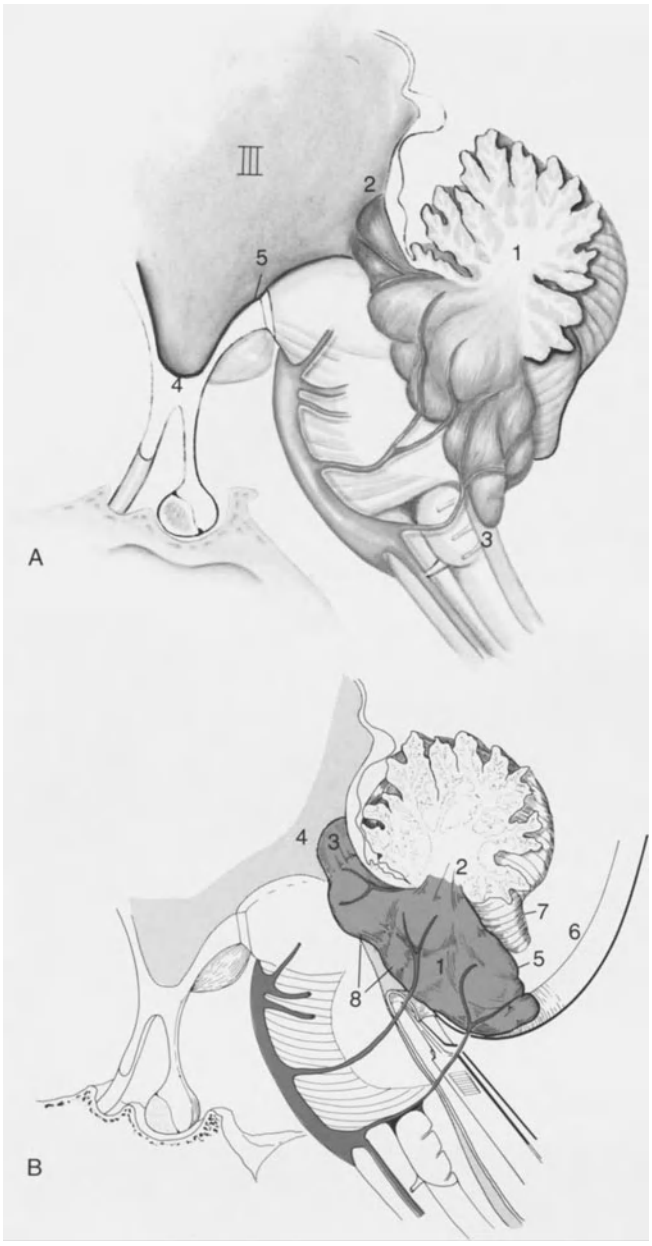


Figure 10.61. Legend see p. 259.



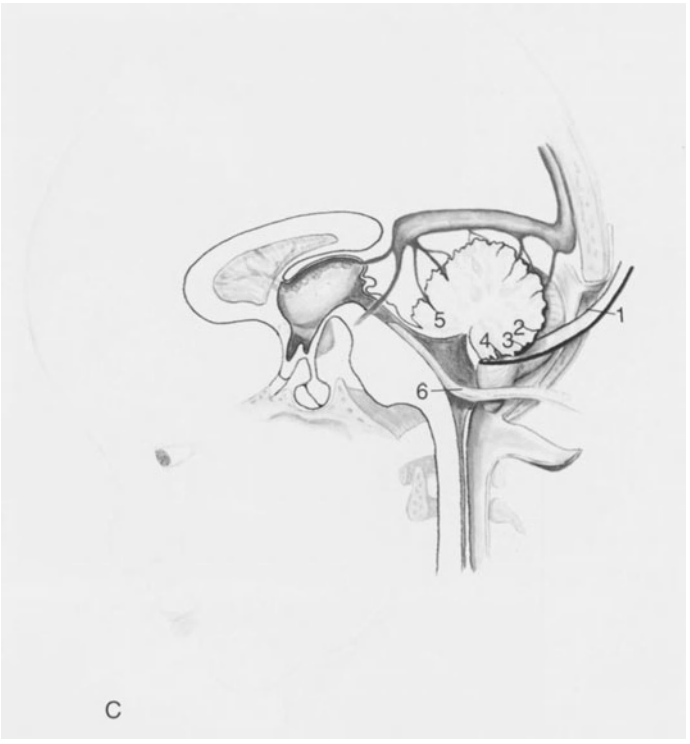


Figure 10.62. (A) Medulloblastoma growing from the region of the nodulus (*1*) fungates into the IV ventricle, often plugging both the aqueduct (*2*) and the foramen of Magendie (*3*). This results in dilation of the III, disappearance of the suprachiasmatic and infundibular recesses, and dilated III compressing both the optic chiasm (*4*) and stretching floor of the III ventricle (*5*). The gradual dilatation of the III ventricle may result in optic atrophy, secondary to compression, and hypothalamic inadequacy. Occasionally, in advanced cases, a bitemporal hemianopsia may occur and, if the hydrocephalus is not treated, blindness. (B) Medulloblastoma (*1*) growing from the region of the nodulus (*2*) the cerebellar vermis. The mass may be seen to fungate (*3*) through the aqueduct of Sylvius and into the posterior portion of the III ventricle (*4*). It has also grown through the vallecula (*5*) to occupy the cisterna magna (*6*) and displace posteriorly the tonsil (*7*). The lobular appearance of the mass, along the floor of the IV ventricle (*8*) makes it difficult for one to distinguish clinically between an ependymoma growing from the floor of the IV ventricle and medulloblastoma bulging into it. Consequently, great care and patient dissection of tumor from the floor are essential. (C) Exploration of the IV ventricle is best achieved by placing a narrow-blade spatula (*1*) beneath the pyramis (*2*) and uvula (*3*) of the cerebellar vermis, and then elevating the spatula so as to open the vallecula. The tip of the blade may then be inserted through the vallecula into the IV ventricle and angled superiorly, elevating the nodulus (*4*) along with uvula and pyramis. This brings the inferior cerebellar vermis superiorly, allowing the surgeon to inspect the inferior portion of the floor of the IV ventricle (obex and vagal triangle). Only a minimal amount of further elevation is necessary to permit inspection of the colliculus facialis, but the operating table must be rolled forward (reverse Trendelenberg maneuver) before one may inspect the aqueduct of Sylvius and the anterior medullary velum (*5*). Once the inferior and superior portions of the floor of the IV ventricle have been inspected, a Telfa (*6*) should be lain along the floor so as to protect it before proceeding to inspect the anterior medullary velum.

◄ **Figure 10.61.** (A) Meduloblastoma: The arachnoid of the cisterna magna is covered by a multitude of seedlike-appearing metastases (*single arrows*). This is common in medulloblastoma. It is not indicative of either a poor prognosis or subsequent central nervous system or peripheral metastases. In this axial image (B) one sees an ovoid mass obliterating the IV ventricle and adherent to its roof. The mass is dishomogeneous, of high signal intensity and has a necrotic area in its posterior portion. (C) The enormous dimensions medulloblastoma may assume are shown here, as is the plugging of the aqueduct of Sylvius by the superoanterior tongue of tumor. (D) This post-gadolinium study reveals very sparse foci of enhancement. (E) This post-gadolinium, early postoperative MRI scan reveals no evidence of tumor, a defect in the vermis, and a now normal IV ventricle.

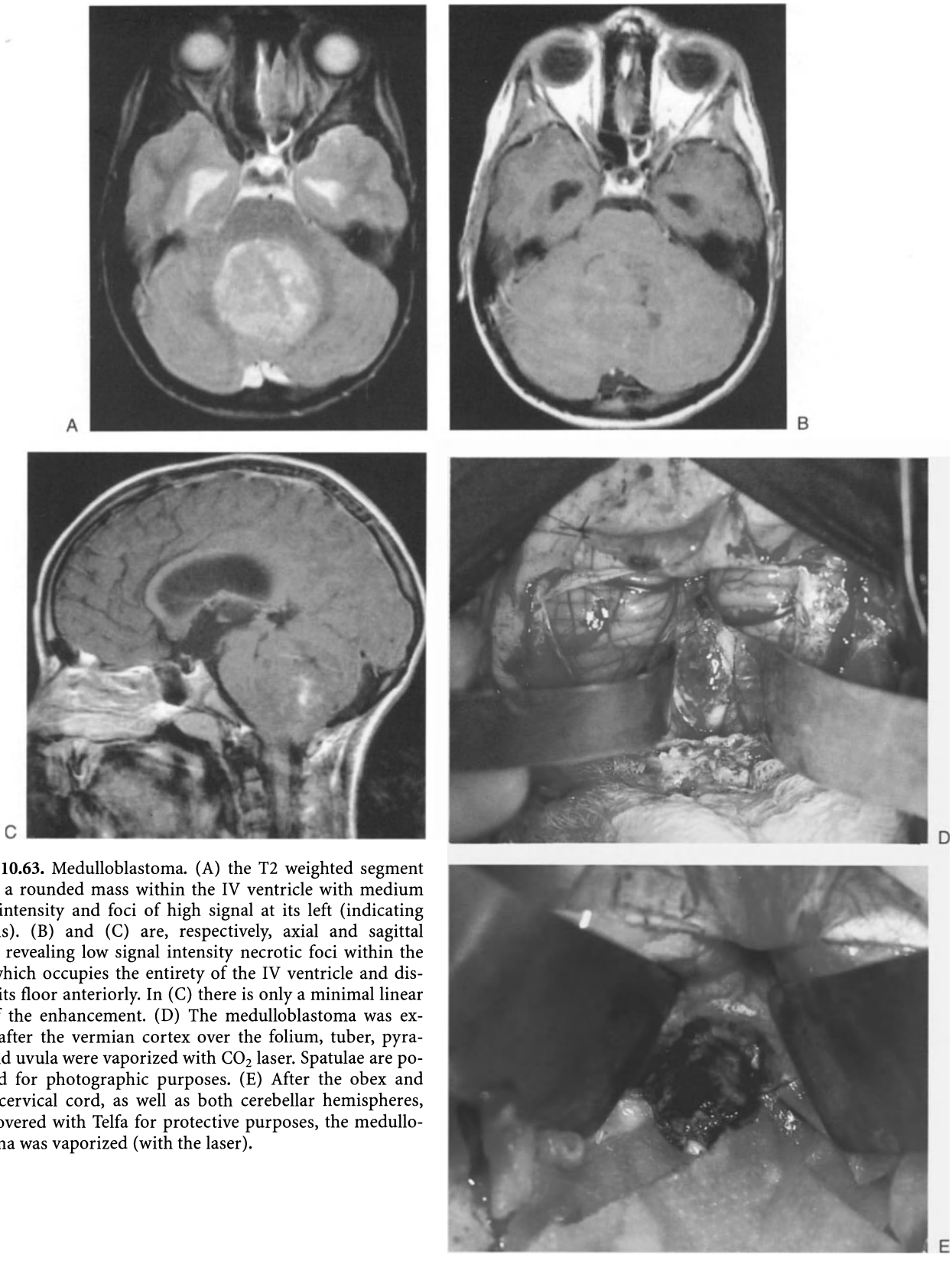
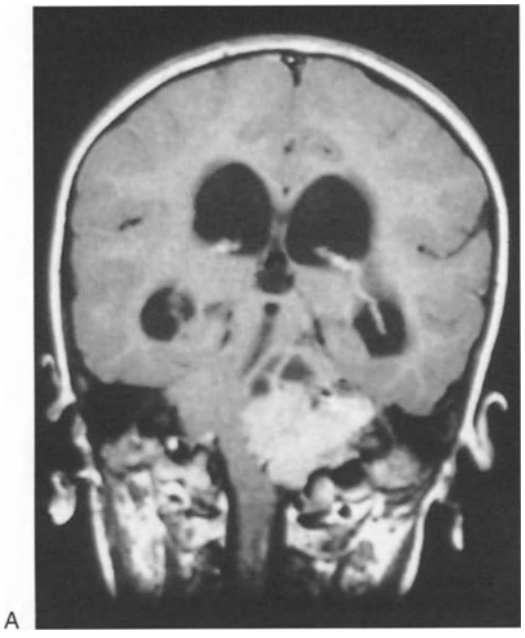
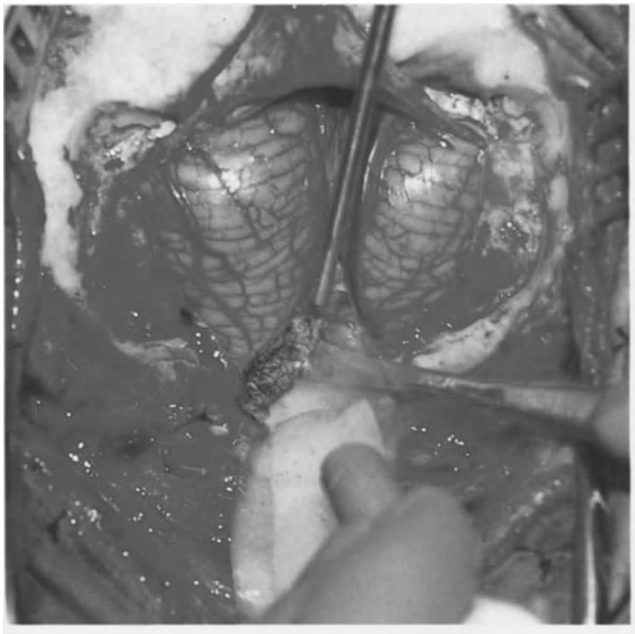


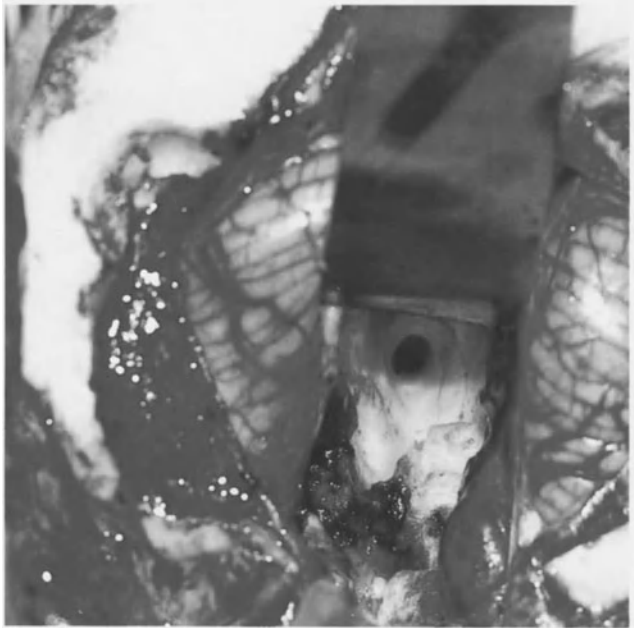
Figure 10.63. Medulloblastoma. (A) the T2 weighted segment reveals a rounded mass within the IV ventricle with medium signal intensity and foci of high signal at its left (indicating necrosis). (B) and (C) are, respectively, axial and sagittal images revealing low signal intensity necrotic foci within the mass which occupies the entirety of the IV ventricle and displaces its floor anteriorly. In (C) there is only a minimal linear area of the enhancement. (D) The medulloblastoma was exposed after the vermian cortex over the folium, tuber, pyramis, and uvula were vaporized with CO_2 laser. Spatulae are positioned for photographic purposes. (E) After the obex and upper cervical cord, as well as both cerebellar hemispheres, were covered with Telfa for protective purposes, the medulloblastoma was vaporized (with the laser).



A



B



C

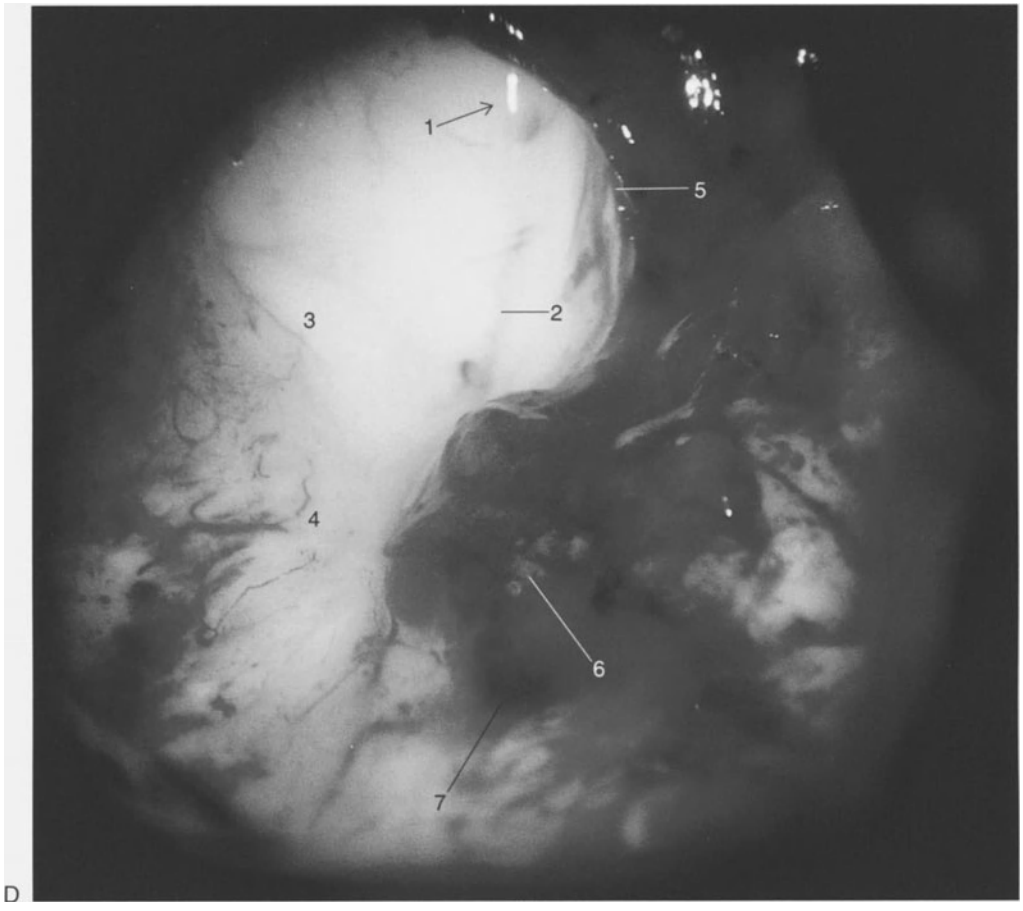
Figure 10.64. This particular medulloblastoma is interesting from two points of view: it occupies and invades the left cerebellar hemisphere and tonsil along with the brachium conjunctivum and brachium pontis; it has metastasized upstream through the ventricular system to the ependyma overlying the head of the caudate nucleus on the right. (A) This coronal post-gadolinium study reveals the polylobulated medulloblastoma within the left cerebellar hemisphere and invading the bulb and pons, via the brachium conjunctivum and the brachium pontis. Varying stages in laser vaporization of a medulloblastoma which was extending primarily into the cerebellar hemisphere and tonsil. (B) The thin layer of normal cerebellar tissue overlying tumor extension into the left cerebellar tonsil is being vaporized. Note the characteristic appearance of cerebellar surface during vaporization. (C) Almost all of the tumor has been removed, with only two small tongues persisting within the medial surfaces of the left cerebellar hemisphere and tonsil. There is no persistent vermian tumor. The floor of the IV ventricle and the remarkably dilated aqueduct of Sylvius were, respectively, flattened and plugged by the medulloblastoma. The tumor was removed from the aqueduct of Sylvius with Takahashi forceps, not with laser, to avoid inadvertent damage to the ependymal surface. (D, E) see p. 262.

Extension of tumor into brainstem is not vaporized, nor aspirated. Laser removal of a tumor from the cerebellar portions of the brachium pontis, restiform body, and the brachium conjunctivum should be performed with the micromanipulator mounted to the operating microscope. One must not extend the resection into the brainstem (medullary or pontine) portions of the cerebellar brachia!

It is not wise to use a laser to vaporize a tumor within the aqueduct of Sylvius! This is best removed using a micropituitary forceps and a Penfield #4 dissector. The same applies for any plugs of tumor that may be extending into the lateral recesses.

Having used both laser and Cavitron, we prefer the latter: it is simpler to use, but very cumbersome to hold and maneuver. The use of Telfas is identical.

After the vermian and intraventricular portions of the medulloblastoma have been removed, one proceeds to work in the retro- and lateral medullary areas, gently separating tumor from these portions of the medulla oblongata and inserting protective layers of Telfa between tumor and normal neural structures.



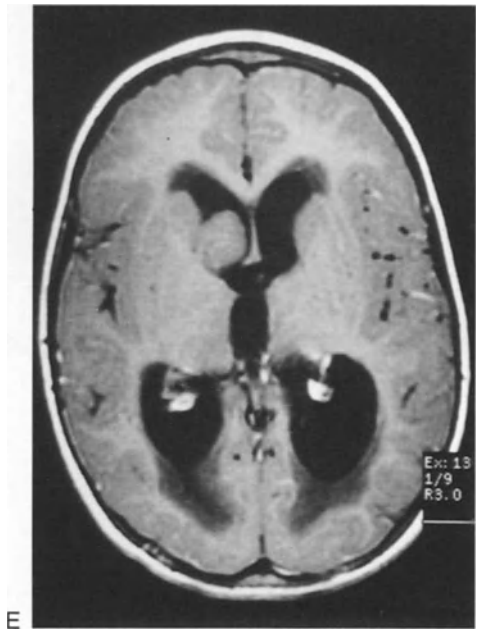


Figure 10.64. (D) After the hand-held laser unit was used for gross removal of the medulloblastoma, the unit was attached to the operating microscope and the micromanipulator used to vaporize all visible residual tumor (considered safe to remove). The aqueduct (*1*) has returned to almost normal size. The central sulcus (*2*), colliculus facialis (*3*), hypoglossal eminence (*4*), and anterior medullary velum (*5*) are all well visualized and intact. There is a miniscule amount of tumor (*6*) at the center of a small blood clot (*7*), at the entrance of the brachium pontis into the pons varolii. In (E) a large tumor nodule is seen growing from the ependymal surface at the head of the caudate nucleus *on the right*.

The hind cranial nerves should be identified along with the PICA. Often, the medulloblastoma invades the medullary surfaces of one or both cerebellar tonsils, giving the surgeon the false impression that the tumor is invading the medulla oblongata. It is best to aspirate or vaporize neoplasm from over the cranial nerves and arterial structures, rather than attempt to pull it away. This minimizes damage.

Once all of the medulloblastoma has been removed, the operating microscope may be used to scan the operative field, inspecting carefully the bed of the tumor for residual patches of neoplastic cells. These should be resected.

In the event Cavitron or laser are not available to the surgeon, the medulloblastoma is best removed *en bloc*, with the dissection beginning superiorly by inserting a spatula into the vermian cerebellotomy. The spatulae are brought over the surface of the tumor and guided forward with the sense of touch, so as to bring them within the plane separating normal from neoplastic tissue. With medulloblastoma this is quite easy, since the consistency of the normal tissue is very different from that of the tumor. The spatula may be guided over the surface of the tumor by allowing its tip to lead the way, as the surgeon holds it delicately and advances it over the dome of the tumor. From time to time, the malleable spatula should be molded to fit the curve of the dome of the tumor, allowing the surgeon to separate, as closely as possible, the cerebellum from the tumor. Long sweeps over the dome, in lateral directions, facilitate this freeing of the tumor from the cerebellum. When the child is in the sitting position this occurs readily because the tumor gradually falls into the operative field as the surgeon frees it. The cerebellum is held in place with a self-retaining retractor. Once the dome of the tumor has been freed, the sweeps are extended further laterally, freeing the tumor from the medial surface of each cerebellar hemisphere prior to attempting to free the inferior portions from the cerebellar peduncles. One will not damage the floor of the IV ventricle, since protective Telfas have already been placed. The lateral extensions of the tumor are now freed, working from below upward, ascertaining that there is no extension into brainstem portions of the restiform body or the brachium pontis. If there is, *then the spatula is passed cleanly across the neoplastic extension into the peduncles, rather than inserted into the peduncles with the intent to enucleate tumor from them.*

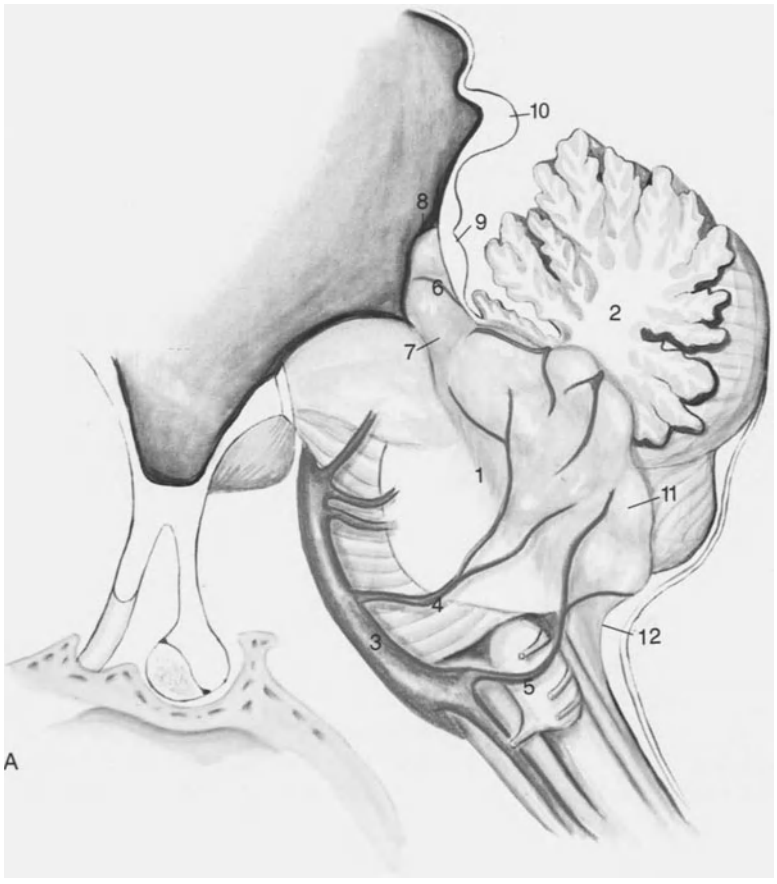
These extensions may also be coagulated and then transected at the cerebellopontine and cerebellomedullary junctions. After the tumor has been removed from the lateral extension toward the restiform body, the same technique is carried out to resect it from cerebellum to brachium pontis. This is done first on one side, then the other.

At this time the tumor mass occupies what is, apparently, the entire operative field, since it will have been freed from all of the cerebellar vermis and hemispheres. Generally, if it does not flow freely into the operative field and out of the IV ventricle, one may assume that it is being held either by strands of arachnoid and cerebellar vermis or by a plug which is extending from the IV ventricle into the aqueduct of Sylvius and, at times, up into the III ventricle. Consequently, the surgeon should inspect carefully the dome to be sure there are no vascular structures within the arachnoidal and cerebellar adhesions to the tumor, and then free the dome until he is able to look at the anterior medullary velum. When this is done, the tumor will be pulled downward by gravity and the neurosurgeon will be able to separate tumor from the anterior medullary velum. Penfield #4 dissectors may be used to tease the tumor away from the ventricular surface of the anterior medullary velum, and then out of the aqueduct of Sylvius. One may do this with impunity, since there are no vascular connections between tumor and ependyma. This will free the tumor, and deliver it from the ventricular system and out of the posterior fossa.

After the medulloblastoma has been completely removed, one inspects the IV ventricle to determine whether there have been neoplastic extensions into its walls or floor. In the event they are identified, no attempt should be made to remove them from the surface of the IV ventricle. Otherwise, the risk of damage to the IV ventricular nuclei is so great as to outweigh greatly any theoretical benefit which may be expected from removal of these last few clusters of cells. Similarly, extensions of tumor into the pontine and medullary portions of the brachium pontis and restiform body should be left in situ.

Because of the tendency of the tumor bed to bleed, it is best to irrigate the field and wait approximately 10 min to be certain that no bleeding persists before proceeding. Complete closure of the dura, in a watertight fashion, and anchoring of the bone flap back into position restores the normal anatomical relationships of muscle, bone, dura, and cerebellum. This averts cerebrospinal fluid leaking, herniation of the cerebellum into the defect, sterile meningitis, suboccipital bulging, and a definite area of weakness in skull protection of the hindbrain.

The great advantage of total resection of medulloblastoma is that it affords the child the greatest opportunity for cure*: a 42% minimum 10-year survival in our series.* There is no greater morbidity associated with radical resection. Indeed, biopsy and partial resection are associated with a greater morbidity than "total" resection. Total resection means *resection of medulloblastoma* in its entirety. When the *tumor extends into the brachia or stem total resection is not possible.*



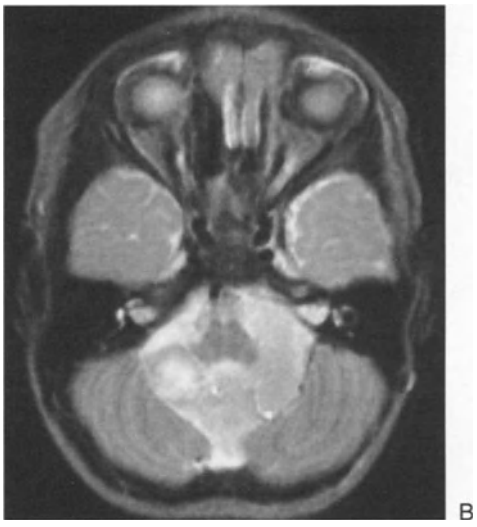


Figure 10.65. (A) The ependymoma growing from the floor of the IV ventricle (*1*) displaces the cerebellar vermis (*2*) posteriorly. It causes the same changes in the IV ventricle and its bordering structures as the medulloblastoma. The basilar artery (*3*) is practically never deformed, though AICA (*4*) and PICA (*5*) may be both stretched and bowed. A plug of tumor (*6*) insinuates itself into the aqueduct (*7*), dilating it and extending into the posterior III ventricle (*8*). The resultant triventricular hydrocephalus deforms the collicular plate (*9*) and displaces the pineal gland (*10*) posteriorly. Similarly, a plug of tumor invades the vallecula (*11*) and enters the cisterna (*12*), deforming the cerebellar vermis. With reference, then, to (A) one sees that the form of ependymoma of the posterior fossa which is considered to be most common is that of the floor of the IV ventricle, with the tumor growing from the entirety or an extended portion of either the facial or hypoglossal trigones. However, careful review of the point of origin of these tumors reveals that they may originate at the superior fovea, the medial eminence, the vestibular area, the ala cineria, the brachium conjuctivum, the brachium pontis, the restiform body, or ...within the hemispherical or vermian components of the cerebellum itself. (B) is a T2 weighted image revealing the presence of a tumor mass filling completely the IV ventricle, fungating out into the valecula through the foramen of Magendie and at both, right greater than left, foramina of Luschka. The lesion shows a diffusely medium high signal intensity with high intensity at the right side, at the junction of the restiform body and brachium pontis along the lateral recess toward the foramen of Luschka. (C, D) see p. 265.

Ependymoma (Figs. 10.65, 10.66)

The ependymoma remains a bur under the saddle of the neurosurgeon and neuropathologist. In the posterior fossa it is difficult to distinguish clinically from the medulloblastoma: the same histological appearance of lesions bordering upon the ventricular system and those embedded in cerebral parenchyma is associated with a malignant clinical course in the former and a benign course in the latter; ependymoma and medulloblastoma disseminate equally through the CSF pathways and the histologically malignant and benign ependymomas behave identically to their biological activity.

The surgical challenges presented by the midline ependymomas, bordering upon the III and IV ventricle, are considerable, since they grow into the ventricle and tend to invade the brainstem structures freely. This combination of invasiveness and extrusion into the ventricles is very likely what underlies the relatively high postoperative mortality and very low 10-year survival in children.

The higher mortality, long-term and postoperative, of posterior fossa ependymomas is probably the result of brainstem invasion. To be emphasized is the surgical pursuit of these tumors with the intent to attain a radi-

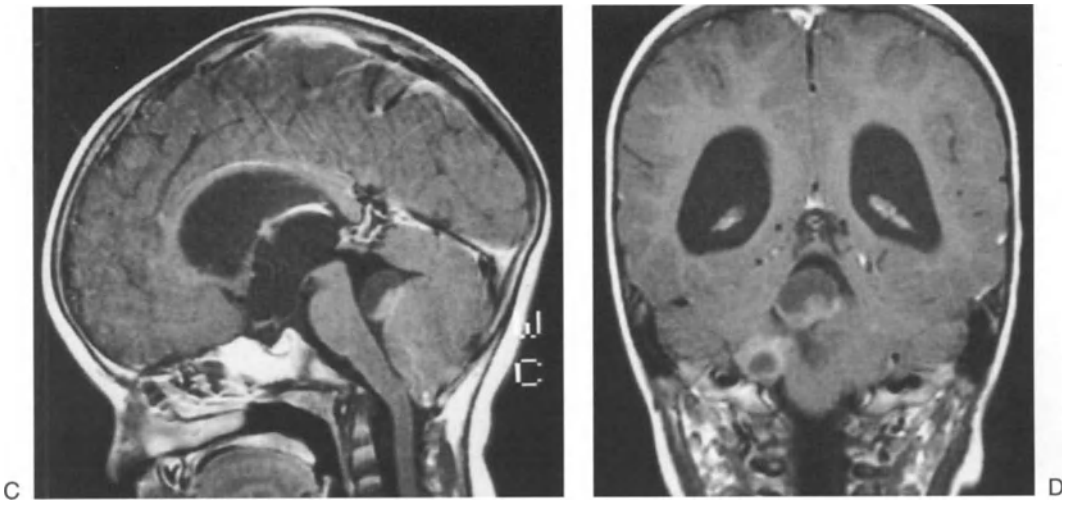


Figure 10.65. (C) The sagittal cut illustrated in (B), a post-gadolinium image, reveals the tumor to fill almost the entirety of the IV ventricle and to have its cysts at its superior extremity. The most superior portion of the IV ventricle is remarkably dilated, the collicular plate is displaced in a horizontal plane and the aqueduct of Sylvius is widely opened, producing triventricular hydrocephalus. One notes scattered foci of enhancement. The post-gadolinium image (D) reveals the dishomogeneous characteristics of this tumor, and two cystic components which enhance. Also, one may appreciate the extension of tumor through the lateral recess with a bulbous dilation at the foramen of Luschka *on the right*. Though more delicately outlined, one sees the excrescence, sessile, of non-enhancing ependymoma extending from the hypoglossal triangle *on the left*, lateralward across the ala cineria and into the vestibular area before fungating out into a partially enhancing and partially cystic mass which pushes across the midline to the right, where it is attached to the area of the stria medullaris. The brachium conjunctivum and brachium pontis are bilaterally involved.

cal resection. From what has been written by Undjian and Marinov in *The Child's Nervous System* (1990), it appears that aggressiveness is justified, for when it is successful children live longer and do not need radiation therapy. If this observation is made by the authors and elsewhere by others, one giant step forward will have been taken: a radically resected posterior fossa ependymoma recurs as rarely (or commonly) as a radically resected intracerebral ependymoma.

By and large the surface appearances of the cerebellum and cisterna magna in ependymoma are identical to medulloblastoma. Consequently, the surgeon will have no cue concerning the histological nature of the tumor until he has split the vermis and attempts to pass Telfa along the floor of the IV ventricle. It must be stated at this point that at times the medulloblastoma may be so intimately adherent to the floor of the IV ventricle – without invading it – as to give one the false initial impression that the tumor is an ependymoma growing from it. The operating microscope permits precise and safe identification of neoplastic excrescence from the floor. If tumor is growing from the floor of the IV ventricle, complete resection should not be attempted. At best, one may debulk the tumor.

The operative procedure consists of identifying the lateral borders of the tumor, separating it from normal tissue by the placement of Telfas between it and medulla oblongata where there is a free plane between it and the floor of the IV ventricle. When the Telfa has been placed, aspiration or vaporization are used to remove the tumor completely, bringing the dissection down to a plane parallel with the floor of the IV ventricle and avoiding attempts to extend the dissection below the plane of the surface of the floor of the IV ventricle. If, however, lobular extension into the aqueduct does not come out easily, with the use of a Penfield #4, it should be left in place.

The operation is considered complete when the IV ventricle is opened, permitting the surgeon to inspect the surface to see that the tumor resection has been performed flush with the surface of the floor of the IV ventricle. Irrigation of the operative field and a 10-min wait precede closure.

Choroid Plexus Papilloma (Fig. 10.60)

Choroid plexus papillomas are rare, but one of the most representative tumors in childhood. They constitute 3.9% of pediatric brain tumors and 10% of infantile brain tumors. While the IV ventricle is the common location for adult papillomas, the lateral ventricle is the more common site for pediatric papillomas. Papillomas of the III ventricle, however, are extremely rare, constituting only 0%–15% of choroid plexus papillomas.

The first surgical resection of a III ventricle papilloma was performed by Dandy [33] in 1922, but the first

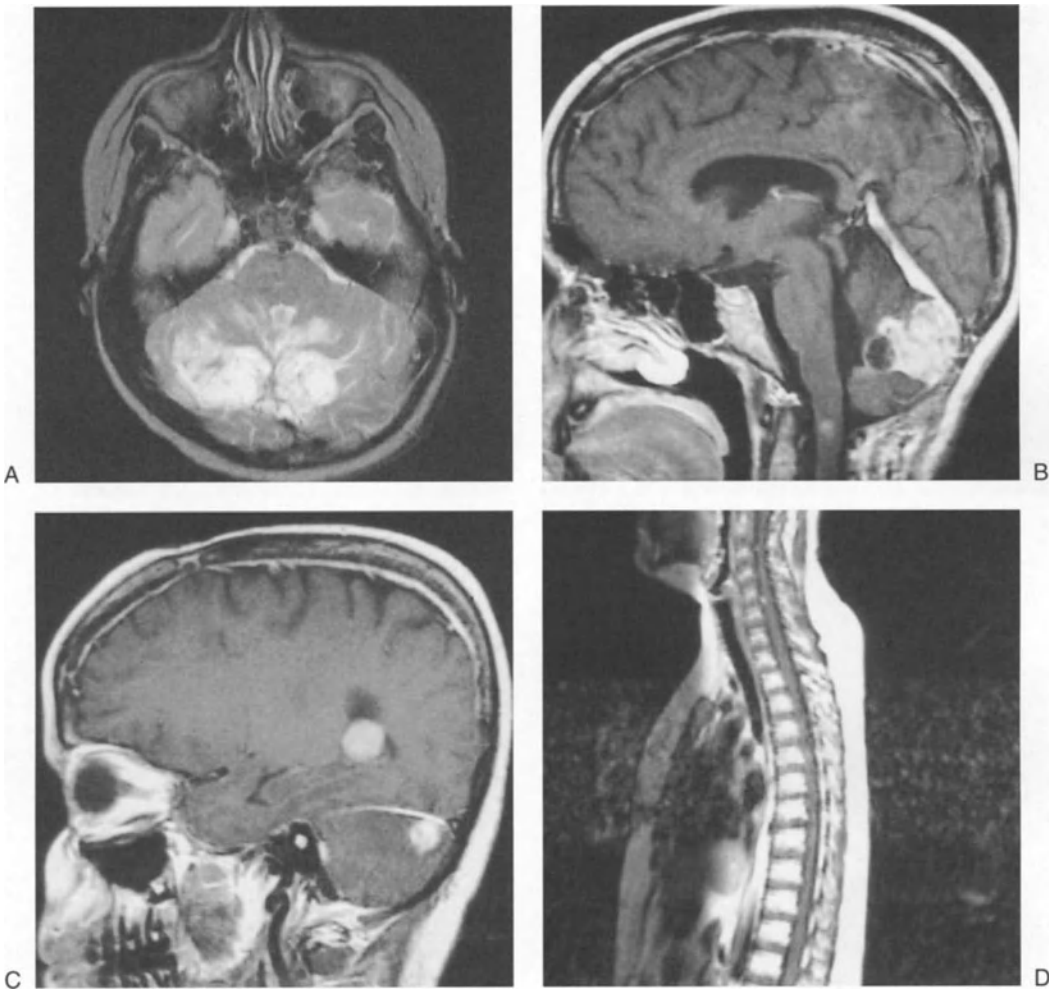


Figure 10.66. This is a recurrent ependymoma in a child who was diagnosed to have had, in addition to this tumor, a *choroid plexus papilloma* and a *neurinoma* of the VIII cranial nerve. Common things are common, and rare things rarely happen, so that most probably this is either a cerebrospinal fluid disseminated choroid plexus papilloma or ependynoma, since both of these tumors may behave in this manner and the two of them are at times quite difficult to separate one from the other histologically. In (A), an axial image, there is hyperintense signal within the cerebellar mass, which is located posterior to the IV ventricle and which enhances after gadolinium injection. Note extension of these masses, flame-like in appearance, toward the periphery of the cerebellar hemispheres. A local ependymoma recurrence is shown in (B), a postoperative study, as is a nodule within the central canal. In (C), one may identify a high signal intensity in the acoustic canal, another at the glomus of the choroid plexus, and a third attached to the cerebellar surface of the tentorium. In (D), drop metastases may be identified at the C3 and C5 levels.

successful resection was reported by Masson [2] in 1934. Since then, various reports concerning choroid plexus papillomas of the III ventricle have appeared in the literature. However, experience in a single institution is limited to only a few cases.

Even though choroid plexus papillomas of the III ventricle are histologically benign, their deep location, vascularity, and tendency to affect young infants and the presence of hydrocephalus pose significant problems in clinical management.

The presenting signs and symptoms are uniformly related to coexisting hydrocephalus. The mechanism of hydrocephalus is due to mechanical obstruction of the CSF pathways, to overproduction of CSF, or to a combination of both. Evidence of overproduction of CSF may be seen on CT images that show enlarged IV ventricle and subarachnoid cisterns distal to the obstruction at the III ventricle, as we observed in four of our patients. This is a reasonable pathognomonic sign for choroid plexus papilloma when it is seen in III ventricle tumors. Blockage of CSF absorption secondary to repetitive hemorrhage from tumor or to higher protein content in the CSF may also contribute to the occurrence of hydrocephalus. Sudden death due to acute ventricular obstruction and increased intracranial pressure as seen in a case of colloid cyst of the III ventricle has also been

reported. Thus, with hydrocephalus being the major cause of the presenting signs and symptoms of choroid plexus papilloma, it is reasonable to decompress hydrocephalus first by V–P shunting when a young child is critically ill. One should keep in mind, however, that there is often no communication between the two lateral ventricles; thus, bilateral shunt placement is needed. One problem of shunting for hydrocephalus due to choroid plexus papilloma is the potential risk of creating abdominal ascites due to the overproduction of CSF. Direct surgical resection of the papilloma would eliminate this concern related to the shunting if the condition of the patient is acceptable for craniotomy.

CT provides sufficient information with regard to tumor location and extension and to associated pathological changes. Papillomas of the III ventricle are manifest with marked contrast enhancement and an irregular caulifowerlike appearance. They are almost invariably solid, but occasionally are associated with cyst formation or calcification. Papillomas often present anterior-superior extension into the lateral ventricle cavity through the foramen of Monro, occasionally reaching the roof of the lateral ventricle. In other instances the direction of tumor growth is anteroinferiorly, where the tumor occupies distended supraoptic and infundibular recesses of the III ventricle. Posterior extension to the quadrigeminal cistern was not seen in our series, but Ausman et al. have reported a case of choroid plexus carcinoma predominantly located in the posterior III ventricle.

Postoperative subdural effusions almost always occur. These are the result of ventricular-subdural CSF fistula draining the CSF directly through the cortical opening. This complication may need drainage by means of placement of a subdural shunt. It can occur even after a transcallosal approach. This may represent active hydrocephalus, which may be externalized to the subdural space, but subdural effusion demonstrated on CT does not necessarily require shunting. Clinical evidence of neurological changes, progressive macrocephaly, or other signs indicative of increased intracranial pressure dictate whether or not a shunt is to be placed in the subdural space. Boyd and Steinbok recommend filling the lateral ventricle with physiological saline and then sewing the pia mater with 4–0 silk over a piece of Gelfoam resection. Cases treated in this fashion did not show subdural effusion postoperatively. The same authors applied a tissue glue to close the corticotomy with good results.

Once choroid plexus papilloma is removed successfully, the patient often enjoys an excellent outcome. This tumor rarely recurs or shows spontaneous malignant transformation. The role of radiation therapy for benign papillomas has been questioned for the following reasons: there is little or no incidence of recurrence after complete resection; there may be no recurrence even after subtotal resection and a high incidence (50%) of recurrence after subtotal resection and radiation therapy is reported. Therefore, one should be cautious in irradiating the developing child's brain, as III ventricle papillomas develop in particular in infancy, and successful surgical resection per se can cure the disease.

This tumor is the only true intra-IV ventricular neoplasm; the others (medulloblastoma and ependymoma) fungate into the ventricle from parenchyma. The papilloma grows from the choroid plexus, is pedunculated from terminal branches of the anterior inferior cerebellar artery (AICA) and the posterior inferior cerebellar artery (PICA), and occupies completely the IV ventricle as one polylobulated, beef-red, vascular tumor. It extends into and occupies completely the cisterna magna, draping over the dorsal aspect of the medulla oblongata.

The primary surgical considerations for IV ventricle papilloma include:

1. The invariable presence of symmetrical, obstructive, triventricular hydrocephalus superimposed upon communicating hydrocephalus, necessitating insertion of a shunt prior to removing the tumor
2. Determining whether there is an extension of tumor into the aqueduct, the cisterna magna, or the pontocerebellar angle
3. Considering the possibility of invasion of the floor of the IV ventricle and cerebellar hemisphere by (malignant) tumor.

Once the dura has been opened, the surgeon has visual evidence of a choroid plexus papilloma in that he may identify superiorly and laterally displaced cerebellar tonsils and vermis by a strawberry-colored, cauliflower-appearing, mass occupying the cisterna magna. The arachnoid over the surface is slightly opalescent, but permits excellent visualization of the tumor mass. Opening of the arachnoid allows one to see the secretory surface of the choroid plexus papilloma.

Review of cerebral angiography of the choroid plexus papilloma reveals that it is nourished primarily by the choroidal branches of the PICA and AICA at the lateral recesses of the IV ventricle! These latter vessels are the most threatening and dangerous to the surgeon if they are overlooked. It also presents the risk of one of the choroidal branches of AICA being opened and retracting into the lateral recess, out of reach of the surgeon. If, for one reason or another, adequate vertebral angiography is not available, the surgeon must assume that the primary feeders, the most dangerous feeders, are coming from the choroidal branches of the AICA at each lateral recess.

After the arachnoid has been opened the tumor will stand out clearly in the interval between medulla oblongata, cerebellar tonsils, and cerebellar vermis. The feed-

ers from the PICA may be identified along the tumor surface, and coagultaed one at a time. It is best to do this in a systematic fashion, starting inferiorly and medially on one side and working one's way around the surface of the tumor in a circumferential fashion, ending at exactly the same point on the other side. Such a 360° turn, with the tumor being gradually delivered into the field, eliminates the risk of traction and compression on the brainstem. It also diminishes the amount of bleeding.

If laser is available, it may be used at this time to vaporize (cutting factors should not be used) a groove within the circumference of the tumor, precisely at the border between tumor and surrounding cerebellum and brainstem. The greater concentration of vessels is located in the midline, immediately beneath the pyramis and nodulus of the cerebellar vermis, with the lesser concentration being located laterally along the border between tumor and tonsil. One-millimeter vessels may be coagulated with the laser, but anything larger than this necessitates bipolar cautery. In fact, bipolar cautery "runs" in the circumferential gutter should be performed with each layer of laser use, so as to coagulate and then transect the bridging vessels. Not only does this diminish considerably the bleeding, but, most helpful, it diminishes flow of blood into the tumor so that the mass gradually collapses as it is being dissected. This affords maximum protection from compression damage to the brainstem and surrounding deep cerebellar nuclei. At varying depths, ranging from 4 to 7 mm of circumferential gutter formation, one may gradually shrink the tumor with bipolar cautery as it is delivered into the operative field. This adds the third element of safety for this technique: shrinkage of tumor for maximizing neural and vascular visualization, the other two being coagulation of bridging vessels and diminution of tumor bulk by diminishing the flow of blood into the tumor. This dissection technique brings the surgeon into a position where he may first identify the floor of the IV ventricle, covering it completely with Telfa, and then begin to separate papilloma from the roof and lateral walls of the IV ventricle. The surgeon then enters the area of the lateral recesses, where he may note tongues of tumor extending from the IV ventricle through a lateral recess and into the pontocerebellar angle. It is best to coagulate one's way across the tongues of tumor, rather than attempt to deliver them into the IV ventricle. If this is tried, branches of AICA may be torn. Coagulating directly across the plane of tissue is followed by sectioning of the coagulated area with microscissors, and then freeing of the tumor from the lateral wall. This is repeated on the other side, and Telfa lain over the floor of the IV ventricle up to the aqueduct of Sylvius. If the pedicle connecting tumor to the roof of the IV ventricle and the supply from PICA have not already been coagulated and sectioned, it is done at this time and the freed tumor lifted from the field.

If laser is not available, one may equally effectively use bipolar to shrink the papilloma, microlobule at a time, *avoiding attempts to occlude the vascular pedicles and remove the tumor en bloc.* The shrinking of these tumors may be tedious and time-consuming, but it is the only technique to be used if there is no laser. Once the tumor has been shrunken, lobule at a time, the coagulated lobule is cut away, and one repeats the procedure until the entire tumor has been removed. Cavitron may be used to remove those portions of the tumor which have been shrunken into an avascular mass with bipolar coagulation.

Wait approximately 10 min to be sure all bleeding has stopped before pursuing the tongues of tumor extending through each lateral recess. Enter the recesses from the ventricular end (coagulating tumor within them), going from one side to the other. Then, the pontocerebellar recess is exposed by elevating the tonsil and proceeding over the IX, X, and XI cranial nerves to the region of the obex. Great care must be taken not to coagulate the internal auditory artery on either side, lest facial paralysis and deafness result. Another 10-min wait, to assure hemostasis, precedes dural closure and bone flap reapproximation.

Brainstem Glioma (Figs. 10.67–10.78)

In this past decade many neurosurgeons, pediatric and general, have dedicated themselves more or less completely to operating more or less radically on tumors involving the brainstem. Initially, there was trepidation, then enthusiasm, and then ever greater numbers of neurosurgeons expanded their indications for operating tumors in this anatomical location. As approaches to the skull base became surgical realities, the next step, entrance into the brainstem, was only natural from a technical point of view. The considerations regarding possibilities of effecting a surgical cure for glioma on one hand and the surgical separation of a neoplasm, voluminous or not, from such medulla oblongata structures as VIII through XII and the medial lamniscus, such pontine structures as V–VIII and the cortical spinal tracts, such mid-brain structures as III and the red nucleus along with the substantia nigra all render entrance into the stem at the very least problematic. Considering refinements in microneurosurgical technique, minimally invasive methodologies, and precision of spatial localization, the difficulties seemed surmountable. *We should do well to stop now to look at what was undertaken, what was achieved, and whether these surgical procedures, all things considered, add up to a statistically greater probability for improvement or cure than the equally dramatic refinements made both in chemotherapy and radiosurgery.*

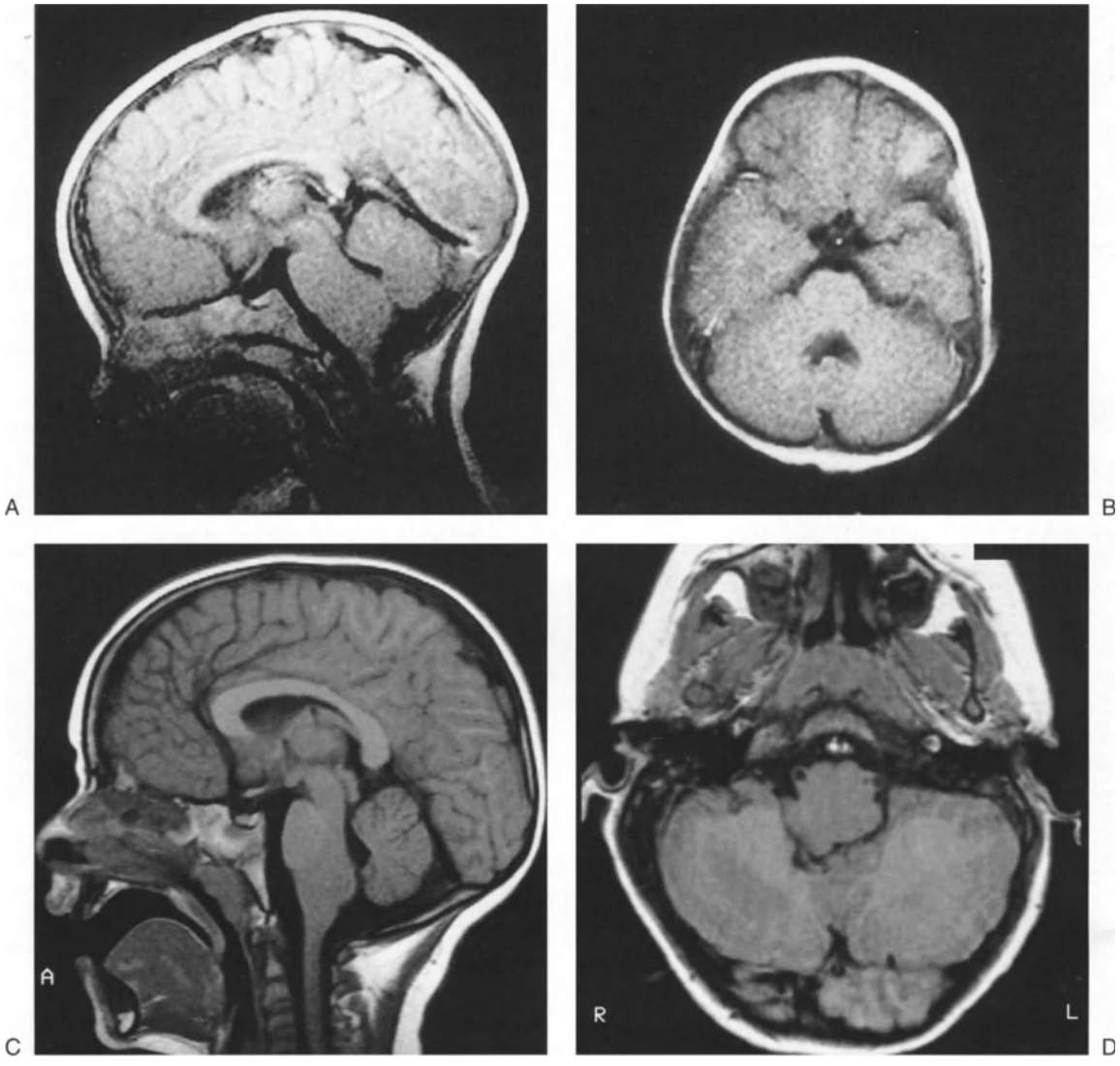


Figure 10.67. Brain stem tumor. These sagittal (A) and axial (B) pre-radiosurgery MRI studies reveal a large mass occupying the posterior pontine area and almost the entirety of the bulbar areas. The child presented with gradual, but progressive VI and VII, X and long tract deficits. Radiosurgery was considered the best approach. In (C) and (D), studies performed after three radiosurgical sittings, at 3-month intervals, in this 10-month-old child at the time of diagnosis, revealed almost total disappearance of the mass. The neurological deficits cleared steadily over approximately a 4-month period of time. (C) This sagittal T1 weighted image, 374/15, reveals, if anything, diminution in volume of the bulge posteriorly at the clava and gracilis, without any evidence of dilation of the cisterna magna, IV ventricle, or supratentorial ventricular system. (D) This T1 weighted image reveals expansion of the area of clava and gracilis with irregular diminution of the volume of the inferior portion of the IV ventricle.

It appears then that some lesions, the cavernoma or the very rare intraparenchymal brainstem cerebrospinal fluid cyst, are amenable to refined microneurosurgical techniques and that the results are so favorable as to indicate surgery; other lesions, such as diffuse gliomas of the midbrain or pons or medulla oblongata, are certainly beyond any reasonable expectation to be able to help significantly the patient. The histological nature, as in lesions of the optic pathways, when benign is equally favorable when the patient is not operated on as when surgery is performed; conversely, when malignant the results are grim. From a review of the major general and the two pediatric neurosurgical journals, one must recognize that as of this time no systematic analysis of surgical indications, results, and comparisons to such other treatments as chemotherapy and radiosurgery have appeared.

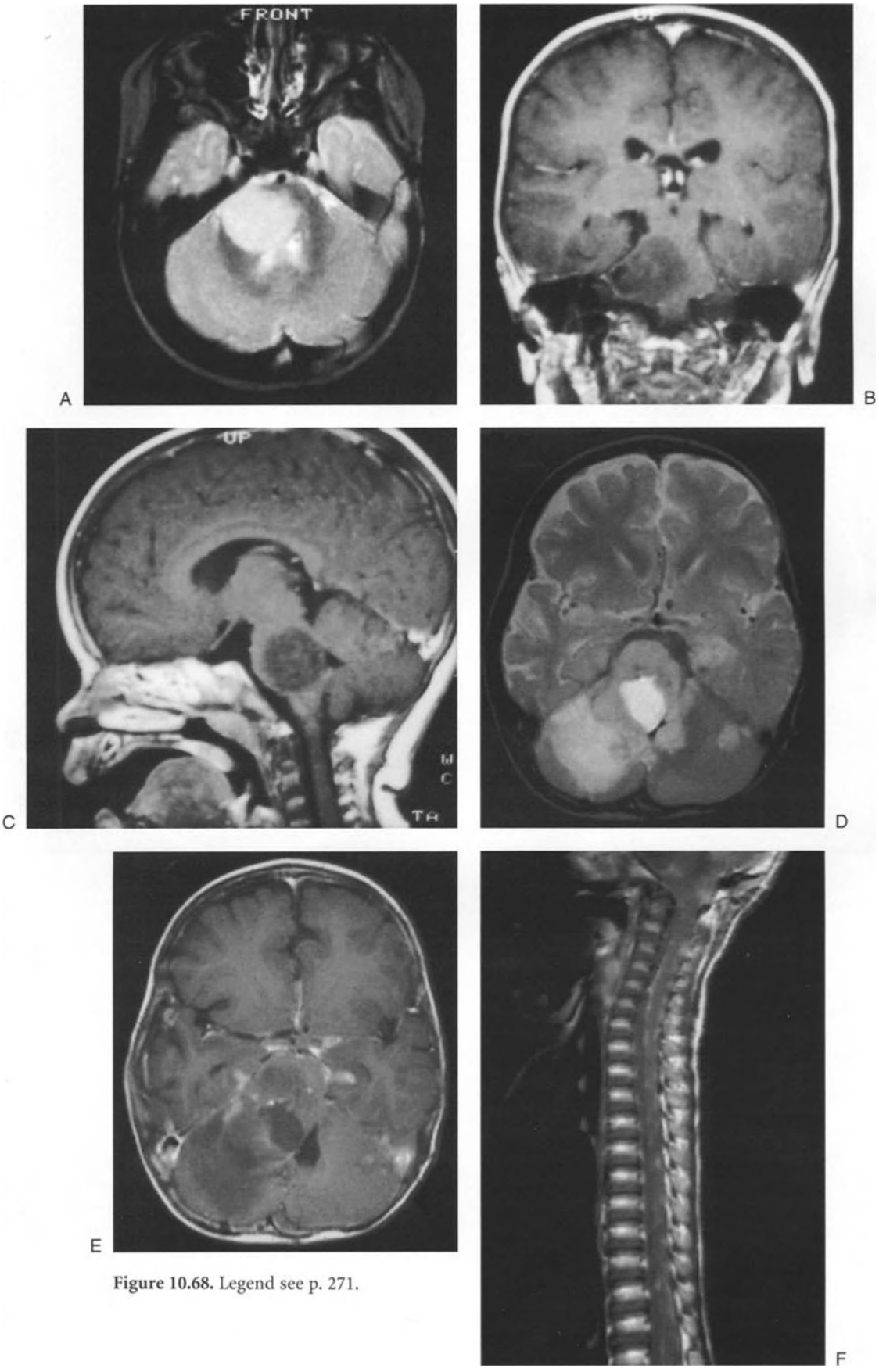


Figure 10.68. Legend see p. 271.

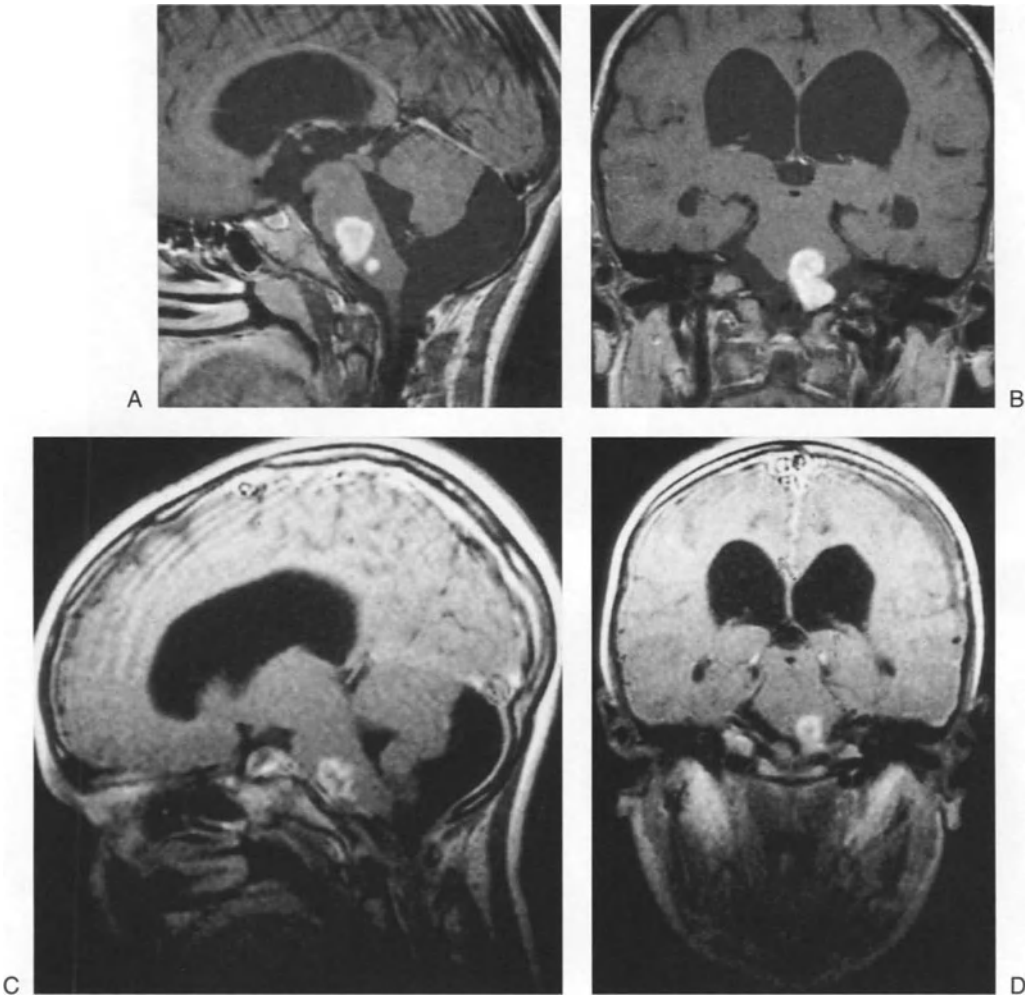


Figure 10.69. Pontine glioma: (A) this sagittal post-gadolinium study reveals the very large mass at the pontomedullary junction. Note the retrograde cerebrospinal fluid chamber dilation secondary to this partially intra-axial and partially exophytic glioma: perimedullary cistern, cisterna magna, IV ventricle, III ventricle, lateral ventricles. (B) This sagittal post-gadolinium study puts into relief the dumbbell shape of the glioma, its major portion being in the pons, and the fact that it has not obstructed passage of cerebrospinal fluid within the pontine cistern. The lateral ventricle dilation is obvious: very probably the pathogenesis of the hydrocephalus is an expression of tumor as a biophysical entity and not as a mechanical obstructive agent. (C) Postoperative sagittal and (D) coronal images revealing subtotal resection: amputation of the exophytic mass, gutting of the necrotic core. Consequently, the child was treated with radiosurgery: three single sittings at 6-month intervals. (E–H) see p. 272.

◄ **Figure 10.68.** This child had a well-circumscribed pontine tumor, ovoid in form, which surfaced at the lateral wall of the IV and VII, and a hemiparesis. Surgery was performed at another center (we considered her inoperable). She was left with bilateral VI and VII, permanent tracheostomy and gastrostomy, and bed-ridden because of a quadriplegia. Early recurrence and seeding were subsequently documented. (A) Homogeneous high signal intesity ovular mass in the right pons with extension into the right cerebellar peduncle. (B) Coronal post-gadolinium study. (C) Post-gadolinium images showing no enhancement of this homogeneous mass which is expanding within the pons. (D) Axial image performed 5 months after subtotal resection, showing necrotic hemorrhagic area of very high signal intensity and tumor regrowth. The hyperintensity in the right cerebellar hemisphere is the result of the surgical approach. (E) Axial post-gadolinium study revealing leptomeningeal enhancement secondary to subarachnoid diffusion. (F) Sagittal spine image post-gadolinium, revealing drop metastasis as high signal intensity lesions.

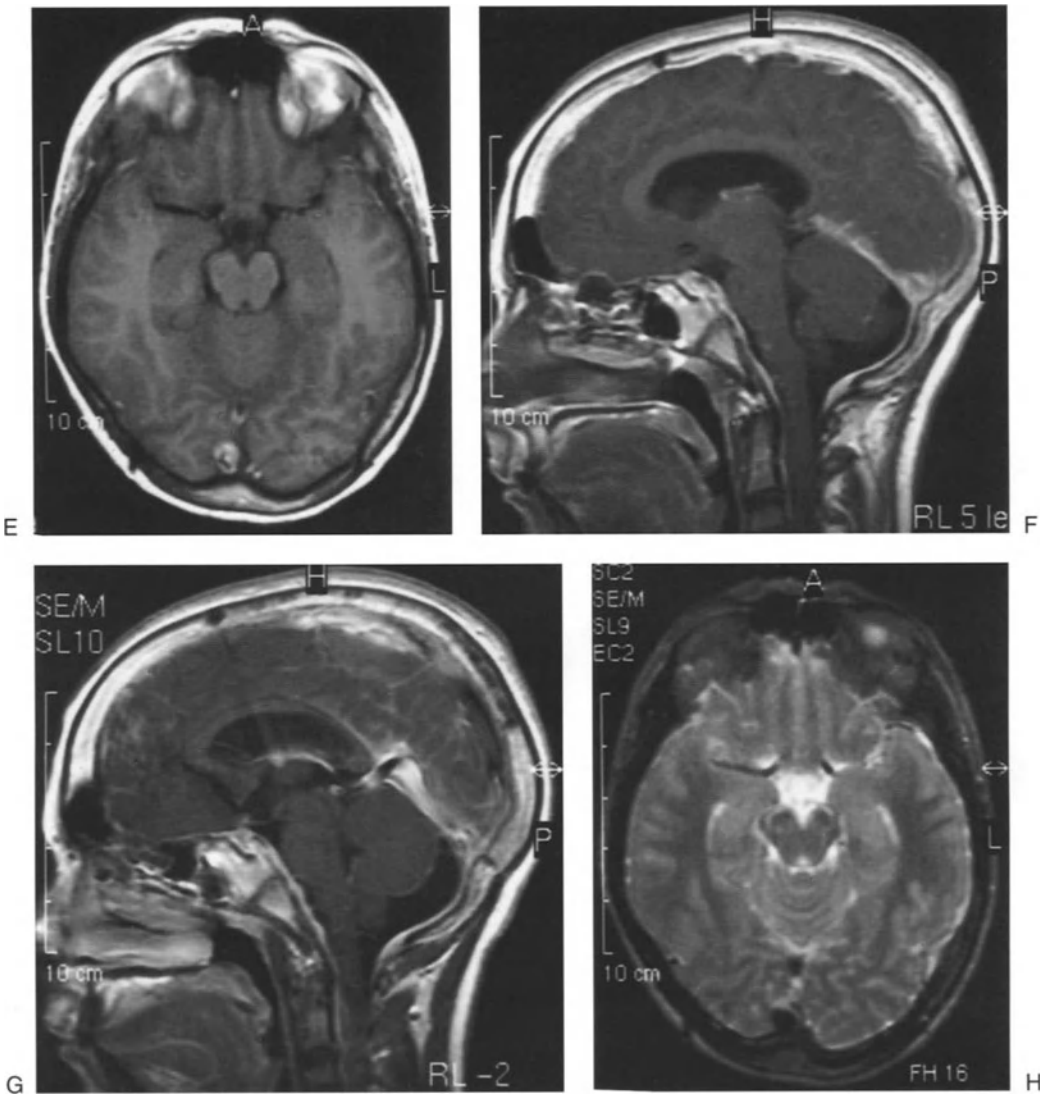


Figure 10.69. The tumor disappeared completely, as shown in (E) and (F) axial. (G) shows the 19-month post-radiosurgery gadolinium study...revealing no signs of residual tumor; and (H) is the T2 weighted image...revealing only CSF in the peritumor cavity.

If an intraparenchymal brainstem glioma, occluding the IV ventricle only because of fusiform tumefaction of the stem, is encountered, biopsy is not recommended because the complications could be severe. However, exophytic gliomas, fungating out of the restiform body or medulla, are not uncommon occurrences in childhood. This type of glioma, occluding completely the IV ventricle because of the size of the exophytic mass, is one which should be subtotally resected, using laser or sharp dissection. Such gliomas have neuroradiological characteristics which are indistinguishable from medulloblastoma or ependymoma.

After the dura has been opened, the cisterna magna is entered and the tonsils separated. This allows the surgeon to visualize the tumor as a glistening, yellowish-white mass which, when fungating, is irregular in outline, having large lobules extending into the IV ventricle, the vallecula, and the cisterna magna. It is not possible to tell immediately whether the tumor is an exophytic mass, growing from a relatively small peduncle, or whether it is a large, fusiform tumefaction of the brainstem. Consequently, dissection over the surface of the tumor should be accomplished first by separating the lateral walls of the IV ventricle from one another, in order to visualize completely the floor of the IV ventricle. If the surgeon observes that the floor of the IV ventricle is uniformly bulging into the operative field, and that he may identify such structures as the facial colliculus or hypoglossal triangle, he should withdraw at this time. If, however, he is able to identify the multilobulated excresences of an exophytic glioma, he should proceed to separate the individual lobules from cerebel-

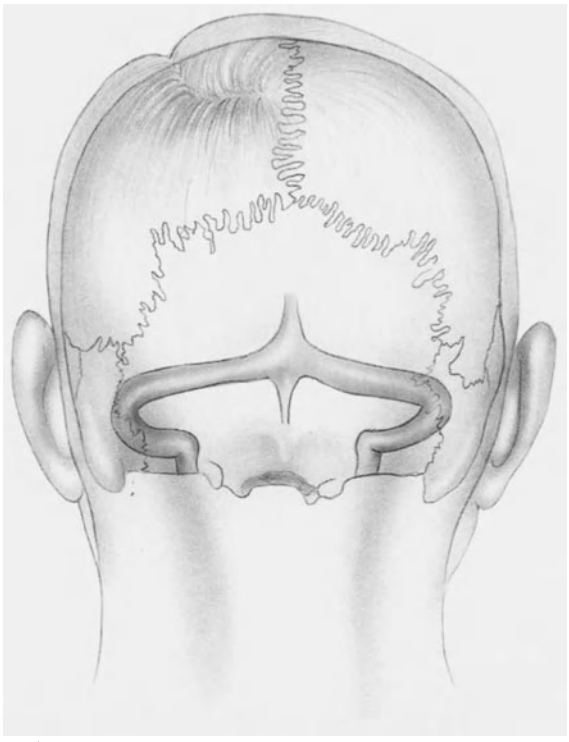


Figure 10.70. Relative positions of the transverse, sigmoid and jugular sinuses with the occipital bone, occipitomastoid sutures, and foramen magnum.

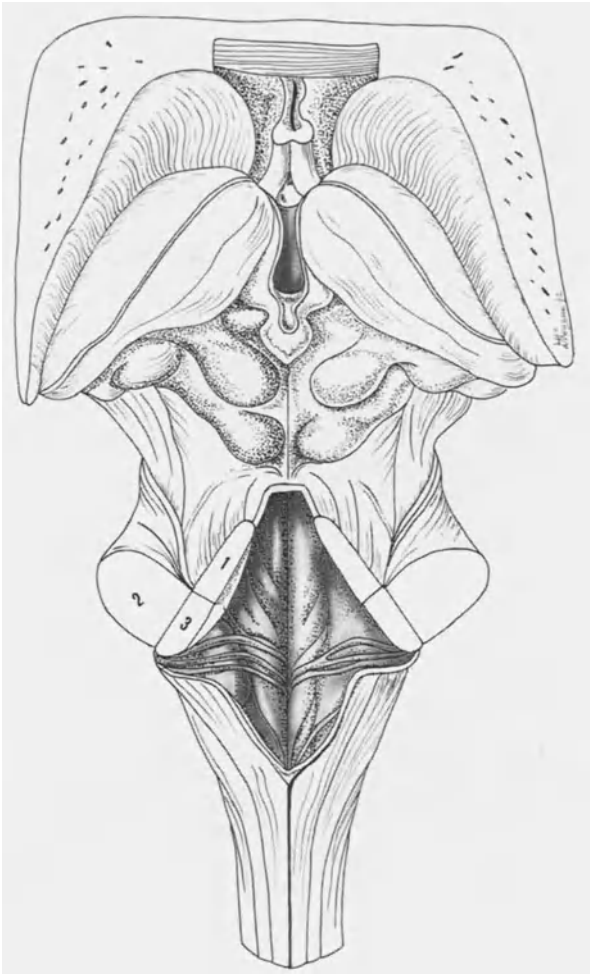


Figure 10.71. Of particular importance here are the relative positions of the brachium conjunctivum (*1*), the brachium pontis (*2*), and the restiform body (*3*). Tumor resection should not be followed into any of these cerebellar peduncles, nor into the substance of the floor of the IV ventricle.

lum and lateral surfaces of the IV ventricle. The individual lobules, once isolated, are amputated either with laser vaporization or sharp dissection. After the Telfa has been placed along the lateral surface of the wall of the IV ventricle, one must dissect the lobules from the main tumor mass to avoid severe pontomedullary damage. For this reason, laser and Cavitron are the resection instruments of choice for removal of the individual lobules, at the point where the glioma becomes exophytic, generally at the restiform body. Since these are bloodless tumors, once this point is identified, the exophytic mass may be lifted away from the brainstem by cutting across the peduncle of the tumor about 3 mm from the surface of the stem.

In the above, the reader will have noted that emphasis was given either to laser or fine dissection, without mention of the ultrasonic aspirator. Though ultrasonic aspiration has real advantages, as yet in the author's experience the disadvantages are decisive in excluding their use in some cases: the instrument handle is much too large to permit effective use and complete visualization of the field, and the tisue aspiration cannot be completely controlled at such a depth through such a small opening.

When cystic lesions have been unequivocally diagnosed preoperatively, free entrance into the brainstem is rewarded with excellent results. The same applies to cavernomas and the extraordinarily rare tuberculoma.

Low-grade gliomas may be partially (and even in some cases totally) removed, but we have not found that "the game is worth the candle": these low-grade tumors, as the high grade ones, are...responsive both to chemotherapy and radiosurgery.

To date, the most systemic appraisal of the postoperative results, long- and short-term relationship between histological characteristics and survival, significance of symptomatological progression and imaging characteristics is the paper by Behnke et al. Their observations and results are not surprising, but they are objectively expressed: 60% 2-year survival, 8% postoperative mortality, no relationship between tumor histopathology and radicality of resection, gross total tumor resection in 50% of operations, all malignant tumors were associated with a survival of less than 2 years, most children suffered a postoperative neurological deterioration, and 20% of the survivors had cavernomas.

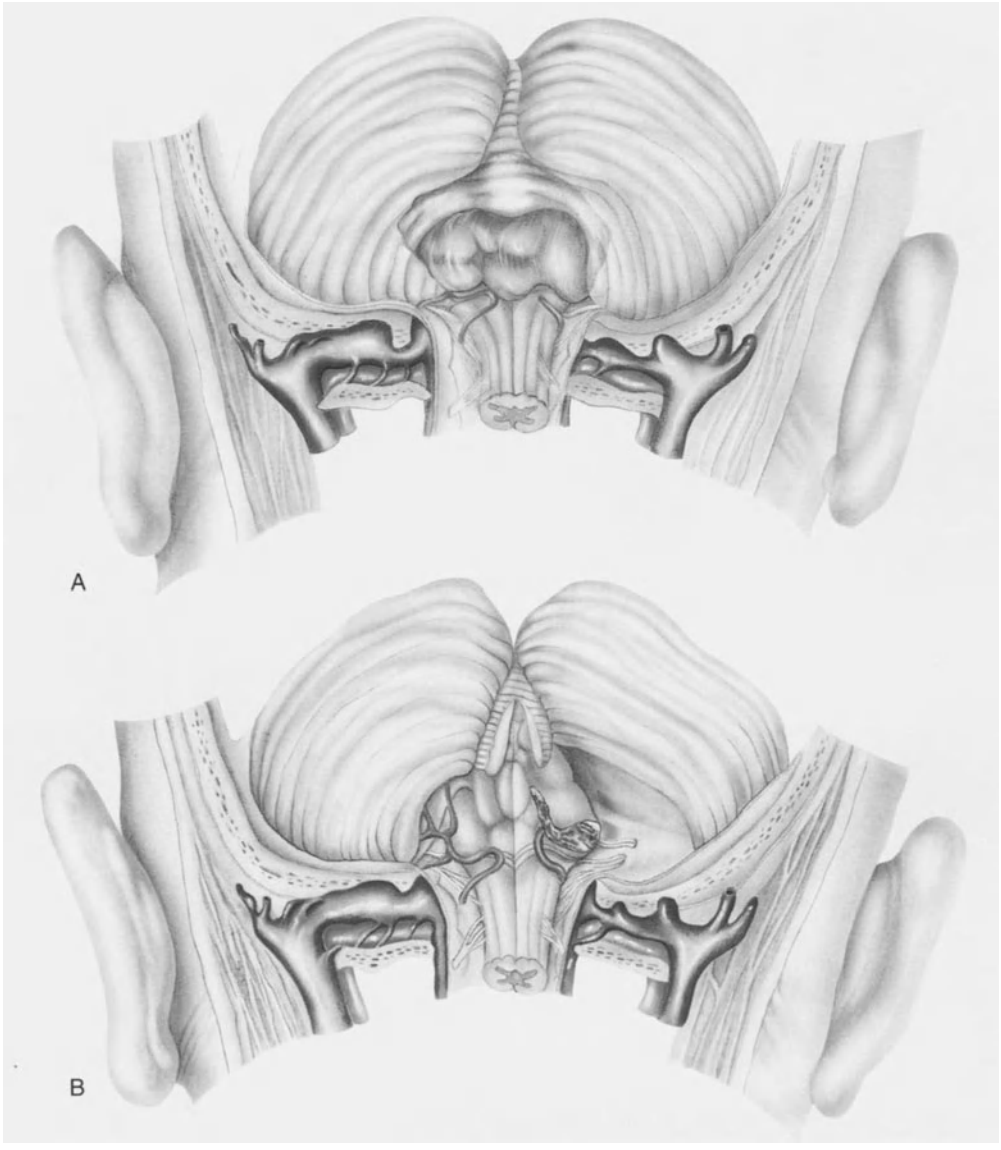


Figure 10.72. A brainstem tumor expanding into the IV ventricle through the right brachium pontis and brachium conjunctivum. In (A) the tumor is seen to bulge through the foramen of Magendie into the vallecula, invading and displacing the uvula of the vermis and the tonsils. It is important to identify the PICA early, then to resect the invaded portions of the cerebellum before beginning an intratumoral resection...which is carried out bit-by-bit, preferably with ulstrasonic aspiration, allowing the surface of the tumor to be easily separated from those adjacent structures it may not have invaded. Then, a Telfa is placed beween normal floor of the IV and collapsing tumor periphery to identify and protect it, until the point, area, of excrescence from the stem is identified. (B) Once the pedicle, pedunculated or sessile, is identified, fluffy cotton is placed around it, "collar-fashion" and then the tumor excrescence resected without entering the peduncles. If the vermis is not invaded it is better to split its inferior one-third, as in this drawing: it gives the tumor space through which it may deliver itself gradually, and diminishes indirectly IV ventricle floor compression. These two drawings assist in the understanding of anatomical relations between the vertebral artery and suboccipital veins, PICA and the hind cranial nerves.

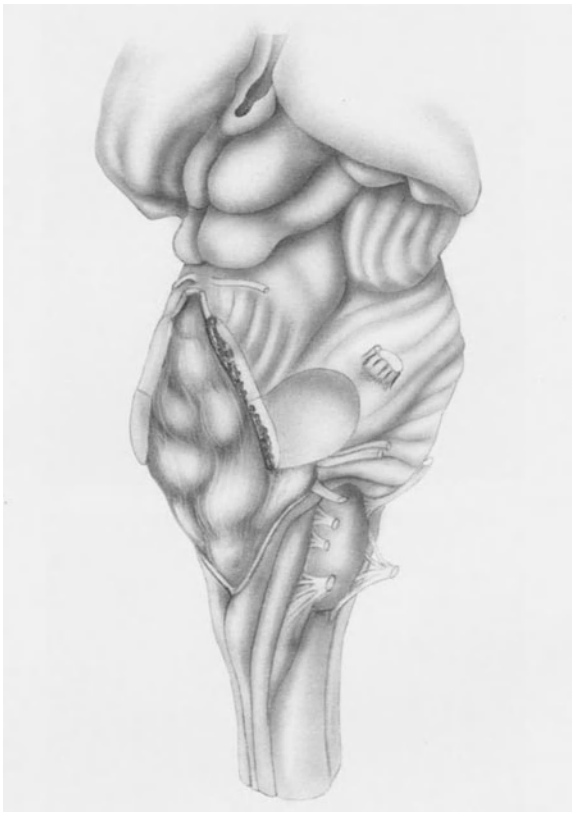
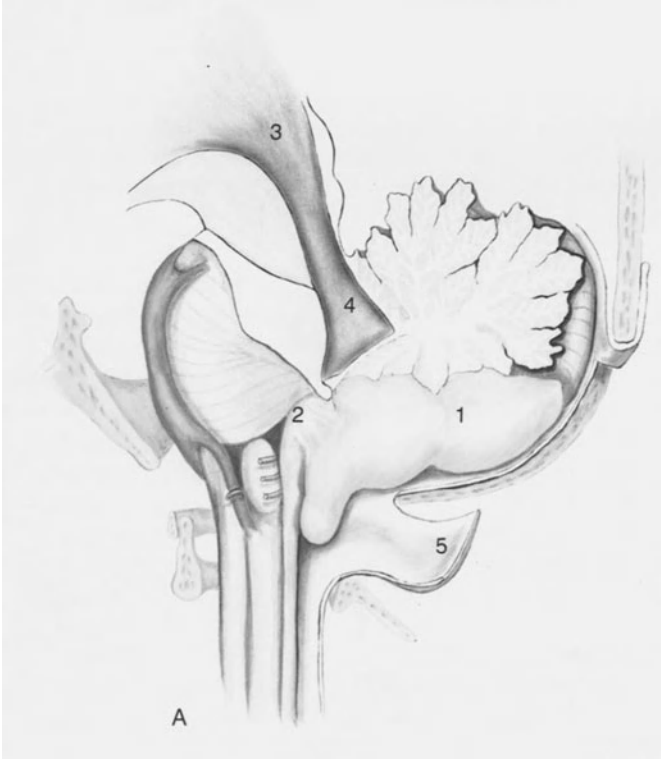
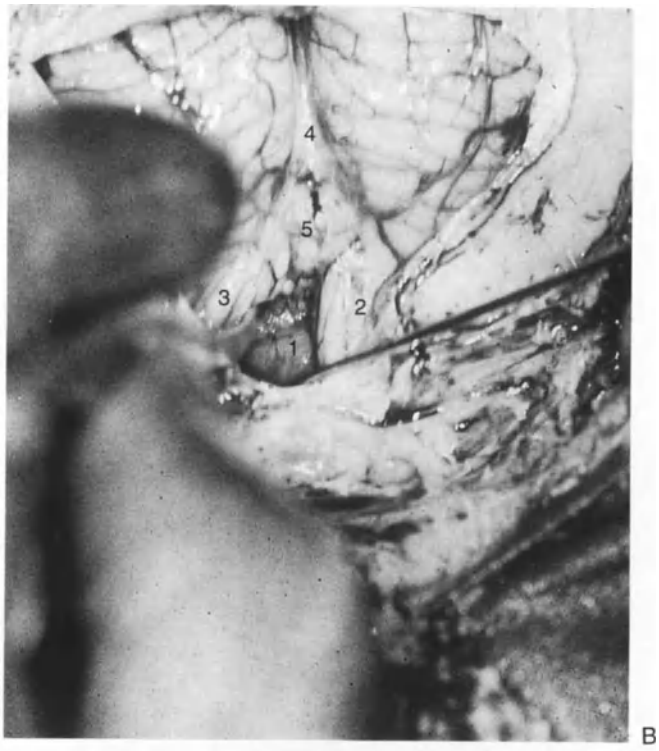
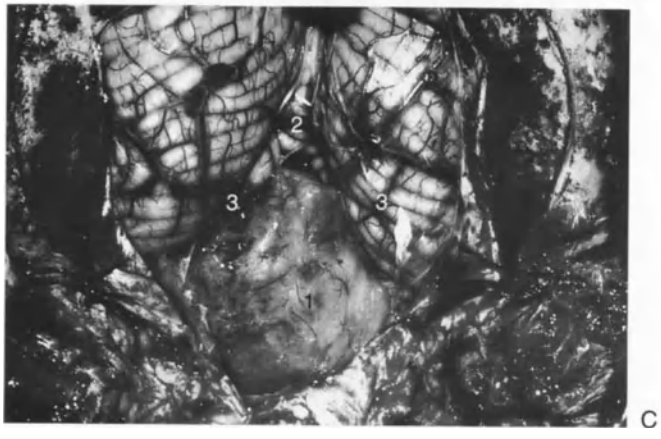


Figure 10.73. The tumor rests within the IV ventricle. Note the invasion of the brachium conjunctivum and the brachium pontis, as well as the passage of the tumor through the lateral recess and towards the pontocerebellar angle.







◀ **Figure 10.74.** (A) Exophytic brainstem glioma. The tumor (*1*) has a pedicle (*2*), from which it extends, fungating into the cisterna magna. The aqueduct (*3*) and IV ventricle (*4*) are dilated, so one may expect a gush of cerebrospinal fluid at some time during the resection. Note the sling (*5*) fashioned from the dura. This should be sewn upward and outward to provide desired support. (B) The exophytic brainstem glioma (*1*) nestles between the normally placed right tonsil (*2*) and the elevated left tonsil (*3*). The tuber (*4*) and pyramis (*5*) are elevated. This is a rather typical presentation for an exophytic brainstem glioma expanding within the vallecula and cisterna magna. (C) This giant exophytic brainstem glioma (*1*) has expanded to occupy the cisterna magna, elevate the vermis (*2*), and displace superolaterally the cerebellar hemisphere (*3*). The mass was attached to the clava by a 3-mm pedicle of neoplastic tissue, from which it was removed.

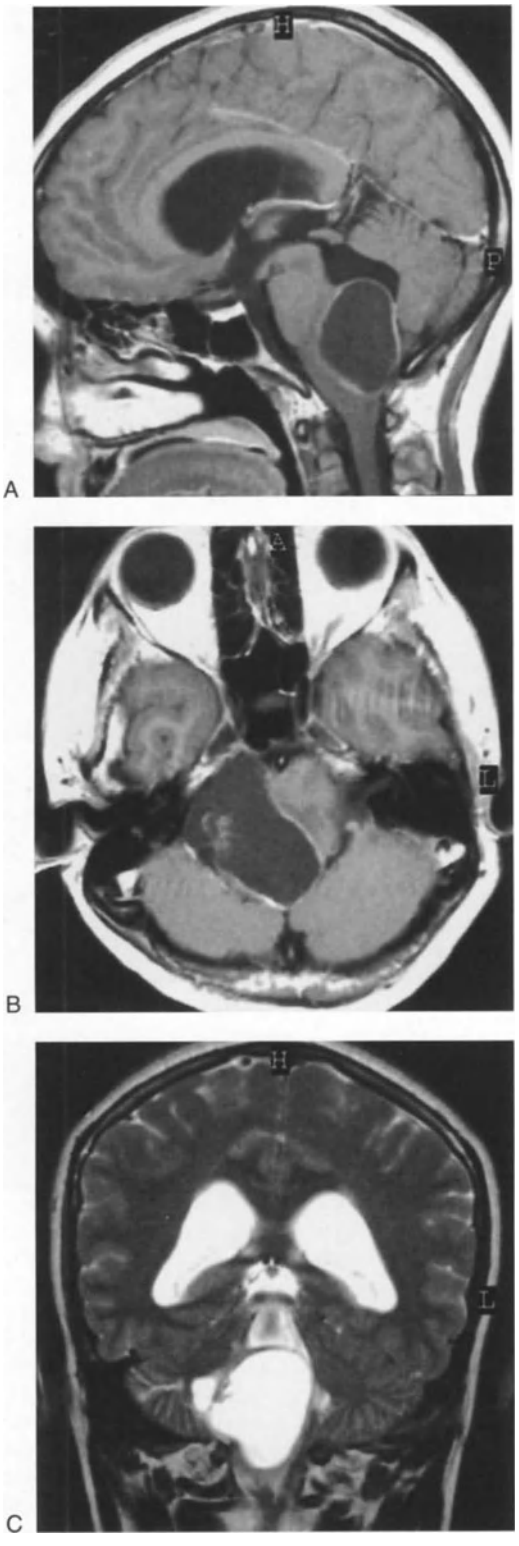


Figure 10.75. Cystic pylocitic astrocytoma involving pons and medulla. (A) is a post-gadolinium sagittal secion, (B) is a post-gadolinium axial section, and (C) is a coronal T2 image. The cystic mass is at the right foramen of Magendie, occupying also the pontocerebellar cistern, elevating the floor of the IV ventricle. There are imaging characteristics of gross calcification

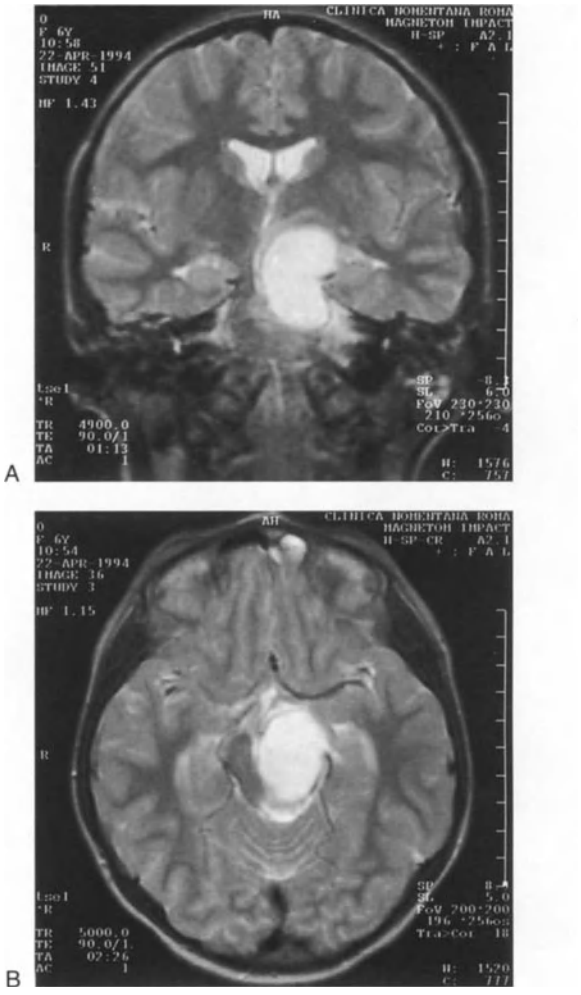


Figure 10.76. Astrocytoma (grade 2) of the mid brain. (A) This image shows a hyperintense lesion in the left cerebellar peduncle, a homogeneous signal, with infra- and supratentorial extensions. Note the wedging in the tumor caused by the tentorial edge. (B) Axial turbo spin-echo image shows expansion of the tumor within the entirety of the left cerebral peduncle.

What is most surprising is the perception of... breakthroughs, when in fact only the terminology has changed. This occurs very often throughout the literature relating to pediatric neurosurgery. For example, and pertinent to brainstem tumors, since the mid-1960s these have been identified as eminently operable lesions – providing one limits oneself to resecting the exophytic mass or emptying the cystic cavity. Lassiter et al. [25] were accepting as a foregone conclusion the fact that entering the brainstem surgically was not a technical problem, but that the resultant neurological deficit(s) was an unacceptable price to pay in the light of: (1) the limited survival time; (2) the high evidence of post- and perioperative complications; and (3) equally "good" results being reported by responsible radiotherapists first and chemotherapists later. Pierre-Kahn et al., in 1993 [135],

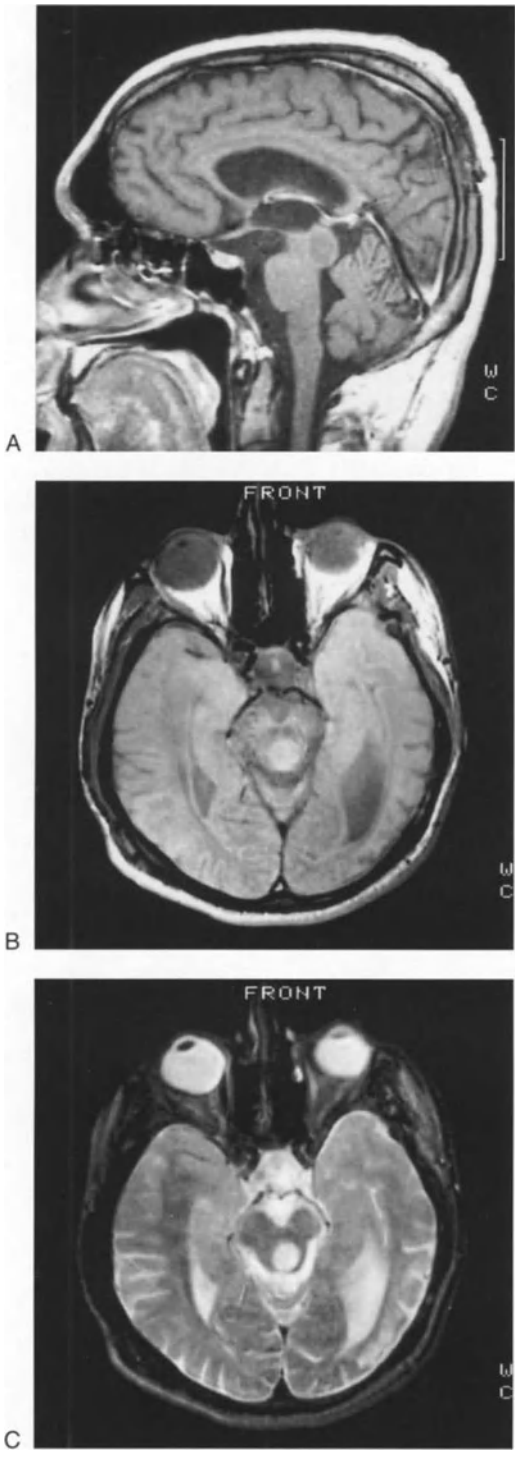


Figure 10.77. This is a tectal glioma with triventricular hydrocephalus. (A) Sagittal post-gadolinium study reveals no tumor enhancement. (A) Axial proton density study reveals the high signal intensity mass in the tectal area; (B), (C) the two T2 weighted images reveal even more clearly the high signal intensity lesion.

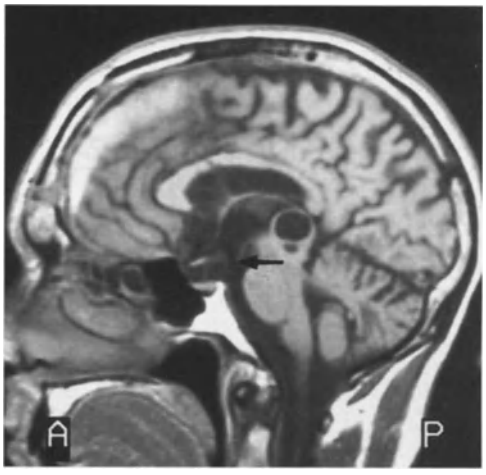


Figure 10.78. This is an adolescent with a cystic glioma of the tectal plate, presenting clinically with a midline syndrome and triventricular hydrocephalus. The treatment was surgical III ventriculostomy (*arrow* indicates the high flow area across the floor of the III ventricle: functioning III ventriculostomy).

returned to this theme, illustrating, with an excellent series of patients treated over a 20-year period, that: (1) surgery for malignant brainstem tumors was of no positive value; (2) removing the *exophytic* components of benign tumors improved survival remarkably; and (3) that if a benign tumor was totally or subtotally resected, postoperative irradiation was not necessary [100]. They, as those before them, make the clear distinction between *intra-axial* and *exophytic* tumors, with regard to postoperative results, as the basis of surgical indications: tumor "surfacing" at some point on the brainstem, and well-defined limits of tumor tissue. These remain very valid criteria, and are best expressed as such. It is confusing to extrapolate from this, suggesting that our instrumentation permits us to deal effectively with other than well-circumscribed and "surfaced" tumors.

Vermian Tumors

Irrespective of whether a vermian tumor is solid or cystic, though the former are very much more common than the latter, there is no difference either in the surgical approach or exposure of the vermis. The only anatomosurgically significant determining factor is whether vermian tumors occupy the superior or inferior cerebellar triangle, since the primary location of the tumor, above (superior triangle) or below (inferior triangle) the great horizontal fissure (horizontal line of the transverse sinus), is decisive. Superior triangle tumors are best approached through a quadrilateral free bone flap, whereas inferior triangle tumors require a (triangular) free bone flap extending across the rim of the foramen of Magnum.

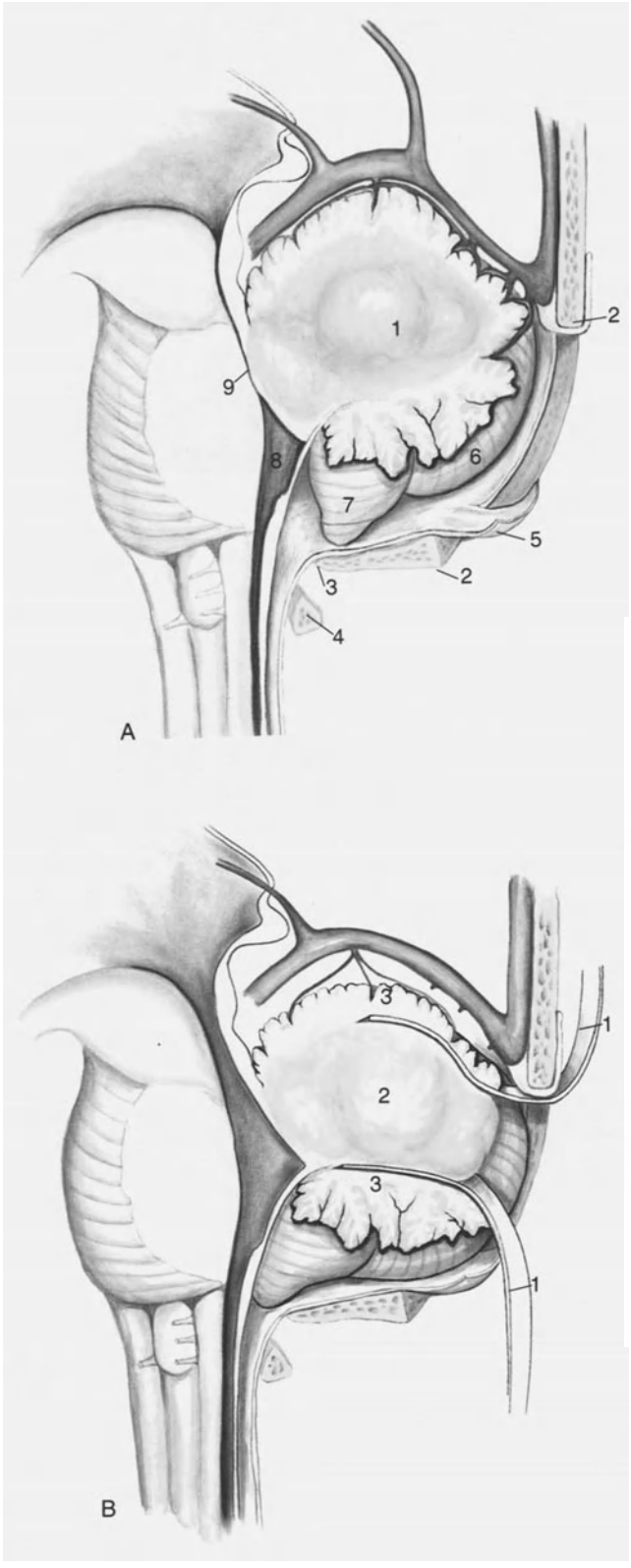
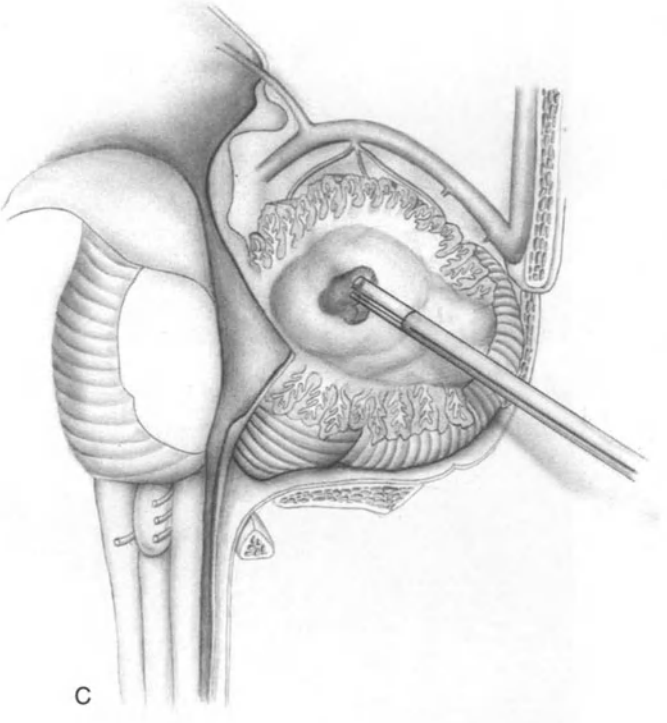


Figure 10.79. (A) Solid, superior cerebellar triangle vermian tumor. The tumor (*1*) is primarily located above the line of the transverse sinus, so a quadrilateral bone flap (*2*) is reflected, neither opening the rim of the foramen magnum (*3*) nor taking away the arch of C1 (*4*). The dura is fashioned into a sling (*5*), which supports the cerebellar hemisphere (*6*). Horizontal

Superior Triangle Tumors (Figs. 10.79–10.82)

The anatomical considerations for superior triangle tumors pertain to the superior cerebellar veins (draining into either the tentorium or the supraculminate system), the anterior medullary velum and brachium conjunctivum, and the deep cerebellar nuclei. Once the cerebellum is exposed, one has excellent visualization of the inferior cerebellar veins draining superiorly into the transverse sinus. The inferior vermian veins are readily identified, and generally observed to be remarkably deformed, splain apart from one another or both shifted to the same side. The region of the folium and tuber reveals widened cerebellar folia. Since vermian masses remain confined to the vermis, but expand into one or both cerebellar hemispheres, symmetrically or asymmetrically, the surgeon will also observe widening of hemispherical folia. It is unusual for superior ver-



squamous occipital bone supports the tonsil (*7*). The IV ventricle (*8*) may remain open, but the aqueduct (*9*) is almost always compressed. (B) If one does not have access to the Cavitron or the laser, then the much less desirable technique of using spatulae (*1*) for removal is used. They are insinuated between the tumor (*2*) and the cerebellar parenchyma (*3*), after having been bent to conform to what the surgeon estimates to be tumor contour, and then slid over the tumor so as to separate it from cerebellum. Delivering tumor through the cerebellotomy takes the compression from the anterior medullary velum and allows the IV ventricle to reexpand. (C) This half-tone drawing of a superior cerebellar triangle vermian tumor represents schematically the intratumoral technique for ultrasonic tumor aspiration: ideal for such solid tumors in such "safe" locations.

mian tumors to cause thinning of the squamous portion of the occipital bone, as commonly occurs in inferior vermian triangle tumor.

The cerebellotomy is perfomed along a vertical line extending parallel to the superior vermian veins, from the pyramis as far superiorly as the folium. Both superior vermian veins are coagulated and transected, so that they may be retracted from the operative field, after the cerebellotomy has been performed and the surface of the tumor approached, without risk of opening into them or tearing them from the transverse sinus during the dissection.

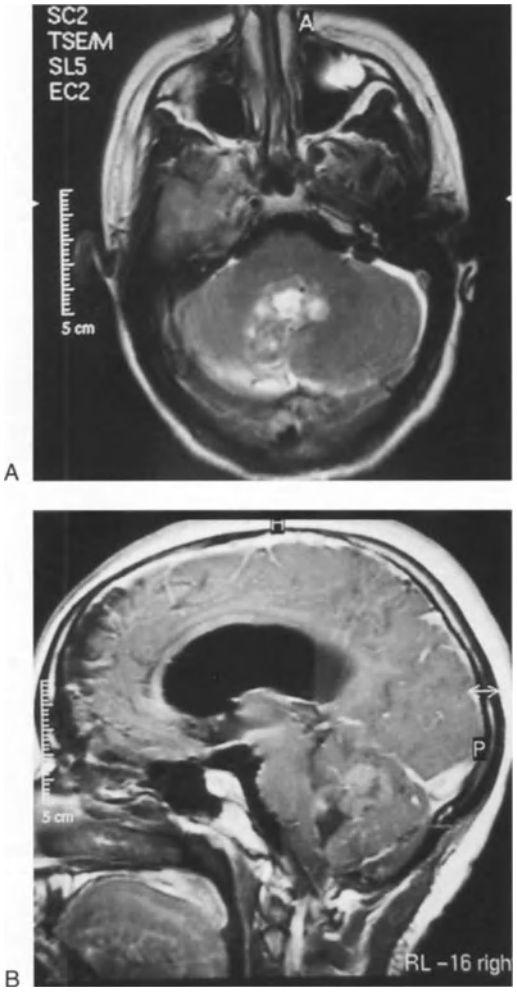


Figure 10.80. Pilocytic astrocytoma is another of the many problems which have existed since the very early classifications of brain tumors were presented: what correlation exists between the morphological classifications and the biological nature of a tumor? Recent works on this type of a tumor have, by and large, concluded that this is a benign tumor, which need not necessarily be totally resected. Some even more recent studies have reported that these conclusions may be a bit too optimistic. In (A), an axial, and (B) a sagittal postenhancement study, one sees a vermian pilocytic astrocytoma which was resected to approximately 90% of its volume, but recurred to almost twice its original size within 6 months...after the completion of three separate sittings at 3 month intervals of radiosurgery.

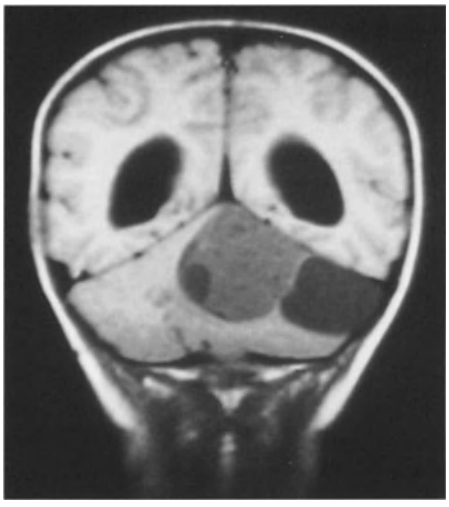


Figure 10.81. This superior vermian tumor has a very large solid component occupying the entirety of the superior vermis ...a cystic component which extends into the left cerebellar hemisphere. Thus, the more convenient approach is directly to the solid component, rather than posterolaterally into the cyst.

Once the most posteroinferior portion of the vermian tumor is encountered, one makes the determination as to whether it is solid or cystic. If it is cystic, the cyst should be punctured and the fluid drained before the tumor cavity is inspected in search of a mural nodule. If a nodule is encountered it should be removed with the Cavitron, aspirating the tumor in situ without making any effort to remove it *en bloc*. Self-retaining retractors are set one on either side of the midline, and the tips of the blades are nestled into the interval between tumor and cerebellum, directed in an horizontal plane.

These bloodless tumors may be aspirated readily. The removal is easy and precise. As tumor bulk diminishes, the retractors are continuously reset, alternating their positions between horizontal and vertical, to accommodate the outlines of the tumor. Progressive debulking of the mass results in delivery of its superior portions into the operative field, so that one need not dissect around the superior or lateral surfaces. Tumor which extends inferior to the horizontal plane, in which the surgeon is working, of course, will not "well up" into the operative field. Dissection of this portion necessitates elevating it into the operative field with the use of either tumor or micropituitary forceps. Suspending the tumor with one of these instruments allows the surgeon to use the Cavitron to aspirate its inferior pole, freeing small lobules one at a time. Attention is given to orientation, to avoid entering the region of the deep cer-

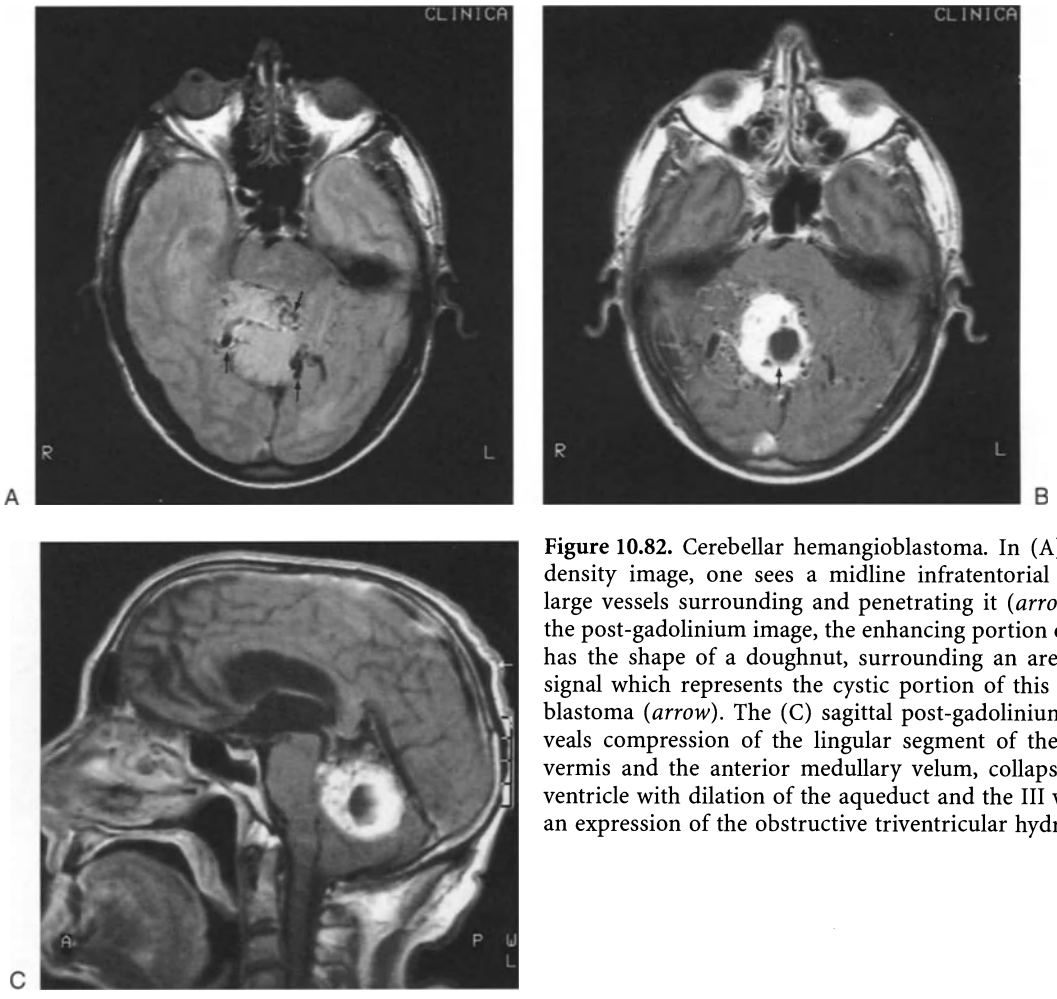


Figure 10.82. Cerebellar hemangioblastoma. In (A), a proton density image, one sees a midline infratentorial mass with large vessels surrounding and penetrating it (*arrows*); in (B) the post-gadolinium image, the enhancing portion of the mass has the shape of a doughnut, surrounding an area of hyposignal which represents the cystic portion of this hemangioblastoma (*arrow*). The (C) sagittal post-gadolinium study reveals compression of the lingular segment of the cerebellar vermis and the anterior medullary velum, collapsing the IV ventricle with dilation of the aqueduct and the III ventricle as an expression of the obstructive triventricular hydrocephalus.

ebellar nuclei at the center of each cerebellar hemisphere, immediately superior to the brachium pontis and the brachium conjunctivum. Care should be taken not to aspirate in the region of the culmen monticuli without having placed Telfa or cotton fluffies behind the target area so as to assure integrity of the supraculminate and galenic systems.

In the event one does not have access to the Cavitron or a laser, then the *classical* technique for removing solid vermian astrocytomas should be used. This consists of inserting dissectors, or small spatulae, along the line of demarcation between tumor and normal cerebellum, gliding the instrument over the hardened tumor mass by using the tip of the dissection blade to feel tumor and then to separate it from the surrounding cerebellum. One is advised, when using the classical technique, to resect solid vermian tumors *en bloc*, rather than attempt to take them out piecemeal. This avoids traction and tugging on the cerebellar peduncles, something which may result in infarction or swelling of the brainstem.

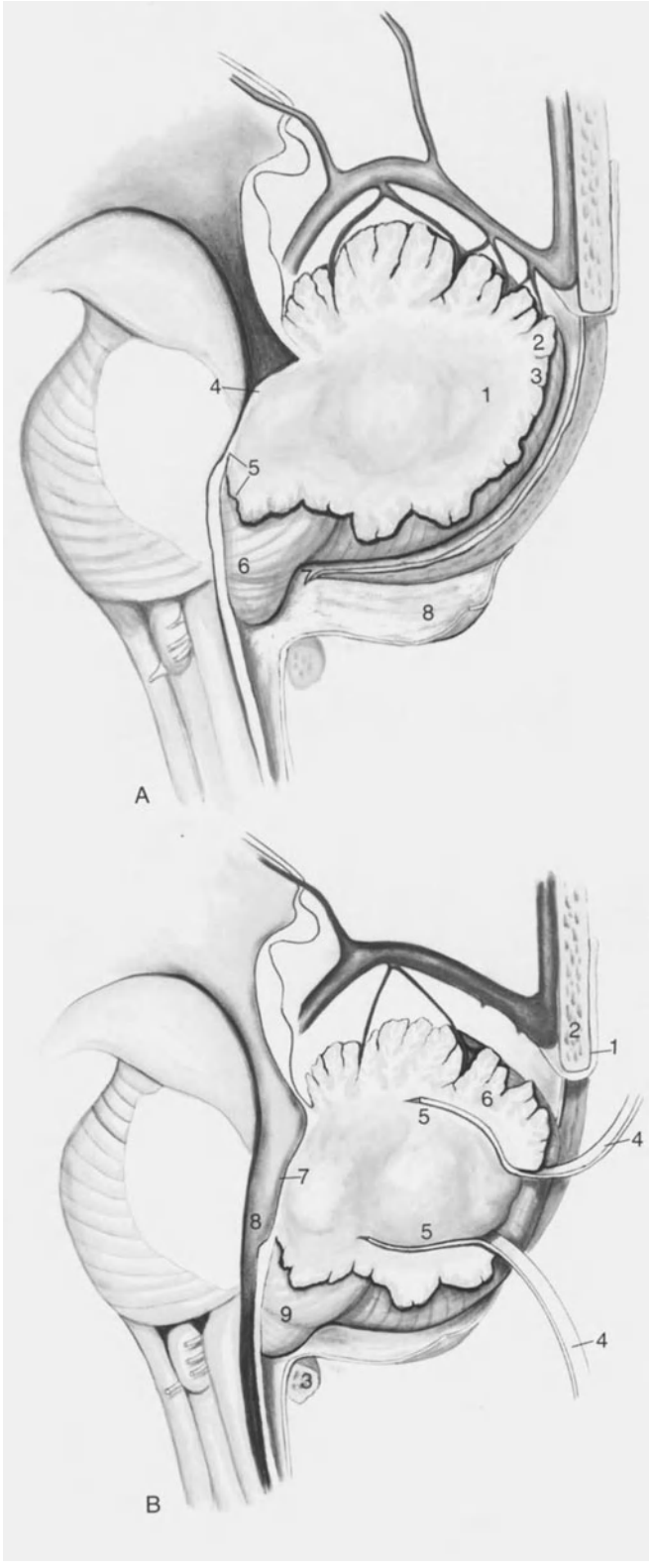
Dissection in the line of demarcation between tumor and cerebellum is best performed from superior to inferior, and from medial to lateral, freeing the superior hemsiphere of the tumor from surrounding brain before proceeding to separate its inferior pole from cerebellar tissue. After the brachium pontis has been identified, on each side, and dissectors swept over the inferior lateral surfaces of the tumor, one will observe that the mass falls from the cerebellum, out of the posterior fossa. Fluffy cotton is inserted into the cavity left by resected tumor. The bridging superior cerebellar veins, going from the remaining portions of the vermis and medial superior cerebellar hemisphere to the tentorium, are individually coagulated and transected. It is not wise to leave these veins intact, since head movement may tear them from the tentorium, resulting in a postoperative clot. This applies indifferently whether the tumor is removed with *Cavitron*, *laser*, or *classical* (dissector spatulae) technique.

Regardless of the technique used for removing the tumor, it is only rarely possible to maintain the integrity of the anterior medullary velum, since the superior vermian astrocytoma is intimately attached to this structure. During dissection one takes care to maintain an anatomical orientation, to avoid entering the bra-

chium conjunctivum. After the tumor has been removed, the surgeon will have an excellent view of the floor of the IV ventricle, particularly the superior portion, visualizing the colliculus facialis, lateral recesses, aqueduct of Sylvius and inferior colliculi.

Inferior Triangle Tumors (Figs. 10.83–10.87)

The basic difference between superior and inferior vermian triangle tumors is that the former appear to be intimately attached to the *superior medullary velum,* whereas the latter are attached to the *inferior medullary velum* in correspondence to the nodulus and the cho-



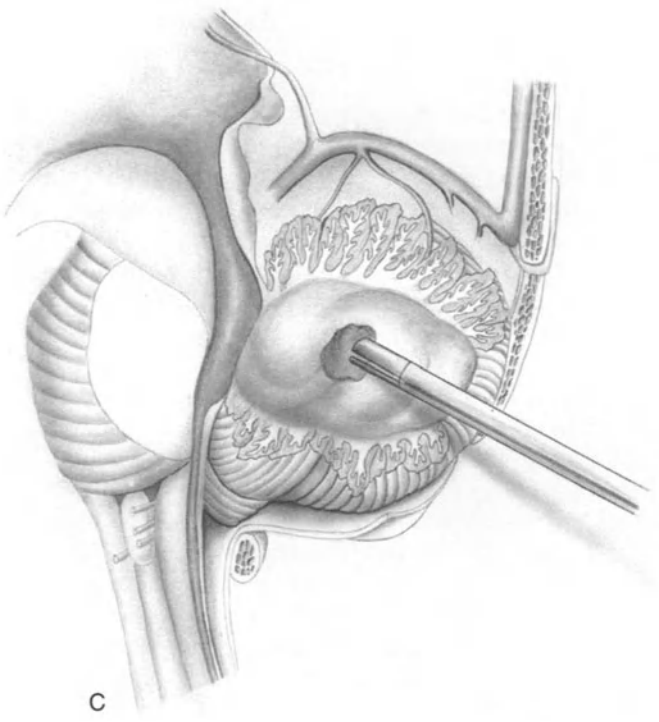


Figure 10.83. (A) Suggested technique for nonlaser resection of inferior vermian astrocytoma. This astrocytoma (*1*) expands within the vermis beneath the level of junction of folium (*2*) and tuber (*3*), extending as far anteriorly as the posterior medullary velum (*4*). This occludes the IV ventricle at the level of the foramen of Magendie and vallecula, pushing the uvula and nodules (*5*) against the floor of the IV ventricle and the tonsils (*6*) through the foramen magnum (*7*). A dural sling (*8*) has been fashioned to give support to the tonsils, and minimize traction on the cerebellar peduncles. (B) This techinque is advised only if Cavitron and laser are not available for "in situ," that is to say intratumoral, resection. The tumor is exposed through an inferior cerebellar craniotomy, and the superior trapdoor of the dura (*1*) sewn over the squamous occipital bone (*2*) and then anchored into place. The opening extends into the cisterna magna, so that dura is exposed as far inferiorly as C1 (*3*). The spatulae (*4*) are modeled to fit the contour of the tumor (*5*) and then used to separate tumor from the surrounding, normal, cerebellum (*6*). As this is done, the tumor is delivered from the operative field, and, consequently, the inferior medullary velum (*7*) comes away from the floor of the IV ventricle, unblocking it (*8*). The tonsils (*9*) do not reenter the posterior fossa immediately. (C) The "intratumoral" technique for resection, probably the oldest used in brain tumor surgery, consists of nothing more than gutting the tumor from its core peripherally. This half-tone drawing represents its use in an inferior vermian astrocytoma.

roid plexus of the IV ventricle. These tumors displace posteroinferiorly the tuber, pyramis, and uvula of the cerebellar vermis. They displace the nodulus anteriorly. Consequently, after the dura is opened one notes remarkable widening of the inferior vermian and cerebellar hemisphere folia. Separating the tonsils permits one to look at tongues of tumor extending between the cerebellar hemsipheres, posterior to the superior spinal cord and obex of the IV ventricle. The lateral and retromedullary segments of PICA are displaced laterally. One observes, within the vallecula, a tumor that is not covered by cerebellum.

The cerebellotomy is extended from the tuber inferiorly to the border between the tumor and uvula, and the deformed and displaced inferior cerebellar vermis separated from the surface of the tumor with spatulae, until approximately a 2-cm surface area of tumor has been exposed. Telfa is then positioned to protect the surfaces of the neural tissue and the vascular structures, before resection is begun. The same considerations apply in the use of dissector-spatulae for removal.

Irrespective of the surgical technique for removing the inferior vermian tumor, one must place Telfa along the floor of the IV ventricle to protect it, since most of the bulge of the tumor is into the inferior medullary velum and, consequently, onto the surface of the floor of the IV ventricle. The danger areas for encountering significant vascular structures are: the lateral surfaces of the tumor (the medial surfaces of the cerebellar tonsils, where the PICA is located), and the choroid plexus of the IV ventricle. The former structures must be preserved. The latter structure may be coagulated and removed with tumor. At the completion of tumor resection, one has visualization of the bulbomedullary junction, the floor of the IV ventricle and its lateral recesses, and the medial surfaces of the tonsils.

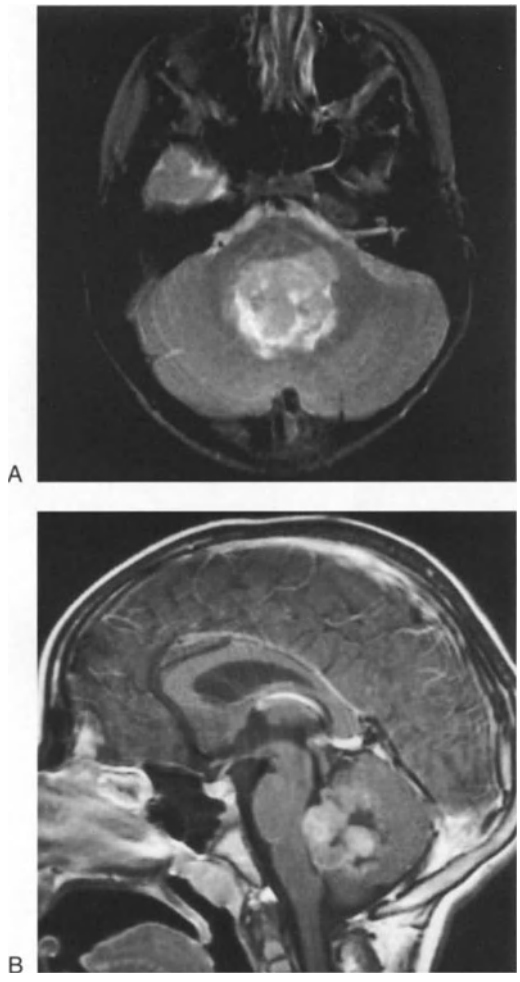


Figure 10.84. Inferior vermian astrocytoma. (A) The image shows a midline mass in the posterior fossa with a dyshomogeneous hyposignal and foci of high signal intensity (necrosis) which infiltrates the inferior portion of the roof of the IV ventricle. In (B), a sagittal post-gadolinium image, the high signal intensity of the mass with necrotic portions located posteriorly is interposed with low signal intensity areas. The lesion is contiguous with the floor of the IV ventricle, but clearly does not invade it. This observation was confirmed at surgery.

Surgical Consideration

In considering the brainstem either for tumors confined to it or those entering into or exiting from it, it is best to state that for the sake of a conventional understanding of the term "brainstem," the thalami and basal ganglia are *not* included. The anatomical limit ends inferiorly at the clava, ventrally at the peduncle; posterolaterally at the laterale geniculate bodies; medially by the III cranial nerves; and anteromedially by the mammillary bodies. A period of study of the figures relating to this section is more helpful than a verbal description. The cerebellar – like the cerebral – peduncles are clinically and surgically considered elements of the brainstem.

In the slightly dorsolateral projection, extending from superior to inferior, the external landmarks of the brainstem are: midbrain, with the peduncles, lemniscal trigone, geniculate bodies, and corpora quadrigemina; and the pons, with the pons Varolii and brachium pontis laterally, IV, V, VII–VIII, and junction of the cerebellar peduncles (superior, middle, and inferior) with the cerebellum. Inferiorly, the external landmarks in this projection are the olive flanked ventrally by XII and dorsally by IX–X–XI.

The dorsal perspective of the brainstem has as external landmarks, from superior to inferior; in the midbrain dorsally the quadrigeminal plate and laterally the geniculate bodies; the posterior surface of the pons has the anterior medullary velum, VI, the point of entrance of the cerebellar peduncles (superior, middle and inferior) into the cerebellum, and that portion of the floor of the IV ventricle superior to the stria medullaris.

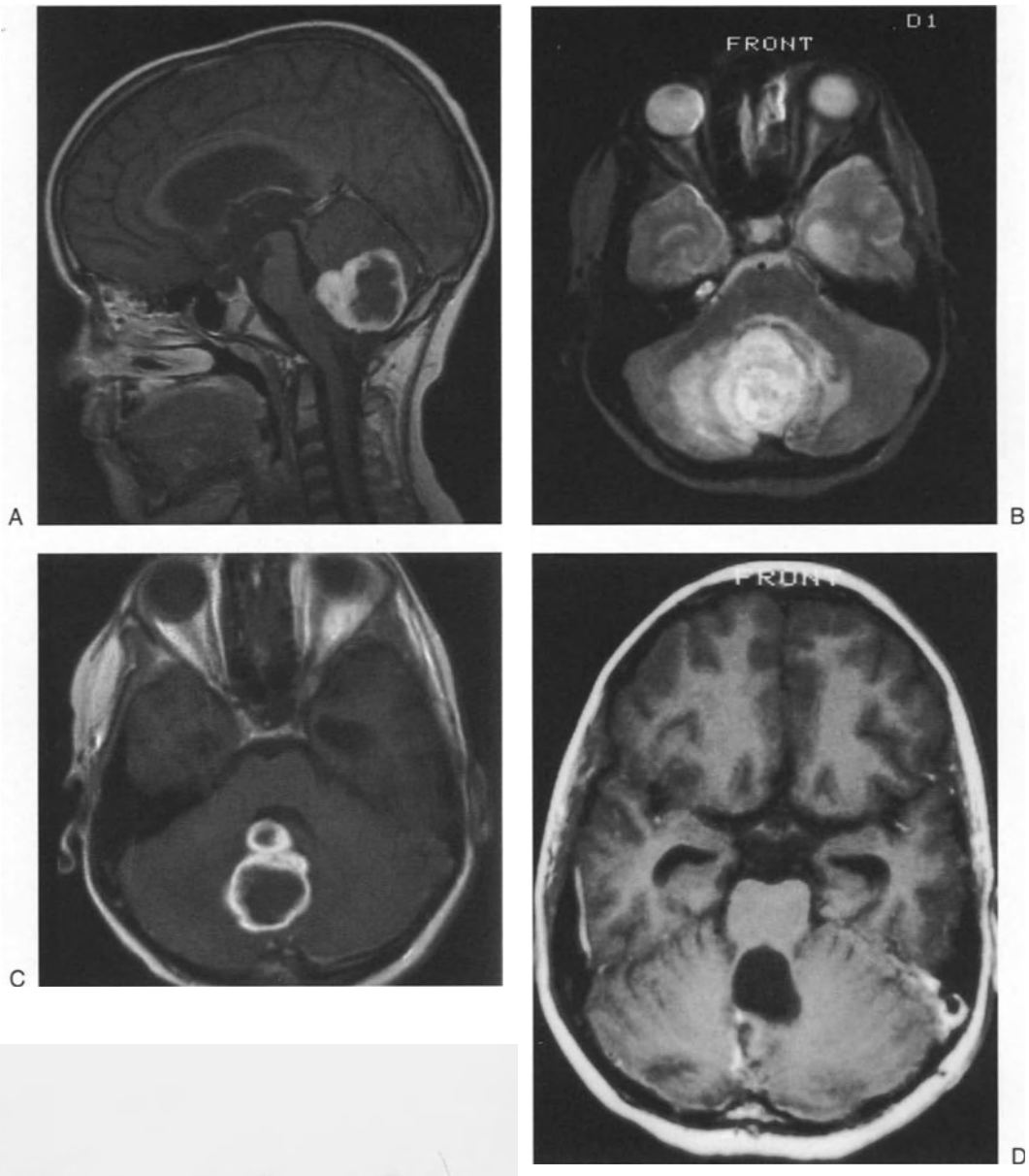


Figure 10.85. Astrocytoma, inferior vermian. In (A) one sees a large mass of high signal intensity, located posterior to the floor of the IV ventricle, which displaces the roof of this chamber. In (B) a sagittal section and (C) an axial section one sees peripheral enhancement in this post-gadolinium study and a central nonenhancing necrotic portion. In (D) the axial post-gadolinium, 4th-day postoperative image reveals the mass almost completely resected, with the exception of a linear enhancement, only minimal surgical defect, expanded IV ventricle.

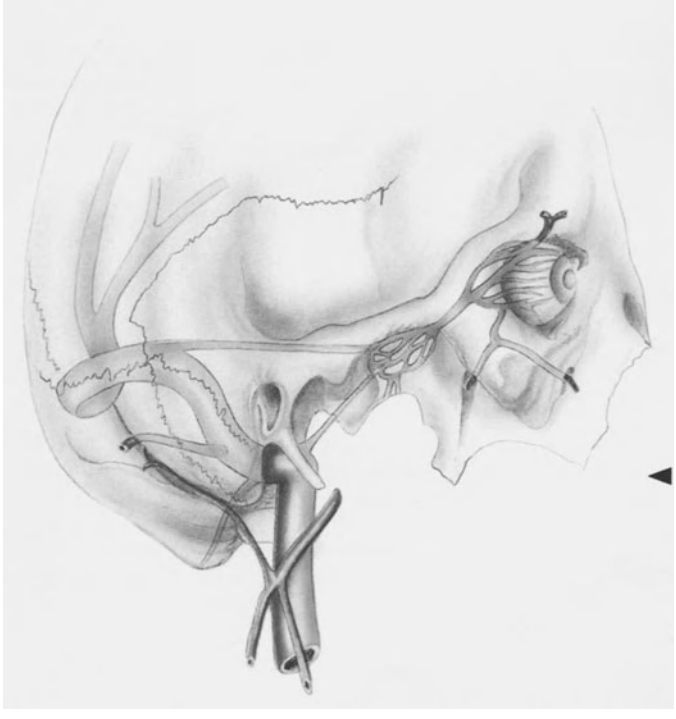


Figure 10.86. The performance of surgery on lateral and ventral foramen magnum tumors requires a full knowledge of venous, sinus, and vein anatomy...and drainage patterns. To perform the surgery, there is no substitute for skull dissections in the laboratory. The key venous structures are the transverse sinus, emissary veins, the two (superior and inferior) petrosal veins which establish flow into the transverse sinus and the jugular bulb, respectively, the internal jugular vein, and the occipital and facial veins.

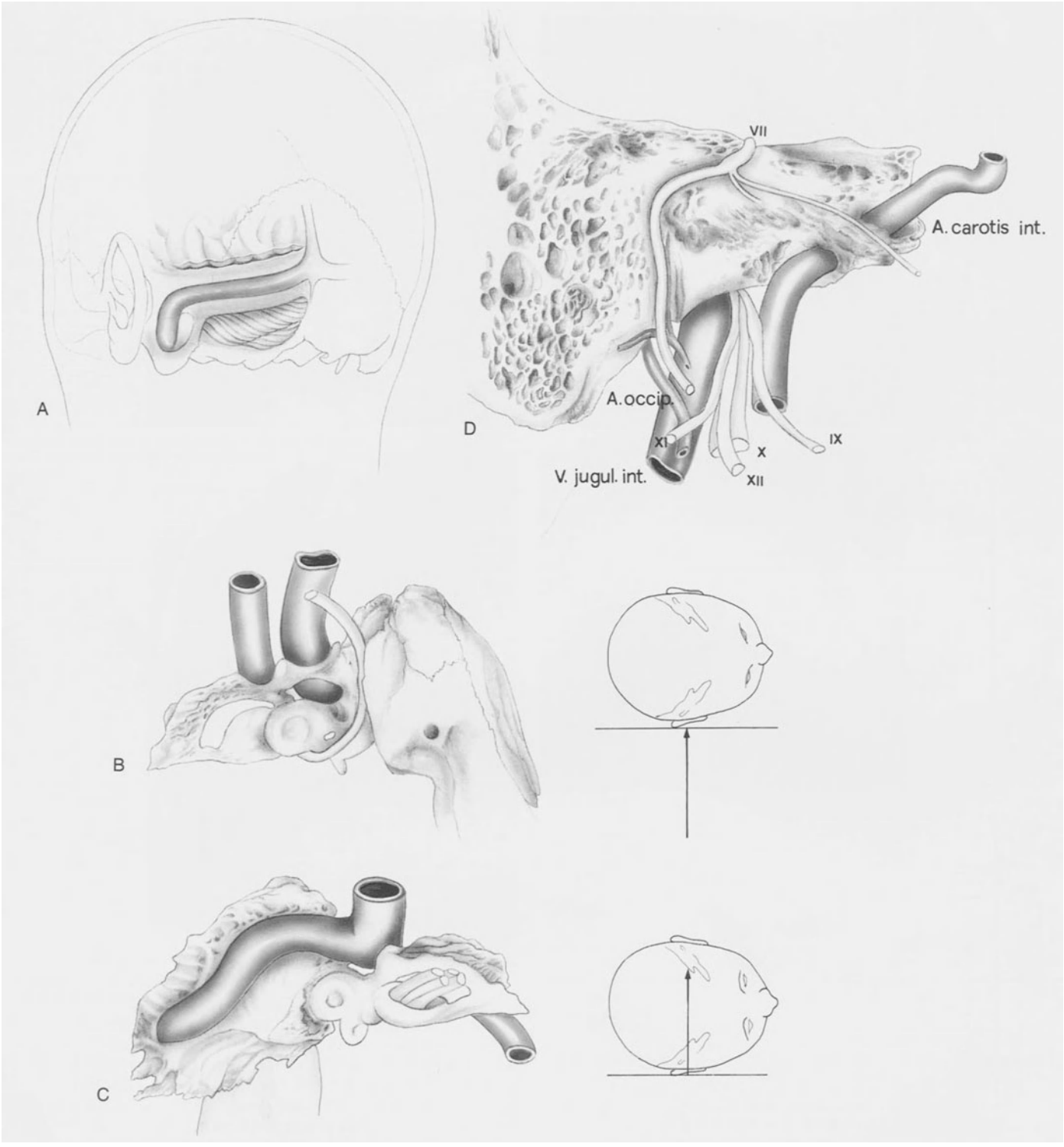
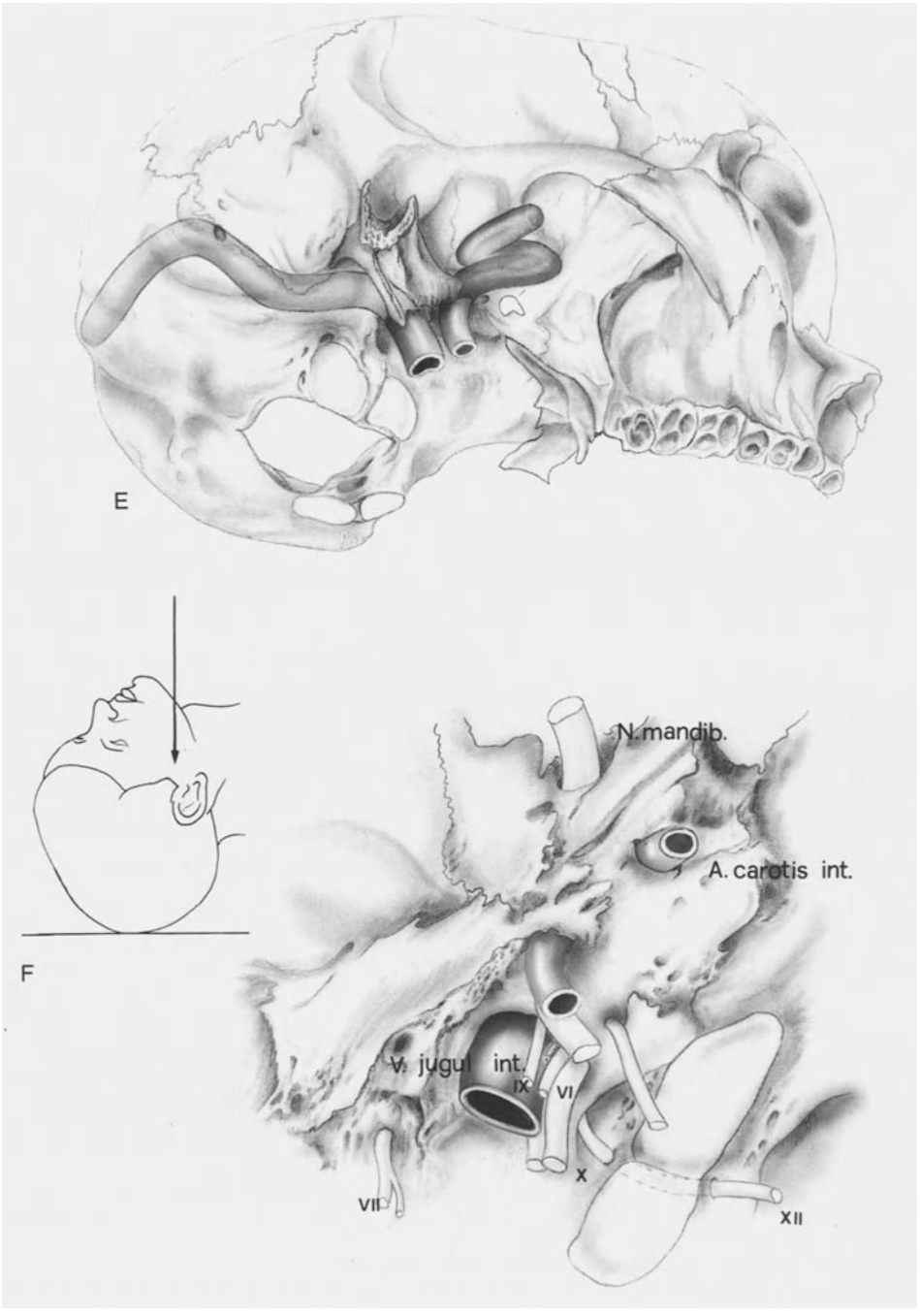


Figure 10.87. (A) For didactic purposes, and to establish anatomical points of reference, a description of the transverse and sigmoid sinuses is best begun with a postero-oblique view of the occipitomastoid area to obtain three-dimentionalized conceptualization of the directions mainly of the sigmoid sinus as it folds upon itself and passes into the jugular bulb. (B) With the surgeon looking directly at the external auditory canal on the involved side, the mastoid bone is posterior, the facial nerve exits from the fallopian canal between it and the internal jugular vein, and the internal carotid artery is immediately anterior (*in this illustration to the left*) of the vein, and at times juxtaposed to this structure. In this illustration the skeletonized semicircular canals are visualized, as is the lateral rim of the jugular foramen and the jugular bulb. (*Continued* on p. 285).



◄ **Figure 10.87.** (C) This illustration is a projection through the patient's head, permitting the reader to envision the view from the intracranial compartment, illustrating the internal carotid artery coursing through the carotid canal, the VII and VIII cranial nerves entering the internal auditory canal, the skeletonized semicircular canals, and the sigmoid sinus passing into the internal jugular vein at a right angle with the jugular bulb being formed at this junction. (D) This sagittal plane through the fallopian canal puts into relief the inframastoid neurovascular bundle, which often is involved in sarcoma, neuroblastoma, etc., of the region of the foramen magnum. (E) Rotating the head around an axis passing from the glabella to the external occipital protuberance permits one to appreciate the anatomical relationships between the foramen magnum, the occipital condyle, the mastoid eminence, and the styloid process on one hand...and the intradural portions of the transverse and sigmoid sinuses plus the carotid artery and its siphon, on the other. The region of the clivus behind the pterygoid plates is that occupied by chordomas. (F) This cone-down view of the interval between the stylomastoid foramen and the foramen ovale in the sagittal plane, and the foramen magnum and the petrous base in the coronal plane, reveals the concentration and compacting of the individual elements of the infrapetrous neurovascular bundle. The mandibular nerve does not enter into the anatomical confines of this bundle. This illustration should be read in the perspective of (E).

The dorsal aspect of the medulla oblongata consists of that portion of the IV ventricle beneath the stria medullaris, the obex, and the medial (gracilis, or clava) and lateral (cuneatus) tubercules.

Thus, depending upon the prominent location of a diffuse or circumscribed intra-axial lesion, or one which is exophytic, the surgical plan may consist of a *straight posterior* (classical) *suboccipital* approach; *posterolateral* or *lateral approach*; or combined *infratemporal/transtentorial* and *posterolateral suboccipital* approach. Though the transoral, or "face splitting" transclival approaches are technically feasible, the pathology for which they may be indicated either does not exist in children or, as in the case of chordoma, permits at best a minimally palliative endeavor. Irrespective of these latter points, the anterior and lateral approaches to the brainstem are not pediatric neurosurgical procedures, unless done with a maxillofacial or skull base surgeon.

Regarding the classical, midline suboccipital approach, the subject is exhaustively dealt with in sections on positioning, bur holes and flaps, durotomy, and cerebral retraction.

Bulbar or pontine tumors surfacing either at the IV ventricle, the clava, or the cuneatus are very conveniently dealt with through a midline skin incision and suboccipital craniotomy. The transverse and sigmoid sinuses are of no particular interest in this approach nor is the jugular bulb...unless the tumor fungates out through the lateral recesses and foramina of Luschka into the pontocerebellar angle.

This approach permits immediate visualization of the IV ventricle and restiform body, but one sees the brachium pontis or brachium conjuctivum only after the entirety of the tumor occupying the IV ventricle has been resected. Often, in resecting the tumor, those portions of the cerebellar peduncles which are involved are so completely invaded that one resects the tumor without the awareness that these structures had been converted into neoplasia. When the anterior medullary velum is involved, it too is resected, at which time the surgeon looks directly into the aqueduct of Sylvius and over the collicular plate. Globs of tumor may extend from the IV ventricle through the aqueduct into the iter of the third ventricle, causing this latter structure to dilate. Brainstem tumors growing from the floor or lateral walls of the IV ventricle, as a rule extend out into the vallecula, passing through the foramen of Magendie, and may even occupy small portions of the cisterna magna. It is unusual for primary brainstem tumors to occupy the entirety of the cisterna magna, something which may occur with medulloblastoma or vermian astrocytoma. The posterior inferior cerebellar arteries generally do not nourish directly these tumors, but are either splain out over them or deformed.

Brainstem tumors, especially infiltrating gliomas, cause cranial nerve damage by destroying the nuclei, not by direct damage to the nerve trunk. However, especially in those brainstem tumors with a large exophytic component fungating into the pontocerebellar angle, or those growing through the lateral recess to expand in the angle beyond the foramen of Luschka, damage may be brought directly upon the nerve trunk. Thus, if VI and VII are involved one may suspect nuclear damage, but if VII and VIII or V, VII and VIII are involved nerve trunk damage is also a possibility.

Posterolateral Approach

In performing a *posterolateral approach* it is necessary to separate from medial to lateral the splenius, semispinalis capitis, splenius capitis, and sternocleidomastoid muscles from their insertions onto the occipital and mastoid bones (Fig. 10.93). The craniotomy is then performed within the occipital bone, at its most inferolateral vertical plane, immediately inferior to the transverse sinus and immediately posterior to the sigmoid sinus, so that its lateralmost third rests posterior to the lateral surface of the cerebellar hemisphere, exposing this structure and the posteroinferior face of the transverse sinus as it descends to the jugular bulb. This permits the surgeon to work at the most extreme lateral surface of the posterior fossa, sliding over the floor of this compartment towards the pontocerebellar angle. The vascular structures which could menace him are the transverse sinus, the sigmoid sinus, the jugular bulb, and the entrance of the superior petrosal sinus into the sigmoid sinus. This latter need not be dissected and transected for a posterolateral exposure (but it must be sectioned for a combined supra/infratentorial approach).

The relationship between the mastoid process (not aerated in very young children and only moderately aerated in juveniles) and the sigmoid sinus is fairly constant, though the location and prominence of the jugular bulb are quite inconstant. Removal of the posterior and lateral portions of the mastoid process is straightforward, but almost invariably punctuated by two bleeding sites: emissary veins. Depending upon one's need to rise higher in the pontocerebellar angle or to begin more laterally so as to look at the lateral surface of the bulb and pons, the extent of mastoid resection is greater or lesser.

The dura is opened, the inferolateral aspect of the cerebellar hemisphere is elevated superomedially, and the spatula is inserted at approximately a 30° angle to the projected line of the petrous apex, with a slight elevation of the tip of the spatula at the flocculonodular lobe and minimal accentuation of the superomedial retraction, bringing into view first the XI, then the X and lastly the IX cranial nerves; and the lateral recess of the IV ventricle...with the pyramis and olive in full view at the pontomedullary junction. This is sufficient for lat-

eral medullary approaches to bulbar tumors. For entrance into the inferior half of the pons, beneath the sensory root of the V, one need only realign the microscope objective along the medial plane: the sensory root of the V is visualized to the gasserian ganglion, both the superior and inferior petrosal veins are in view, and the pons Varolii from just lateral to the median raphe to the base of the brachium pontis is within working distance. Visualization of the superior half of the pons is clear, but there is no room to work; access to the inferior half of the pons is excellent for visualization and work.

Combined Supra/infratentorial Approach
(Figs. 10.88–10.94)

In children this surgical approach is very seldom used since (1) the brainstem is a vertical structure and the basal cisterns are large; (2) the foramen of Bichat is a still-to-be-completely developed structure, remaining a large opening and only a virtual foramen; (3) the temporal lobe is relatively small compared to the parietal lobe, rendering retraction easy and atraumatic; and (4) the multistructure (bone/tentorium/sinus/cranial nerve) pathology, with the exception of NF-1, does not occur. Consequently, in the almost totality of cases it is sufficient to use the subtemporal approach to the mid-brain and superior half of the pons, simply sectioning the still hypoplastic tentorium to improve effective working space. However, for the sake of completeness of the section, a description of the combined approach will be given first, and then that limited to the infratemporal/transtentorial approach.

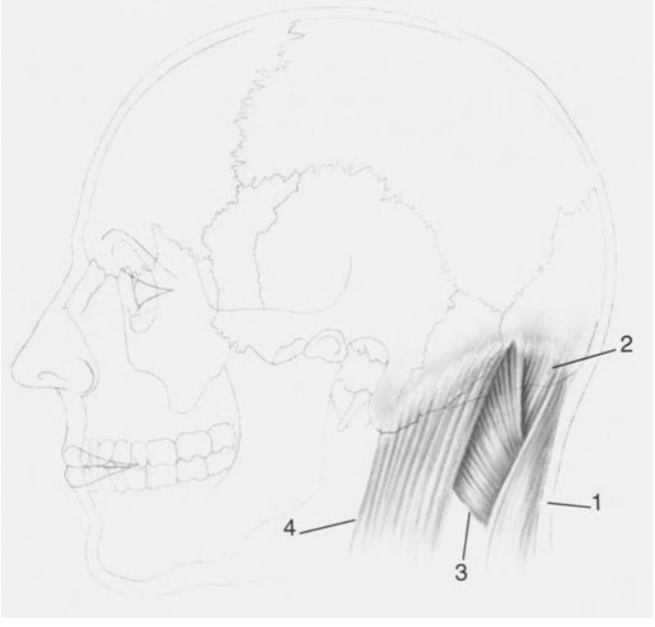


Figure 10.88. Insertion of (*1*) trapezius, (*2*) semispinalis capitis, (*3*) splenius capitis, and (*4*) sternocleidomastoid muscles onto the occipital and mastoid bones.

The child is put into the supine position, as described in Chap 1, "Positions," with its head rotated contralateral to the side of the pathology so that the sagittal plane of the skull is parallel to the coronal plane of the body. It is extended, and dropped slightly, as illustrated in the section on temporal flaps (see Chap. 3, "Bur Holes and Flaps"). In adults, maxillofacial and

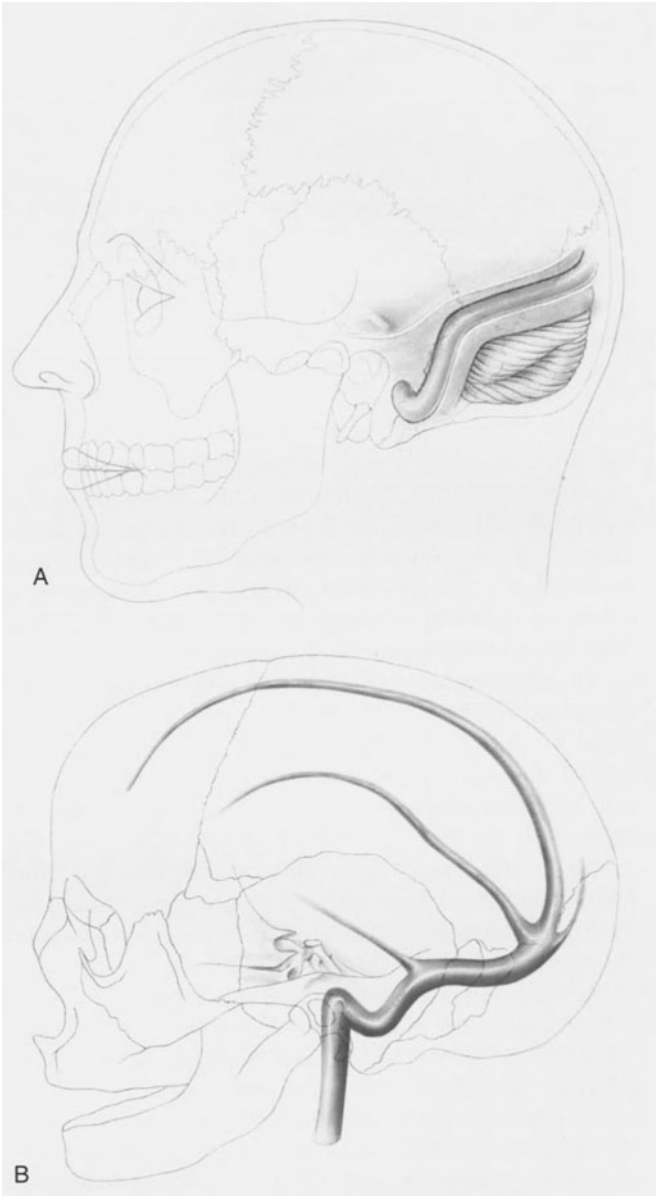


Figure 10.89. (A) The key anatomical structures at the dura/bone/brain interfaces are well seen when projected as lateral images. The transverse sinus courses inferiorly, laterally, and anteriorly into the sigmoid sinus and then this latter into the jugular bulb. (B) This very schematic projection of the child's skull onto the posteroinferior dural sinus illustrates the descent and lateral course of the transverse sinus, the characteristic shape of the sigmoid sinus and the entrance, at right angles, of the superior petrosal sinus into it, the jugular bulb pointing upward, and then the internal jugular vein.

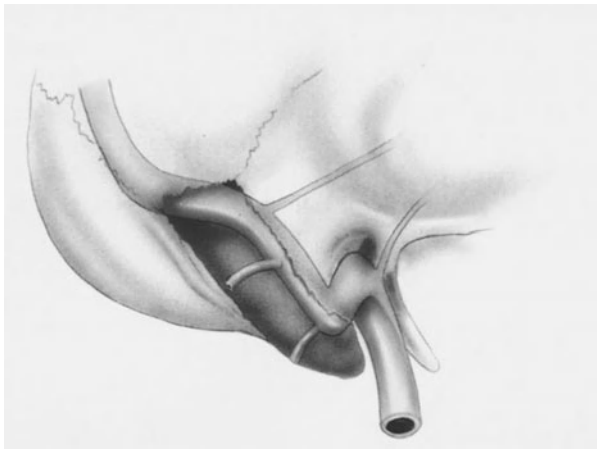


Figure 10.90. The posterolateral portions of the mastoid bone have been removed (drilled away). One notes the two emissary veins crossing the shadowed area (image of the pre-resection mastoid process). The entrance of the superior petrosal vein into the sigmoid sinus and the inferior petrosal vein into the jugular bulb is shown. The internal jugular vein is slightly medial to the styloid process.

otorhinolaryngologic surgeons are accustomed to flexing the head, bringing the mandible to rest upon the contralateral shoulder, so as to accentuate visualization of the mastoid bone and process. In children this is both unnecessary (the mastoid is small and the sternocleidomastoid and splenius capitis muscles are poorly developed) and dangerous (the mandible may compress the internal jugular vein). The skin incision used is that illustrated in Chap. 2, "Incisions," the external auditory canal is not exposed (much less transected), and then a partial mastoid resection is performed as described in this section under posterolateral suboccipital craniotomy.

A posterior temporal osteoplastic bone flap and a left posterolateral suboccipital craniotomy are then perfomed one right after the other, opening the dura mater along a line perpendicular to the center of the zygomatic arch anteriorly and the posteromedial surface of the sigmoid sinus posteriorly, so as to visualize the entrance of the vein of Labbé into the sigmoid sinus. The inferior dural flap is turned down at the floor of the middle fossa, exposing the middle and inferior temporal lobes. The surgeon now has an excellent line of vision along the tentorium, retracting the inferior temporal lobe towards himself as he dissects his way to the tentorial edge. At the same time, a retrosigmoid posterolateral approach as previously described is effected. The presigmoid approach is not indicated.

For details concerning the dissection along the tentorium, transection of the tentorium, and structures which may be visualized, consult the following section on the infratemporal transtentorial approach to the brainstem.

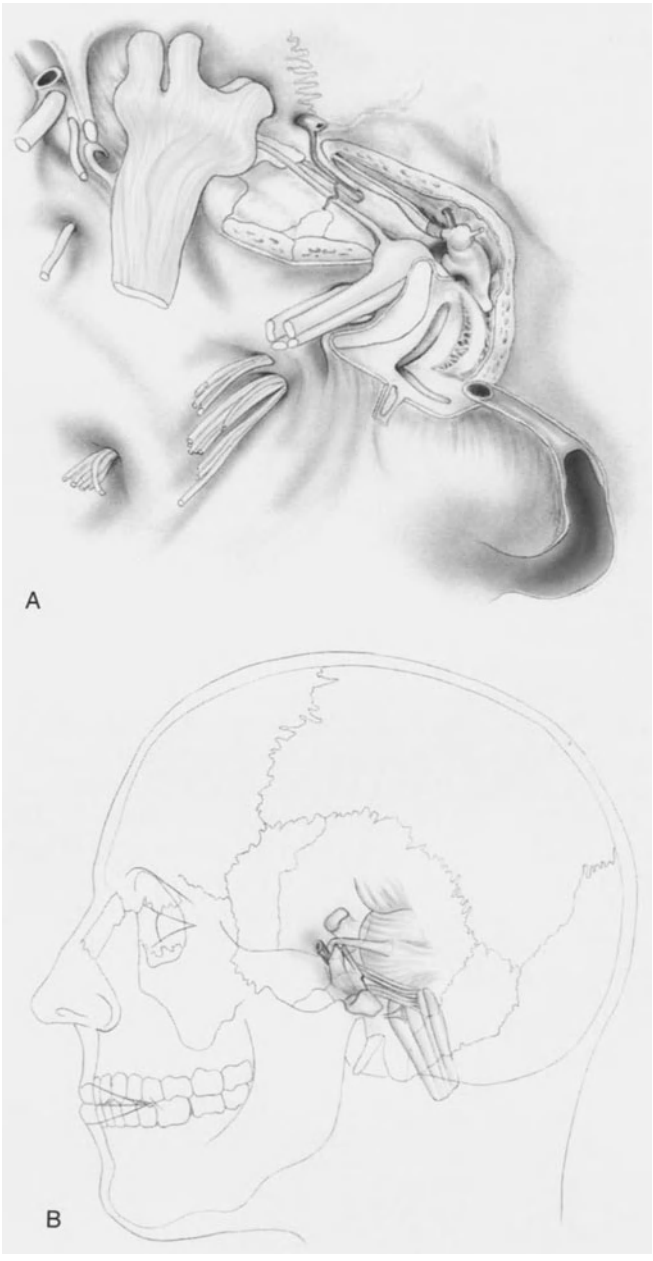


Figure 10.91. (A) Few views of the line of vision along the gutter of the base of the petrous pyramid, just medial to the jugular bulb, into the lateral recess and skirting the flocculonodular lobe and olive have ever been projected as clearly as the original dissection illlustrated in Pernkoff's atlas, from which this study was prepared. The brainstem from pons down to medulla is cut away from the nerves, which need not be labeled: the figure is for the reader to contemplate. (B) This is the view one has of the brainstem when the exposure described in the text is effected, after entering the angle along the line of vision set out in (A). Realizing that the opening is through the squamous occipital bone, across the posterolateral portion of the base of the mastoid, assists one to visualize that the effective working area is from the olive up to the sensory root entry zone...especially in children (who have a very vertical brainstem).

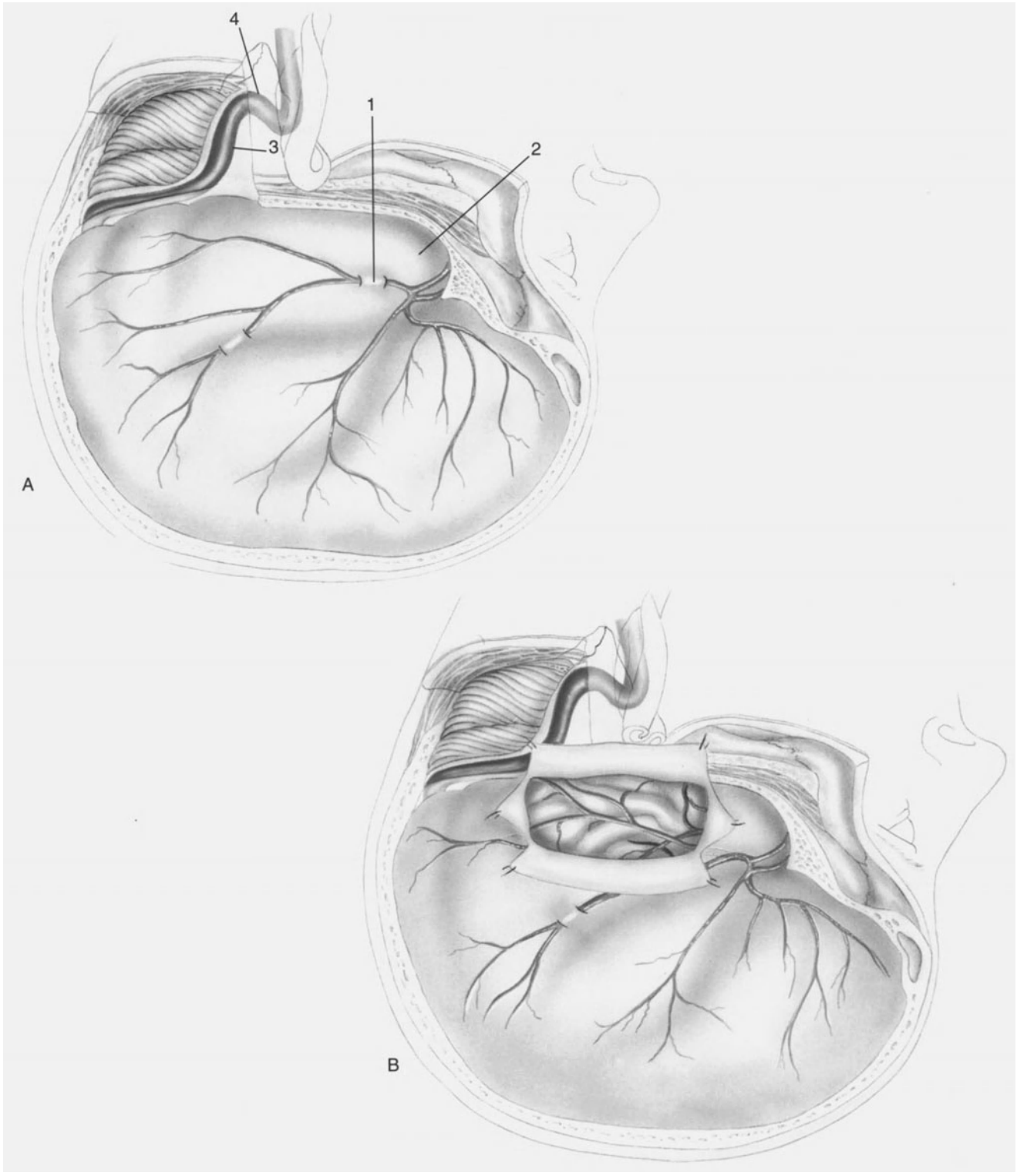


Figure 10.92. (A) The key elements illustrated by this color drawing are: (*1*) the intradural course of the branches of the middle meningeal artery; (*2*) the temporal pole adjacent to the anteriormost portion of the floor of the middle fossa but not (in children) nestled beneath the lesser sphenoid wing; (*3*) the point of transition of transverse into sigmoid sinus is far enough posteriorly for the supratentorial approach; and (*4*) the sigmoid sinus is a barrier separating retro- (to be used invariably) from pre- (not to be used) sigmoid routes to the lateral brainstem. (B) The dural opening need only expose the middle and posterior thirds of the temporal lobe, but it must be brought down to the floor of the middle fossa. It is preferable not to expose the vein of Labbé. This position of the child's head facilitates retraction and minimizes compression damage to the temporal lobe...but one must take care not to permit stretching of the vein of Labbé.

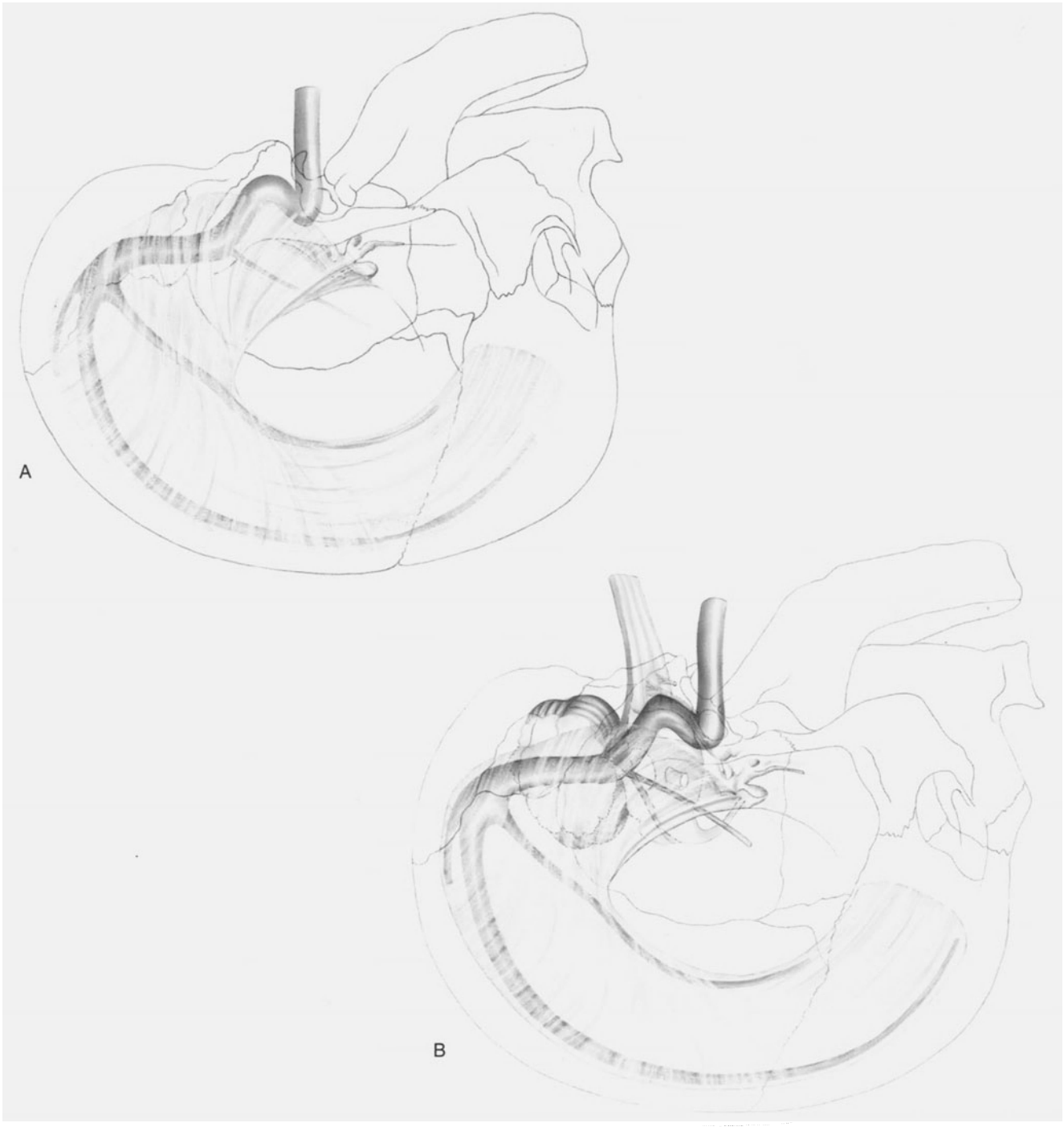


Figure 10.93. (A) This drawing illustrates the continuity of falx cerebri and tentorium, and the venous sinuses contained within the two dural layers. The foramen of Bichat is not yet a formed structure in that the opening in the tentorium is too large to permit one to refer to it as a "foramen." The entrance of the superior petrosal sinus into the transverse sinus where this latter structure passes into the sigmoid sinus is important in that a full view of the flocculonodular lobe; VII, VIII, and V; and the bulb cannot be attained from above without doing so. (B) The brainstem (from III ventricle floor to the cervical roots of XI) is seen through "the transparent" tentorium, permitting the reader to realize that mid-brain may be worked on effectively without opening the tentorium, but that the upper portion of the pons wants tentorial opening...and that the lower pons wants sectioning posteriorly to, and across, the superior petrosal vein: The tentorial flap may then be reflected, exposing the stem as shown in (C). (C) see p. 291.

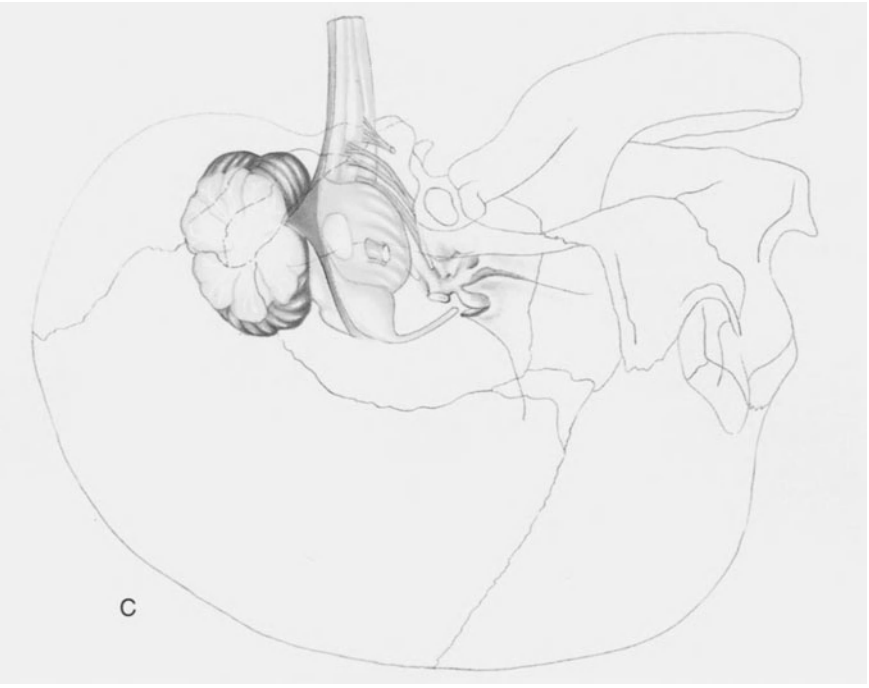


Figure 10.93. (C) With the child supine, head dropped minimally below the horizontal axis of the trunk, the brainstem is fully visible after the tentorium has been opened and its lateral flap brought to anchor either to the periosteum or dura at the edge of the temporal flap. The sigmoid sinus is now posterior to the area of access: out of the way.

Foramen Magnum Tumors

Foramen magnum tumors may be pedunculated extensions of brainstem (as already described) or spinal cord tumors expanding within the confines of the foramen magnum, above and below its rim. Lobules occupy the cisterna magna and subarachnoid spaces around the upper cervical cord (and the tonsils). At times, they may enter the vallecula. The peduncles from which these tumors expand are narrow, *so that, in essence, the tumor may be completely removed up to its attachment.*

Another foramen magnum tumor encountered in childhood is mesenchymal in origin, occurs in children with von Recklinghausen's disease, and grows from a nerve root: the perineurial fibroblastoma. Of course, meningioma and dural sarcoma may be located in this region, but they are so rare as to render comment superfluous.

The glioma, that is the exophytic tumor growing from the medulla oblongata or cervical cord, expands dorsally, whereas mesenchymal tumors expand lateral and ventral to the cervical medullary junction. Both have a tendency to fill completely the cisterns around their point of origin. Commonly, the dilated cistern or subarachnoid space resulting from such obstruction may be the causative factor for presenting neurological deficits.

Inferior vermian tumors, those growing from the region of the pyramis, expand within cisterna magna, cross the foramen magnum, and lobulate dorsal to the spinal cord, presenting as typical foramen magnum tumors.

Irrespective of whether the foramen magnum tumor expands dorsal, lateral, or ventral to the cervical medullary junction, at the rim of the foramen magnum, the craniotomy performed is for an inferior cerebellar triangle tumor. The dura is opened in the classical manner after the arch of C-1 has been osteotomized at its lateralmost portions, bilaterally, and dislocated posteriorly onto the spinous process of C-2. This permits opening of the dura down to the C-1/C-2 interspace. A cut perpendicular to the linear dural opening is then made at this point, extending as far laterally as possible on both sides. This permits one to sew the dura back without "tenting" it along the ventral surface of the upper cervical cord, minimizing risks of compressive damage (as described in the section on spinal dural opening).

This exposes the inferior cerebellar triangle, the cerebellar tonsils, and the upper spinal cord. Opening of the arachnoid of the cisterna magna reveals the tumor, and permits one to determine whether it is dorsal, lateral, or ventral to the spinal cord. If it is dorsal, one may be relatively certain that the mass is a glioma. Lateral or ventral location does not preclude a glioma, but does suggest that one consider a mesenchymal mass.

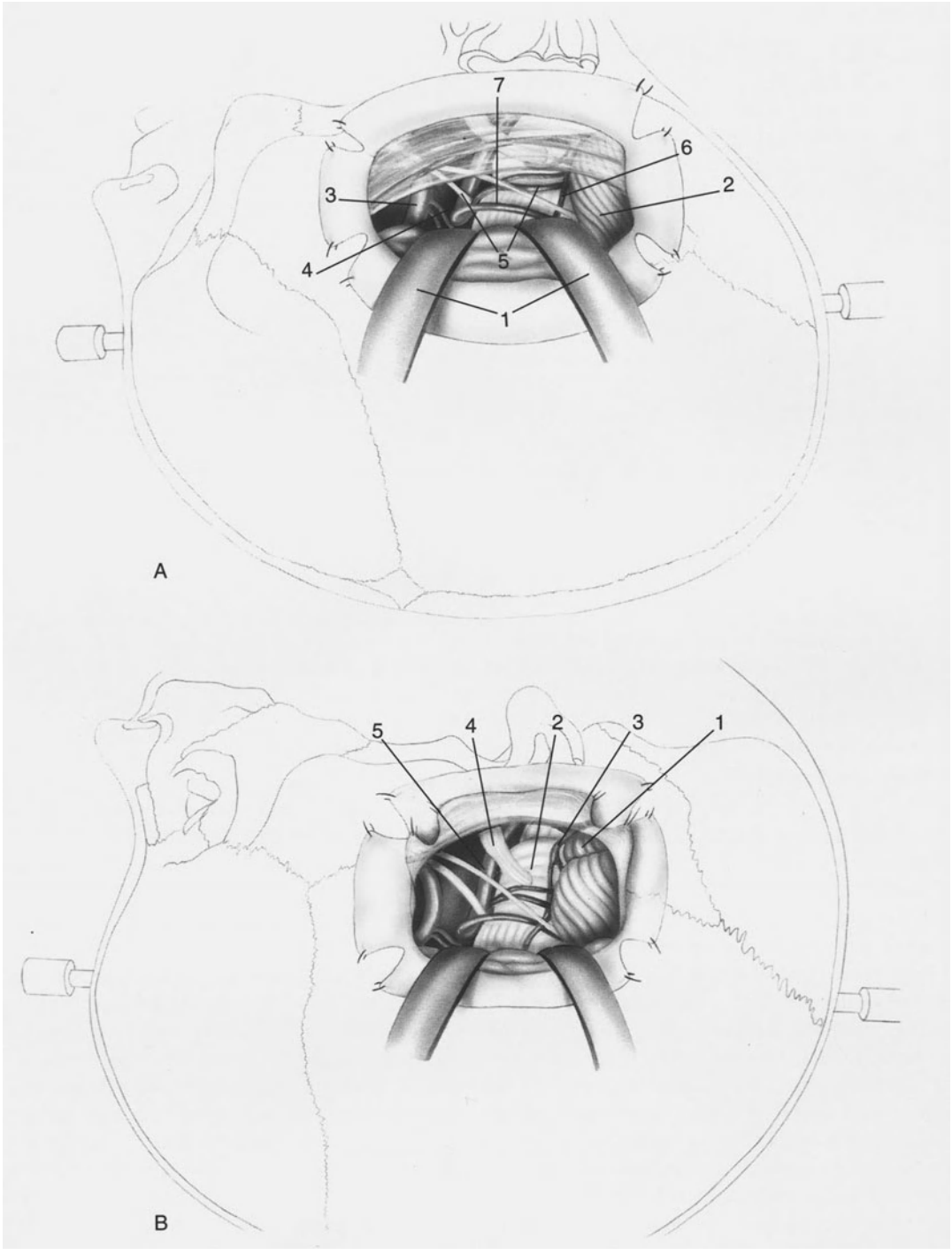


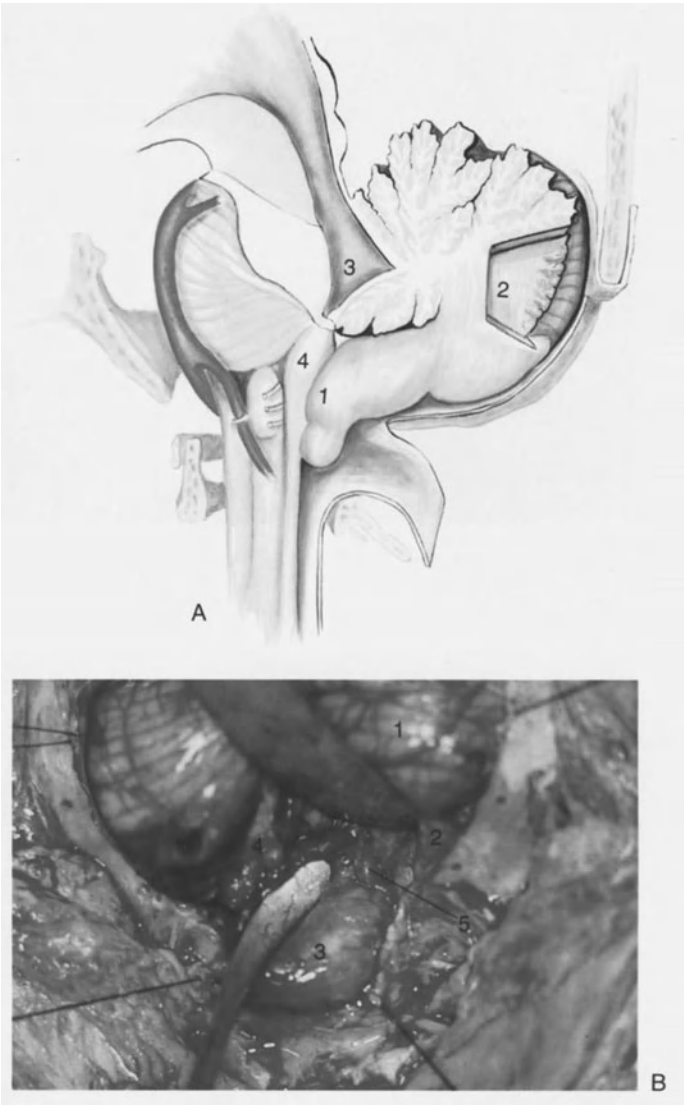
Figure 10.94. (A) This drawing was taken directly from an intraoperative procedure on the pons. It reveals (*1*) the ease of retraction of the temporal lobe; (*2*) the lateral surface of the cerebellar hemisphere protruding above the tentorial edge; (*3*) the cartotid artery with its anterior choroidal and posterior communicating artery branches; (*4*) the basilar fundus with its posterior cerebral and superior cerebellar branches; (*5*) the III and IV cranial nerves; (*6*) the transverse pontine vein; and (*7*) the superior portion of the pons...all above the line of the tentorial edge. (B) Once the tentorium has been cut and sewn out of the direct operative field, one sees (*1*) the flocculus and lateral recess of the IV; (*2*) the inferior pons and superior portion of the bulb (olive); (*3*) the inferior extension of the transverse pontine vein; (*4*) the sensory root of V at the entry zone; and (*5*) the inferior portion of the trunk of the basilar artery.

Dorsal Foramen Magnum Tumor (Fig. 10.95)

After the arachnoid has been opened and the tumor exposed, the surrounding neural and vascular structures should be covered with Telfa. One then separates the lobules of tumor from bridging bands of arachnoid tissue, which attach it to the underlying cervical cord and the borders of the IV ventricle. It is separated from the laterally lying tonsils.

No attempt is made at this time to determine where the peduncle of the foramen magnum tumor is, since even minimal manipulation causes respiratory or cardiac arrest. It is preferable to vaporize the tumor with a laser (5 W, defocused beam, pulse wave) so as to diminish considerably its bulk. The vaporization should extend into all lobules of the tumor on the dorsal surface prior to proceeding to either lateral surface. As tumor volume diminishes, one may bring Telfa cotton more and more between the ventral and dorsal surfaces of the cervicomedullary junction, delivering the residual tumor into the operative field where it may be vaporized. Ultrasonic aspiration with the Cavitron is very effective for resections of these tumors.

Once all tumor lobules have been vaporized or aspirated, one may inspect the surface of the cervicomedullary junction, or inferior vermis, to identify the pedicle of the tumor. If the pedicle is coming from the inferior cerebellar vermis, it should be followed into the vermis and resected completely. If, on the other hand, the pedicle is at the junction of cervical cord and medulla oblongata, the resection should extend precisely to a line which is flush with medulla and/or spinal cord. If Cavitron or laser are not available, bipolar/microscissor resection technique should be used. The resection should



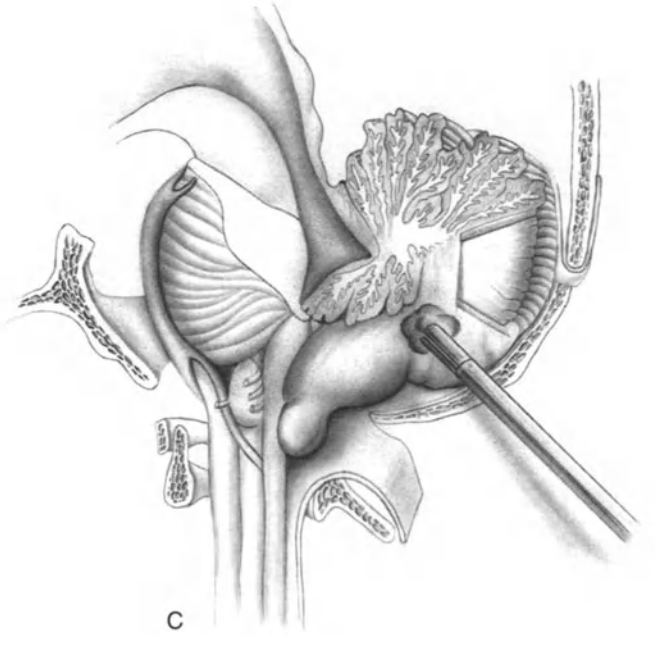


Figure 10.95. (A) Foramen magnum tumor (*1*) pedunculating from the inferior vermis (*2*). Gliomas may become exophytic, so that one may expect to see signs of cerebellar dysfunction, obstructive hydrocephalus, and spinal cord compression in foramen magnum tumors. Here, the exophytic tongue of tumor has obstructed the cisterna magna, resulting in dilation of the IV (*3*) and compression of the cervicomedullary junction (*4*). The value of laser in the removal of these tumors is readily appreciated when one considers the degree of adherence between the exophytic tumor and medulla, the compression of the latter, and the traumatic effects of traction. (B) Elevation of the cerebellar hemisphere (*1*), resection of all but a small fragment of the right tonsil (*2*), and depression of the exophytic tumor (*3*) reveal the pedicle of tumor (*4*) extending from the cerebellar hemisphere to the exophytic mass. Enormous branches of PICA (*5*) enter the tumor. (C) The foramen magnum component of pedunculated inferior vermian tumors is ideally resected by the "intratumoral" ultrasonic aspirator technique, since the peduncle of tumor may be manipulated about with the tumor forceps, allowing the surgeon to gain almost 360° vision of the tumor circumference as he proceeds with the resection.

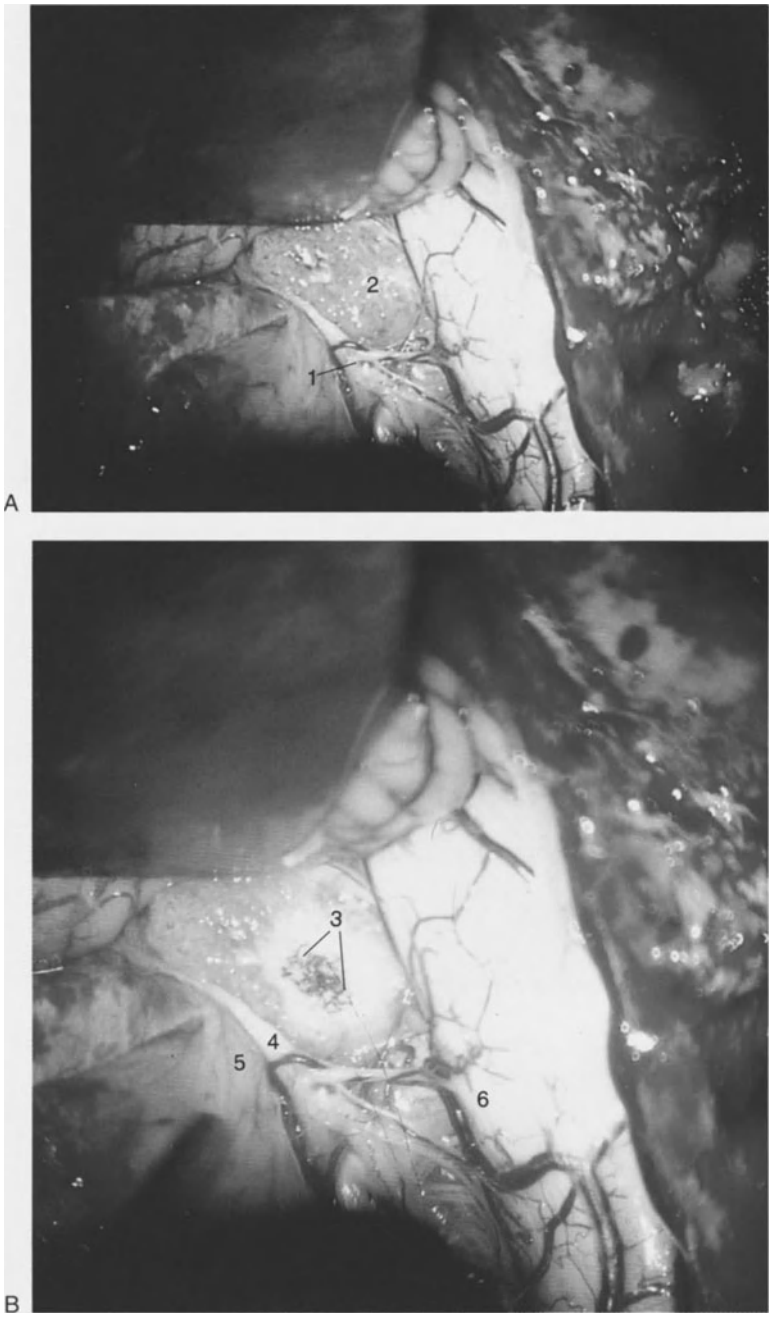
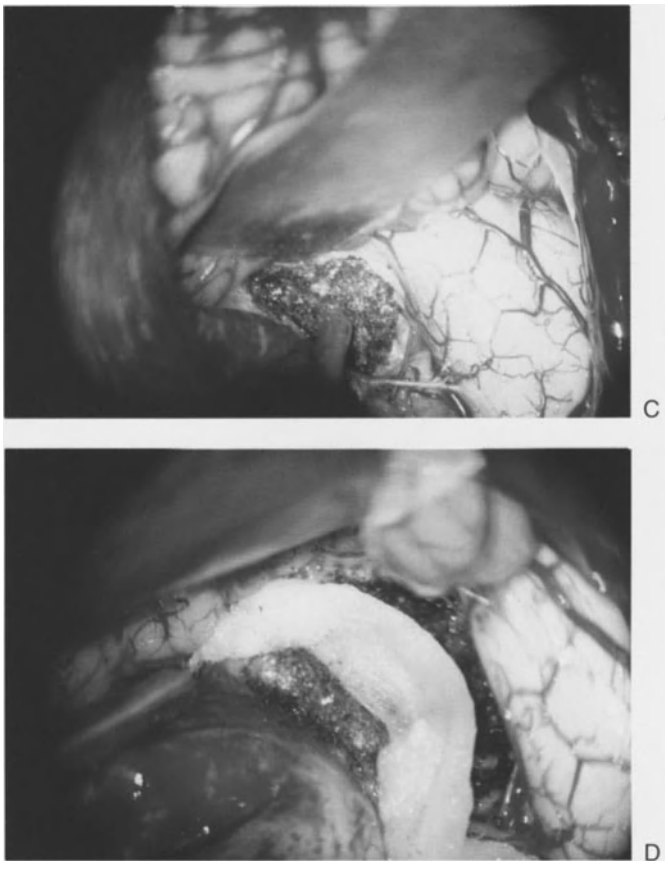


Figure 10.96. Legend see p. 295.

not extend to a level which is flush with the cervicomedullary structures: it should be stopped approximately 2 mm distal to this point!

Lateral Foramen Magnum Tumors (Fig. 10.96)

Lateral foramen magnum tumors expand within the interval between the IX, X, and XI cranial nerves dorsally. The ventral, lateral, and retromedullary segments of the PICA are splain out over the expanding mass *and* feed it, rendering access to it tedious and potentially dangerous. There is no need to enlarge the craniotomy for removal of these tumors. Resection of the tonsil exposes the dorsal cervicomedullary junction and brings the inferior pole of the tumor into view. This facilitates exposure of the mass and identification of the neurovascular structures engulfed within it. Very small, thin strips of Telfa are used to protect these structures. As each neurovascular structure is freed, it is surrounded by either Telfa or strips of Teflon. Similar protection is afforded the olive. The vertebral artery should be identified and covered with Telfa.



◄ **Figure 10.96.** (A) Lateral foramen magnum tumor. This is a chordoma extending from the clivus, lateralward, to expand within the interval between IX, X, and the spinal accessory (*1*) cranial nerves dorsally, and the XII cranial nerve ventrally. The tumor (*2*) extends below the foramen magnum and superiorly to the level of pontomedullary junction. The cerebellar tonsil and hemisphere are being elevated. Note that the spinal cord is displaced to the contralateral side and posteriorly. (B) This series of photographs illustrates the technique for tumor removal by laser, but either shrinking it with bipolar or, preferably, ultrasonic aspiration with the Cavitron are very effective. Laser vaporization has begun at the center of the tumor, resulting in a central area of destruction and a peripheral area of edema. Vaporization of the tumor shrinks it considerably in size by converting the edematous area to an area of dehydration. The carbon dioxide laser beam, which is invisible to the human eye, is vaporizing tissue between the two neon laser dots (*3*), which project red to the human eye. These neon laser dots serve as guides for direction of the carbon dioxide laser beam. The spinal accessory (*4*) is wedged between tumor and the rim of the foramen magnum (*5*), and its spinal rootlets appear to be tethering the spinal cord (*6*). (C) Continued vaporization of the tumor converts the dehydrated tissue into "clom," laser termionology for smoke. The tumor literally disappears. Note the improved vascularization of the spinal cord, just to the right of the remarkably shriveled tumor, and the slackening of the spinal rootlets of the XI cranial nerve. (D) As that portion of the chordoma which is engulfing the posterior inferior cerebellar artery is vaporized with the laser, fluffy cottons are brought in to protect vascular and parenchymal structures.

Ventral Foramen Magnum Tumors (Fig. 10.97)

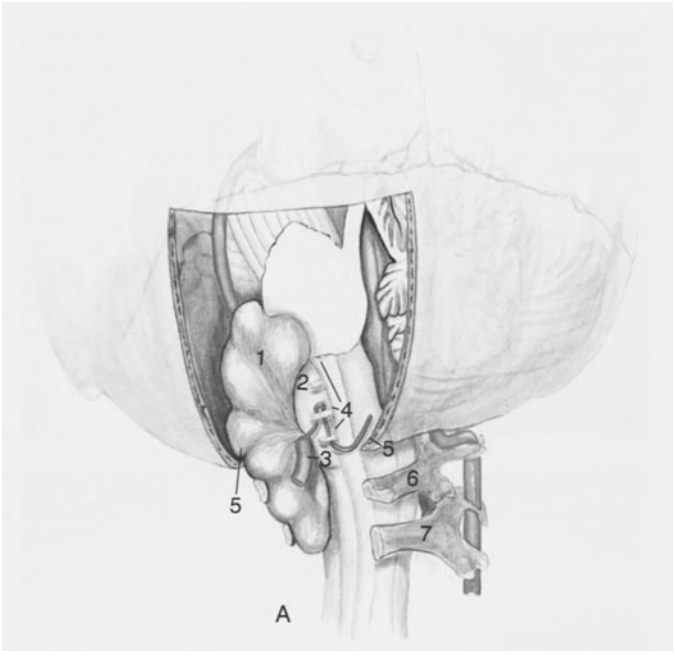
Both purely ventral tumors and a ventral extension of lateral tumors are more threatening in thought than they are in fact. Ventral tumors displace the cervicomedullary junction and vertebrobasilar system dorsally and laterally, but do not extend between cranial nerves, as do lateral foramen magnum tumors. Therefore, they may be vaporized in situ. The residual fragments are gently delivered from the interval between clivus and ventral cervicomedullary junction with the use of either a Penfield #4 dissector or micropituitary biopsy forceps. The laser should be used only sparingly, and with *great caution for tumors in this location, since the course of the vertebral basilar system is very variable and, consequently, one may not be sure that one has protected it completely.* Differing from lateral foramen magnum tumors, at times, it may be necessary to enlarge the osteotomy of the rim of the foramen magnum, extending it to the condyle, so as to obtain a more lateral view of the ventral tumors. It is never necessary to enlarge this more than 3 mm.

Cerebellar Hemisphere Tumors

The cerebellar hemsphere tumor, as the vermian tumor, may be solid or cystic. In either event, it may remain within the confines of one cerebellar hemisphere, or, most commonly, extend into vermis. There is no significant difference in incidence of extension into the superior or inferior vermian regions. Similarly, there is no difference in the incidence of extension into the brachium pontis or restiform body, though *the hemispherical tumor more often extends into one or more cerebellar peduncles than does the vermian tumor.*

Solid Cerebellar Hemisphere Tumors (Fig. 10.98)

The removal of solid cerebellar hemisphere tumors is effected with the same laser or Cavitron techniques as used for vermian tumors. The same applies to the use of the dissector/spatula techniques. The differences have to do with anatomical references: VII, VIII, IX, X, and XI cranial nerves; PICA and AICA; clava, and olive. Similar anatomical considerations apply to removal of cystic astrocytoma of the cerebellar hemisphere.



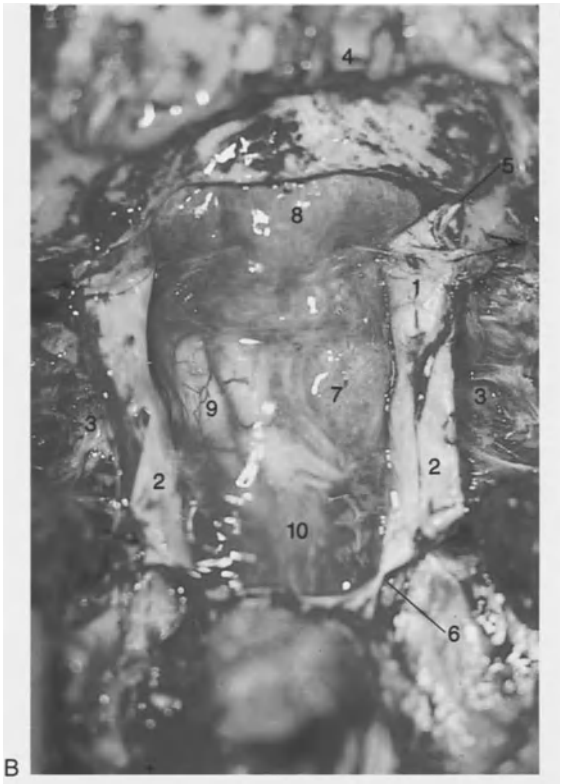


Figure 10.97. (A) This is a postero-oblique drawing of a mesenchymal foramen magnum tumor (*1*) expanding ventral to the medulla oblongata. The olive (*2*) is deformed, the vertebral artery (*3*) is engulfed and displaced, and the hind cranial nerves (*4*) are stretched. The tumor expansion above and below the foramen magnum (*5*) necessitates resection of the arch of C1 (*6*) and, at times, C2 (*7*). (B) The dura mater has been opened just above (*1*) the rim of the foramen magnum and then the durotomy extended from superior to inferior in the midsagittal plane, *reflecting spinal dural flaps (2) over the osteotomized medial rim of C1 (3) and the craniotomized medial rim of the foramen magnum (4)*. Note that the dural flaps have been incised superiorly (5) and inferiorly (6) bilaterally, so as to avoid tenting its ventral surface and compressing the ventral surface of the spinal cord even more. The herniated cerebellar tonsils (*7*) have the reddish appearance of tumor shining through from beneath the intact arachnoid. In fact, the tumor (*8*) is ventral to the spinal cord and medulla oblongata, pushing lateralward, and the spinal cord (*9*) is displaced. Take note of the very large accumulation of cerebrospinal fluid within the subarachnoid space inferior to the level of the tumor (*10*). (C) The arachnoid has been opened, giving egress to the cerebrospinal fluid, and allowing one to appreciate the remarkable dilation of the subarachnoid space (*1*) inferior to the tumor (*2*) and the posterolaterally displaced spinal cord (*3*). Upper cervical roots are splain over the inferior and superior domes of the tumor. The tonsils (*4*) may now be seen above the rim of the foramen magnum.

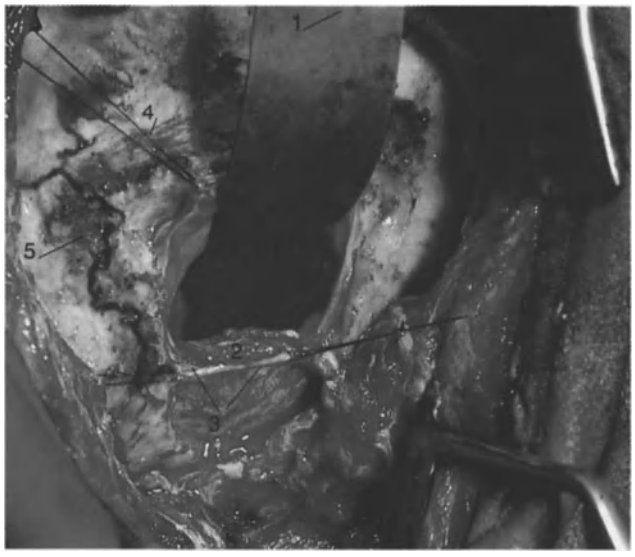


Figure 10.98. Nonlaser ("classical") technique for removal of solid cerebellar hemisphere astrocytoma. This procedure is used either when a laser is not available or when, as in this case, the solid tumor occupies the cerebellar hemisphere at the pontocerebellar angle, either adjacent or attached to the tentorium and walls of the sigmoid sinus. The cerebellotomy has been performed and a spatula (*1*) inserted into the cavity, elevating the superior portion of the cerebellar hemisphere. The inferior portion (*2*) is suspended by a sling of dura (*3*) fashioned in such a manner as to offer support and, thus, avoid herniation through the craniotomy site. For orientation purposes, the transverse sinus (*4*) and the sigmoid sinus (*5*) have been drawn in.

In sum, cerebellar hemisphere tumors may occupy the entirety of the hemisphere, being "coated" only by cortical tissue, so that removal of the tumor brings one onto cortex and adherent arachnoid; traction may stretch cranial nerves; dissection or vaporization may damage them directly. The same applies to the cerebellar arteries and brainstem. Consequently, for both solid and cystic hemispherical tumors one is advised first to elevate the inferior cerebellar hemisphere, then to identify VII, VIII, IX, X, and XI cranial nerves, and, finally, to place Telfa cotton between the hemisphere and cranial nerves. The same should be done for the PICA and AICA, prior to laying a piece of Telfa along the lateral surface of the medulla oblongata and pons. Subsequent to this, and throughout the resection, one repeatedly returns to inspect these protected neural and vascular structures.

Cystic Cerebellar Hemisphere Tumors (Fig. 10.99)

Cystic cerebellar hemisphere tumors accumulate large volumes of fluid. After the cerebellotomy has been perfomed the cyst wall should be coagulated and incised, draining completely the cystic fluid before placing the self-retaining retractors along the cyst wall. This suspends the superior portion of the cerebellar hemisphere, and permits a search for a mural nodule. If one is found, it is resected along with the capsule of the cyst: the same techinque is used in all cystic cerebral and cerebellar tumors.

Parasellar Tumors

General Anatomical Parameters

In childhood, the parasellar tumors are craniopharyngioma, optic pathway glioma, and germinoma. Though dermoid, epidermoid, and chordoma are generally included in this category, their incidence in childhood is almost nil.

Anatomy (Fig. 10.100)

Parasellar anatomy is quite intricate in that osseous, arterial, neural, ventricular, venous, and cisternal structures interrelate with one another in a complicated three-dimensional manner. This anatomy, in the normal case, requires a great deal of time and effort to master. In the abnormal case, especially for extraparenchymal tumors such as craniopharyngioma, the matter is compounded.

The parasellar area holds the following structures:

1. Body of the sphenoid bone: anterior and posterior clinoid processes, sella turcica, planum sphenoidale, basisphenoid
2. Optic pathways: two optic nerves, optic chiasm, optic tracts
3. Pituitary: stalk, gland
4. Anterior circle: internal carotid bifurcation, A-1 of the anterior cerebral artery, anterior communicating artery, M-1 of the middle cerebral artery, posterior communicating artery, anterior choroidal artery
5. Hypothalamus: anterior-inferior portion of the III ventricle, infundibulum, tuber cinereum, mammillary bodies
6. Basal cisterns: suprachiasmatic, infundibular, mamillary, interpeduncular, pontine
7. Cranial nerves: oflactory, oculomotor, abducens, trochlear, trigeminal

The anatomical limits of the hypothalamus are, anteroinferiorly, the optic chiasm; posteriorly, the mammillary bodies and interpeduncular fossa; laterally, the internal capsule and inferior portion of the thalamus; superiorly, the III ventricle (the lateral walls of the lower part of the III ventricle form part of the hypothalamic area). The hypothalamus extends inferiorly to join the pituitary stalk.

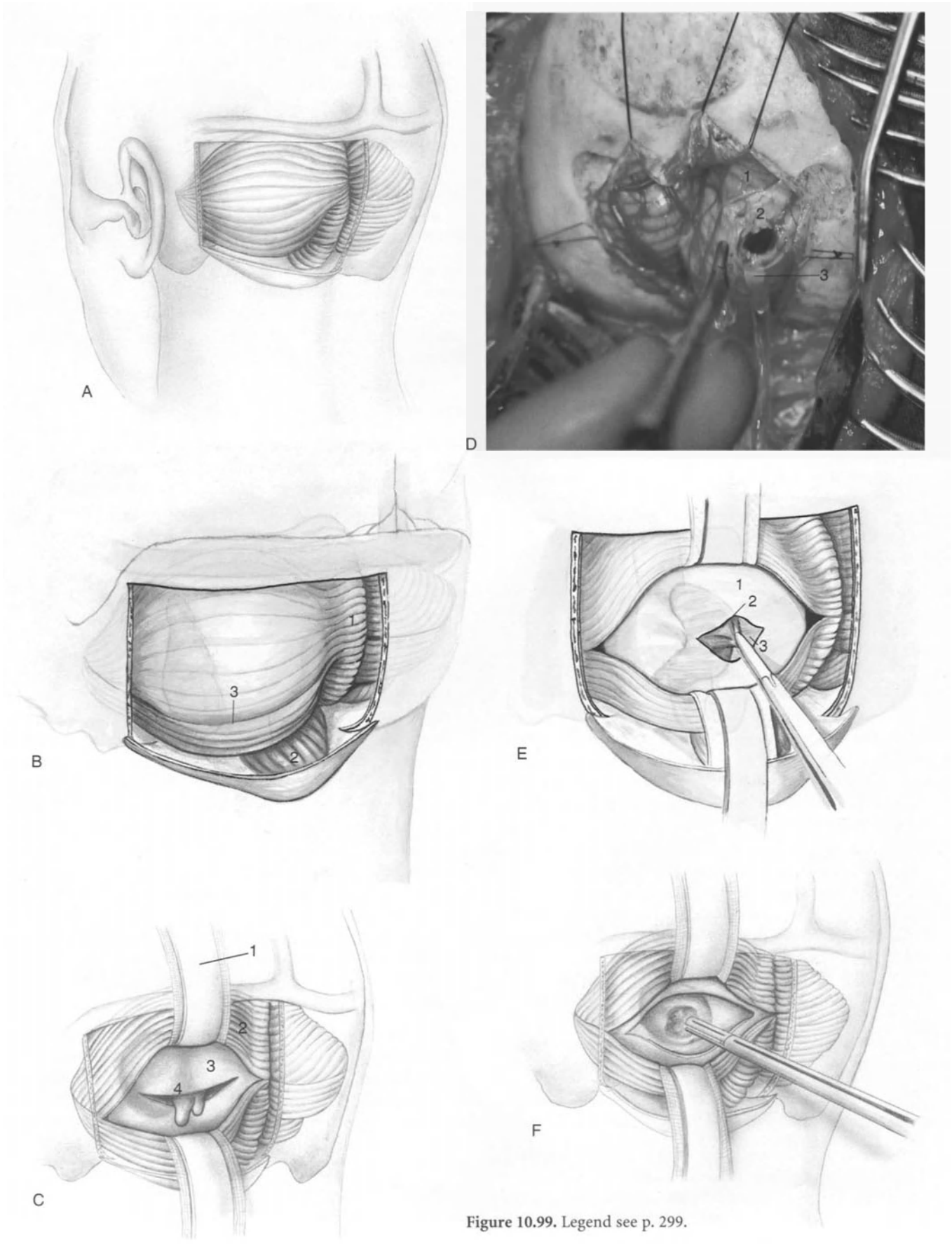
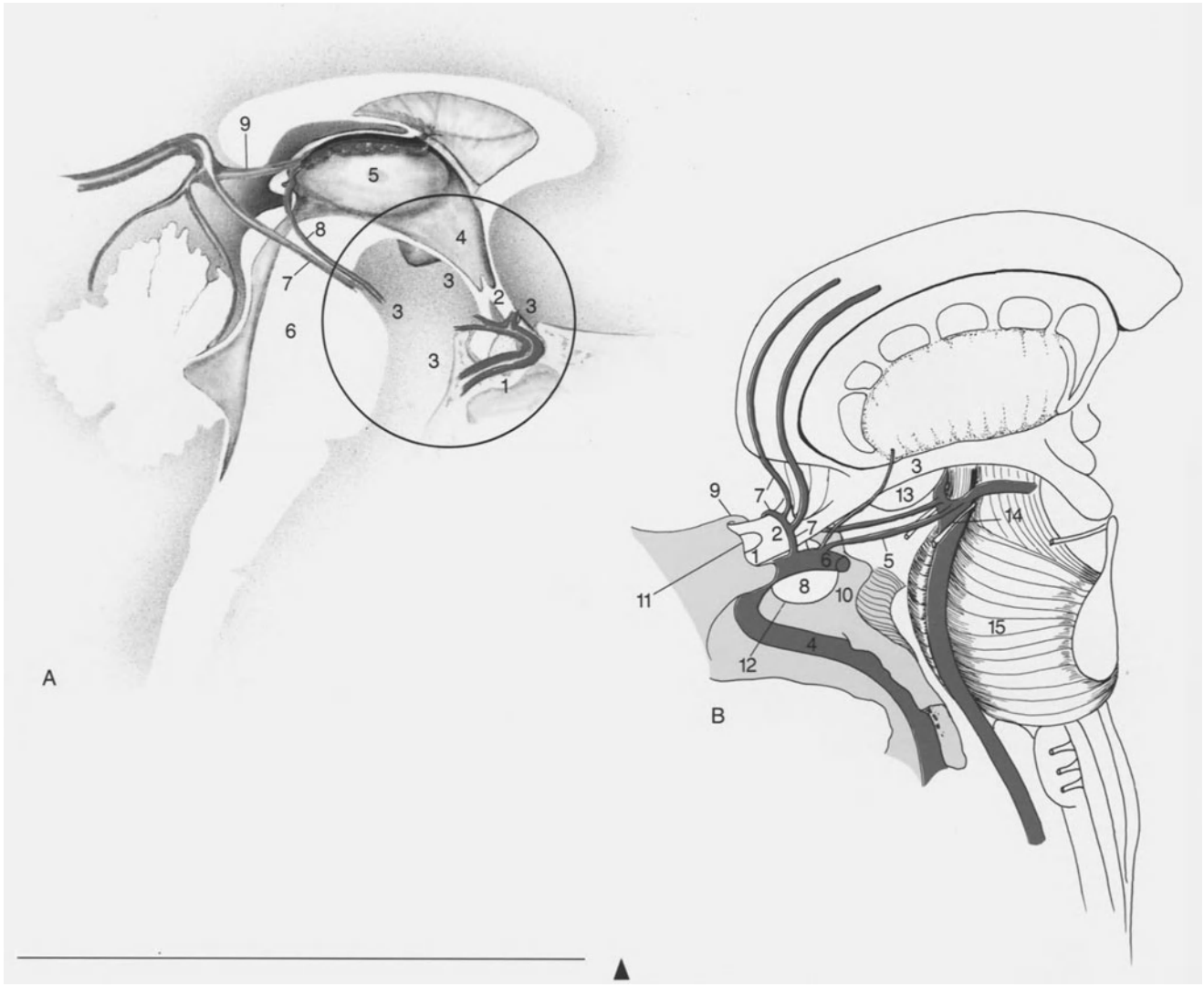


Figure 10.99. Legend see p. 299.



◄ **Figure 10.99.** (A) For a cerebellar hemisphere tumor, the lateral occipital flap is preferable. The surgeon may choose to bring the medial limb of his exposure to the midline, as in this drawing, but for the lateral limb he need not go past the line of the digastric groove. (B) Cystic tumor occupying the left cerebellar hemisphere, displacing the vermis (*1*) medially and the tonsil (*2*) inferomedially. The folia over the superior surface of the hemisphere are widened and flattened. The cerebellotomy (*3*) has been extended to the junction of hemisphere and vermis.(C) After the cerebellotomy has been made, self-retaining retractors are set with blades (*1*) placed between cerebellum (*2*) and cyst wall (*3*), allowing the cyst to bulge out of its bed. An incision (*4*) is then made in the cyst and capsule and the fluid drained. (D) The cerebellum (*1*) "capping" the cyst wall (*2*) has been opened along with the latter, allowing the viscid, yellow cyst fluid (*3*) to flow from the cavity. (E) As the fluid is drained, the cyst wall (*1*) collapses, falling from its surrounding cerebellar bed. The tumor nodule (*2*) is grasped with forceps and pulled from the inner capsule wall (*3*), facilitating delivery of the entire capsule from the cerebellum. (F) After the cyst has been drained, one may resect the nodular component with the ultrasonic aspirator if this is available. This is a much preferable technique since it avoids pulling, etc.

Figure 10.100. (A) This is a descriptive, not a spatially proportioned, illustration of the midline structures which may be involved by a parasellar tumor, depicting relative location of the sellar (*1*), chiasmatic (*2*), cisternal (*3*), hypothalamic (*4*), superior III ventricular (*5*), and brainstem (*6*) areas. Note the relative location of the basal vein (*7*), medial posterior choroidal artery (*8*), and internal cerebral vein (*9*). The reader sees readily that opening the lamina terminalis brings him immediately into the III ventricle; and that from this chamber passage into the interpeduncular cistern and area of Lilleguist's membrane necessitates opening the floor of the III ventricle. Though this may be done without causing damage to the hypothalamic nuclei when a tumor is not present, in this latter circumstance the neoplasm so deforms it that almost invariably panhypopituitarism results. (B) The signicant anatomical structures within the parasellar area, from the point of view of surgery of a craniopharyngioma, are the optic nerves (*1*), optic chiasm (*2*), and optic pathways (*3*); the internal carotid (*4*), posterior communicating (*5*), middle cerebral (*6*), and anterior cerebral (*7*) arteries; the pituitary stalk and gland (*8*); the anterior (*9*)and posterior (*10*) clinoid processess, as well as the planum sphenoidale (*11*) and the sella turcica (*12*); the mamillary body (*13*); III cranial nerve (*14*); and pons (*15*).

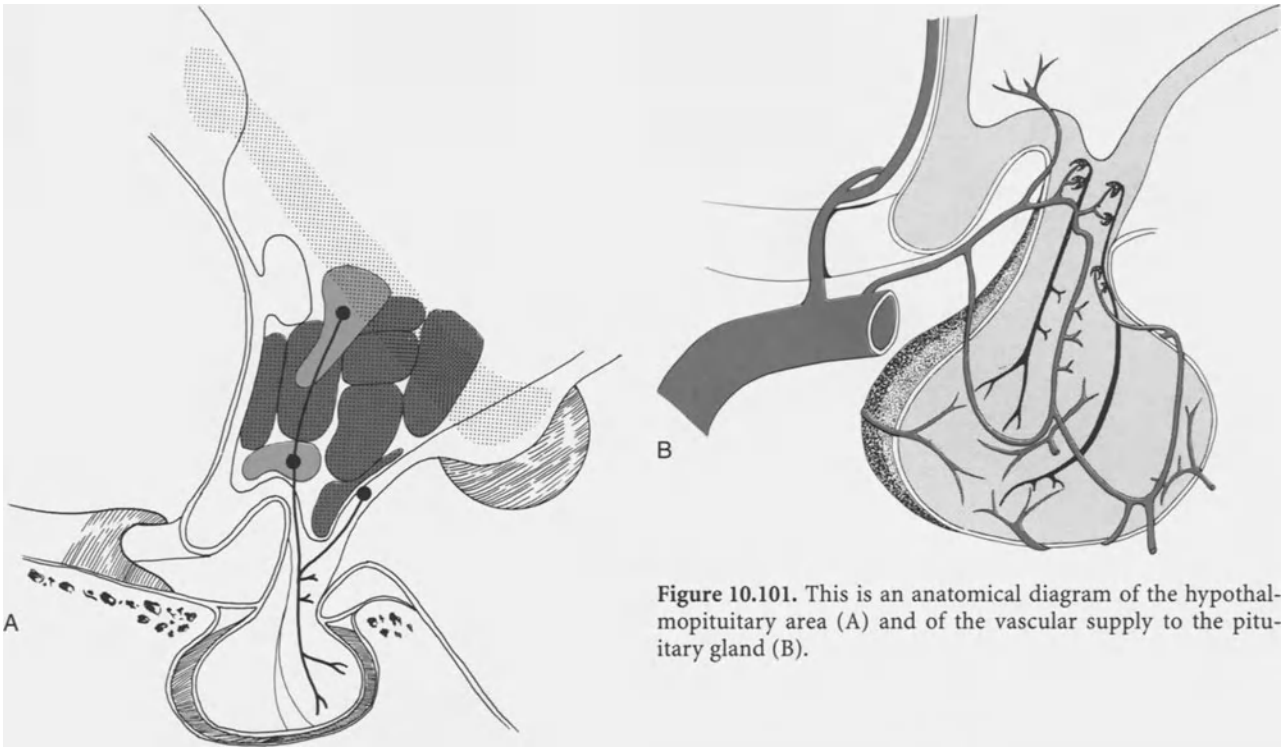


Figure 10.101. This is an anatomical diagram of the hypothalmopituitary area (A) and of the vascular supply to the pituitary gland (B).

The hypothalamus, infundibular stem, and posterior lobe of the pituitary gland (pars nervosa) are derived embryologically from one anatomical and functional unit (Fig. 10.101).

Certain groups of hypothalamic neurons cluster to form nuclei with the supraoptic (vasopression) and paraventricular (oxytocin) being the most easily identifiable, having well-defined neural connections with the neurohypophysis through the axons forming the supraoptic/hypophyseal tract. The pars distalis, pars intermedia, and pars tuberalis – the adenohypophysis – form the anterior portion of the pituitary gland. The pars tuberalis together with the infundibular stem make up the pituitary stalk. The infundibular stem extends inferiorly from just anterior to the tuber cinereum (bulge into the posterior floor of the III ventricle) directly into the neurohypophysis (pars nervosa).

The adenohypophysis consists of anterior and posterior lobes (pars intermedia), which are separated by Rathke's cleft, as well as the pars tuberalis.

The superior hypophyseal artery, which arises (on either side) from the internal carotid artery, in the superior loop of the siphon, extends superiorly to the upper portion of the hypophyseal stalk before dividing into anterior and posterior branches. The upper portion of the infundibular stem and median eminence are supplied by anastomosing branches which are formed from the superior hypophyseal artery. The trabecular artery, a large branch of the superior hypophyseal artery, follows an anteroinferior course along the anterior surface of the pars tuberalis to the point at which it penetrates the pars distalis to gain access to the infundibular stem.

The intracavernous portion of the internal carotid artery on either side gives origin to an inferior hypophyseal artery, which divides into medial and lateral branches anastomosing with their homonyms from the opposite side, thereby forming an arterial ring that surrounds the pars nervosa. One notes that the pars nervosa, fed primarily by the inferior hypophyseal arteries and anastomotic branches from the trabecular artery, has a distinctly separate arterial system from the pars distalis, which has a caudal system fed primarily by branches of the superior hypophyseal artery.

Patterns of Growth

Intraparenchymal tumors of the parasellar area grow either from the optic pathways or the hypothalamus. Consequently, they expand within these structures, growing along the neural pathways and into the surrounding III ventricle and parasellar cisterns. Extraparenchymal tumors, on the other hand, deform, stretch, and compress all structures within the parasellar area, as they expand within the sella turcica, parasellar cisterns, and III ventricle. Invasion of parenchyma by craniopharyngioma is not the exception. It is, however, unusual for craniopharyngioma to expand beyond the confines of the anterior III ventricle as it grows within the basal cisterns.

The structures normally damaged by craniopharyngioma are the optic pathways, hypothalmopituitary system, mammillary bodies, inferior and anterior III ventricle, vein of Rosenthal, posterior cerebral artery, and anterior circle. The basilar artery may be deformed by craniophyngioma, generally being displaced posteriorly, but, with the rarest of exceptions, it is not engulfed by the tumor. Neoplastic calcifications may be adherent to it. These occur in the "*formes geants*" [136].

Craniopharyngioma

The craniopharyngioma most commonly encountered in children is the suprasellar, calcified, extension of a tumor mass which enlarges greatly the sella turcica, compresses the optic nerves and chiasm, and bulges into the inferior portion of the III ventricle. It may present as a highly calcified (80% of the children) or a purely cystic mass, or as a Rathke's cleft cyst, occupying the region of the sphenoid sinus and the sella turcica. Craniopharyngioma may exist *in coincidence* with Rathke's cleft cyst: a typical craniopharyngioma in the intracranial compartment and a Rathke's cleft cyst extending from the floor of the sella turcica into the region of the sphenoid sinus. Neither craniopharyngioma nor Rathke's cleft cyst occur, except in the rarest of circumstances, under 2 years of age.

Optic Pathway Gliomas

Optic pathway gliomas growing within the subarachnoid spaces in childhood are commonly symptomatic before the 2nd year of life, though they may not be diagnosed until a later age. They do not limit themselves to one optic nerve, very often occupying the optic chiasm as well as one or both optic nerves, extending posteriorly along the optic tracts or superiorly into the lamina terminalis. *Because of the fact that optic pathway and hypothalamic gliomas present in the same manner clinically, have the same natural history irrespective of treatment modality, and are indistinguishable from one another neuroradiologically,* they are herein considered from the surgical point of view as identical tumors and will *not*, accordingly, be described separately.

Germinoma (Fig. 10.102)

The germinoma is a very distinct clinical entity, differing remarkably from the optic pathway and hypothalamic gliomas on one hand, and the craniopharyingioma on the other. Specifically, this tumor presents clinically with endocrine dysfunction, does not cause changes which may be identified with plain skull X-rays, and most commonly becoming symptomatic long before attaining a size large enough to be diagnosed neuroradiologically as a mass lesion. It responds immediately and dramatically to roentgen therapy.

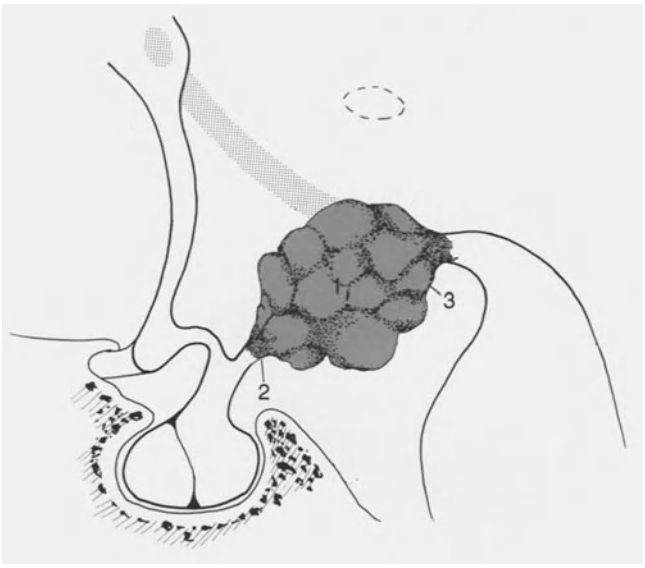


Figure 10.102. Hypothalamic *germinoma* (*1*) grows from the region between the pituitary stalk (*2*) and the mammillary bodies (*3*). It may fungate into the III ventricle, but, generally, remains quite small.

Regarding the most general and telegraphic of messages regarding surgical indications and techniques for parasellar tumors, one may state that:

1. Bifrontal craniotomy is recommended for resection of craniopharyngioma when the surgeon has evidence to suggest that he may attain a surgically "complete" resection.
2. A unilateral, anterior frontal craniotomy is recommended for partial resection of a craniopharyngioma.
3. There is no indication for surgery for germinoma, and only limited indications for optic pathway or hypothalamic gliomas, or hamartomas.
4. Debulking of these latter three tumors (though at times necessary) has not been demonstrated to provide benefits superior to those of chemotherapy and roentgen therapy alone.
5. Hypothalamic hamartomas are not amenable to radiation therapy, and only occasionally require surgery. Indeed, the operative morbidity for children with tumors within the hypothalamus, children who are either already suffering from hypopituitarism, or children who may develop a hypopituitary syndrome from the operative procedure is much greater than for any tumor, including IV ventricle lesions.

Clinical Characteristics of the Two Most Common Parasellar Tumors

Parasellar Glioma (Figs. 10.103–10.111)

Optic pathway (subarachnoid) and hypothalamic gliomas should be considered the same tumor clinically, and referred to as parasellar glioma, since attempts to resect either carry prohibitive risks, without offering any benefits. Surgery is performed *only* when one is unable to distinguish preoperatively between a craniopharyngioma and a parasellar glioma, something which is now rare indeed, or when there is a giant globular mass extending from the region of the hypothalamus/optic chiasm. The criteria for distinuishing between craniopharyngioma and parasellar (optic pathway/hypothalamic glioma) are:

1. Parasellar glioma
   a) Is most common under 2 years of age
   b) Causes roving nystagmus or spasmus mutans
   c) Rarely causes diabetes insipidus, commonly causes precocious puberty
   d) Invariably enlarges or deforms optic foramina and planum sphenoidale
   e) Very seldom calcifies
   f) Enhances diffusely in CT and MRI scans, showing lucent areas within its center
2. Craniopharyngioma
   a) With the *rarest* exception does not occur under 2 years of age
   b) Causes visual field cuts
   c) Commonly causes diabetes insipidus and panhypopituitarism, never causes precocious puberty
   d) Almost invariably calcifies
   e) Very rarely enlarges optic canals
   f) Never deforms planum sphenoidale without enlarging sella turcica

One may encounter a parasellar tumor (often a hamartoma) which has grown to enormous size, with necrotic center, and which, consequently, may be causing difficulty because of its volume. These tumors (as stated above) should be debulked surgically: they are exophytic gliomas which are amenable to subtotal resection just as the exophytic glioma of the hindbrain.

Biopsy of Parasellar Glioma (Fig. 10.112)

After the anterior frontal craniotomy has been performed, and the dura opened, self-retaining retractors are placed over the right frontal lobe along the line of the lesser wing of the sphenoid. This exposes the right olfactory and optic nerves, and avoids the necessity of transecting the right olfactory nerve. The perioptic arachnoid is opened, exposing the optic nerve from the anterior clinoid to the optic chiasm. One may, in this manner, inspect all of the anterior portion of the optic

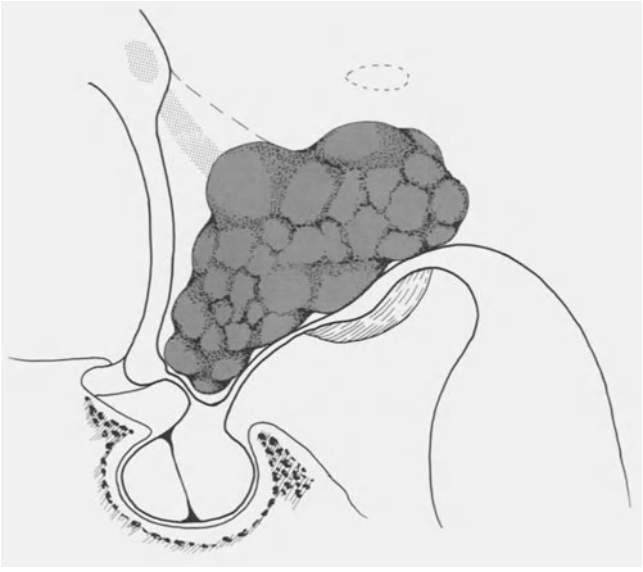


Figure 10.103. Hypothalamic *glioma*, different from the germinoma, grows from the region of the optic chiasm, between the supraoptic and infundibular recesses of the III ventricle. The mass invariably fungates into the III ventricle, often becoming giant in size, and lobulates into the intrapeduncular cistern.

pathways, to determine whether the tumor involves just one optic nerve, or the entire structure. Good visualization of the optic pathway suffices for "biopsy," rendering surgical opening of the pathways and removal of a small portion of the neoplasm unnecessary. In fact, one is adivsed to avoid biopsy if the visual observations are diagnostic of optic pathway glioma. If it is decided to take the biopsy, this should be done at the most expanded, avascular, necrotic or cystic appearing area.

A #15 blade is used to incise the tissue, and approximately 2 mm^3 neoplasm is removed with micropituitary biopsy forceps.

Hypothalamic Glioma

This tumor, as almost all of those involving cerebral and ependymal parenchyma in children, has a tendency to degenerate from benign to malignant. The tendency seems to be reported more often recently, but there are no data to permit us to conclude that this is a rule. About 10% of the opticochiasmatic-tumor children treated by Dicks et al. developed anaplastic changes over 15 years. Radiation therapy may be the cause.

There is no reason to biopsy a glioma within the hypothalamus. Also, there is no justification for opening the lamina terminalis, entering the III ventricle, and inspecting visually the hypothalamus. Both of these procedures present unacceptable risks of damage to the hypothalamic nuclei, and no advantages.

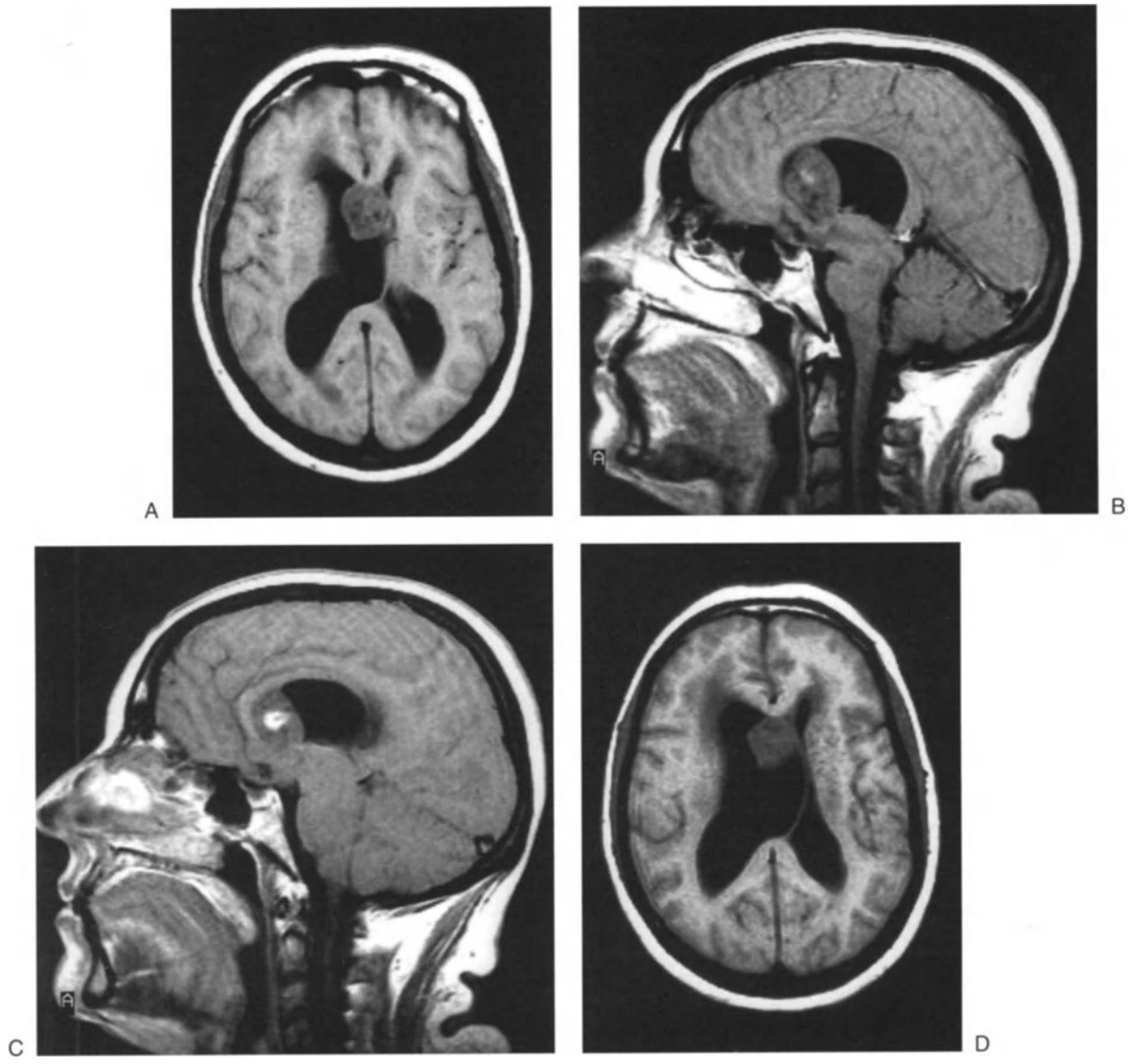


Figure 10.104. This figure (a series of axial and sagittal sections, T1 and T2 weighted sequences pre- and post-gadolinium injections) reveals almost the entirety of the above-described variables. In (A) one may identify the point at which the spongioblastoma is growing from the lamina terminalis, but only abutting upon the ventricular surface of the genu of the corpus callosum, causing asymmetrical hydrocephalus, as it expands within the right lateral ventricle; (B) this section, however, reveals the tumor to be growing both from the ventricular surface of the genu of the corpus callosum and the septum pellucidum; (C) in this slightly off the midline parasagittal cut the tumor is seen to grow from the hypothalamus but to be independent of the lamina terminalis; (D) in this cut, 1.3 mm to the left of the mid-sagittal plane one sees that the tumor is growing from the ventricular surface of the area olfactoria and the lamina terminalis as it expands posterior to the rostrum and genu, involving the columns of the fornix. (E–G) see p. 304.

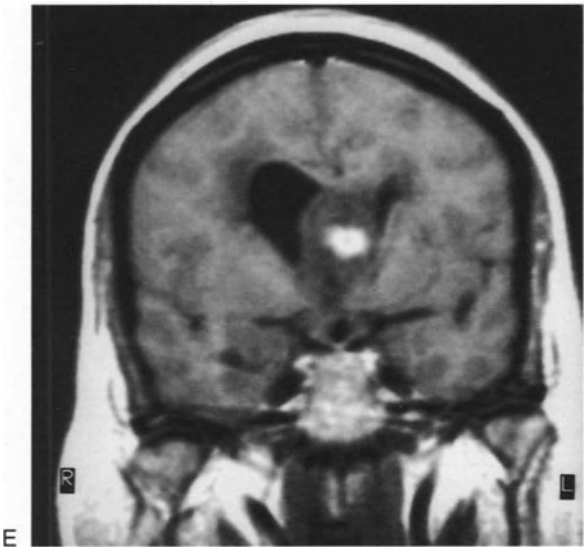


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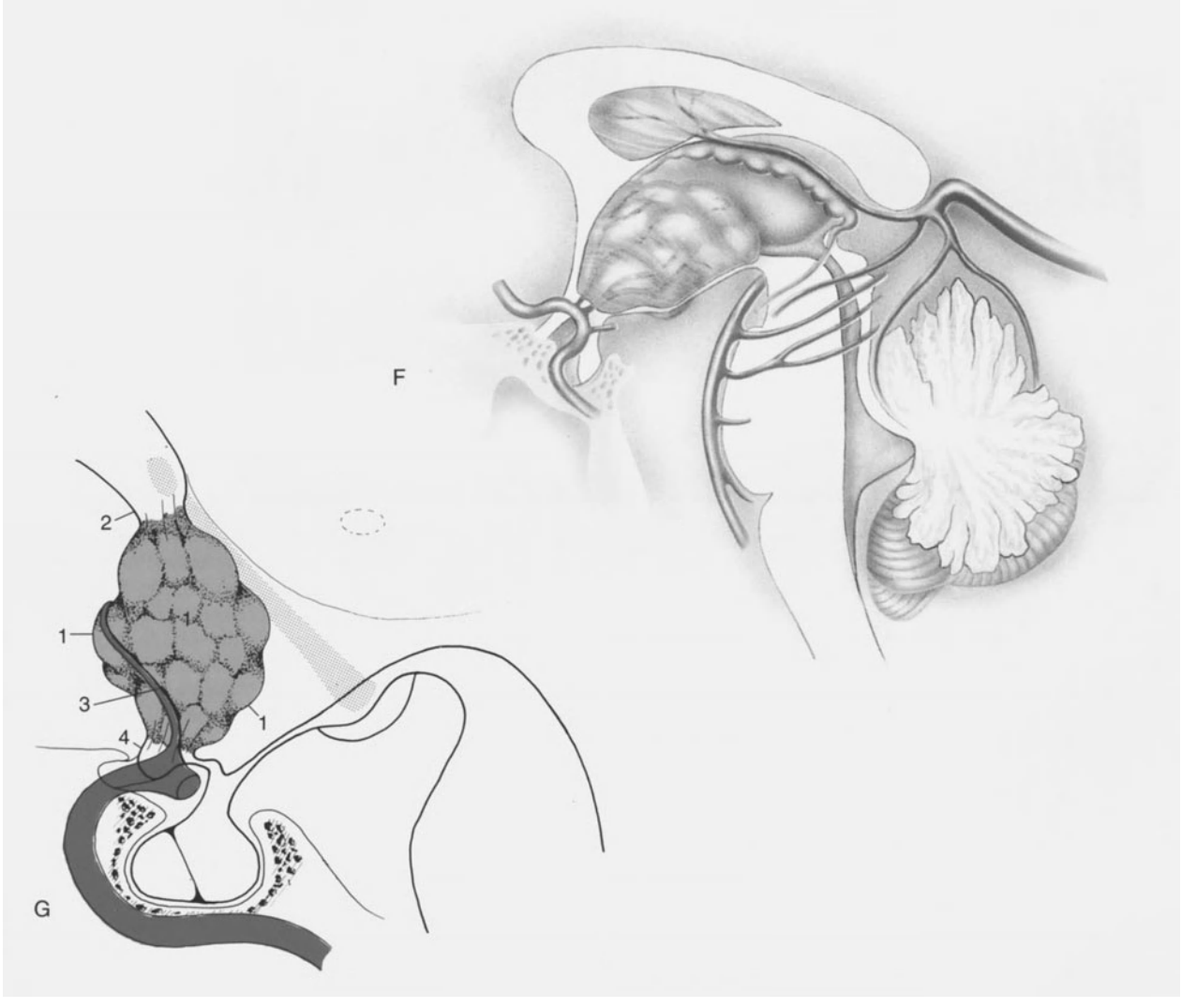
Figure 10.104. (E) This coronal section reveals the tumor growth along the septum pellucidum, inferiorly at the region of the mammilothalamic tract and the mammillary bodies, and superiorly at the inferior surface of the corpus callosum. In sum, tumors involving the septum pellucidum and lamina terminalis by definition may involve the columns of the fornix, the optic nerve, the optic chiasm, the intraventricular portion of the anterior perforated substance, the hypothalamus, the mammillary bodies, etc. In (F) such a tumor is schematically represented in the sagittal section, to allow the reader to identify the above possible contiguous relationships of such a neoplasm and the vascular supply from perforating branches of the anterior circle and the choroid plexus of the III ventricle. (G) The glioma (*1*) expanding from lamina terminalis bulges posteriorly into the III ventricle and invades the rostrum of the corpus callosum (*2*). The A-1 segment of the anterior cerebral artery (*3*) is elevated and deformed, as the tumor extends inferiorly into the region of the optic chiasm (*4*).



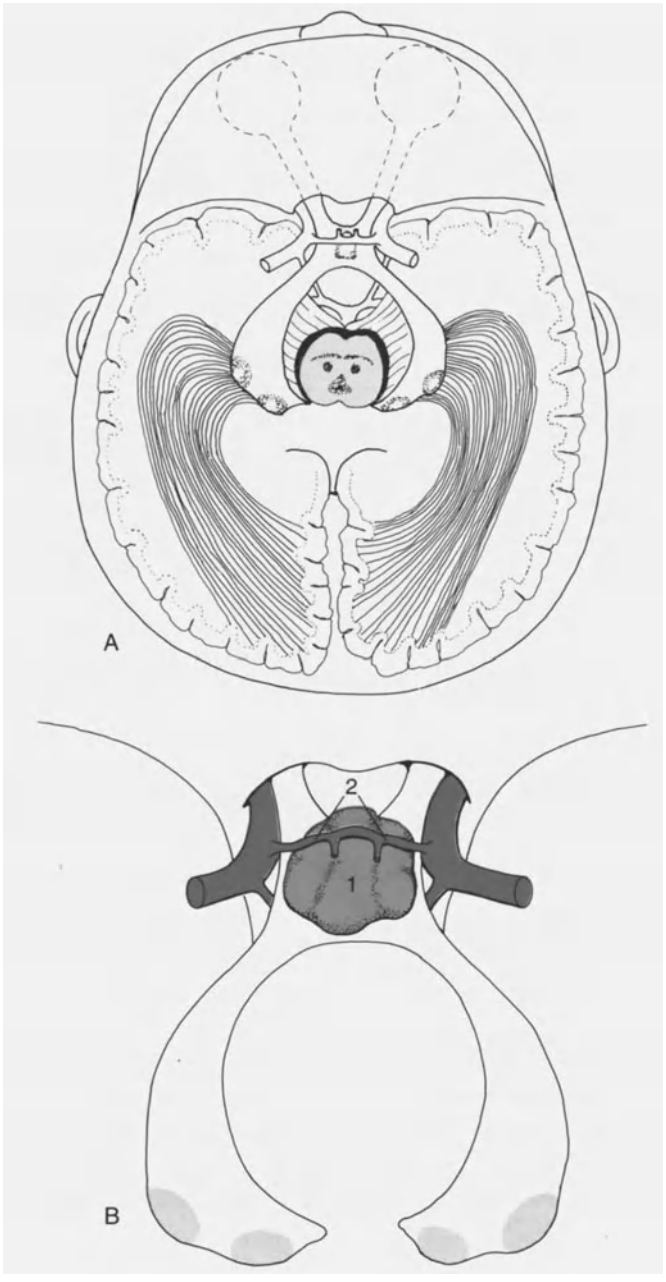
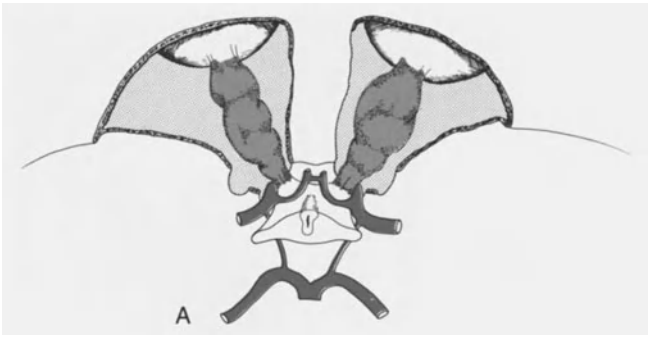


Figure 10.105. (A) The optic pathways are reproduced here to permit a visual point of reference for discussion of gliomas of the optic pathways. (B) This is a coronal drawing of a glioma of the lamina terminalis (*1*) illustrating how its anterior and superior bulge deforms and stretches the A-1 segment of the anterior cerebral arteries (*2*).



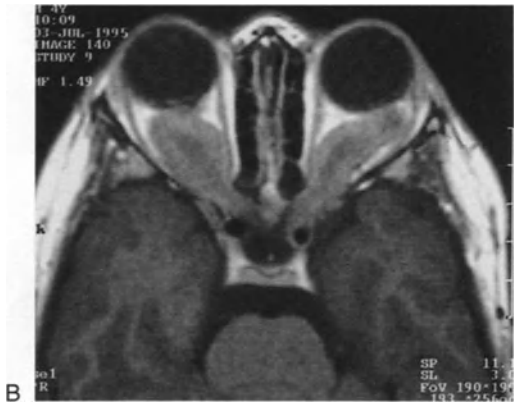


Figure 10.106. (A) Bilateral optic pathway gliomas are not uncommon in children with von Recklinghausen's disease. Here, tumor invasion of the intraorbital portions of the optic nerves is illustrated. (B) Bilateral optic nerve glioma. This axial spin-echo post-gadolinium image reveals medium high signal intensity lesions occupying both optic nerves, extending through the optic foramina and into the optic chiasm.

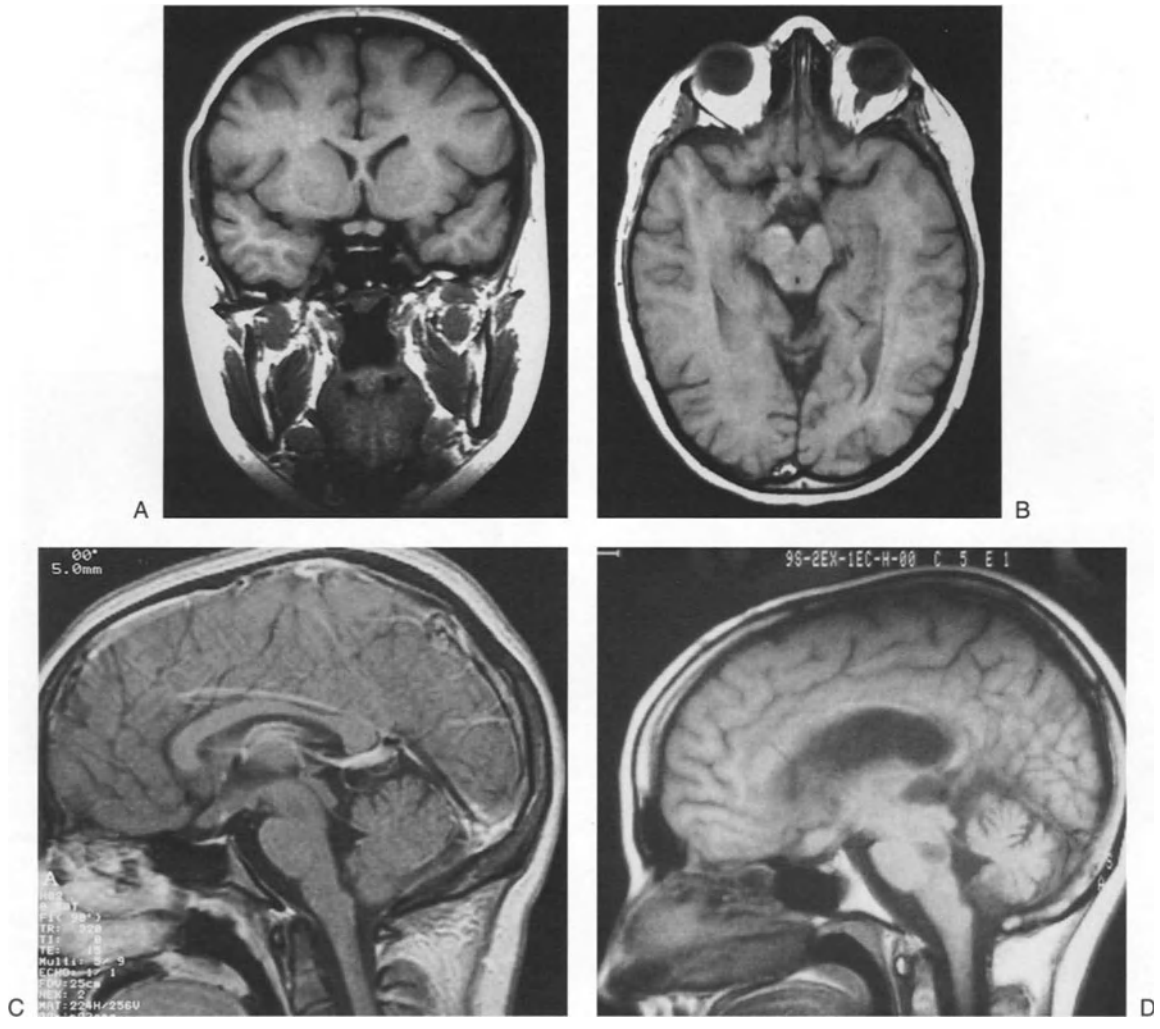
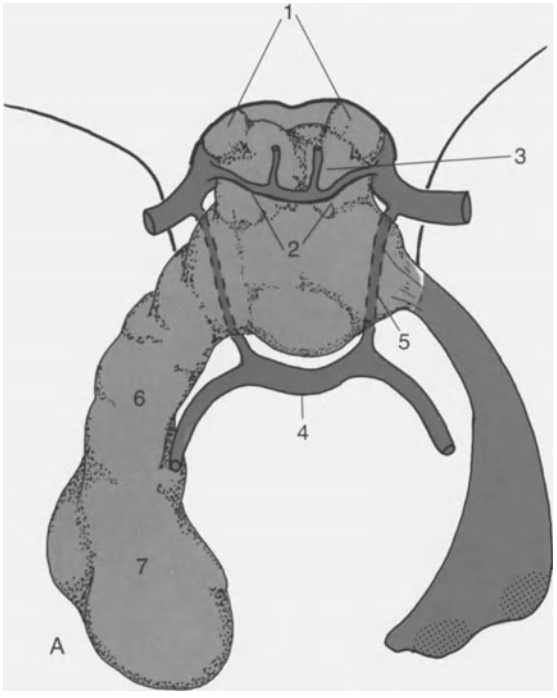
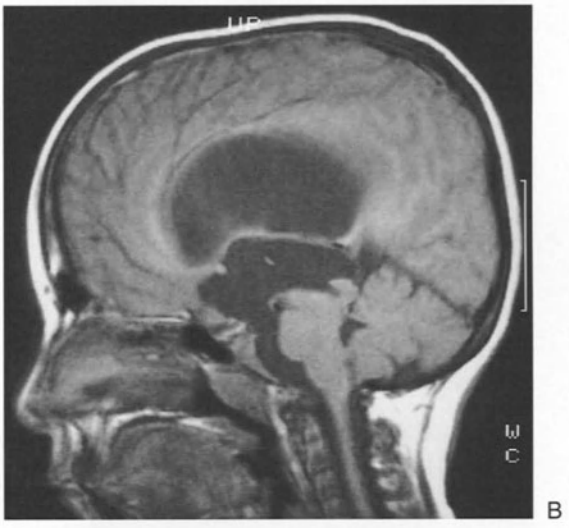
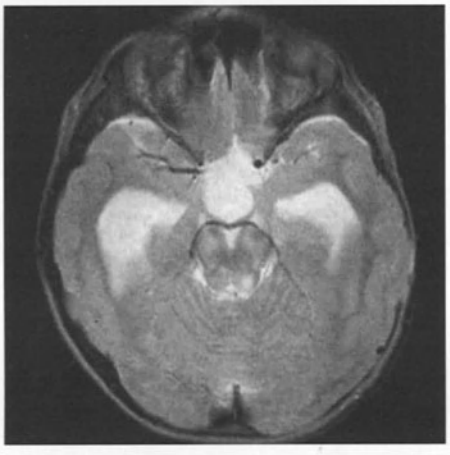


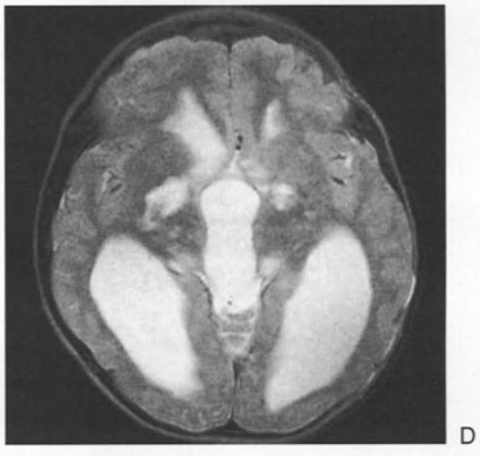
Figure 10.107. (A) This coronal section reveals the uniform circumferential neoplastic expansion of both optic nerves, and (B) the axial section puts into relief neoplastic involvement of the optic nerves, the optic chiasm, and the optic tracts. (C) In the sagittal post-gadolinium study one sees uniform neoplastic expansion of the optic chiasm and the nonenhancing mass in the tegmentum of the mesenchephalon. The aqueduct of Sylvius is patent and the ventricles are not dilated. In (D), following chemotherapy, the chiasmatic mass has diminished somewhat, and there is aqueduct occlusion (secondary most probably to periaqueductal neoplasm, triventricular hydrocephalus, and a relatively large pontine mass). In both (C) and (D), one sees neoplastic involvement of the lamina terminalis.

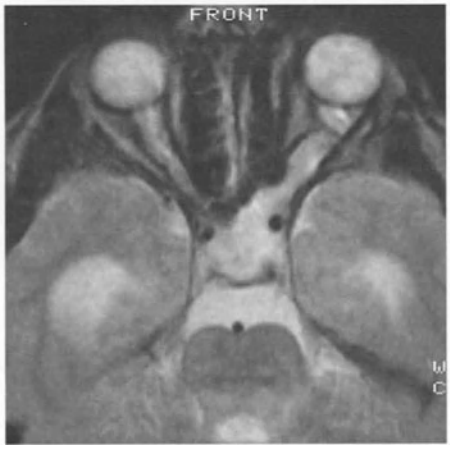
Figure 10.108. (A) Optic pathway gliomas may involve the entirety of the intracranial optic nerves bilaterally (*1*), elevating and deforming the A-1 segments of the anterior cerebral arteries (*2*) at the transition point between optic nerve and optic chiasm (*3*). Tumefaction of the optic chiasm may fungate posteroinferiorly into the interpeduncular cistern and subsequently displace the basilar fundus and both posterior cerebral arteries (*4*) posteriorly, stretching the posterior communicating arteries (*5*) and displacing them inferiorly. It is not unusual for optic pathway gliomas to extend along the geniculocalcarine tract (*6*) and to expand within the temporal lobe (*7*). This child has neurofibromatosis – *1*, with glioma involving the entirety of the optic pathways (from the optic nerves through to the optic radiations), dysplasic lesions in the periaqueductal area causing aqueductal stenosis, and secondary triventricular hydrocephalus. In (B) the aqueductal stenosis is illustrated, and the triventricular hydrocephalus in (C) and (D). In (C) one also sees multiple areas of high signal intensity on the T2 weighted images within the periaqueductal regions. These are typical of the dysplasias one sees in neurofibromatosis patients. The left optic nerve is enlarged, and there is a high signal intensity in the post-gadolinium studies. In (E) and (F) one sees that the lesion involves the entire left optic nerve and the left half of the chiasm. ►











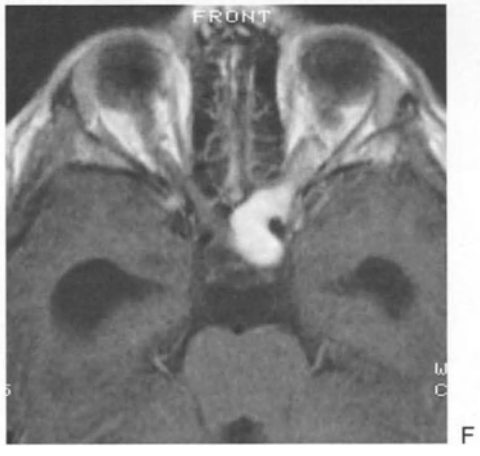
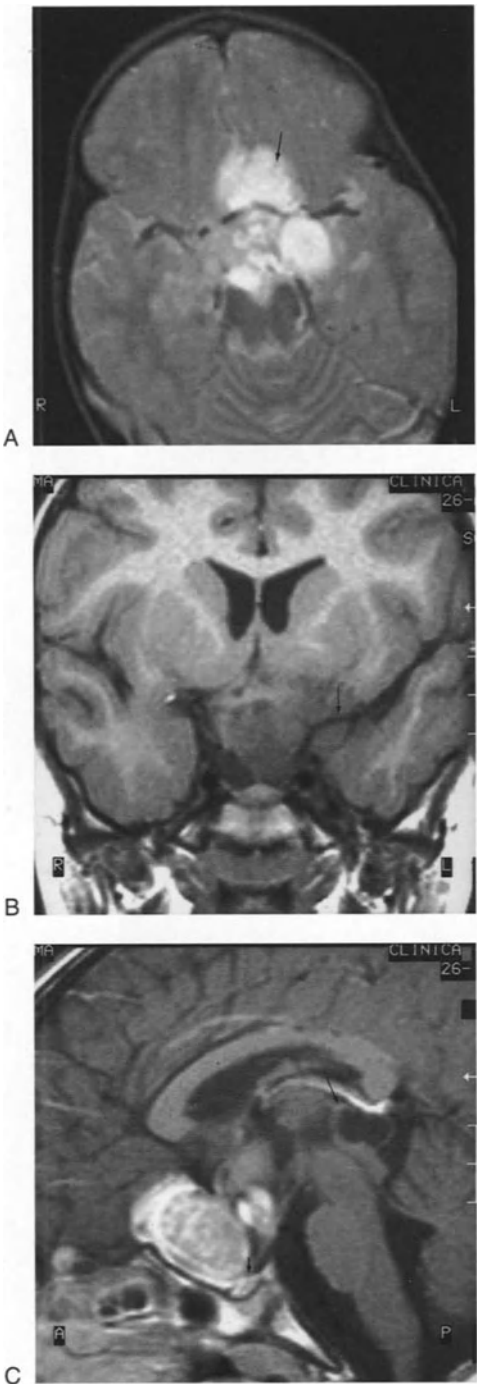


Figure 10.108. Legend see p. 306.



◀ **Figure 10.109.** This is a pilocytic astrocytoma of the optic chiasm. (A) Axial spin-echo image revealing a very large mass of iso-high signal intensity occupying the suprasellar area and the interpeduncular cistern. A portion of the mass (*arrow*) extends anterior to the A1 segments of the anterior cerebral arteries. (B) This coronal section reveals low signal intensity mass, not identifiable chiasm, and tumor encircling both the left internal carotid and left middle cerebral arteries. (C) Sagittal post-gadolinium image revealing a large, lobulated, enhancing mass which compresses the pituitary gland (*small arrow*). There is no enhancement of the superior portion of the tumor, which is pushing the anterior wall of the III ventricle posteriorly. The *long arrow* indicates a cystic tumor of the pineal gland, which is compressing the anterior colliculi.

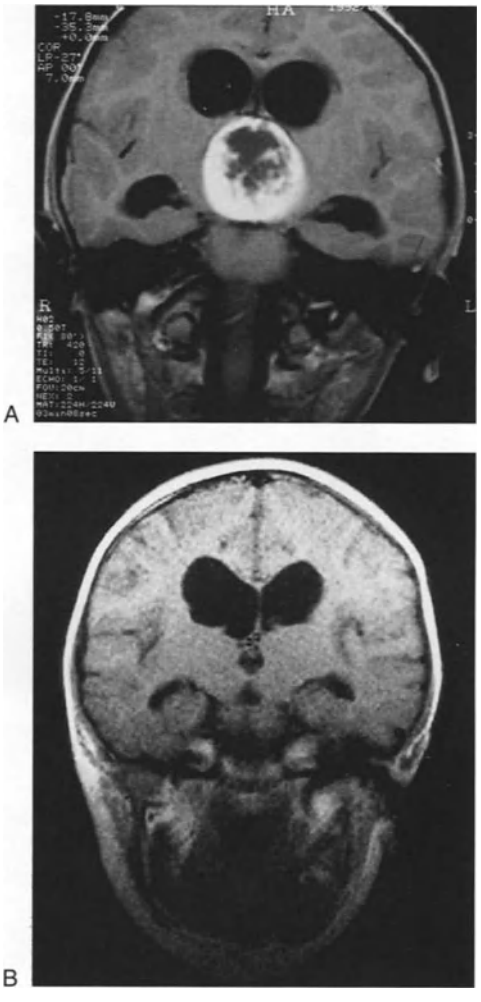


Figure 10.110. Chiasmatic glioma: (A) This coronal post-gadolinium MRI study reveals the presence of an enormous hypothalamic mass extending from the supra- and retrochiasmatic (floor of the III ventricle structures) areas. The tumor was treated (by Prof. V. Valentino) in three sittings, roughly at 3-month intervals, of radiosurgery. (B) This coronal post-gadolinium study reveals almost complete tumor disappearance.

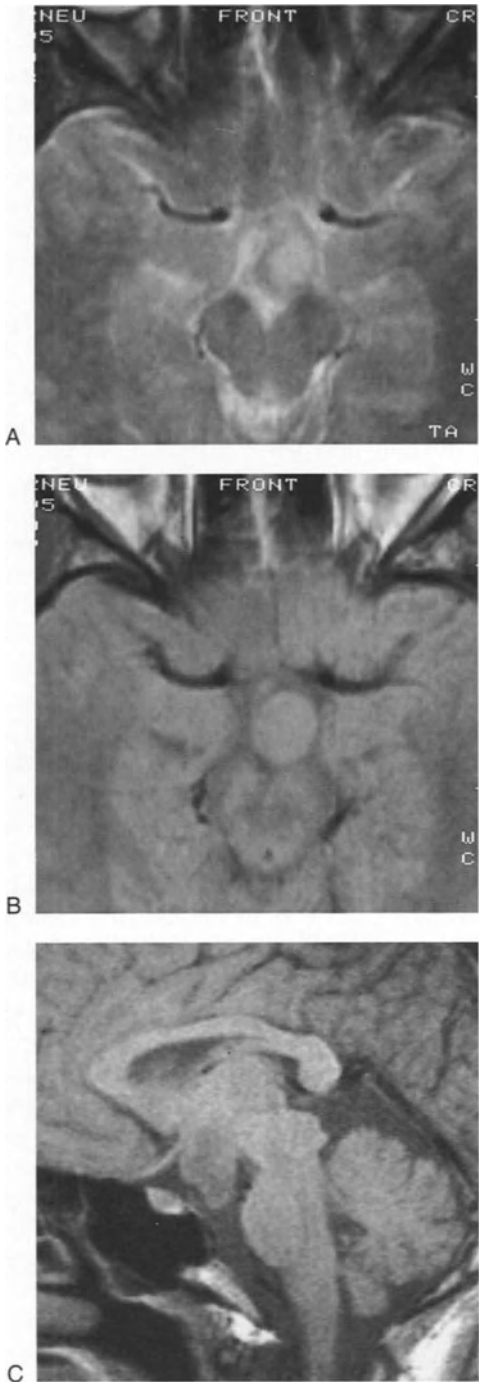


Figure 10.111. Hypothalamic hamartoma. This very characteristic tumor is illustrated in the axial spin-echo 2,400/90 and 2,400/15 (A and B, respectively) to have the same signal intensity as gray matter, and sharp borders. In the sagittal section (C), the tumor hangs from a rather sessile base into the interpeduncular and pontine cisterns.

Figure 10.112. Hypothalamic glioma. Again, the hypothalamic glioma, as the optic pathway glioma, really should not be operated on, since the diagnosis may be made preoperatively. However, in the event either of an inability to make this diagnosis (because of inadequate clinical information) or uncertainty concerning its anatomy and nature, it should be approached through a unilateral frontal craniotomy. The optic nerve (*1*) is normal, but the internal carotid artery (*2*) is displaced remarkably from this structure, by a hypothalamic glioma (*3*), which is expanding anteriorly beneath the optic chiasm and opening up the interopticocarotid space.

Craniopharyngioma

Introduction

The clinical picture, imaging characteristics, demographic incidence, and neuropsychological complications of *craniopharyngioma* are well understood and agreed upon by all. Unfortunately, the therapeutic modality of choice, operative results, and complications of ionizing therapy remain matters of great disagreement among experienced clinicians treating craniopharyngioma. With regard to operative indications, there are further disagreements concerning whether the cystic portions of the tumor, most common in childhood, should simply be drained, whether a subtotal resection is sufficient, or whether one should "push" for a total (?) resection.

Following this line of reasoning, there is still further disagreement as to whether, whichever of the previous three surgical procedures is selected by the surgeon, radiation therapy should follow the operative procedure, with a wide swing between the purists who only operate and the eclectics who operate and then ask for an ionizing therapy consultation. At the other end of the spectrum stand those who maintain that the proper treatment in childhood is cyst drainage or partial resection

followed by radiation therapy. Intermediate between the two are those who recommend cyst drainage and intracavitary injection of radioactive isotopes (brachytherapy) for the cystic or primarily cystic tumors. The heat in this debate comes from the fact that all three groups are reporting more or less identical results.

The radiotherapists and brachytherapists are fairly well able to understand one another's position in that both endeavor to drain the cystic lesion and then treat the neoplastic area with ionizing therapy. It is not clear why the proponents of radical resection routinely fail to refer their patients for some form of X-ray therapy.

Those tumors which do not show calcification on skull X-ray films carry a significantly better 5-year survival, are generally characteristic of adults, and the association "absence of calcification: craniopharyngioma" is more expressive of the squamous papillary than the adamantinous type tumor is valid. The predominantly solid, noncalcified squamous papillary tumor occurs mostly in the adult, whereas the usually cystic and calcified adamantinous tumor occurs in adults and children. It is rather evident, therefore, that the squamous papillary tumor which occurs in approximately one-third of adult cases is truly a rarity in childhood, only one case having been reported, by Zülch in a 15-year-old boy.

The postoperative outcome for squamous papillary tumors is very much better than for adamantinous tumors, patients with the squamous papillary type having been reported by Kahn to be free from recurrences [137]. In fact, there have been no recurrences in the patients with squamous papillary tumors presented in his series; but, again, one must be cautious regarding the matter of follow-up in light of the fact that 50% of craniopharyngioma recurrences occur after 5 years [138, 139]. The absence of islets of tumor in brain tissue adjacent to adamantinous craniopharyngiomas, so often suggesting tumor recurrence in the adamantinous type, has not been observed in the 15 squamous papillary craniopharyngiomas the authors examined. This is of great help in allowing the neurosurgeon operating on childhood craniopharyngioma to understand *that there are islets of tumor invading the surrounding brain tissue*. Hence the worse prognosis for adamantinous tumors in children than adults, and the greater risk of recurrence.

There are two types of craniopharyngioma, the squamous papillary and the adamantinous, allowing the reader to speculate that the adamantinous type may originate from ectopic embryonic cell rests of enamel organs. There is a difference in the biological activities of these two tumors, as well as in their ability to provoke calcification and to form intraneoplastic cysts. If nothing else, this may help greatly to explain the very different postoperative clinical courses followed by adults and children, the relative ease with which adult craniopharyngiomas are resected, the great difficulty in separating childhood craniopharyngiomas from the cisternal surface of the floor of the III ventricle, the "absence" of craniopharyngioma invasions of parenchyma in the adult, and the high incidence of parenchymal islets of tumor in the child.

It is surprising, indeed, to realize that until 1977, when the use of computed tomography became commonplace in the Western world, only 35 cases of symptomatic *Rathke's cleft cysts* had been reported. These cysts, considered to be vestigial remnants of Rathke's pouch, vary considerably in size from almost microscopic to gigantic dimensions, thus being reported in 4.2%–26% of routine autopsy series, but becoming clinically obvious only when expansion of the mass is such as to cause endocrine disturbances, headache, or visual field defects. This wide discrepancy between the high incidence as an incidental finding at autopsy and the extreme rarity as a clinically diagnosable entity is more than just a curiosity: Rathke's cleft cysts have much in common with a somewhat more frequent and certainly much better known developmental space-occupying lesion, the craniopharyngioma.

Although the terms "Rathke's cleft cyst" and "Rathke's pouch cyst" are intuitively understood to indicate the same pathoanatomical lesion, the other names used to describe this "malformation/tumor"– "pituitary cyst," "mucoid epithelial cyst," "intrasellar epithelial cyst," "colloid cyst of the pituitary gland"– are not as readily understood to indicate the same cystic pathology derived from true remnants of Rathke's pouch: a dorsal diverticulum, lined by (ectodermal) epithelial cells, extending from the stomadeum toward the end of the 3rd week of embryonic life. At approximately the same time, the diencephalon extrudes as a downgrowth of neuroepithelium, the infundibulum. When the infundibulum and Rathke's pouch come into contact with one another, the neck of the latter becomes occluded at the buccal-pharyngeal junction. Approximately 1 week later, during the 6th week of embryonic life, the oral epithelium and Rathke's pouch separate from one another, and then the pars distalis of the hypophysis develops from the anterior wall of the pouch as the posterior wall remains as a poorly defined pars intermedia. The lumen of the pouch is reduced to the residuum, which is Rathke's cleft, the structure generally considered to be the origin of Rathke's cleft cysts. As is most common in biology, there are different theories: some consider Rathke's cleft cysts to take their origin from the neuroepithelium, others see the origin as in the endoderm, and still others regard the cysts as a metaplasia of anterior pituitary cells.

From a light microscopic point of view, these cysts are composed of a single layer of cuboidal or columnar epithelial cells which often, but not invariably, are ciliated. The epithelium itself may be simple or stratified,

and may have squamous components, in which case one may identify the pathogenetic cellular element for symptomatic cysts. The range between cuboidal, columnar, and stratified squamous, ciliated epithelium, is wide and continuous, leading many to suggest that Rathke's cleft cysts and craniopharyngiomas may share a common cellular origin from Rathke's pouch.

What is most interesting for a lesion which has some developmental and histological characteristics in common with the craniopharyngioma is the almost nonexistent, or at least the very low, incidence of recurrence, if the lesion is resected rather than simply aspirated. There appears also to be a very low complication rate resulting from a direct surgical approach and –a pleasant surprise – good reason to expect some postoperative recovery of neurological or endocrine deficits.

In sum, Rathke's cleft cyst is a rare clinical entity, a rather common anatomical variant found at autopsy, a nonfatal, intracranial space-occupying lesion, a tumor which may be treated equally well by either trans-sphenoidal or subfrontal approaches, an almost impossible lesion to diagnose histologically preoperatively, and a congenital tumor which has a low incidence of postoperative recurrence.

Surgical Considerations Regarding Craniopharyngioma Classification

In 1910 Dean Lewis [140] was the first surgeon to remove a craniopharyngioma. The longest recorded survival following craniopharyngioma surgery was in a patient operated on by Harvey Cushing [141] in 1923, a 50-year survival. This case puts into relief what may well be the most telling expression concerning craniopharyngioma surgery: since scarcely any two craniopharyngiomas are precisely alike and since those of similar size and character show such differences of behavior, the course of any given tumor is impossible to foretell.

Raimondi and Rougerie, in their work on the clinical and surgical management of this tumor, cited Trippi and coworkers as having suggested that inoperability be determined by any one of the following conditions: prefixed optic chiasm, marked upward extension into the floor of the III ventricle, extensive unilateral or bilateral temporal lobe involvement, firm adhesions to the optic chiasm. Consequently, one concludes that the most important factors in evalutating a child with a craniopharyngioma for surgery are anatomical location of the tumor and the resultant morbidity.

The vascular and parenchymal changes permit an appraisal as to whether the tumor mass should be partially or totally excised, and the predictability of ultimate prognosis regarding survival and quality of survival. This is true because the craniopharyngioma, though it may invade cerebral parenchyma, causes its major damage by *growth* (resulting in displacement of the optic pathways and hypothalamus) and *stretching* (resulting in attenuation and occlusion of the perforating branches of the circle of Willis).

A major portion of craniopharyngioma growth results from accumulation of fluid within multilobular cavities. Both solid tumor and calcifications comprise a significant, though generally minor, percentage of the total tumor. To a certain extent, CT allows one to evaluate the presence of calcifications, solid neoplasm, and cystic compartments within the "tumor" mass, but does not provide precise information because there is overlap of densities of highly liquified cellular aggregates and highly cellular cystic chambers. MRI answers these questions.

Classification of Craniopharyngioma

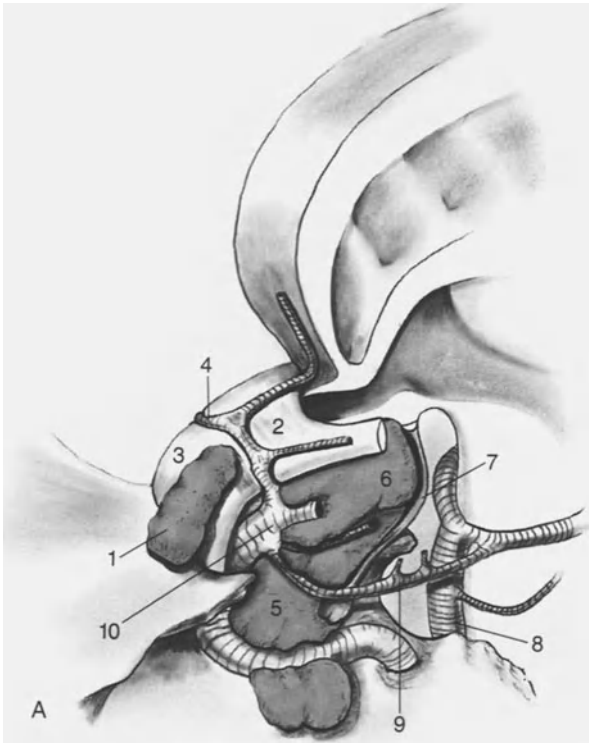
The recommended surgical classification of craniopharyngiomas, dividing them into five distinctly different anatomical forms (locations), is that proposed by Jacques Rougerie [136]:

Prechiasmatic Craniopharyngioma
(Figs. 10.113, 10.114)

The tumor may be intra- and suprasellar, or almost exclusively suprasellar, but the primary expansion of the mass is into the prechiasmatic area. It grows anterior to the optic chiasm, between it and the optic nerves superiorly and internal carotid arteries inferiorly, in its extension over the planum sphenoidale and anterior to the lamina terminalis of the III ventricle. Very rarely, it grows first over the planum sphenoidale and then turns posteriorly to expand along the suprachiasmatic surface, engulfing this structure, and displacing posteriorly the lamina terminales.

Craniopharingyomas that rupture through the *diaphragma sellae* to expand anterior to the optic chiasm extend superiorly, anterior and inferior to the chiasm.

This tumor growth elevates the optic chiasm and, with it, the A-1 segments of the anterior cerebral arteries. The prechiasmatic cistern is obliterated as the tumor grows anterior to the chiasm, and then superior to it, displacing it posterosuperiorly, stretching and verticalizing the two optic nerves. The supraoptic recess of the III ventricle is obliterated and the lamina terminalis is displaced posterosuperiorly. The optic nerves and chiasm are damaged primarily because of the direct compressive force, but also because of stretching and attenutation of branches from A-1 to the optic chiasm. For these reasons, funduscopic examination reveals optic atrophy, and visual field examination reveals defects indicative of optic nerve and anterior chiasm involvement. The visual field defects are often "explosive" in oneset and progression.



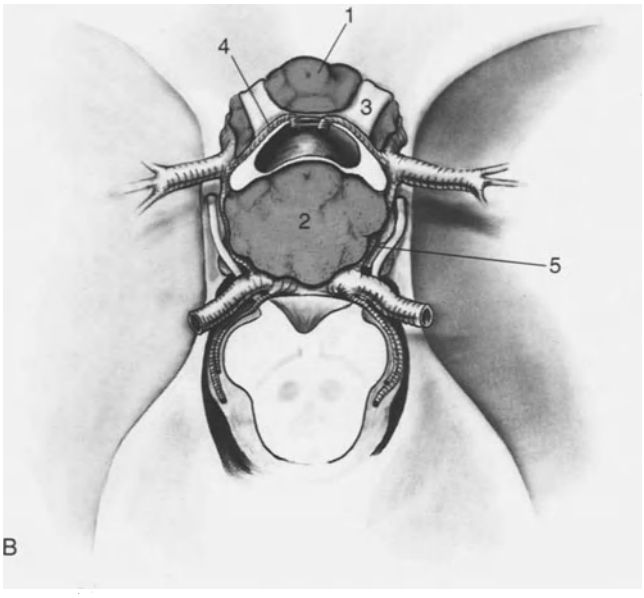
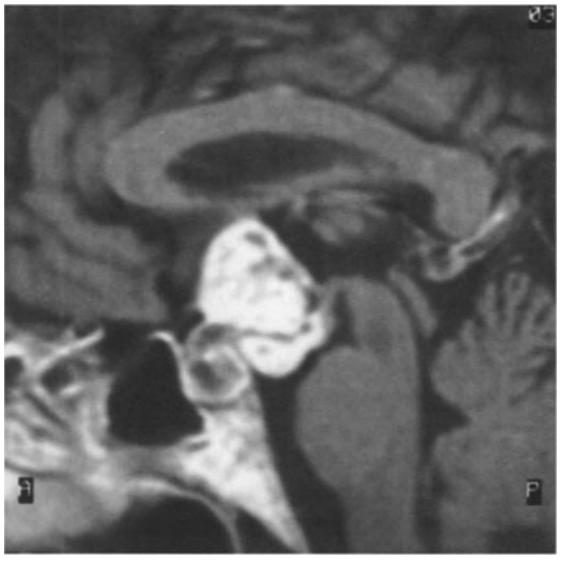


Figure 10.113. (A) The prechiasmatic extension of tumor elevates the optic chiasm (*2*) and verticalizes the optic nerve (*3*). A-1 (*4*) is elevated with the chiasm, so that it may be used as an angiographic indicator of whether there is prechiasmatic extension of tumor. There is always infrachiasmatic (*5*) and retrochiasmatic (*6*) tumor present, since rupture through the diaphragma sellae is first followed by expansion of tumor within the infrachiasmatic area, then anterior and posterior growth, with the latter displacing the pituitary stalk (*7*) backward and then rupturing into the III. The basilar artery (*8*) is not deformed, although the posterior comunicating artery (*9*) is stretched as the tumor elevates the internal carotid (*10*) and insinuates itself between carotid and optic nerve. Tumor may grow into the cavernous and sphenoid sinuses. (B) This is an axial representation of prechiasmatic (*1*) tumor extension, putting into relief the fact that this represents the smallest tumor lobule, with the major portion elevating the chiasm and expanding posterior to it (*2*). The stretching of the optic nerves (*3*), A-1 (*4*), and the posterior communicating artery (*5*) is illustrated.



◀ **Figure 10.114.** The craniopharyngioma illustrated in the drawings (Figs. 10.113, 10.120) and MRI study is most unusual: it has grown out of the prechiasmatic space and then extended posterosuperiorly, displacing the lamina terminalis posteriorly by insinuating itself between the subcallosal gyrus and the lamina terminalis. The anterior cerebral artery is visible immediately anterior to the subcallosal gyrus. To understand better the pathological anatomy resulting from the expansion of this craniopharyngioma, a drawing of the normal suprachiasmatic anatomy and one of the extensions of the tumor are included. The important structures illustrated are A1, A2, the optic chiasm, and the lamina terminalis, as shown in Fig. 10.120. The extension of the tumor revealed in the subsequent MRI studies is illustrated in Fig. 10.113A. The optic chiasm is elevated by the infrachiasmatic portion of the tumor, the olfactory nerve is displaced downward by the prechiasmatic extension, the lamina terminalis is displaced posteriorly by the suprachiasmatic extension, and tumor is seen infiltrating itself between the optic chiasm and the posterior communicating artery. This post-gadolinium sagittal image reveals a hyperintense mass occupying the suprasellar area, extending from the sella turcica, in front of the optic chiasm, displacing the lamina terminalis posterosuperiorly. There is tumor mass on either side of the internal carotid artery.

Carotid (or MRI) angiography reveals elevation and posterior displacement of the A-1 segment of the anterior cerebral arteries, permitting the specific diagnosis of the prechiasmatic location of the tumor. The rationale for, and attempt at, complete resection is indicated by the venographic studies which reveal elevation of the veins of Rosenthal at their origin in the anterior perforated substance.

All tumors of significant enough size to bulge through the prechiasmatic space also expand widely *beneath and behind* the elevated optic chiasm. Therefore, one must be prepared to remove retrochiasmatic lobules. The enlarged prechiasmatic area, made ample by partial tumor removal, facilitates this. However, no single exposure technique applies to any single anatomical displacement events.

Intrasellar Craniopharyngioma

The intrasellar craniopharyngioma, by definition, is located entirely within the sella turcica, beneath the *diaphragma sellae*, and causes enlargement of the sella turcica's volume, partial or total destruction of the clinoids, invasion of the sphenoid sinus, or compression and displacement of the cavernous sinuses. When visual field deficits are identified, one should consider the possibility that the *optic nerves are being compressed in their course along the superolateral borders of the sphenoid sinus, not in their subarachnoid location.*

The most common clinical presentation for intrasellar tumors results from endocrinologic deficit. Significant portions of the pituitary gland may remain intact, justifying every effort to identify it at the time of surgery. Previously, these tumors have been considered to present with signs indicative of pituitary dysfunction, giving cause to believe that some, or all, of the pituitary function could be preserved since the gland may not be totally destroyed. More recently, we have come to learn that intrasellar tumors may extend into the sphenoid sinus, fungating within it and filling it, compressing the medial and inferior portions of the optic nerves in their course along the superolateral aspect of the sphenoid sinus.

Retrochiasmatic Craniopharyngioma (Fig. 10.115)

The tumor mass extends posterior to the optic chiasm, between it and the optic tracts, displacing and, at times, invading both the hypothalamus and III ventricles.

Approximately one-third of the patients with retrochiasmatic craniopharyngiomas are under 10 years of age. Less than 25% of these tumors may be "totally" removed *biologically*, e.g., permanent cure. The early and late combined mortality rate is high, ranging from 20% to 30%, the morbidity, higher still.

Posterior and superior growth of the craniopharyngioma from the sella turcica allows it to expand within the area delimited by the posterior half of the circle of Willis. The obstruction of the basal cisterns and the posterior portion of the III ventricle results in *hydrocephalus*, an increase in *intracranial pressure*, and optic atrophy or *papilledema*. Optic atrophy is less, and papilledema more, common. The increase in intracranial pressure, compounded by *biventricular hydrocephalus*, may be compensated by a ventriculoperitoneal shunt, which should be bilateral if it appears that the tumor mass is occupying all of the III ventricle, obstructing one foramen of Monro. *Hydrocephalus is an ominous sign in children with craniopharyngioma.*

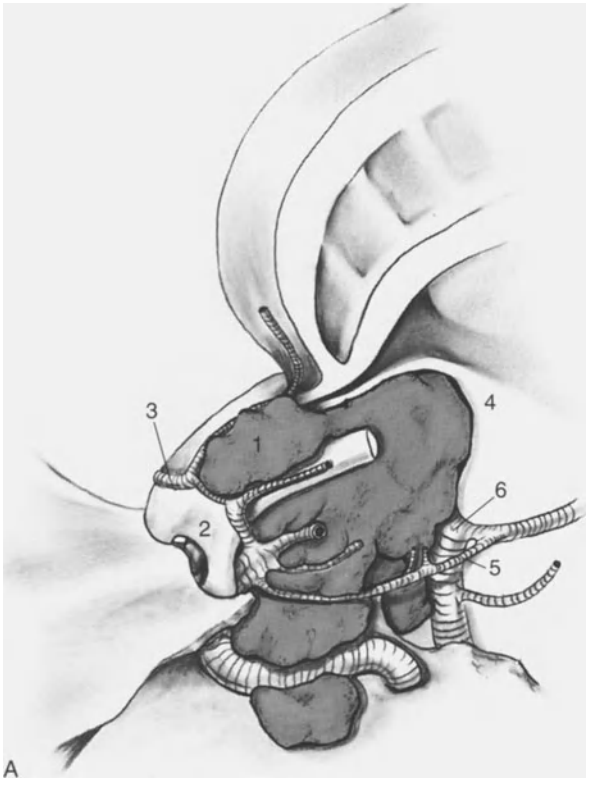
The location of these tumors behind the optic chiasm is particularly grave in that it renders surgical removal more difficult and risk of damage to the optic pathways and hypothalamus greater. Since these patients suffer an increase in intracranial pressure, compressive effects on the optic pathways, varying degrees of destruction of hypothalamic nuclei, and attenuation or occlusion of the thalamo-perforating arteries, surgical manipulation may be further complicated by permanent damage to already severely compromised neural structures.

Retrochiasmatic extensions more often adhere to, or invade, the hypothalamus. The tumor may engulf the posterior communicating arteries and their thalamoperforating branches. The combination of damaged hypothalamus plus a severe increase in intracranial pressure may be complicated by postoperative gastrointestinal hemorrhage. Third, IV and VI cranial nerve palsy results either from expansion of the tumor mass within the circle of Willis or surgical manipulation for its removal. Pure retrochiasmatic extension with either a prefixed chiasm, or an optic chiasm displaced anteriorly by the tumor mass, presents the neurosurgeon with one of the most challenging of operative procedures, with *regard more to judgement than technique.*

Removal of the tumor encumbers one to do so "piecemeal," through anatomical planes between the oculomotor nerves and carotidobasilar systems. *Total resection of these tumors should not be pursued at all costs.* Partial resection may result in dramatic clinical improvement and prolonged remission periods, especially if the complicating hydrocephalus is successfully and continuously compensated by a functioning shunt system. Radiosurgery is very effective.

Les Formes Géantes

These giant forms "explode" from the sella turcica, expanding into the prechiasmatic region, directly superiorly, into the retrochiasmatic region, and at times into the posterior fossa and sylvian fissures. The clinical evolution is punctuated with rapid and multiple recurrences. The sella turcica is completely destroyed and



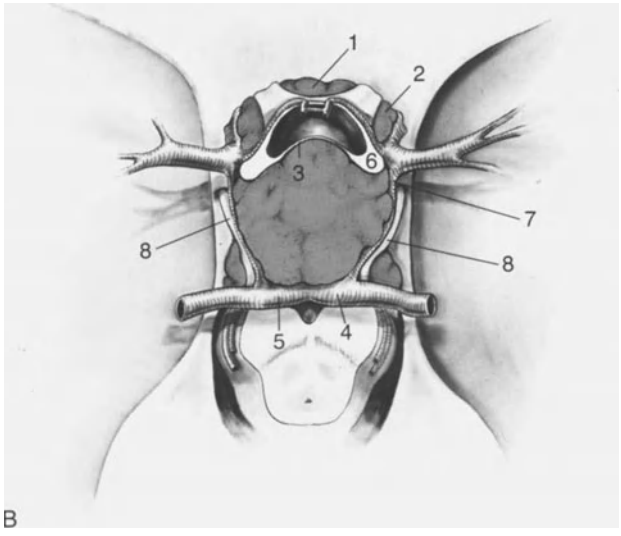


Figure 10.115. (A) Retrochiasmatic tumor extensions (*1*) result in forward displacement of the optic chiasm (*2*), an anatomical change illustrated angiographically by forward and, at times, remarkably downward displacement of A. (*3*) The tumor elevates remarkably the floor of the III, at times fungating into it (*4*), and engulfing the pituitary stalk so the surgeon may not identify it. There is depression and stretching of the posterior communicating artery (*5*), secondary to lateral tumor expansion and posterior displacement of the basilar artery (*6*). (B) Retrochiasmatic tumor. This axial view illustrates that there is very little prechiasmatic space (*1*), and that the tumor is opening the interopticocarotid space (*2*), as well as deforming the hypothalamus (*3*) and displacing the basilar (*4*) and posterior cerebral (*5*) arteries backward. The difficulties in resecting it completely stand out. It may be partially removed and debulked, by working through the interopticocarotid space and the retrochiasmatic region between the optic tract (*6*) and posterior communicating artery (*7*). The III cranial (*8*) nerve is displaced laterally.

the tumor extends into the anterior, middle, and posterior fossae. The apparently single globular mass, in fact, is generally polylobular with discreet excresences occupying the prechiasmatic and retrochiasmatic areas. It bulges into the middle and posterior fossae, deforming completely the hypothalamus and midbrain, stretching and engulfing the individual components of the circle of Willis. Extensive areas of calcification are most commonly located along the periphery of the tumor, at times subjacent to the extremely dense capsule, and at times adhering to the walls of major vessels.

They are more abundant in volume and greater in extent in children who have had repeated operations and courses of X-ray therapy, as well as in the few reported instances in newborns. The dense capsule adheres to the pia arachnoid and often blends imperceptibly into areas of gliosis within the hypothalamus, medial surfaces of the temporal lobes, and the midbrain. Dense capsule, extensive areas of calcification, and new bone formation are often in continuity with one another, separating cystic compartments from those containing solid tumor. Frontal and temporal lobe involvement accompanies destruction of the optic pathways, as well as invasion of the ventricular system, and compressive occlusion of arteries and veins. Damage to the oculomotor nerves loses its clinical significance because of the invariably present blindness. The basal cisterns are obliterated by the tumor mass, but, curiously enough, though there is often a severe increase in intracranial pressure, hypertensive ventricular dilation – hydrocephalus – is generally not present.

Atypical Craniopharyngioma

The atypical forms of craniopharyngioma may be located within the pharynx, the posterior fossa, limited entirely to the sphenoid sinus, even in the pineal region. When craniopharyngioma is not located in the parasellar area, but within the sphenoid sinus, clivus, pharynx, posterior fossa, or pineal region, it does not behave clinically as a parasellar tumor. It may be a polycystic mass extending freely within the basal cisterns (a not too difficult tumor to resect). Accordingly, it is of embryologic and histological interest. The histological diagnosis is invariably a surprise to the clinician when it is made, since the pharyngeal, clival, and sphenoidal

forms appear histologically as adamantinomas; but the cystic forms, squamous epithelial, are a pleasant surprise.

High morbidity predictors are "severe hydrochephalus, intraoperative adverse events, and early age at presentation"; and predictors of tumor recurrence are "large tumor size, early age, and severe hydrocephalus...whereas complete tumor resection (as determined by postoperative neuro-imaging) and radiotherapy given electively after subtotal excision were less likely to be associated with recurrent disease." Since, for the present at least, neuro-imaging remains a macroscopic modality, not able to identify microscopic infiltrates or characteristics, it seems justified to prescribe postoperative irradiation therapy (preferably radiosurgery) even in those children with negative neuro-imaging. A review of the not insignificant number of recurrences after total (neuro-imaging documented) resections supports this treatment suggestion.

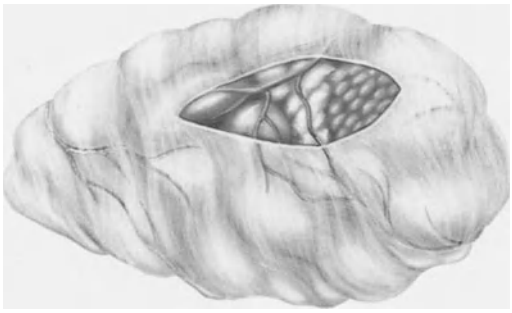


Figure 10.116. It is the gross pathology of the craniopharyngioma which is of particular interest to the surgeon, though the histological characteristics (adamantinomatous or papillary) are those which correlate more directly with age of onset and prognosis (papillary type more common in adults and better prognosis, adamantinomatous type more common in children with greater tendency for local recurrence). The matter of a capsule has assumed controversial proportions, since those surgeons supporting every effort for a "total" resection sustain a capsule is present, protecting surrounding structures. Quoting Schiffer, "the tumor has, in general, well-defined borders, but may present clear adhesions to surrounding nervous tissue, in which it elicits marked reactive gliosis. It is therefore debatable whether a limiting capsule is present." This illustration represents a composite of my own surgical experiences with 65 craniopharyngiomas in childhood. The outer glistening surface, which has the visual appearance of a capsule, is often partially arachnoid membrane and partially proteinacious film formation. I have never seen a craniopharyngioma, in surgery or at autopsy, with a capsule, in the true sense of the term. The gross appearance of the tumor may be macrolobular around cysts of varying size, brilliantly smooth, or microlobular. By and large, the vascularization of these tumors, arterial and venous, is along the surface and one sees direct arteriovenous shunting.

General Comments on Craniopharyngioma Surgical Anatomy and Technique (Figs. 10.116–10.121)

After the bifrontal craniotomy has been performed, the dura is opened over both frontal lobes and the superior sagittal sinus transected and cut at the crista galli. The falx cerebri is separated from the crista galli, and both olfactory nerves are coagulated and transected unless one chooses to use the already described technique to preserve them. This brings the surgeon's line of vision over the retracted frontal lobes, along the cribriform plate and orbital roofs, and beyond the lesser wings of the sphenoid and anterior clinoid processes to the anterior circle and lamina terminalis. Spatula retraction is discouraged: gravity suffices in children. The frontal lobes fall slightly posterolaterally, putting some stretch upon the arachnoid of the sylvian fissure and the middle cerebral arteries.

The optic nerves exit from the cranial portion of the optic canal, running posteromedially (they rest at almost right angles to one another throughout their entire course from the globe to the optic chiasm). Internal carotid and anterior cerebral arteries are covered by the same arachnoidal membranes as the optic nerves and chiasm. Each optic nerve rests upon the internal carotid artery as the former leaves the optic canal and the latter leaves the cavernous sinus. The internal carotid then turns slightly lateralward and inferiorly, running in the direction of the anterior perforated substance, where it bifurcates into the anterior and middle cerebral arteries. A-1 passes over the lateral surface of the optic nerve and runs along the superior surface of the optic chiasm, anterosuperiorly, to join its homonym of the other side via the anterior communicating artery.

It is important to envision the relationship between the arachnoid membrane, optic nerves, and components of the anterior circle. It is also advisable to study the relationships between the posterior communicating and posterior cerebral arteries, the oculomotor nerves, the hypothalamus, and the midbrain. Removal of the arachnoid permits one to separate safely optic nerves from the individual components of the anterior circle, and to identify microvasculature going from these vessels to the optic pathways.

The tentorium inserts onto both the anterior and posterior clinoid processes, with the intervening slip of dura permitting entrance of the III and IV cranial nerves into the cavernous sinus. Figures 10.113, 10.116, and 10.121 allow the reader to understand how very difficult it is to remove tumor from beneath the optic chiasm and hypothalamus, and to evaluate the merits and risks of working between the carotid artery and optic nerve anteriorly, and the optic tract and posterior communicating artery posteriorly. Tumor expanding posterior to the hypothalamus, as in retrochiasmatic tumors, not only pushes superiorly into the III ventricle, but also expands posteriorly into the mammillary cis-

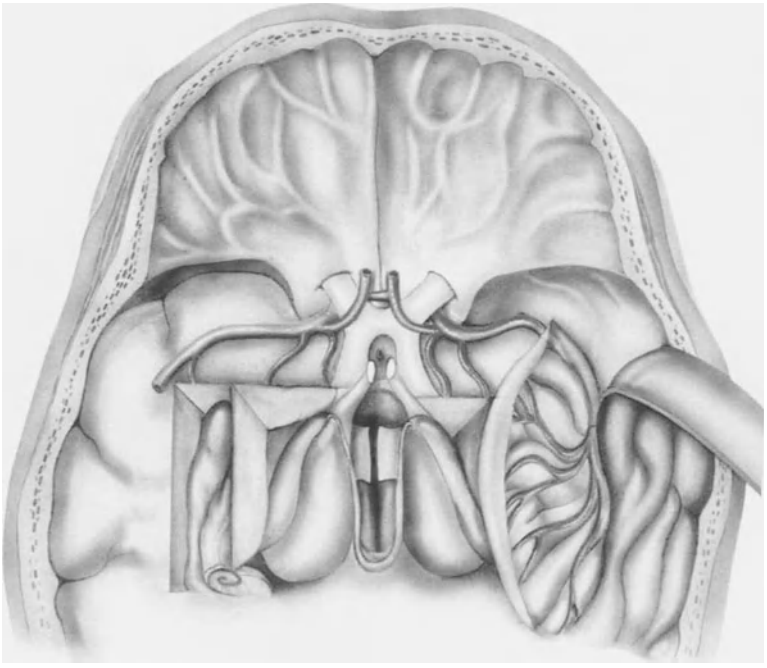


Figure 10.117. The "eloquent" parasellar structures which may be involved, to a greater or lesser degree, by craniopharyngioma or surgical manipulations associated with its partial or total resection, are herein illustrated schematically. For purposes of highlighting each significant structure, the relative sizes vis-à-vis other structures in this drawing vary from real anatomical relationships. In the suprasellar area are the optic nerves, the internal carotid artery A-1, and anterior communicating artery. The retrochiasmatic structures are the optic tracts, the mammillary bodies (not illustrated in this drawing), and the columns of the fornix. The suprasellar structures are the pituitary stalk, the suprachiasmatic and infundibular recesses of the III ventricle, and the hypothalamic nuclei. The lateral perichiasmatic structures are the anterior choroidal artery, the posterior communicating artery, the paraolfactory area, the bifurcation of the internal carotid artery, and the amygdala and pes hippocampus. Also illustrated in this drawing, and of importance, are the temporal lobes, retracted off the insula on the right and resting posteroinferiorly on the sphenoid wing on the left.

tern, displacing the posterior cerebral and basilar arteries posteriorly and the posterior communicating artery and III cranial nerve laterally. Retrochiasmatic tumor extensions take their vasculature from the medial surfaces of the posterior communicating arteries and, most rarely, the anterior surfaces of the posterior cerebral arteries.

When a retrochiasmatic craniopharyngioma expands posterosuperiorly into the III ventricle and posteroinferiorly over the basilar fundus, removal through the prechiasmatic space entails manipulating the tumor downward, forward, and then upward, taking it from the posterior floor of the III ventricle, the surfaces of the posterior cerebral and basilar arteries, beneath the optic chiasm, away from the posterior clinoid processes, and through the prechiasmatic space. This manipulation may be greatly faciliated by working through the three routes: prechiasmatic, interopticocarotid, and retrochiasmatic. One may choose to take a fourth route: *translamina-terminalis and through the floor of III.*

Surgical Management of Children with Craniopharyngioma

Supplemental Surgical Management of Craniopharyngioma

Hydrocephalus complicating craniopharyngioma may result from tumor obstructing the basal cisterns, invasion of the III ventricle and occlusion of the foramina of Monro, and, very rarely, posterior displacement of the brainstem with occlusion of the aqueduct of Sylvius. *It is an event which indicates a poor prognosis.* A unilateral ventriculoperitoneal shunt is recommended if one is certain that both foramina of Monro are open. Bilateral ventriculoperitoneal shunts (two completely independent systems) are recommended if one or both foramina of Monro are occluded. Compensation of the hydrocephalus immediately diminishes the increase in intracranial pressure and provides relaxation of cerebral volume, so as to facilitate exposure of the parasellar area for surgical removal of the tumor. Mannitol and lasix are not recommended as adjuncts to diminsh intra-

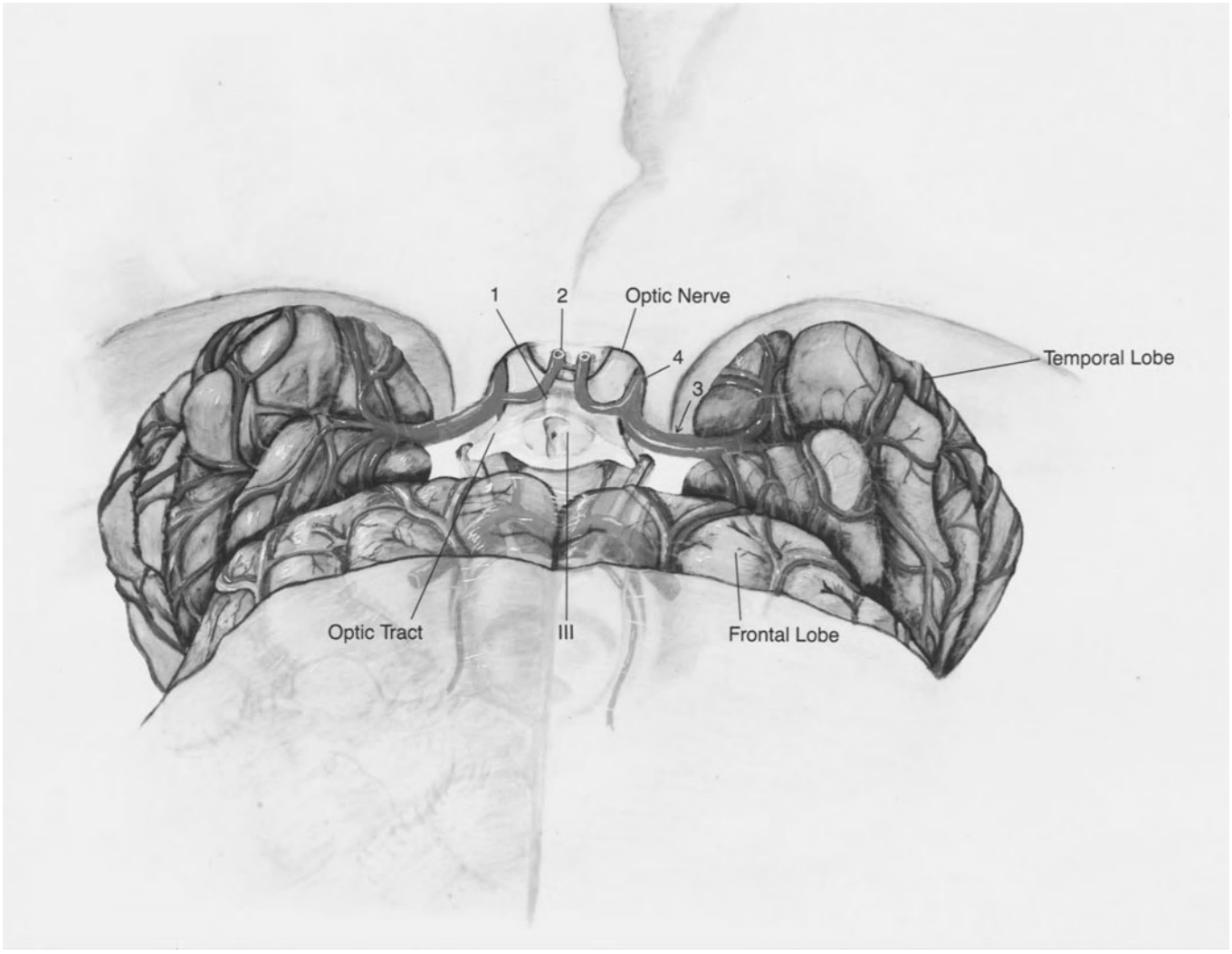


Figure 10.118. The surgeon's view of the parasellar area after the falx cerebri has been cut from the crista galli and both olfactory nerves transected. In this drawing the III ventricle above the hypothalamic sulcus and both optic tracts has been removed to allow the reader to conceptualize intraventricular and retrochiasmatic extension of craniopharyngioma. The depiction of the course of A-1 (*1*) over the optic chiasm and of A-2 (*2*) anterior to the subcallosal gyrus and lamina terminalis is spatially represented, as are those of the middle cerebral (*3*) and internal carotid (*4*) arteries. This allows one to understand the need to open cisterns so as to mobilize the vessels, separating them from the adjacent parenchyma.

cranial fluid volume. The III ventriculostomy is not a treatment alternative.

Evacuation of cystic fluid contained within portions of the tumor, by means of stereotaxically positioned tubes or needles, has been advocated by some authors, who have also recommended injecting radioactive substances or bleomycin into the cystic cavity, so as either to diminish the secretion of cystic fluid or destroy the secretory cells lining the cavity. These procedures are theoretically attractive but have not been widely accepted, very likely because they are not as easy and safe as direct surgical exposure of the tumor mass. It is difficult to identify with precision where within the tumor area the cyst is located, and whether there are multiple cyst cavities. Intraoperative exposure of the tumor, on the other hand, allows the surgeon to identify visually the location and extent of cystic cavities, to drain cystic fluid and remove soft tumor tissue, and to instill under direct vision radioactive substances of one's choosing. The most significant advantage of direct exposure of the tumor mass is decompression, and its removal if feasible.

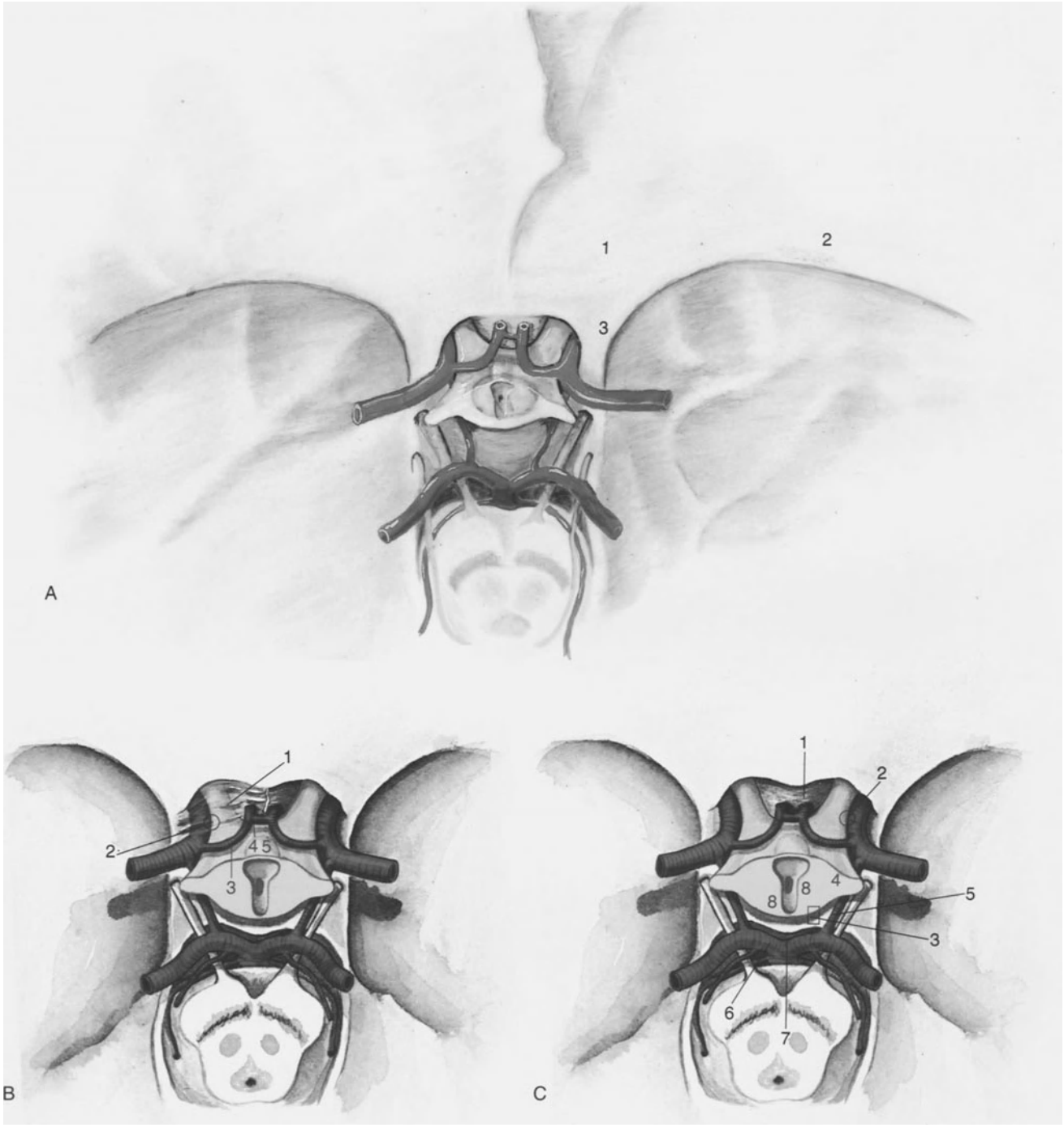


Figure 10.119. (A) The cerebrum and basal ganglia have been removed, allowing one to appreciate access to the parasellar area along the cribriform plate and planum sphenoidale (*1*), or the lesser wing of the sphenoid (*2*) and anterior clinoid (*3*). (B) The arachnoid membrane is intact over the prechiasmatic cistern (*1*), and the interopticocarotid space (*2*) *on the left*, but has been removed *on the right*. A-1 (*3*) is adherent to the chiasm, nourishing it through a multitude of arterioles which, in children with suprasellar extensions of craniopharyngioma, are obliterated from stretching and compression. The junction of A-1 and A-2 is also subarachnoid (*4*): located at the most inferior portion of the lamina terminalis (*5*). (C) With the arachnoid removed, one may identify the four basic approaches to craniopharyngioma: prechiasmatic (*1*), interopticocarotid (*2*), retrochiasmatic (*3*), between the optic tract superiorly (*4*) and the posterior communicating artery (*5*) inferiorly, and translamina terminalis (*)...noting immediately in this latter approach that at the base of the lamina terminalis one comes immediately upon the area of the supraoptic and paraventricular nuclei. One should note how very close the posterior cerebral (*6*) and basilar (*7*) arteries are to the hypothalamus (*8*).

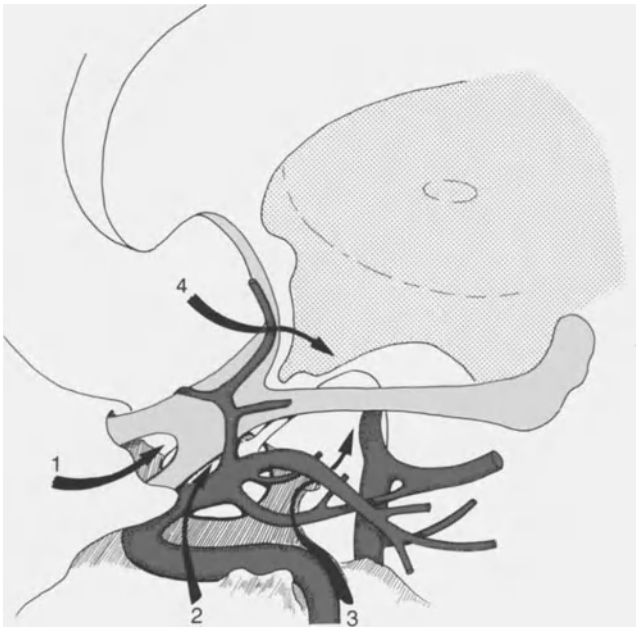


Figure 10.120. The three access (*arrow*) routes to intra- and retrochiasmatic craniopharyngioma: (*1*) The first route, between the two optic nerves, allows one to probe beneath the optic chiasm. It is unsatisfactory because one is obliged either to extend blindly his dissector along the optic chiasm or to pull tumor from it. Either alternative risks further damaging vision (*2*). The interopticocarotid route, elevating the optic nerve and displacing inferolaterally the internal carotid artery, allows one access to the infrachiasmatic area and direct vision of most of it. One may work within this interval anterior and posterior to the internal carotid artery. (*3*) Posterior access to the infrachiasmatic area may be gained by working over the tentorial edge, opening the ambient cistern, identifying the III cranial nerve and posterior communicating artery, and then covering these structures with tefla in order to protect them (*4*). The translamina terminalis route brings the surgeon directly into the III, but he must then go though the floor of the III if the tumor has not already ruptured into this chamber. Resection of the anterior 2 cm of the temporal lobe allows one to work one's way superiorly along the internal carotid artery to its bifurcation without compressing temporal lobe or internal capsule. This provides excellent visualization of the infundibular and interpeduncular cisterns, the basilar and posterior cerebral arteries and the peduncles.

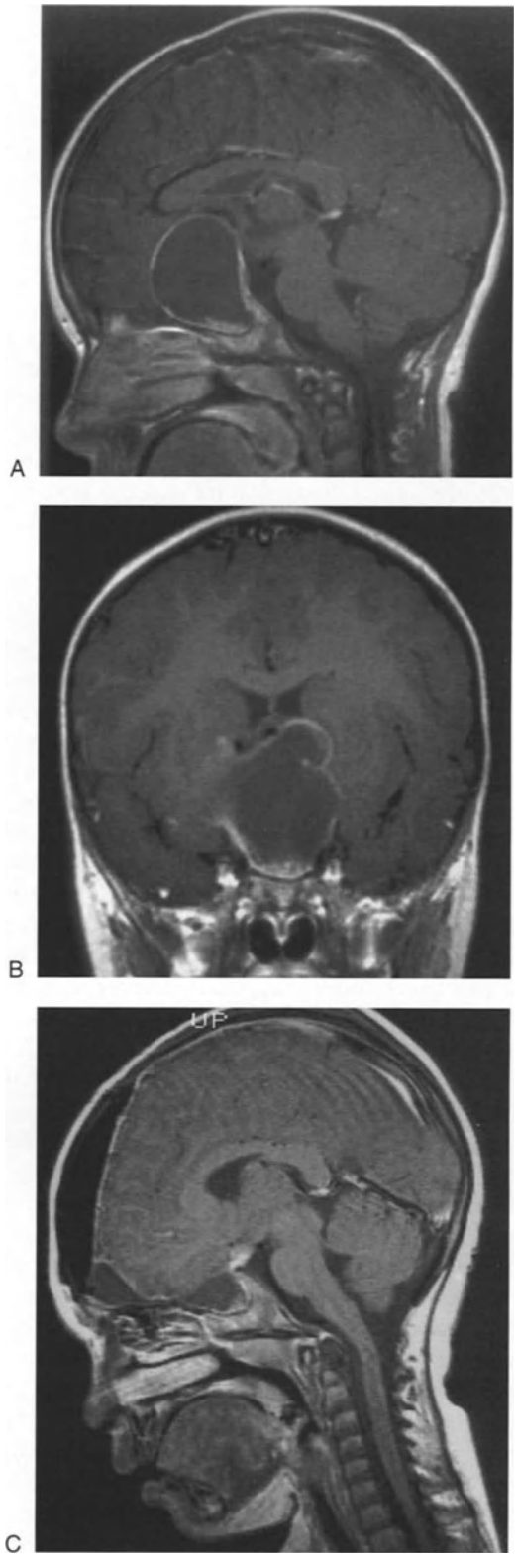


Figure 10.121. Craniopharyngioma: This craniopharyngioma ► is the most favorable of all, except for the pure intrasellar type, for ease of resection: it is entirely prechiasmatic, being composed of a very large cyst (permitting drainage immediately before resection). (A) This sagittal post-gadolinium image shows precisely the location of the mass: the sella turcica is enlarged, there is upward displacement of the frontal lobe and the anterior cerebral arteries, and the anterior commissure is displaced posteriorly as is the III ventricle. (B) This sagittal section reveals the superior extension of the unilobar cyst and its suprachiasmatic lateral bulging. (C) The postoperative, post-gadolinium sagittal section shows only falling away of the dura from the inner table of the skull, and subfrontal fluid collections. No residual tumor is present.

Surgical Approach to Craniopharyngioma: General Comments

Irrespective of the surgical approach to the craniopharyngyoma and its intrasellar, prechiasmatic, retrochiasmatic, or other locations, the technique for removing the tumor is always the same, *but not the anatomical approach.* The technique is predicated upon the fact that craniopharyngioma may be solid, cystic, solid/cystic, well-encapsulated, and poorly encapsulated with areas where tumor tentacles extend into the parenchyma, densely adherent to the cranial nerves and vessels. In addition to this, and adding great complications, is the fact that the craniopharyngioma is variably calcified, that extensive areas of calcified tumor may be adherent to parenchyma or vessels. Lastly, the vascular supply to the craniopharyngioma capsule consists of minute arteries coming from the internal carotid, posterior communicating, and basilar systems, so that separation of the tumor capsule from surrounding tissue may be complicated by brisk bleeding or tearing of a larger branch from a major artery.

Initially the tumor is identified, a portion of its capsule no longer than 3 mm should be coagulated and the capsule opened by the blades of the bipolar forceps. MRI angiography will already have excluded the *remote* possibility that the "tumor" is an aneurysm, so the surgeon need not concern himself with the archaic habit of puncturing the tumor with a spinal needle in order to learn whether it is an aneurysm. Once the capsule is opened, varying amounts of fluid may be removed by placing a microsuction tip into the cystic portion of the tumor. This gives some immediate decompression, relaxes the tension on the optic nerves, and allows the surgeon to inspect the tumor cavity to evaluate the extent of solid or cystic tumor, and the degree of calcification. Then microsuction is used both to aspirate more cystic fluid and to serve as a dissector, to tease the tumor capsule away from surrounding structures by sucking upon it from its luminal surface.

All arachnoidal tissue covering the tumor or the surrounding parenchymal or vascular structures, where one is working, should be removed, using bipolar cautery to shrink it or microscissors to cut it free. Either a #4 Penfield dissector or a microdissector is then used to separate the capsule from surrounding tissue by pressing on the capsule and pushing it toward the sella turcica, away from the optic pathways, thereby identifying bridging strands of capsule, arachnoid, neoplastic vessels, which are coagulated and transected as one proceeds. It is advisable to work in one area at a time, freeing all visible tumor capsule from surrounding structures, rather than moving from right to left, anterior to posterior, superior to inferior. The mobilization of the capsule in individual, discreet areas, allows the surgeon to remove portions of tumor, a bit at a time. The Cavitron is very helpful in *this "bit-by-bit" intratumoral resection technique. Removal of the tumor by grasping a portion of the capsule in a pituitary forceps and continuously and slowly applying gentle traction is discouraged. It continuously puts traction on the surrounding neural structures to which the tumor is adherent. When the tumor is freed, this traction may result in delivering a mass of tumor which is larger in volume than the interval between the optic nerves through which it is being pulled.* This could damage the optic nerves, chiasm, or optic tract. One need not be concerned with "losing" a portion of the tumor in the infra- or retrochiasmatic areas, to which the surgeon is "blind." Simply inserting a microsucker into these areas and irrigating allows him to bring these floating lobules of tumor into view by applying the suction tip to the capsule and gently withdrawing the sucker.

It is advisable to use all three openings (prechiasmatic, interoptico/carotid, retrochiasmatic) to manipulate the tumor, irrespective of the surgeon's decision to use one of the three as the major portal, but use the prechiasmatic and retrochiasmatic openings for insertion of a microdissector so as to manipulate the tumor into the interopticocarotid space for piecemeal removal. The translamina-terminalis approach should be used only if the others prove to be inadequate.

It is difficult to separate thin capsule from thickened arachnoid, so one must take care to inspect closely for the presence of vessels, and ascertain that it is possible to get around the capsule with a dissector before delivering it through the operative portal. If it is not possible to determine whether one is working on capsule or arachnoid, it is preferable to use the microbipolar forceps to coagulate the tissue in question, destroying it, rather than to attempt to pull it through.

Calcified tissue is both tedious to dissect and dangerous to remove, until it is floating freely within the tumor bed, since it may be adherent to a vessel. Consequently, granules or nubbins of calcification should be freed only under direct vision, teasing them away rather than lifting them out in the jaws of a pituitary forceps. Calcifications which are adjacent to surrounding structures or, more importantly, vessels, should be irrigated under direct vision to see whether they float away from their bed. If not, they should be left in place.

Perhaps only with the exception of certain well- delimited intrasellar and prechiasmatic forms, the surgeon's primary mission should not invariably be to perform a complete resection of the tumor. Surgical removal of the intrasellar and prechiasmatic craniopharyngiomas is benign, and attempts at total removal of retrochiasmatic and giant forms subjects the patient to very high morbidity and mortality risks.

Surgical Technique: Specific Procedures

Rhinoseptal Transsphenoidal Approach

Equipment for X-ray control utilizing intraoperative image intensification provides the surgeon with the means for monitoring the intrasellar location of the operative procedure. The operating microscope allows him to distinguish between tumor and compressed pituitary tissue. Attention is given to identifying the leaflike extension of tumor immediately the dura is opened and the sella turcica entered. This portion of the tumor is separated from the compressed residual tissue of the pituitary gland, taking care when working in the lateral sella turcica to avoid entering the cavernous sinuses. Avoiding traction, and maintaining the *diaphragma sellae* intact, one follows the neurohypophysis into the pituitary stalk, which is separated from the tumor if at all possible. If not, it should be clipped or coagulated and sectioned.

The high incidence of recurrences of intrasellar craniopharyngioma speaks eloquently of the difficulty of being certain that one has resected all of the tumor and its capsule. This should not detract the surgeon from taking care to assure the integrity of the *diaphragma sellae* and to close carefully, with bone graft, the sella turcica, especially since a subsequent intracranial procedure may become necessary.

Subfrontal Approach

Either a unilateral right frontal or a bilateral frontal craniotomy may be reflected. Use of the operating microscope has encouraged us to extend our procedures slightly more lateralward (*the bifrontoperional flap*) necessitating reflection of the pterion with the frontal flap, so as to obtain a straight line of vision to the lateral aspects of the internal carotid arteries and the optic nerves, as well as full visualization of the posterior communicating arteries.

The subfrontal approach permits three basic variations:

1. Prechiasmatic Approach (Fig. 10.122). After the cerebral hemispheres have been retracted and the CSF drained from the supraoptic, sylvian, and basal cisterns, the self-retaining retractor is put into place and the right optic nerve exposed. The left optic nerve is exposed after the arachnoid between the two optic nerves has been dissected, freeing the optic nerves from the underlying tumor. This is followed by gutting solid tumor or draining the cystic portion from between the optic nerves, allowing the capsule to fall away. Dissection of the optic nerves is thereby facilitated. At this time one attempts to identify the pituitary stalk. Some surgeons prefer to conserve its anatomical integrity, but this subjects the child to risks of severe immediate postoperative diabetes insipidus from traction on the hypothalamus. It is suggested that blind dissection not be

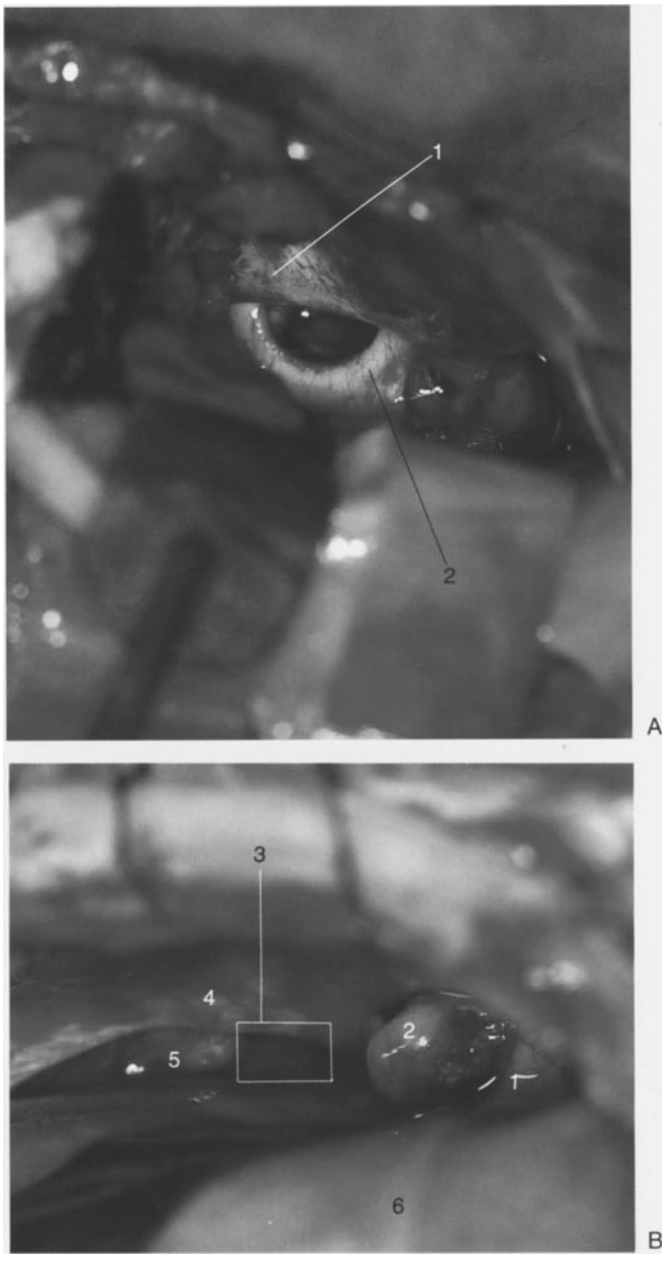


Figure 10.122. (A) Prechiasmatic extension of craniopharyngioma presents no particular surgical problems. There is an adequate amount of tumor protruding between the planum sphenoidale (*1*) and the optic chiasm (*2*) that the surgeon may immediately enter the tumor, drain the fluid within it so as to attain decompression, and then proceed to separate the arachnoid from the optic pathways and carotid arteries. Subsequent to this, piecemeal removal of solid and calcified tumor is accomplished through the large opening between the planum sphenoidale and optic chiasm. (B) The interopticocarotid space provides the surgeon a second portal for access to the craniopharyngioma expanding beneath the optic chiasm (*1*). This photograph illustrates a craniopharyngioma (*2*) which dissected its way through the interopticocarotid space, elevating the optic nerve and depressing the internal carotid artery as it expanded into the middle fossa (*3*). The lesser wing of the sphenoid (*4*), temporal lobe (*5*), and Telfa-covered frontal lobe (*6*) are included in the field.

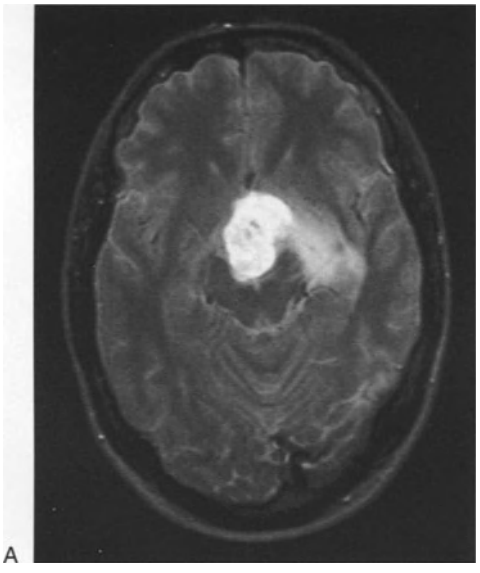
performed behind the tumor *with the intention to deliver too soon the posterior portion of the tumor through the prechiasmatic space.* The next step is dissection of the tumor and removal of its retrochiasmatic component. In this portion of the procedure, the surgeon stays within the subarachnoidal membranes of the basal cisterns in and around the sella turcica. At times, it is necessary to go through the lamina terminalis to access intra-third ventricular extensions of tumor.

The difficulties encountered in this approach arise from the internal carotid arteries and their branches, the oculomotor nerve and the optic pathways, the hypothalamus and basilar artery. These large structures, however, generally do not present a great problem. When necessary, and only for a blind eye, sectioning of the optic nerve facilitates the anatomical exposure of the area.

Problems in the intrasellar area include precise separation of tumor tissue from residual pituitary gland, which is always difficult, and identification of the inferior leaf of the tumor, which may be most difficult to distinguish visually from the dura mater along the sellar floor.

When the surgeon encounters a tumor that is primarily retrochiasmatic, he may find it advantageous to enlarge the subfrontal approach by resecting a portion of the planum sphenoidale, creating an artificial interopticocarotid triangle. This provides direct access to the sella turcica, visualization of the anterior portion of the sella, and a view into the retrochiasmatic area and superior portion of the interpeduncular and pontine cisterns.

2. Interopticocarotid Approach (Figs. 10.122–10.125). This approach permits entry into the space between the optic nerve and the internal carotid artery, on the lateral surface. It was actually described by Dandy [143], and has been used by various authors since that time. The only additional contribution suggested here is the extension of the opening into the sylvian fissure. The interopticocarotid approach gives excellent visualization of the lateral surface of the tumor, especially when



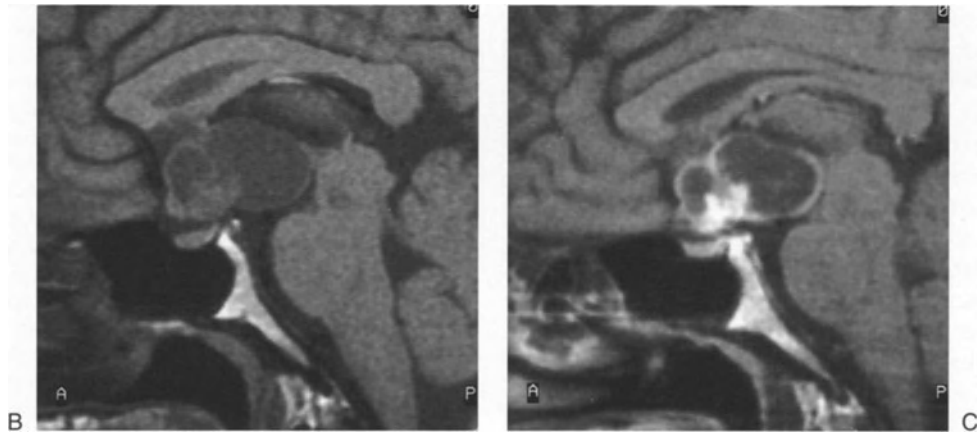


Figure 10.123. This is a large, partially solid, but predominantly cystic infra- and retrochiasmatic craniopharyngioma occupying most of the third ventricle, and pushing the midbrain posteriorly. The pituitary gland is intact. This series of MRI images is best studied along with the drawings of retrochiasmatic tumor extension. (A) There is a mass occupying the suprasellar and interpeduncular cisterns, whose cystic portion has a high signal intensity on the T2 weighted image. (B) The cystic portion of the mass pushes the lamina terminalis posteriorly, jamming the mammillary bodies into the peduncles. The anterior lobule of the mass expands within the space beneath the optic chiasm, which is pre-fixed. (C) In this post-enhancement study one sees to advantage the upward displacement of the optic chiasm and its wedging between the infrachiasmatic and the retrochiasmatic cystic chambers. The capsule enhances. There is a solid component, irregular in outline, at the inferior portion of the tumor.

it extends into the retrochiasmatic region, permitting the surgeon to reach the posterior pole of the tumor with ease. Difficult problems arise in delivering the tumor since the superior pole is out of sight, obliging the surgeon to deliver blindly this portion of the tumor.

3. Retrochiasmatic Approach (Figs. 10.126, 10.127). One may choose to dissect completely the optic nerves, passing along their superior surfaces to the optic chiasm posteriorly to the infundibulum, entering the retrochiasmatic space by separating completely the optic pathways and the hypothalamus superiorly from the posterior communicating artery inferiorly and laterally. This approach may be of value in dissecting suprachiasmatic nodules of tumor. It separates the optic chiasm and tract from the tiny vessels coursing to it from the anterior cerebral arteries, so one runs a real risk of impairing vision.

Unilateral Anterior Subtemporal Approach

One may perform the anterior subtemporal approach (called "*pterional*" by some) in either one or two stages. Cerebrospinal fluid drainage and self-retaining retractors are essential, and sectioning of the tentorium is at times necessary to provide adequate visualization of the parasellar area. After freeing the oculomotor nerves, one has access to the inferior pole of the tumor. By freeing the internal carotid and posterior communicating arteries, access to the lateral surface of the tumor mass is gained. In some instances, when the tumor is small and well circumscribed, it is possible to extend the dissection from one side to the other, being able, thereby, to go from the III cranial nerve on the right to the III cranial nerve on the left. This approach offers two real difficulties to the surgeon: the necessity of dissecting blindly the superior pole of the tumor, and the extreme difficulty in freeing the prechiasmatic extensions of tumor.

Direct Transventricular Approach

The transventricular approach may be made through the right lateral ventricle and the foramen of Monro, or through an anterior transcallosal approach by going between the two columns of the fornix and entering the III ventricle. *From a practical point of view, one must consider the problems in identifing a plane of dissection between the floor of the hypothalamus, which is being displaced by the tumor, and the tumor capsule!* This is a difficult undertaking, one which presents risks of damage to the hypothalamus. It may be undertaken as a supplementary approach, for mutilobular tumors, to be used when the surgeon finds it necessary to free intraventricular extensions of tumor, after he has already delivered significant portions of the mass through the prechiasmatic space.

Recurrences

Apart from growth of macroscopic residual tumors, the case reported by Piepgrass [144] is extraordinarily interesting, since it is the first documented instance of tumor recurrence along the line of surgical approach to the parasellar area, although it is true that Barloon et al. [145] reported in 1988 that a patient they treated with repeated needle aspirations through the frontal lobe suffered implantation of craniopharyngioma along the line of the needle tract. The 47-year-old man treated by the authors underwent a right frontotemporal craniotomy and gross total removal of his craniopharyngioma through the right sylvian fissure and opening of the lamina terminalis, and did not receive ionizing therapy postoperatively.

It is well known that craniopharyngioma may assume an enormous size, being multilobulated in form with digit-like extensions (into the III ventricle, the interhemispheric fissure, the sylvian fissures or the interpeduncular or pontine cisterns), and that it not uncommonly may expand into the III ventricle, occupying this potential cavity completely. Less common sites of development of craniopharyngioma have been reported: foramen magnum, medullary cistern, maxillary sinus. What seems to be a common denominator is a CSF-containing chamber. Therefore, one may suppose that there is a propensity for the squamous epithelial cell rests of the craniopharyngioma to pass along the CSF pathways and to implant themselves within the pia arachnoid, where they then grow.

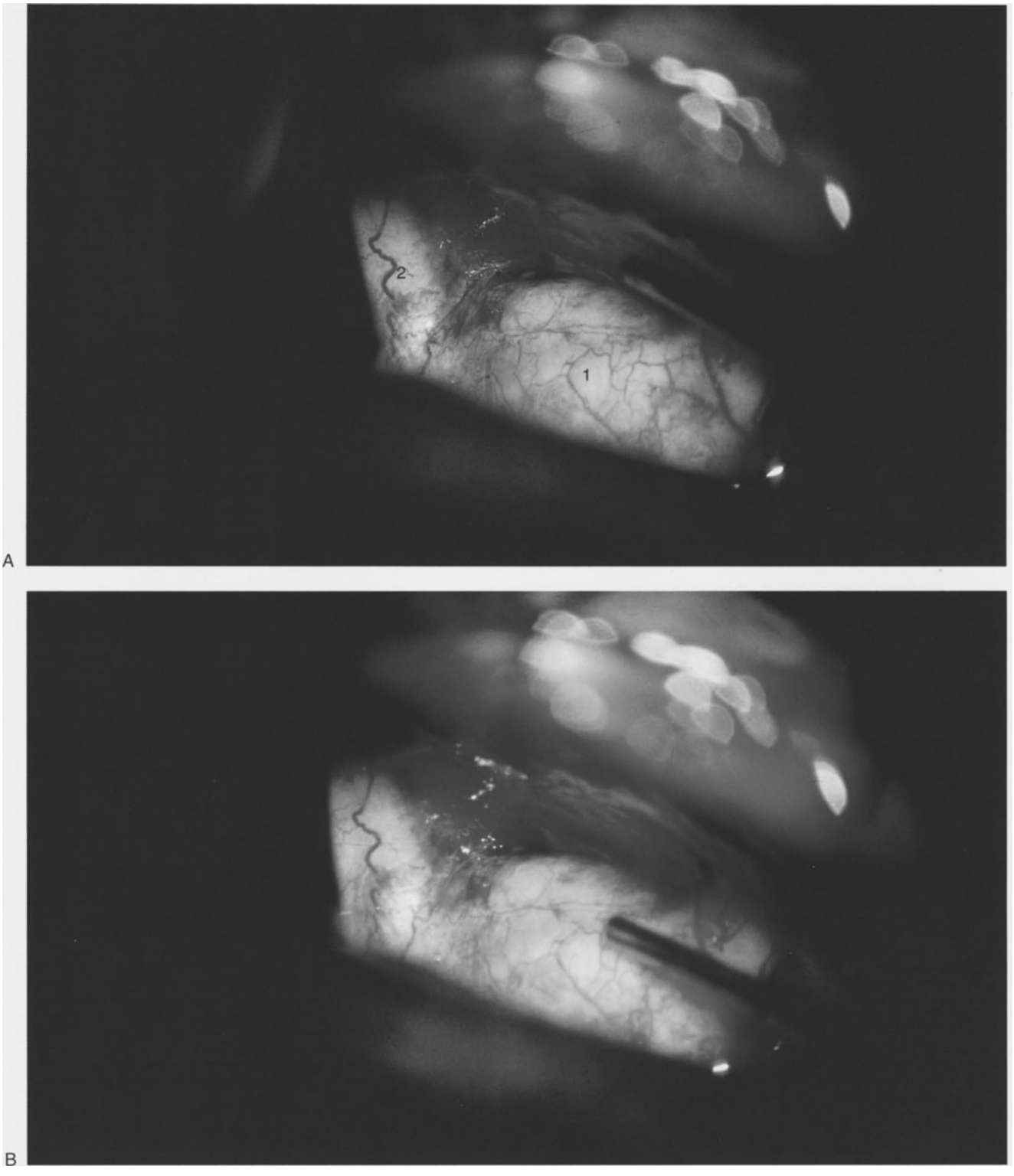
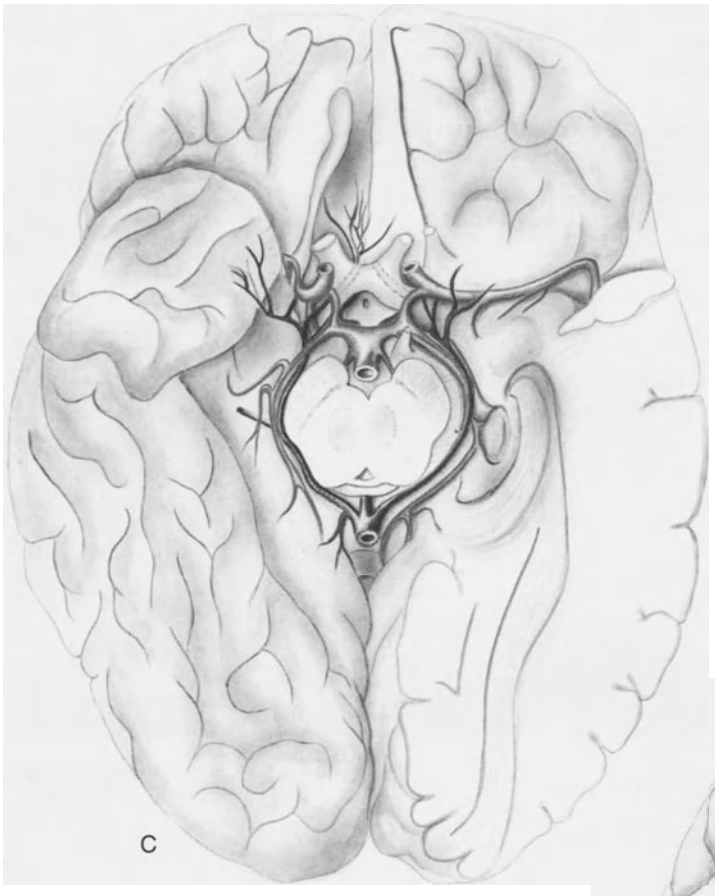


Figure 10.124. (A) This is an example of a child whose craniopharyngioma expanded beneath the optic chiasm (*1*), elevating it and displacing its most anterior surface onto the planum sphenoidale, elevating and flattening the left optic nerve (*2*). One sees a small amount of craniopharyngioma protruding between the left optic nerve and the optic chiasm. (B) The anteriorly displaced optic chiasm has been dissected posteriorly from the planum sphenoidale, exposing the capsule of the craniopharyngioma. Because of the anterior location of the chiasm, it is not feasible to attempt to remove (completely) a craniopharyngioma through this limited opening. (C, D) see p. 325.



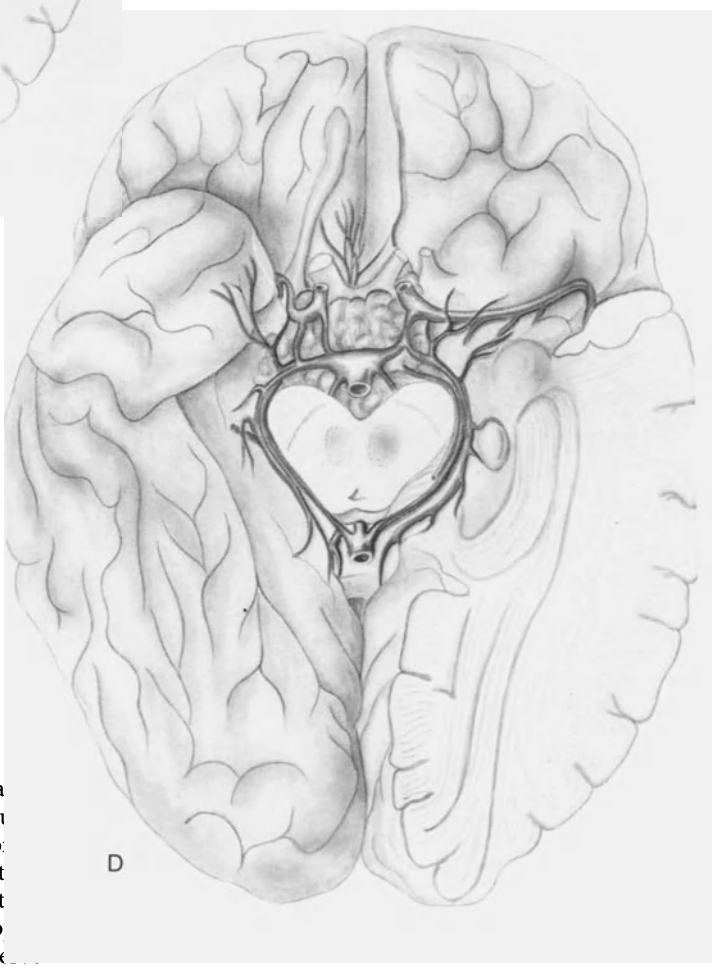


Figure 10.124. (C) This axial projection of the parasellar a
in color, illustrates relative locations of eloquent structı
which may be involved by a large infrachiasmatic extensio
a craniopharyngioma: olfactory nerve and frontobasal art
olfactory trigone and bifuration of the internal carotid art
optic chiasm and trunk of the internal carotid artery, o
tract and posterior communicating artery, III cranial ne
and basilar fundus. (D) Comparing this illustration to (C) al-
lows one to correlate anatomical with neoplastic realities in
the infrachiasmatic space when the intraoperative observa-
tions are those illustrated in (A) and (B).

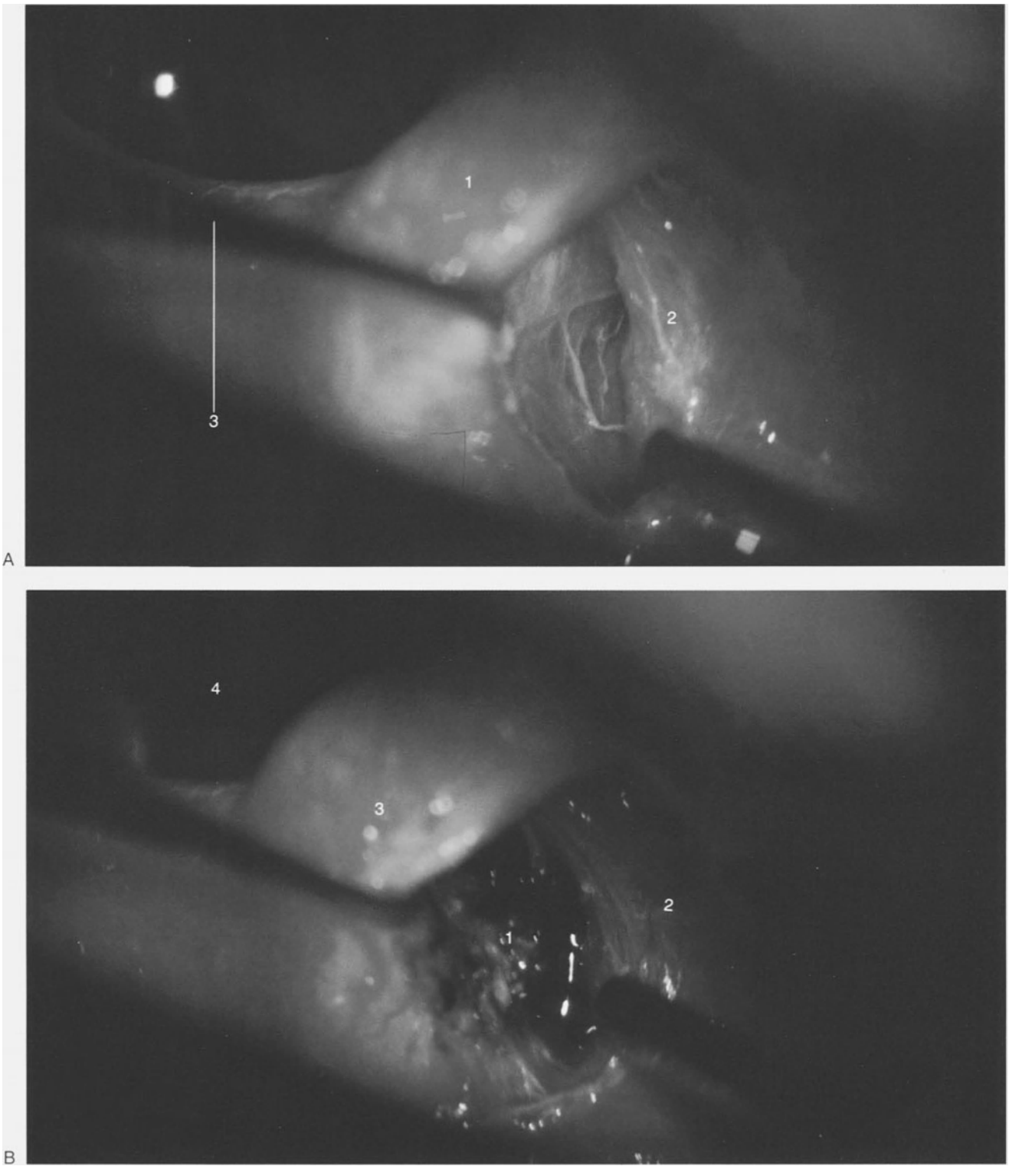


Figure 10.125. (A) One may open the interopticocarotid space. The arachnoid between the optic nerve (*1*) and the internal carotid artery (*2*) is dissected away and the interval between these structures opened by elevating the optic nerve with a Penfield #4 dissector (*3*), freeing the internal carotid artery from both arachnoid and craniopharyngioma capsule. One may safely retract the optic nerve, but it must be done gently and moderately. (B) Once the capsule of the craniopharyngioma (*1*) bordering upon the medial surface of the internal carotid artery (*2*) has been opened, one enters the cavity of the tumor and resects whatever neoplasm may be removed through this opening. (*Continued* on p. 327).

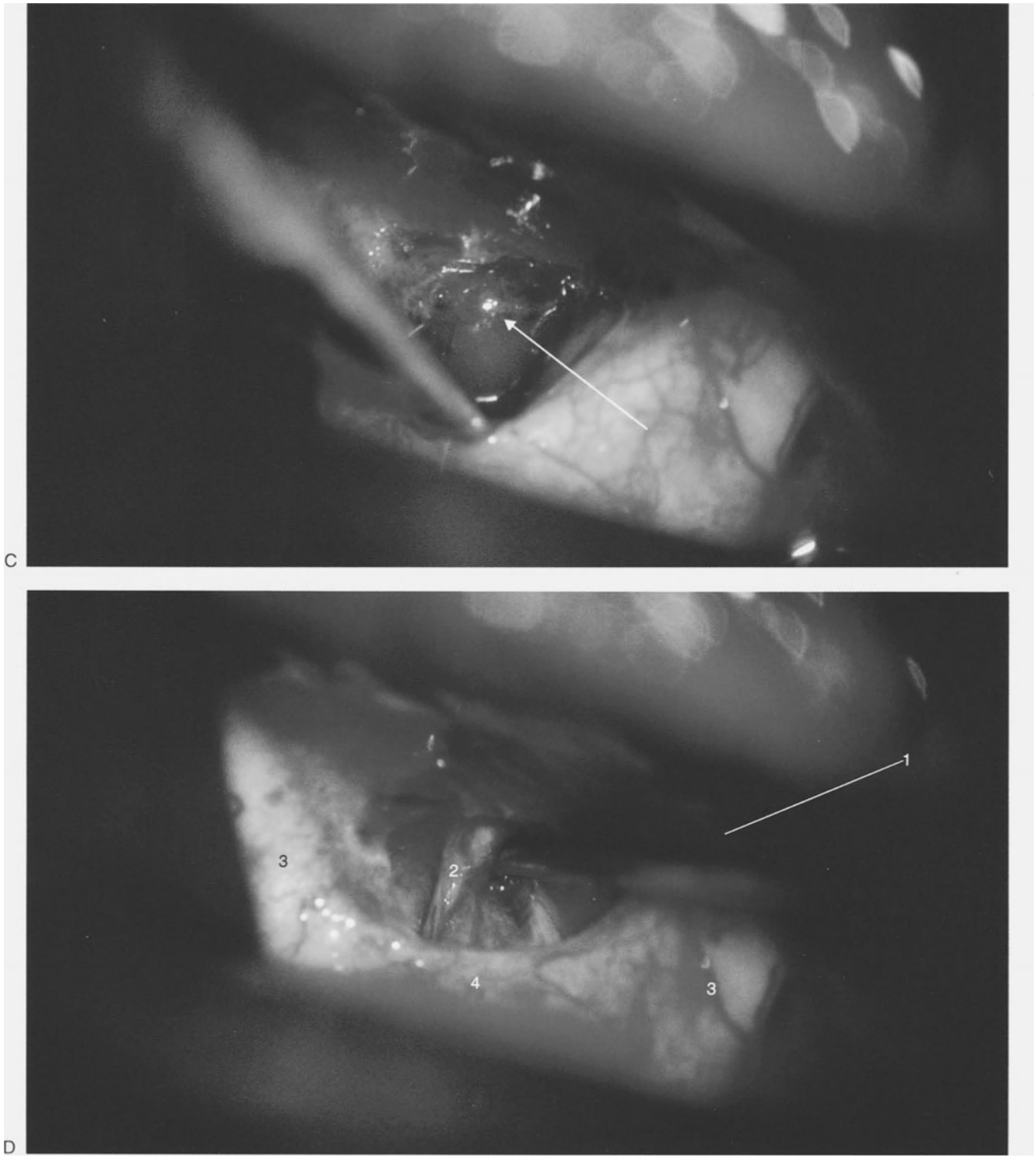


Figure 10.125 (*continued*). Then, either a dissector or the suction is used to "feed" tumor from the interopticocarotid opening beneath the optic nerve (*3*) anteromedially to the prechiasmatic opening (*4*). (C) The tumor in the prechiasmatic space (*arrow*), pushed there through the interopticocarotid opening, may now be lifted from the field. (D) After the gelatinous and semisolid tumor are removed from the prechiasmatic space, the microsuction (*1*) is inserted to draw capsular strands (*2*) into vision so that they may be sectioned and removed, taking care not to damage the optic nerve (*3*) or chiasm (*4*). (E) see p. 328.

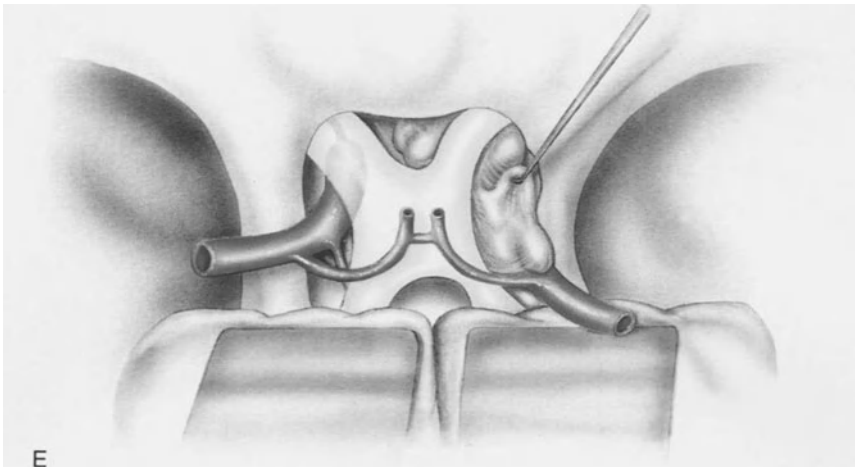


Figure 10.125. (E) This is a schematic drawing of the intraoperative realities illustrated in A–D

Craniopharyngioma in Children: Long-Term Effects of Conservative Surgical Procedures Combined with Radiation Therapy

Different Treatment Modality Approaches with Long Follow-up

Information

The following is a quotation from Harvey Cushing:

Until some method is devised whereby the usual multilocular epithelial lesion can be destroyed or inactivated *in situ*, mortality will doubtless remain high.

Thirty-seven children, treated consecutively for craniopharyngioma between 1972 and 1981 at the Children's Hospital, Boston, were followed by Fischer et al. [146] for more than 10 years, 20 now being beyond high school age. Only 2 of the 34 surviving patients were not contacted directly. In the others information was obtained concerning recurrences and their eventual treatment, social activities, school participation, and engagement in the work market. The authors observed that even when the growth of the tumor was successfully controlled and endocrine problems minimized, major impediments to successful independent adult life resulted from psychosocial factors. This group of patients was treated with "conservative operations" followed by irradiation, or by radical excision of the tumor. Of particular interest is the fact that in ten of these patients the diagnosis was made radiologically and treatment instituted without either analysis of cyst fluid or biopsy of tissue.

The cohort of 37 patients was divided into three groups: group I: 8 patients whose tumor was considered by the surgeon to have been resected totally; group II: 6 patients who were subjected initially to surgery with the intent to perform a total resection, but in whom this was not possible, so partial resection and postoperative radiation therapy were perfomed; group III: 21 patients treated per primum with radiation therapy preceded by either biopsy, cyst drainage, or a shunting procedure. Ten of the patients in the entire cohort were irradiated solely on the basis of radiographic criteria indicative of craniopharyngioma, the radiation being performed on a 4-MV photon beam delivering a tumor dose ranging from 5000 to 5600 cGy with utilization either of an arc wedge rotation or a 3–5 field technique. In the evaluation of their patients the authors used the level-of-function scale proposed by Katz in his 1975 article "Late results of radical excision of craniopharyngiomas in children" [61]. In this classification, level I indicates fully independent patients performing at a level appropriate for their age; level II indicates independence, but with some difficulties; level III indicates an ability to function at age-level, but with partial dependence on others; and level IV indicates full dependence on others.

There was also a grading of school activity, work history, emotional stability, and behavior problems.

There were three deaths, one in each of the three treatment groups. Tumor regrowth or recurrence occurred in 14% of the original 37 children, with 4 occurring in the 7 group I survivors and 1 in the 20 group III survivors, bringing the authors to observe that if deaths and tumor recurrences are combined as treatment failures there were 5 (62.5% among the 8 patients initially treated with radical resection) and 3 (11%) among the 27 initially treated with subtotal resection or conservative operations and irradiation. Thus, it appears to the authors that successful treatment, which they define as no recurrence or death, occurred in 37.5% of those patients treated aggressively but in fully 89% treated conservatively and with irradiation.

Concerning quality of life, the authors noted that the highest level of function was obtained by those patients

treated by surgery and irradiation, and that the lowest level of function was obtained by those treated with radical resection of the tumor, and, additionally, that only three with radical resection did not have radiation therapy. With regard to school performance, the authors noted no difference among the three treatment groups. The same observations were noted for psychological problems, no correlation being noted between psychological problems and school performance. Similarly, with regard to endocrine deficits among 35 patients, in group II there was an 87.5% rate of diabetes insipidus, in group I a 66% rate, and in group III only 1 of 21 patients (5% suffered diabetes insipidus).

The weighting of treatment in these patients was definitely toward conservative operations and radiation therapy, with the 8% and 14% mortality and recurrence rates respectively at a mean follow-up time of 10.5 years being at least comparable, and generally superior to aggressive surgery. There is a suggestion, but no explicit statement, by the authors that the mean follow-up times of 3.1 and 2.6 years, reported respectively by Symon and Sprich [147] and Hoffman [148], do not have the same significance as the 10-year follow-up in their study or in the paper of Manaka et al. [149].

Of real interest is the syndrome of unexplained coma/high fever/cardiovascular collapse as a well-described complication in about 10% of the patients treated with radical surgery, most probably caused by injury to the hypothalamus. This serves as an additional basis to the conclusion that resection attempts should not be pushed to the point of risking hypothalamic injury, as does the greater operative mortality which accompanies attempts at radical reoperation, cited by Katz [61], Matson and Crigler [150], and Sweet [151].

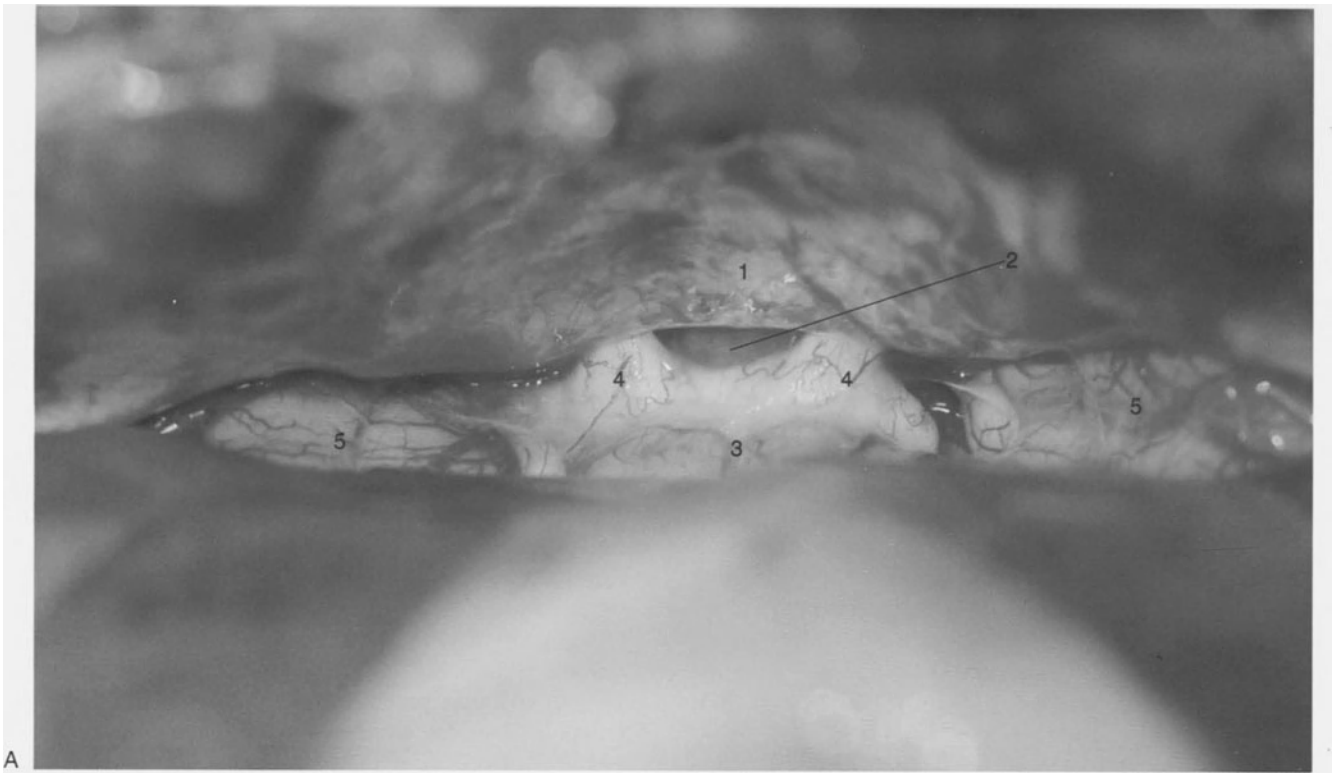
With regard to quality of life, the demands upon cerebral function (neurological, intellectual, psychosocial) increase greatly as the child grows into adolescence and adulthood. Patients treated with conservative surgical management and radiation therapy did very much better than those treated with aggressive surgery, clustering in the highest level of function on the modified CAT scale. Patients tend to drop from "good" categories to "fair" categories over a 10-year period of time. It may very well be that the psychological and social deterioration results from disturbance of hypothalamic connections to the thalamus, frontal lobes, and cortical areas, in addition to the hypothalamic pituitary complexes which are so essential to endocrine function. What, then, is most striking to the reader is the statement that "patients had uncontrollable anger or aggression, the presumed defect on the hypothalamus of an overly aggressive surgeon," to paraphrase Sweet's quotation (op.-cit.) of Shillito.

Van den Berge et al., in their 1992 paper [152], came to equally positive conclusions from results obtained through still different modalities of treatment. Structuring a basis around the now almost universally accepted concept that the cystic component of craniopharyngioma most often predominates over the solid, they reiterate the equally well-known fact that the surgery to remove the cyst carries with it significant mortality and morbidity. In the light of these two well-established anatomopathological and clinical observations, they pose the conjecture that "resolution or reduction of the cyst(s) may be sufficient to initiate lasting improvement." In order to attain this goal, i.e., the resolution or reduction of the cyst(s), they undertook the treatment of 31 patients with mainly cystic craniopharyngiomas by stereotactic injection of colloidal yttrium-90 into the cyst cavities. The 31 treated patients were selected, always on the basis of a primarily cystic component, from a total of 38 craniopharyngioma patients managed in Rotterdam or Haarlan during a 6-year period. Their patients ranged in age from 4 to 64 years at the time of treatment; the follow-up of the survivors ranged from 2 to 80 months.

The authors divided their treated patients into two subgroups depending upon whether the intracavitary brachytherapy was "primary treatment" or whether the patients had been treated elsewhere with some other accepted therapeutic modality, in which case they were classified as undergoing "secondary treatment." Of the 31 treated patients, 3 underwent multiple procedures *and in each instance were regarded as a new case;* 13 presented with raised intracranial pressure, and 9 required CSF shunting.

The Leksell frame was used in the Phillips Tomoscan to perform the stereotaxic placement of the colloidal yttrium-90 into the craniopharyngioma cysts. When stereotactic respiration of the cyst became necessary, it was never performed within less than 14 days, so as to allow five times the isotope's half-life to elapse. Two hundred Greys were delivered to the cyst wall by injected doses ranging from 0.63 to 36.46 mCi, and no external radiotherapy was used as adjunctive treatment. There were five tumor-related post-treatment deaths, occuring at 2, 11, 27, 35, and 65 months after intracavitary brachytherapy.

The clinical parameters which the authors used to evaluate their results were visual function (visual fields, visual acuity, funduscopy), endocrinological status, and radiologic outcome. The modified Rankin scale was used to grade the degree of handicap, taking into account neurological deficits, daily activities, school performance, and social integration as measurable parameters.



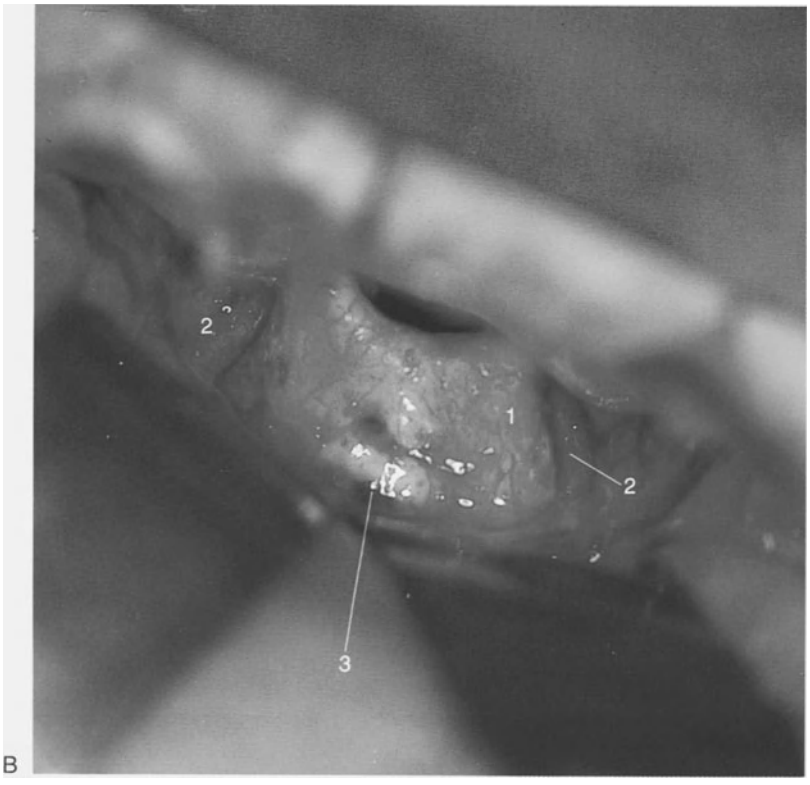


Figure 10.126. Retrochiasmatic tumors expand almost entirely behind the chiasm, though certainly there are both infrachiasmatic and, at times, small amounts of prechiasmatic extensions. (A) This child had an entirely retrochiasmatic tumor. The planum sphenoidale (*1*), prechiasmatic space (*2*), optic chiasm (*3*) and nerves (*4*), temporal lobes (*5*), and arachnoid bridging from the chiasm to the right temporal lobe has been opened, as has the prechiasmatic arachnoid. (B) This is another child with a retrochiasmatic tumor. After all of the arachnoid has been freed from the optic pathways (*1*) and later, in turn, freed from the internal carotid arteries, (*2*) one proceeds to identify the A-1 segments of the anterior cerebral artery so as to dissect the lamina terminalis (*3*) from the chiasm. (C, D) see p. 331.

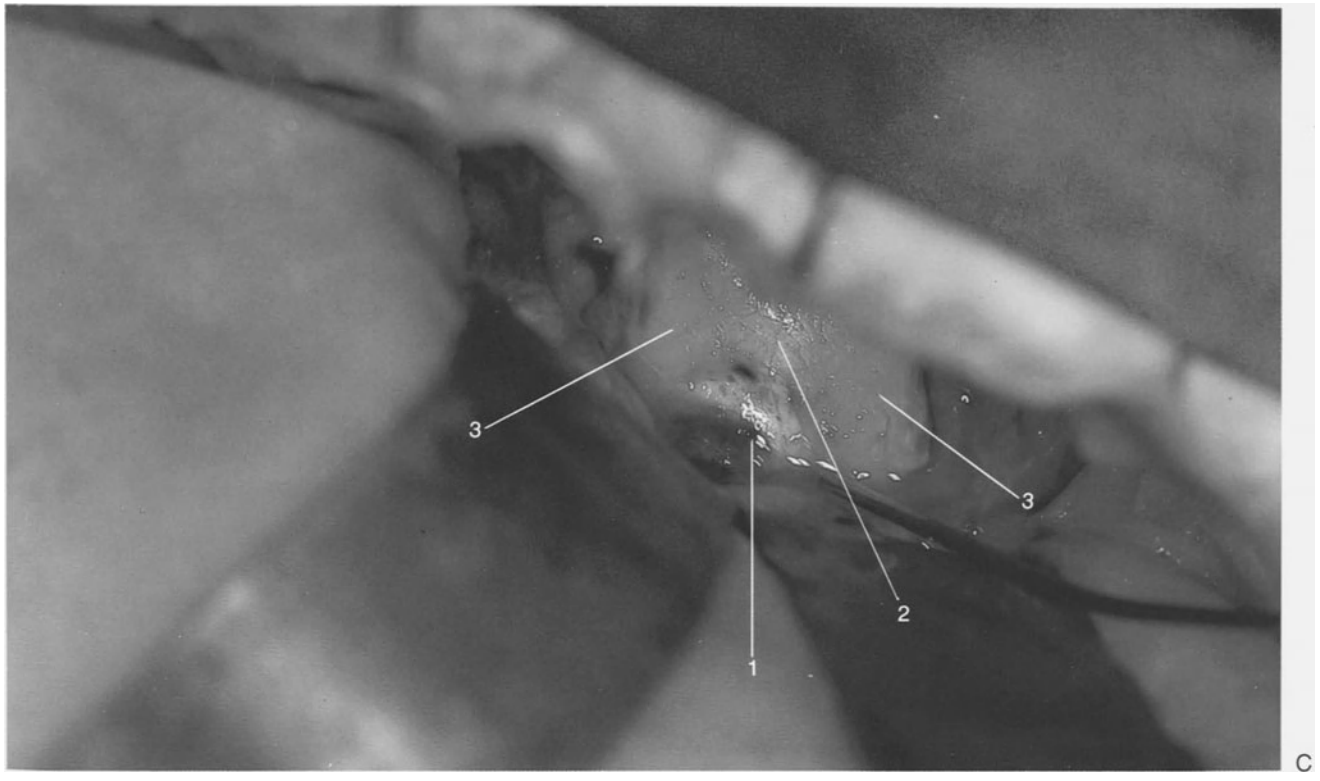
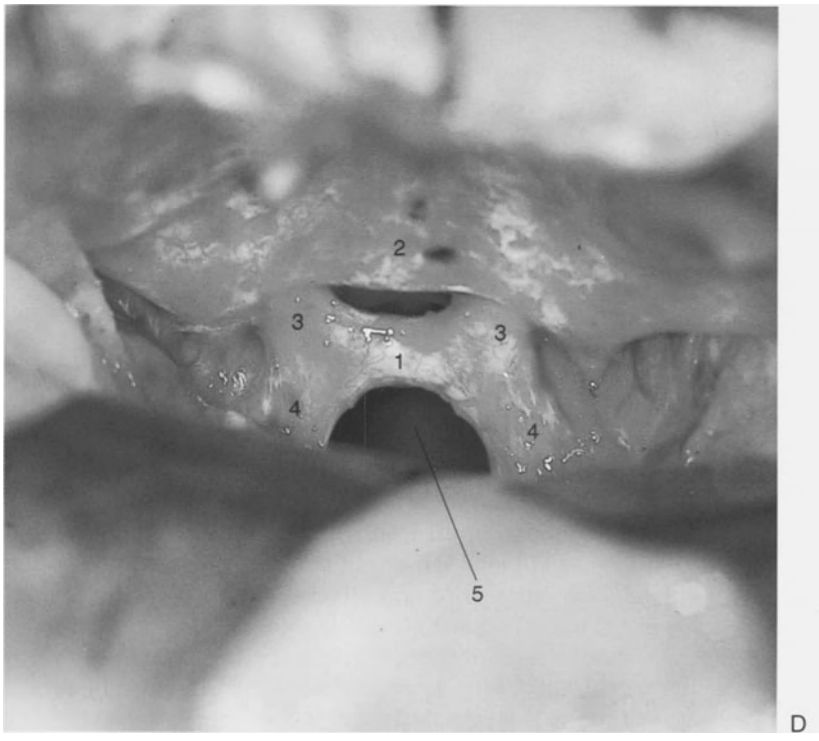


Figure 10.126. (C) Once the lamina terminalis (*1*) has been identified, one may dissect it from the posterior surface of the optic chiasm (*2*) and medial surfaces of the optic tracts (*3*), so as to approach the retrochiasmatic tumor through the III ventricle. (D) The anterior location of the optic chiasm (*1*), almost abutting upon the planum sphenoidale (*2*), and the shortness of the optic nerves (*3*) is well shown in this photograph of the same child who had had a retrochiasmatic craniopharyngioma. After the lamina terminalis was dissected from the optic chiasm and optic tracts (*4*), the retrochiasmatic tumor was removed through the interval between the optic chiasm and the optic tracts (*5*).



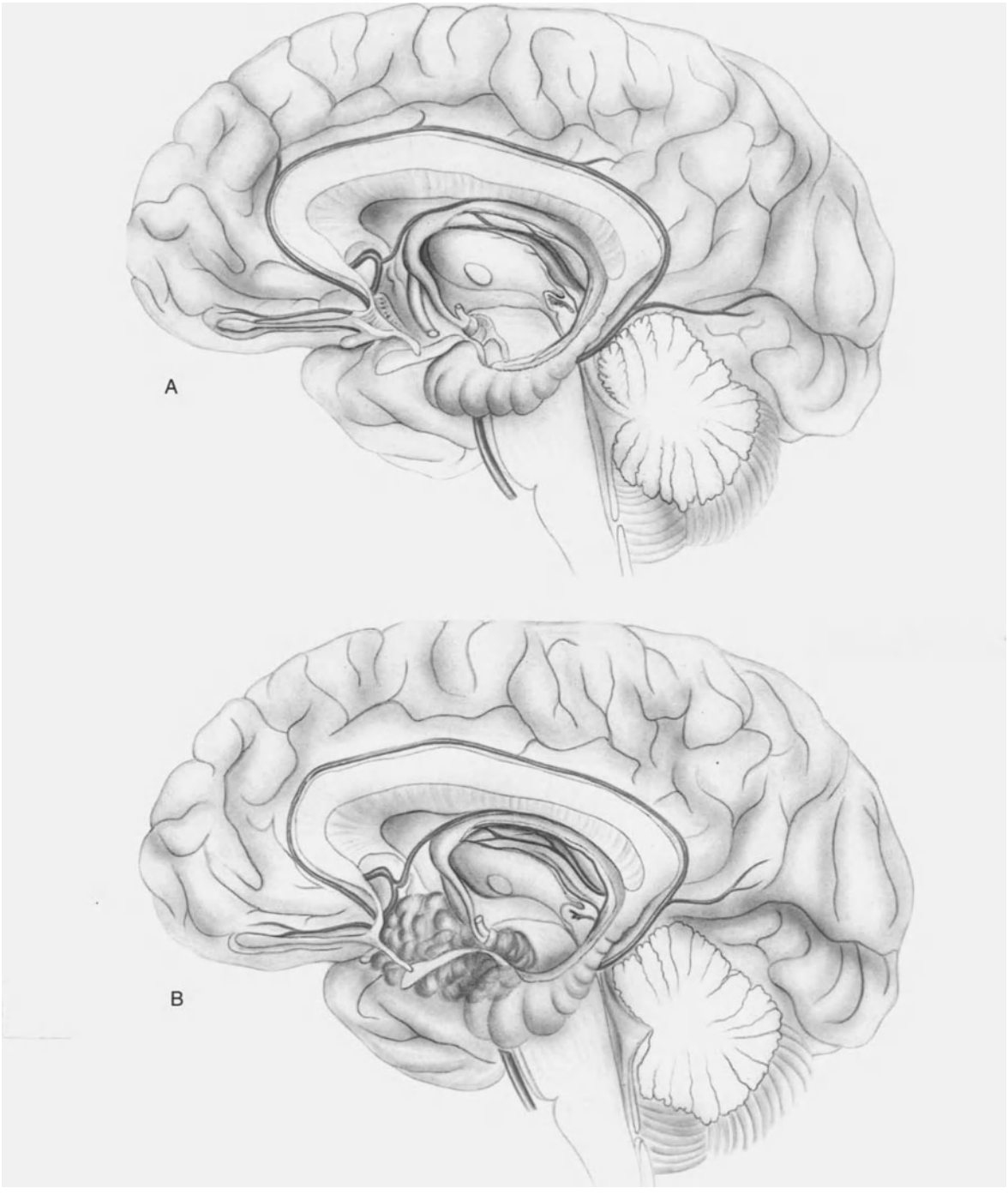
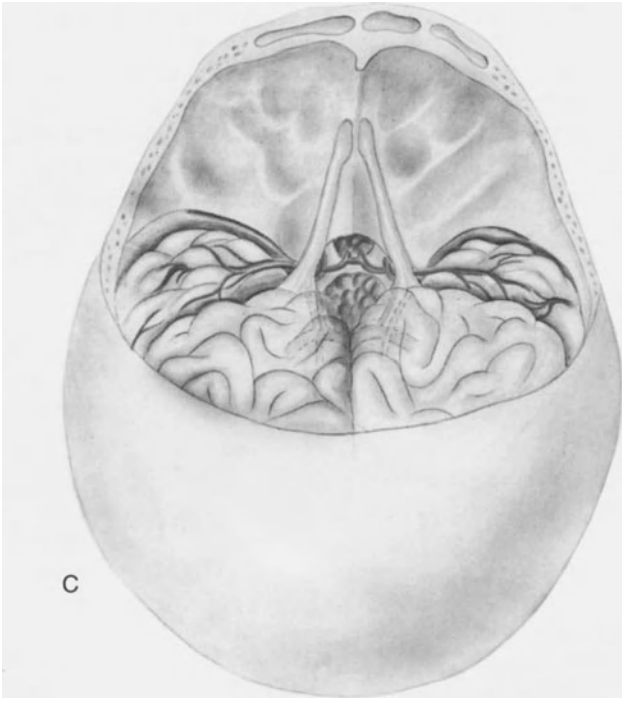


Figure 10.127. A series of five illustrations make up this figure, putting into relief, in the sagittal plane, "eloquent" anatomical structures which may be involved by retrochiasmatic tumor extension, and in the axial plane (C–E) the surgeon's perspective. In (A), from anterior to posterior, one sees the olfactory bulb and nerve, the optic nerve and chiasm, the lamina terminalis taking origin from the posterior portion of the chiasm and passing superior and anteriorly to become continuous with rostrum, the anterior perforated substance, the anterior commissure, the columns and body of the fornix, the mammillary body, and the III ventricle with the massa intermedia at its center. The pes hippocampus, a parasagittal structure, is also illustrated. (B) A primarily intra III ventricular extension of a retrochiasmatic tumor is depicted, as are the deformities and involvement of "eloquent" structures described in the previous figure. (C–E) see p. 333.



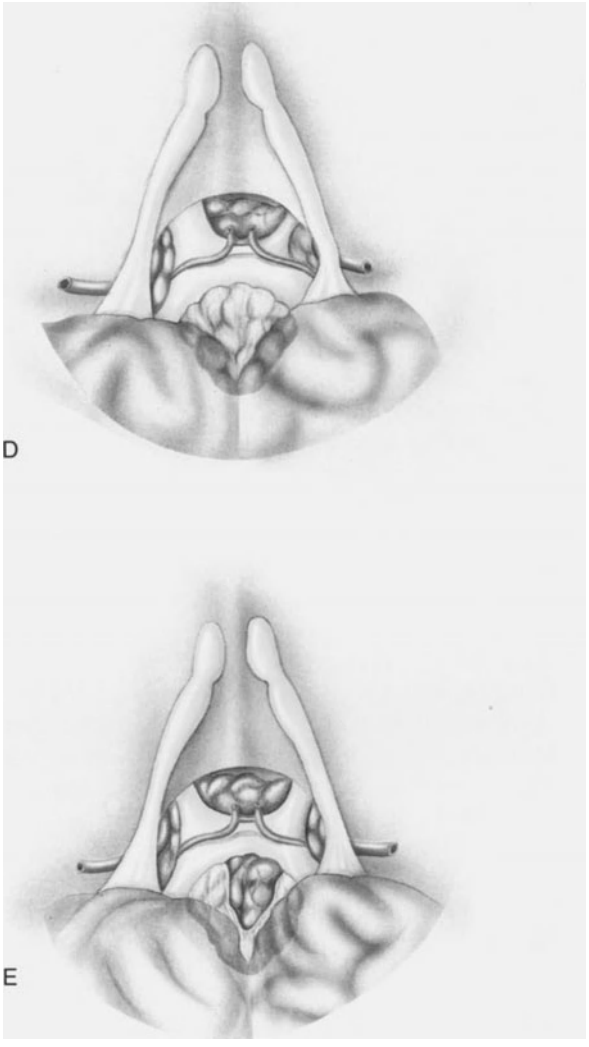


Figure 10.127. (C) With the olfactory nerves preserved anatomically, the frontal lobes fallen away from the anterior fossa (this is not an artistic exaggeration, it occurs in young children), one sees a diagrammatic representation of a craniopharyngioma which has expanded primarily in the retrochiasmatic compartment, invading the III ventricle. This is the appearance when the tumor occupies the III ventricle, not when it occupies the interpeduncular space. Represented as shadows extending from the internal carotid artery are the posterior communicating and posterior cerebral arteries and, crossing over them, the III cranial nerves. (D) and (E) are coned-down views of the operative area before and after opening of the lamina terminalis.

The observations led the authors to suggest that in cystic craniopharyngioma there may be a direct relationship between volume and prognosis, since four of the nine patients with cysts larger than 30 ml eventually died. Additionally, three of the total of five deaths which occurred were in patients with multicystic craniopharyngioma. Eighteen of the 31 patients were gainfully employed, performing well in school, or active in the home environment. Eight patients were seriously disabled. Concerning visual function, there was a correlation between improved or preserved function in the "primary treatment" group on one hand, and deterioration in the "secondary treatment" group on the other: "improvement of visual fields was significantly less, and deterioration of both acuity and visual fields occurred considerably more often in the secondary treatment group."

Regarding cyst size, there was complete post-treatment resolution in 10 patients and a decrease in volume in 12 patients. In three patients there was new cyst formation, requiring a second intracystic injection of yttrium-90 in two and a third injection in one; in six patients the cyst volume remained stable; in three patients the cyst volume increased; and in two patients the solid component of a primarily cystic tumor increased.

The authors were successful in maintaining normal pretreatment pituitary function in nine patients and in normalizing pretreatment deficiences, with "disappearance of diabetes insipidus and normalization of growth hormone levels" in one patient. On the other hand, the total of 10 patients with three or more pretreatment pituitary insufficiencies increased to 17 following intracavitary brachytherapy. There was no instance of excessive production of pituitary hormone following treatment.

Here, now, is proposed an alternative, in a supplemental or supportive treatment modality! The authors stand four-square in front of the neurosurgical reader proposing intracavitary brachytherapy alone or in combination with external radiation (they do not specify whether they mean standard radiotherapy or radiosurgery) as an alternative to radical surgery in those cases in which the craniopharyngioma is mainly cystic. Referring to the 82% of patients with cystic tumors, they state that "brachytherapy often obliterates the cyst or effectively reduces its volume, making major surgery aimed at total removal superfluous."

In evaluating our results, we would do well to include those patients lost to follow-up with our tumor- or surgery-related deaths.

Regarding comparative results between surgical resection and brachytherapy, it is not possible to draw hard and fast conclusions concerning visual or endocrine outcome, but it appears that social reintegration and overall quality of survival are superior in the brachytherapy patients. In this latter group, furthermore, it appears that the "primary treatment" patients do better than the "secondary treatment" patients – but this, then, is intuitive, since the "secondary treatment" patients are those, for surgery as for brachytherapy, whose lesions in and of themselves were refractory to the primary treatment. One may not, tempting though it may be, attribute treatment failure in the first instance to lesser qualified surgical or brachytherapy groups.

In sum, the current literature regarding treatment modalities for craniopharyngioma in children and adults shows that those advocating aggressive surgery with the intention to resect all of every craniopharyngioma all the time are not obtaining the same quality-of-life results as those approaching the problem from a multidisciplinary point of view: resecting that amount of tumor which comes away easily and collaborating with brachytherapists in selected cases, or with radiosurgeons postoperatively to treat the residual tumor.

Corpus Callosum and Septum Pellucidum Tumors

Corpus Callosum: Surgical Anatomy

(Figs. 10.128, 10.129)

Corpus callosum tumors should not be operated on since they are either highly malignant or completely benign. The former are represented by astrocytoma diffusum, primitive neuroectodermal tumor, diffuse intraparenchymal ependymoma, glioblastoma multiforme, lymphoma, and atypical teratoma. All of these extend into the corpus callosum from surrounding parenchymal structures. Though partial or "total" resection is technically possible, the invasion of the parietal lobes and fornices, present to a greater or lesser degree, renders surgical removal more damaging than beneficial. Roentgen therapy, on the other hand, is palliative in some tumors, and may be curative in others. Consequently, invasion of the corpus callosum by a malignant tumor is, in and of itself, a contraindication to surgery.

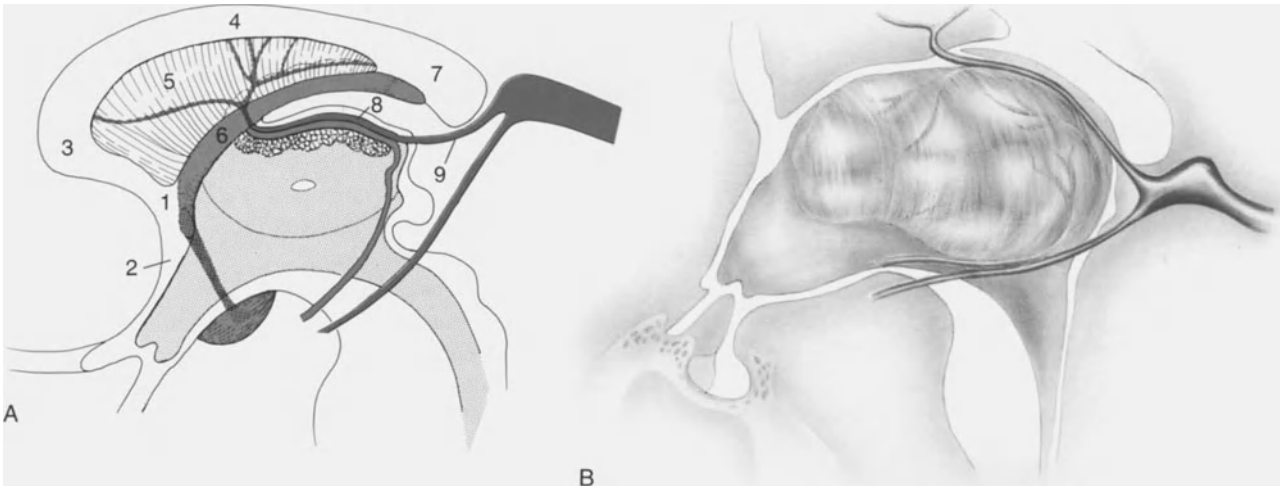


Figure 10.128. (A) The rostrum (*1*) of the corpus callosum is in continuity inferiorly with the lamina terminalis (*2*) and superiorly with the genu (*3*). The genu and body (*4*) give purchase to the septum pellucidum (*5*), which also attaches inferiorly and posteriorly to the body of the fornix (*6*). The splenium (*7*) overhangs the internal cerebral (*8*) and galenic (*9*) veins. In (B), on the other hand, the III ventricular tumor is illustrated to extend inferiorly from the junction of the septum pellucidum and the bodies of the fornix, in a manner similar to that which occurs in III ventricle choroid plexus papilloma.

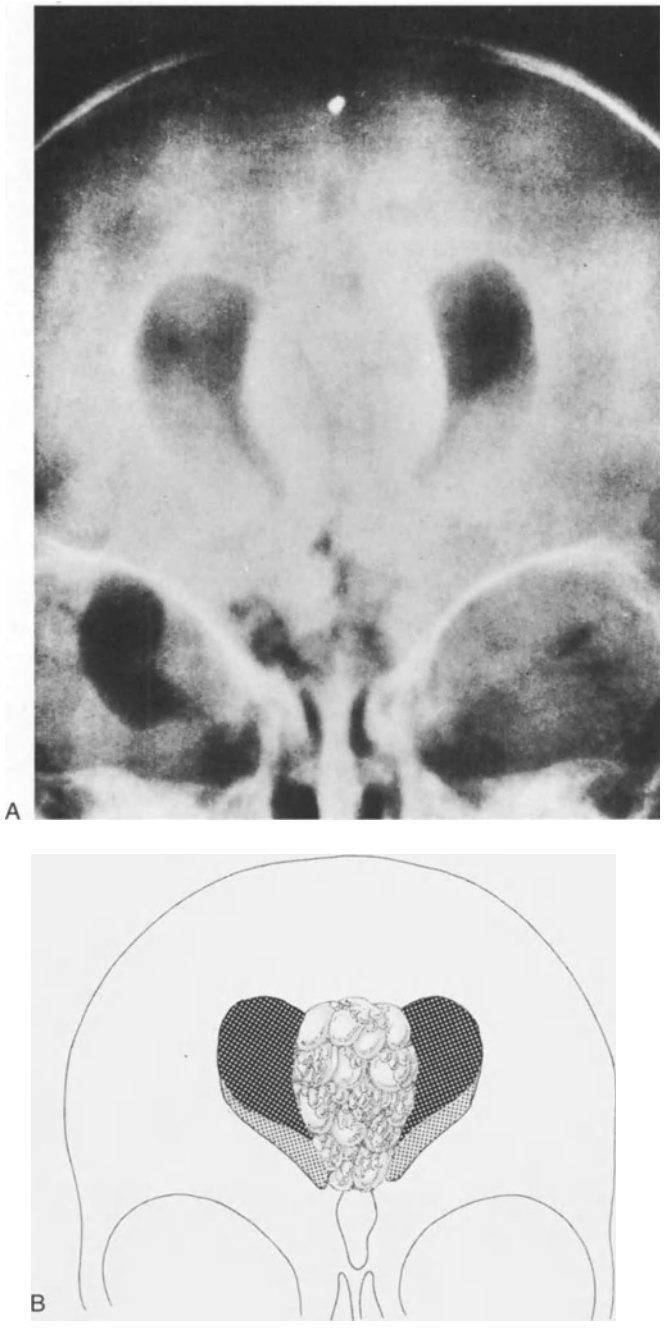


Figure 10.129. (A) Lipoma of the corpus callosum revealed in a pneumogram. (B) a *lipoma* extending also into the corpus callosum.

Benign tumors such as lipoma or typical teratoma need not be resected since they are nonexpansile and have no potential toward malignancy. Resection of these tumors carries with it the operative and postoperative risks, but no benefits.

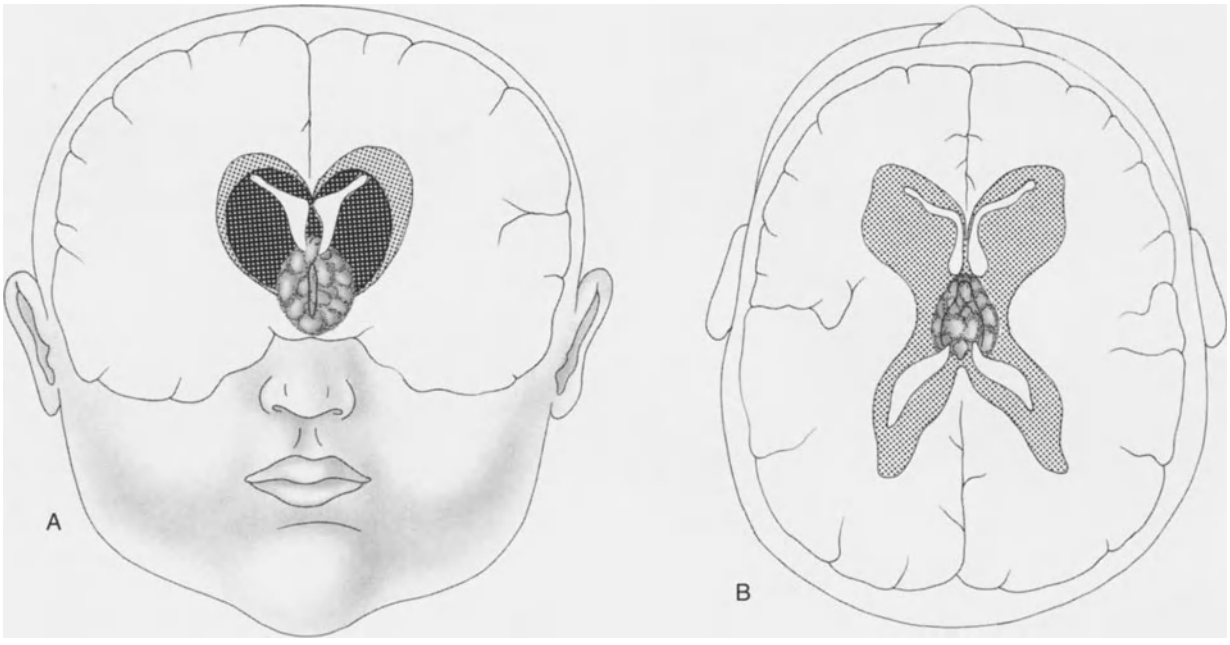
Septum Pellucidum: Surgical Anatomy

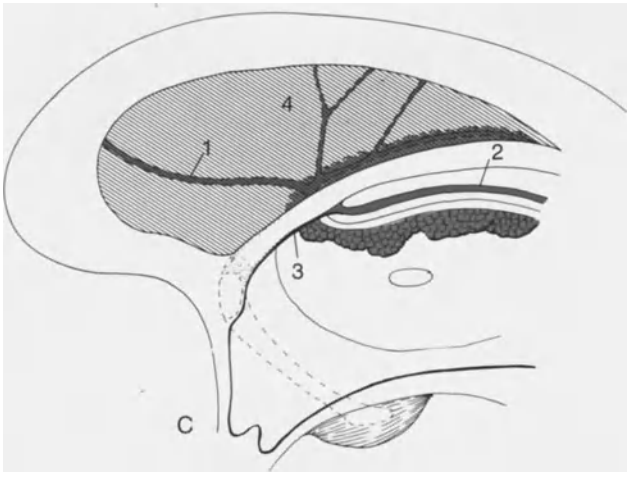
(Figs. 10.130–10.132)

The septum pellucidum is a double membrane strung tautly between the genu and body of the corpus callosum anteriorly, and the body of the fornix and transverse fissure posteriorly and inferiorly. The anterior septal vein runs within it posteriorly from the genu of the corpus callosum to the internal cerebral vein, to which it is tributary. There is a potential space which, on occasion, communicates with the ventricular system, becoming a cyst of the septum pellucidum. This may extend into the transverse fissure as a cavum Vergae. The cavum Vergae and cyst of the septum pellucidum communicate only with the ventricular system. These are to be distinguished from the cavum veli interpositi, a subarachnoid cyst of the tela choroidea of the roof of the III ventricle. The cavum veli interpositi is filled by subarachnoid fluid percolating up through the ambient and quadrigeminal cisterns. Consequently, the distinction beween cavum Vergae and cavum veli interpositi is that the former fills with ventricular fluid and the latter with subarachnoid fluid.

Though intraventricular tumors, such as subependymal astrocytomas, tubers in association with tuberous sclerosis, hamartomas, and ependymomas may adhere to the septum pellucidum, they do not grow from this structure. No specific surgical technique applies to their removal. They should not be resected, and generally the child need not be shunted. One does, however, encounter cysts of the septum pellucidum which progress to such dimensions as to cause hydrocephalus by obstructing the foramina of Monro and the III ventricle, or to act as expansile masses. Surgical treatment of such cysts of the septum pellucidum consists either of a craniotomy, transventricular approach to the cyst wall, and marsupialization of both surfaces of the cyst wall with both lateral ventricles, or of inserting the proximal end of a shunting system into the cyst, thereby collapsing it. Marsupialization is ideally performed with a laser since vaporization of the septum pellucidum, theoretically, will be less often associated with scarring, fibrosis, and cyst reformation. *Shunting sounds simpler than it is*, since the complicating factors are imponderables: the resistance of the cyst wall, and the reliability of inserting the proximal end of the tubing into the cyst cavity. These cysts, whether treated surgically with marsupialization or shunting, tend to recur.

Increased intracranial pressure and papilledema associated with intraspinal tumors are now well documented, though there has not been unanimity concerning their causes. To our knowledge, all heretofore reported cases (the majority being of adults) were not complicated by hydrocephalus. The pathogenesis of the papilledema has been discussed only from the point of view of increased intracranial pressure, a broad entity of different etiologies.





◀ **Figure 10.130.** The two schematic drawings which compose this figure permit the reader to orient himself concerning septum pellucidum masses in axial and coronal sections. (A) and (B) represent a neoplasm compatible with spongioblastoma or giant cell subependymal astrocytoma. (C) The anterior septal vein (*1*), a tributary to the internal cerebral vein (*2*), generally at the foramen of Monro (*3*), is located within the septum pellucidum (*4*).

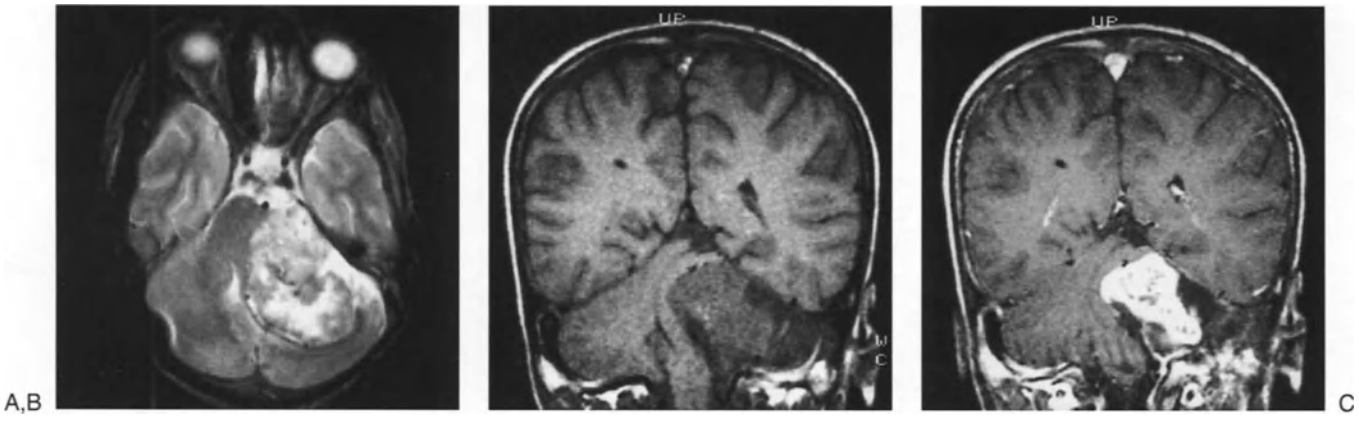


Figure 10.131. Schwannoma, type A, involving the left VIII cranial nerve, in a 14-year-old boy. (A) This axial spin-echo 24,000/90 image reveals the presence of a very large mass with a dishomogeneous signal, occupying the entirety of the cerebellopontine angle, displacing the brainstem and the IV ventricle to the contralateral side. (B) This is the pre-gadolinium study. (C) This post-gadolinium image reveals the dishomogeneous enhancement characteristic of this type of tumor, as well as the foci of low intensity signal indicative of necrotic areas.

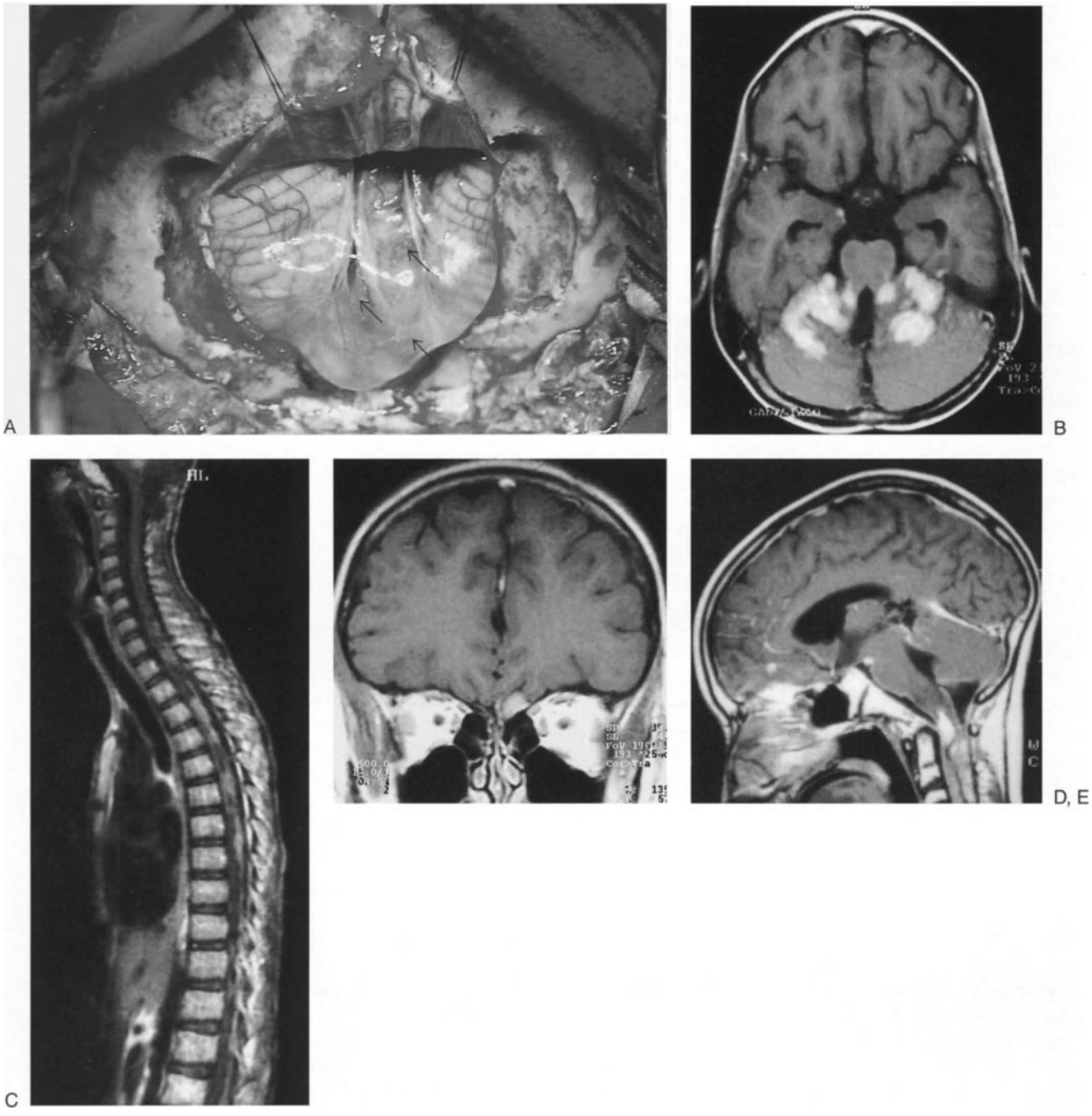


Figure 10.132. (A) Medulloblastoma: The arachnoid of the cisterna magna is covered by a multitude of seedlike-appearing metastases (*single arrows*). This is common in medulloblastoma. It is not indicative of either a poor prognosis or subsequent central nervous system or peripheral metastases. The microseeding of medulloblastoma illustrated in (A) is an extraordinarily common observation along the arachnoid of the cisterna magna. On the other hand, macro seeding very rarely occurs in the early stages of medulloblastoma evolution. In (B), an axial post-gadolinium image, one sees hyperintense leptomeningeal masses around the inferior surface of the cerebellum and toward the pontocerebellar angles. There is also seeding at the uncus of the hippocampus, at the pole of the right temporal lobe, and along the left frontal lobe. In (C), a sagittal post-gadolinium study of the medulla spinalis, hyperintense lesions – "drop metastases" speckle the subarachnoid spaces. (D) This is a coronal post-gadolinium image of another child with medulloblastoma, illustrating another common post-radiotherappy site of medulloblastoma metastases, subfrontal (gyrus rectus). (E) Diffuse micro arachnoidal seeding, along the frontal lobe, inferior surface of the occipital lobe, midbrain, pontine and medullary cisterns and craniovertebral junction is illustrated in this post-gadolinium study as high signal intensity areas.

Teng et al. [153] described two children, 11 and 12 years of age. The papilledema was considered to result from either mucinous material produced as a result of degenerative changes and hemorrhages from the tumor's alteration of CSF absorption; or increased CSF viscosity due to elevated protein level and obstruction of the CSF absorptive site.

Sixty-four cases of childhood primary intraspinal tumors were treated surgically by us (Table 10.12). Hydrocephalus complicated nine cases (14%), two had Recklinghausen's disease (3%), and subarachnoid hemorrhage was the initially observed event in one (1.6%).

The nine patients with hydrocephalus were analyzed further. All except one were under 3 years of age.

Of the three intramedullary tumors associated with hydrocephalus, the astrocytoma involved the entire spinal cord from C-1 to L-4; the mixed glioma extended from the upper cervical cord into the medulla; and the other glioma (histology unknown) occupied the cervicothoracic cord (C-4 to T-10). The two intradural dermoid cysts were well circumscribed, and in the lumbar area the teratoma extended from low cervical to thoracic cord, and the malignant intradural neurofibroma occupied L2–3. One of the primary extradural sarcomas was at the lower lumbar area and the other between T-5 and T-12.

Increased intracranial pressure associated with intraspinal tumors is a well-documented, though rare, syndrome. Arseni and Maretsis [154] reported three cases from a series of 289 tumors of the lower spinal cord and reviewed 23 cases from the literature. Raynor [155] found only 36 cases after reviewing the literature. According to his analisys, the predominant tumor type was ependymoma (56%); 69% of the known lesions were intramedullary gliomas. Sixty-two percent of the patients with papilledema were female and 45% were under 30 years of age.

Factors other than CSF absorption and viscosity may predispose to increased intracranial pressure and papilledema in patients with spinal tumors. Findings in our cases and those of others suggest that arachnoiditis [154], intraspinal tumors, and tentorial [156] or cisternal [157] block, may be significant causative factors. Leptomeningeal infiltration by epithelial or mesenchymal tumors, with elevated CSF protein level and hypoglycorrhachia [158], are additional possible causative entities. Maurice-Williams and Lucey [159] described two patients with intraspinal malignant schwannomas who had raised intracranial pressure secondary to neoplastic arachnoiditis. Subarachnoid hemorrhage with intraspinal tumors also can be a cause of this syndrome. Nassar and Correll [160] reported four cases of subarachnoid hemorrhage due to spinal cord tumors. Eleven of their patients had elevated pressure and six papilledema. Eight of these were ependymoma. Papilledema with elevated CSF protein level has been seen also in patients with Guillain-Barré syndrome. Glasauer [161] postulated that the papilledema in patients with spinal cord tumors is secondary to the elevated CSF protein level, though Joynt [162] came to the opposite conclusion. Bamford and Labadie [163] reported a case of neurilemoma, with normotensive hydrocephalus, concluding that the leakage of fibrinogen into the CSF may be the cause for the hydrocephalus.

From the history and clinical findings, an attempt was made to identify the pathogenesis of hydrocephalus in our nine patients with intraspinal tumors. The brainstem tumor extended down to the level of C-5, and three spinal cord tumors only as high as C-4 and C-2. This makes it unreasonable to postulate that the hydrocephalus results from an obstruction of CSF at a level between the fourth ventricle and the foramen magnum.

In children with a dermoid cyst, chemical or bacterial meningitis, via the dermoid sinus, could be the cause of the increased pressure. The teratoma cases were associated with spina bifida, Arnold-Chiari malformation, and aqueductal stenosis being diagnosed in the neuroradiologic investigations. Both extradural sarcomas had

Table 10.12. Intraspinal neoplasms in infants and children (operative cases) 1963–1979

Type	No.	No. with hydrocephalus
Intramedullary (n + 16)		
Astrocytoma	7	1
Ependymoma	5	0
Mixed glioma	2	1
Oligodendroglioma	1	0
Unknown (glioma)	1	1
Intradural, extramedullary (n = 30)		
Lipoma	12	0
Dermoid cyst	7	2
Schwannoma	3	0
Teratoma	2	1
Medulloblastoma[a]	2	0
Ependymoma[a]	1	0
Sarcoma[a]	1	0
Lymphangioma	1	0
Neurofibroma	1	1
Extradural (n = 18)		
Undifferentiated sarcoma	4	2
Neuroblastoma	3	0
Ewing's sarcoma	2	0
Teratocarcinoma	2	0
Osteoblastoma	2	0
Ganglioneuroma	1	0
Osteogenic sarcoma	1	0
Rhabdomyosarcoma	1	0
Wilm's sarcoma	1	0
Malignant lymphoma	1	0
Total	64	9

[a] Possible dissemination from the intracranial lesion.

complete blockage on myelography. However, the total CSF protein level above the lesion was not high in one case. Autopsy in the other case revealed leptomeningeal tumor infiltration.

It is evident that the occurrence of hydrocephalus in children with spinal cord tumors may not be attributed to any single etiologic or pathogenetic factor, since our cases varied widely with regard to both location (spinal level and intramedullary or extramedullary) and nature (benign or malignant) of the tumor. Quite the contrary, we may exclude spinal cord block, leptomeningeal infiltration by tumor cells, or remarkably elevated CSF protein level as single causative factors. One should, consequently, consider a spinal cord tumor when papilledema and hydrocephalus are present and no intracranial causes may be identified.

Spinal Tumors

Intramedullary tumors are rare in children, with an annual incidence of about one case per million children [164]. As in adults, the most frequent oncotypes are low-grade astrocytomas and ependymomas, which account for about 80% of all intramedullary tumors (74.6% in the present series). The relative frequency in adults is reversed in children, astrocytomas being more frequent than ependymomas, 44.8% and 29.8%, respectively, in our series.

Astrocytomas and ependymomas are the most frequent intramedullary tumors, accounting for about 80% of all tumors in this site [164–171]. The incidence of these two oncotypes is age-related: in children and adolescents astrocytomas (predominarly low grade) are two to four times as frequent as ependymomas; in adults ependymomas are more frequent; in patients over 60 years of age the incidence of these two tumors is similar [164, 165, 171–174].

Although the great majority of astrocytomas and ependymomas behave as slow-growing tumors, they have different prognostic characteristics and present different problems in decision making (Tables 10.13–10.15) [165, 168, 175–177].

Total removal of ependymomas has been reported to vary from 76% to 100% [168, 173, 175, 176], and is followed by good functional results and a recurrence rate ranging from nil to 4.3% [173, 178]. There are, however, differing opinions on the possibility of total removal of ependymomas localized at the cervicobulbar junction or at the conus/cauda equina [175, 179]. Similarly, opinions on radiotherapy vary; some workers consider its value doubtful [173, 176], while others endorse it [164, 180–182].

Table 10.13. Preoperative and postoperative functional status in patients with astrocytoma

Karnofsky functional status	Before operation (no. of cases)	At discharge (no. of cases)	At follow-up (no. of cases)
Grade 1	5	5	7
Grade 2	17	10	13
Grade 3	7	14	9

Table 10.14. Preoperative and postoperative functional status in patients with ependymomas

Karnofsky functional status	Preoperative (no. of cases)	At discharge (no. of cases)	At follow-up (no. of cases)
Grade 1	5	4	7
Grade 2	8	7	5
Grade 3	3	5	4

Table 10.15. Actuarial survival in months

Oncotype	Total removal	Subtotal removal	Biopsy
Low-grade astrocytoma	114	109	84
Ependymoma	219	130	–

Table 10.16. Extent of resection

Oncotype	Total removal	Subtotal removal	Biopsy
Astrocytoma	10 (34.4%)	11	8 (27.5%)
Ependymoma	13 (81%)	3	–

Reports of total removal of intramedullary astrocytoma also vary considerably: between 14.3% and 50% in most series [165, 183], up to 88.2% in a more recent one [177]. However, the value of an aggressive surgical strategy is debatable, since the extent of resection has been shown to have little influence on outcome [166, 179].

Extent of Resection

The possibility of obtaining total removal differs radically between ependymoma and astrocytoma, the figure for the latter being 34.4% (Table 10.16). Our results are practically identical to those reported by other authors for both children and adults [165, 175, 184, 185]. The only exception is the series reported by Epstein and Epstein [172], consisting of 19 children in whom gross total removal was performed.

The different anatomopathological characteristics of these two oncotypes explain why the possibility of radical surgery varies so widely. In fact, in ependymoma there is almost always a plane of cleavage with the healthy tissue, which is rarely observed in astrocytoma.

With regard to total removal of conus ependymomas, we have not found this possible in children, though one group [176] has accomplished it in an adult.

Radiotherapy

In the management of intramedullary tumors one very controversial matter is the usefulness of radiotherapy. In the literature two opposing lines of thought are found. On the one hand, there are those who believe that radiotherapy is indicated for palliative purposes in high-grade tumors only. They suggest it as adjuvant therapy in patients with astrocytomas or ependymomas with a surgical recurrence, basing their opinion on surgical and biological considerations. From a surgical viewpoint, as mentioned above, the great majority of ependymomas and at least a third of astrocytomas can be removed totally. Biologically speaking, both ependymomas and benign astrocytomas are slow-growing tumors with a low cellular growth rate and, therefore, are not very susceptible to radiotherapy. On the other hand, there are recent reports in which radiotherapy is shown to have increased survival times and prolonged the interval before recurrence of intramedullary tumors [164], especially ependymomas.

In view of the risks connected with radiotherapy, we believe this type of complementary treatment should be limited to high-grade tumors and to recurrences of astrocytoma and ependymoma in which histological examination indicates a tendency towards malignancy.

Neurological Status

The data in our series and those reported in the literature clearly demonstrate a close relationship between pre- and postoperative clinical conditions. As observed by Epstein et al. [176], postponement of surgical treatment until the appearance of a significant motor dysfunction invariably leads to an increase in the size of the tumor and thinning of the adjacent spinal tissue. This increases the risk of postoperative neurological deterioration. In other words, the longer the spinal cord is compressed by the tumor, the more susceptible it is to surgical trauma and the less its potential for long-term recovery.

In our experience, and as reported in other series, only patients whose preoperative condition was good or only partially impaired (groups 1 and 2) have experienced long-term functional improvement.

Patients subjected to surgery while in poor condition (group 3) have invariably worsened postoperatively and not improved again. This poor outcome not only represents a further reduction of patient autonomy but is also associated with a greater incidence of infectious, trophic, and respiratory complications.

Taking into account the young age of these patients and the long survival expected for patients with most intramedullary tumors, we believe it is necessary to perform surgery early, while the neurological condition is still good.

Most patients with intramedullary tumors, low-grade astrocytomas and ependymomas alike, generally have a long survival, although this is often accompanied by some degree of neurological dysfunction, particularly in the case of astrocytomas.

While for ependymomas there is a clear relationship between the length of survival and the extent of resection, in the case of astrocytomas no such relationship between survival and total removal or subtotal removal or biopsy was observed. Similar results have been reported in other studies [179].

It seems that this characteristic of astrocytoma stems from its biological features, which are those of a slow-

growing lesion; the fact that it is not uncommon to perform operations for astrocytoma on adult patients whose initial symptoms, usually mild, began 15 and 20 years earlier seems to corroborate this.

Furthermore, the greater risk of postoperative neurological deterioration observed in the case of astrocytomas warrants a more cautious surgical approach to this oncotype than to ependymomas.

In conclusion, the majority of intramedullary tumors in children are slow growing. The most significant prognostic factors in these tumors are the histological type (ependymoma vs. astrocytoma) and grade.

Total removal is possible in the great majority of ependymomas, and in a third of astrocytomas. However, in the latter the extent of resection does not significantly influence survival.

Early surgical treatment is crucial, because the larger the tumor and the poorer the patient's condition, the greater are the risks connected with operation. The indications for radiotherapy and its value remain controversial.

Such vertebral column tumors as the osteoma, hemangioma, and aneurysmal bone cyst have no neurosurgical implications once the diagnosis is made. The interested reader is referred to standard texts on pediatric radiology for the diagnosis of such conditions, and pediatric orthopedics for their treatment. Here, it suffices to state that the aneursymal bone cyst should be treated with roentgen therapy, and that one must avoid the "temptation" to biopsy these lesions since they bleed so profusely as to challenge the most gifted surgeon.

The epidural tumors of childhood are metastatic, either hematogenous, renal, or neuroblast in nature. Almost never seen by a pediatric neurosurgeon, they are treated with roentgen therapy and chemotherapy.

Metastatic Spinal Tumor (Fig. 10.133)

Metastatic spinal cord tumors are extraordinarily rare in childhood, though systemic tumors (such as lymphoma or leukemia) are common. The latter are successfully treated with chemotheapy, so that seldom is a pediatric neurosurgeon called upon to decompress the spinal cord because of epidural metastases. The surgical technique for decompression consists simply of laminectomy, taking care to limit the extent of the laminectomy so as to minimize postoperative scoliosis. We recommend radiation therapy rather than decompressive laminectomy.

The majority of metastatic spinal cord tumors seen by neurosurgeons are truly CSF dissemination of primitive neuroectodermal tumors, regularly treated by che-

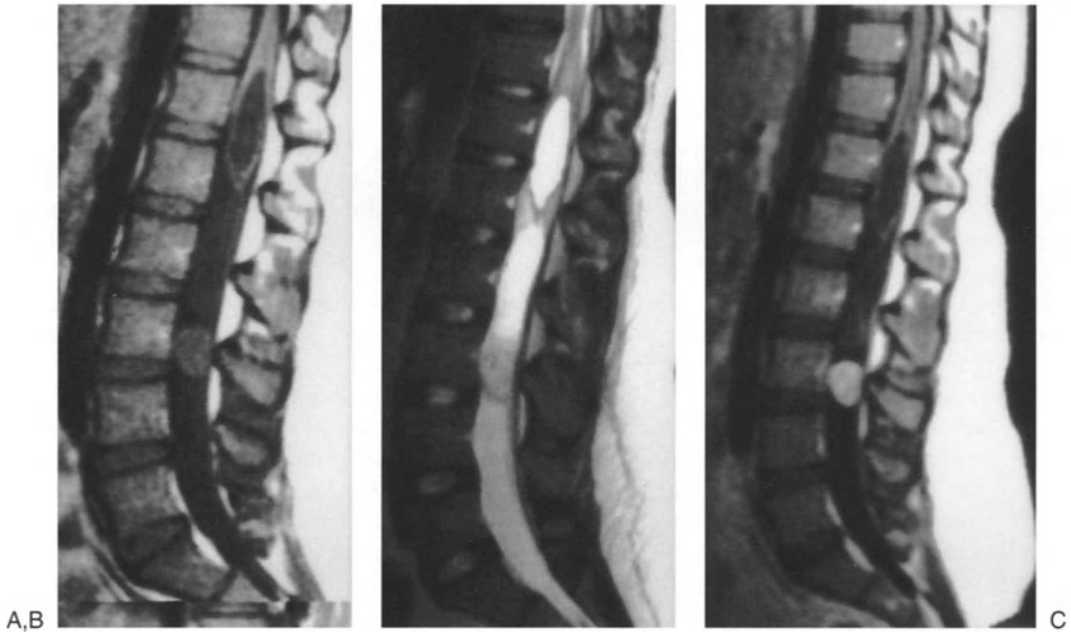


Figure 10.133. This ependymoma of the cauda equina is typical in appearance, and most interesting in that it is associated with (probably the cause of) cystic dilation of the terminal (ventricle) portion of the central canal within the conus medullaris. (A) This sagittal image reveals an enlarged conus medullaris and the cystic lesion at its center (terminal ventricle). One sees the intradural, cauda equina, soft tissue mass at L3. (B) This sagittal turbo 3,000/150 image reveals the high signal intensity of the cyst (identical to CSF) and the hypointense intradural L3 level lesion in this T2 weighted image. (C) This post-gadolinium injection reveals tumor enhancement and no intensity modification of the associated conus medullaris cyst.

motherapy and/or radiation therapy. Moreover, this seeding is most often downstream *and* upstream, into subarachnoid spaces of the brain and spinal cord, intracerebral or/and intramedullary.

Intradural-Extramedullary Tumors

Only the arachnoial cyst and neurofibroma occur in this location in childhood.

Arachnoidal Cyst

The arachnoidal cyst is now easly diagnosed preoperatively with MRI. Surgery consists of resecting as much of the cyst wall as possible, and then reparing the dura around the perimedullary or periradicular area involved.

Neurofibroma

The neurofibroma of childhood extends from the extramedullary/intradural space, along a nerve root, through the intervertebral foramen and then into either the chest or abdomen. Consequently, removal of these tumors is best performed with two surgical teams (one neurosurgical and the other general surgical). The resection should not be carried out in stages. It is not advisable for either the neurosurgeon or general surgeon to remove the tumor from, respectively, the spinal canal or paravertebral space, leaving a clip in place so that the other surgeon, working from the opposite direction, will know when to stop, since it is of primary importance to remove tumor from the intervertebral routes. Consequently, these hourglass neurofibromas should be removed in a single sitting, with the neurosurgeon working first to remove tumor from the spinal canal and the general surgeon then proceeding to remove the tumor from the prevertebral space.

Different from all other pediatric spinal neurosurgical procedures, removal of an hourglass neurofibroma is accomplished by using a "hockey-stick" skin incision with the short limb along the midsagittal plane (behind the spinous processes) and the long limb extending either over the thorax or abdomen, depending upon whether the tumor extends into the thoracic or abdominal cavities. This incision permits both the neuro- and general surgeons access to the spinal canal and prevertebral spaces in a single sitting. A laminotomy permits exposure of the spinal canal, and a costotransversectomy (for thoracic) and transverse process resection (for abdominal) permit exposure of the interverterbral foramen so that the neurosurgeon may follow the tumor extending along the nerve root. This permits him to remove all of the intraspinal tumor from the subdural space and the intervertebral foramen well past the dorsal root ganglia. After the dural defect has been repaired, either by simple suturing or placing a dural graft, the general surgeons may proceed to remove the intrathoracic or intra-abdominal tumor. Repositioning the laminotomy flap may be performed before the general surgeons proceed, or after they have completed their work.

Intramedullary Tumors

Cystic Intramedullary Astrocytoma (Fig. 10.134)

Since it is impossible to distinguish an intramedullary cystic astrocytoma from syringomyelia, if the cavity is single and nonseptated, laminotomy and myelotomy, but not resection of the cyst wall, are performed. The classical forms of syringomyelia are so very typical as to leave no doubt on MRI imaging. However, the astrocytoma cyst may be multiloculated. The treatment is the same.

Intramedullary cysts may form within astrocytomas, or extend cephalad and caudad at either pole of a solid intramedullary tumor. At times the cysts take on the appearance of links of sausages – as in syiringohydromyelia associated with meningomyelocele or the Chiari I malformation. At others, the cysts extend as a single cavity, more or less irregular in form. However, the wall of cystic tumors is always yellowish in tint, unless there has been recent bleeding.

With MRI one may generally know the length, form and content of the cyst, as well as the location and volume of the solid component of the tumor.

The surgery is directed to removing the solid component, draining the cyst, leaving the cyst wall intact, and closing the dura mater "watertight." Laminotomy should be performed over the site of the solid component, the dura opened as already described, the arachnoid opened the full length of exposed tissue and reflected on 6 × 0 or 7 × 0 retention sutures (which are used at the end of the procedure to close this transparent membrane).

The medulla spinalis is opened at its thinnest point, closest to the solid component of the tumor, preferably with a #15 blade, respecting as much as possible the integrity of the surface vessels and arteries. We prefer entering at the medial longitudinal fissure as soon as possible and then extending the opening superiorly and inferiorly with a microdissector, pressing it gently (with the effort to separate fibre tracts rather than cut them). Once in the cyst, inserting soaked fluffy cottons, and using them to extend the opening affords gentle separation.

Microdissectors are very effective in separating the tumor from both its adhesion to the parieties of the cavity and its border with normal spinal cord, the plane of

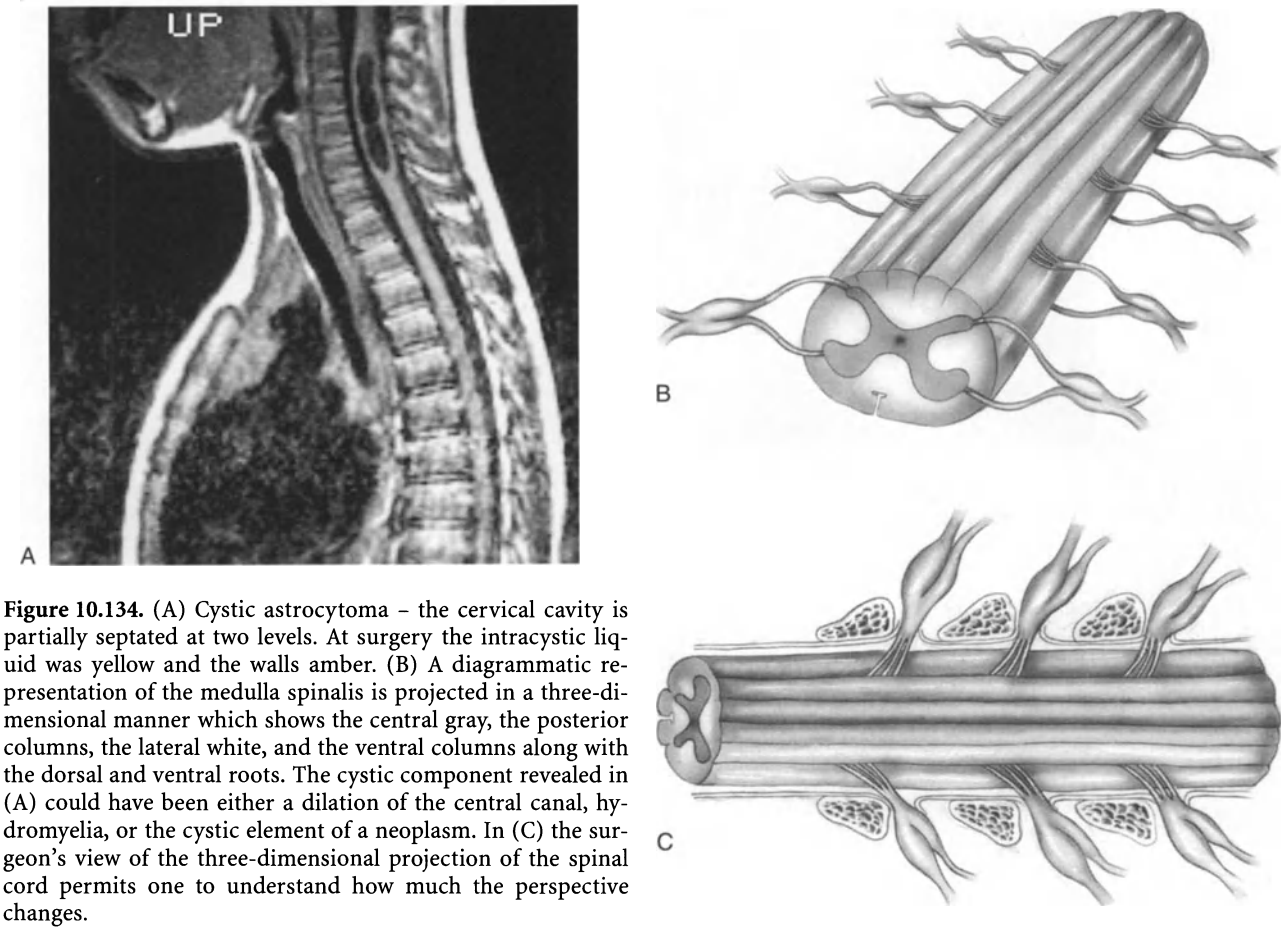


Figure 10.134. (A) Cystic astrocytoma – the cervical cavity is partially septated at two levels. At surgery the intracystic liquid was yellow and the walls amber. (B) A diagrammatic representation of the medulla spinalis is projected in a three-dimensional manner which shows the central gray, the posterior columns, the lateral white, and the ventral columns along with the dorsal and ventral roots. The cystic component revealed in (A) could have been either a dilation of the central canal, hydromyelia, or the cystic element of a neoplasm. In (C) the surgeon's view of the three-dimensional projection of the spinal cord permits one to understand how much the perspective changes.

cleavage. The tumor itself may be "brushed" away with the Cavitron, as it is separated from adjacent normal spinal cord. The Cavitron does not "see" blood vessels. The cyst wall's yellowish tint and the whiteness of adjacent normal tissue are generally readily seen with the operating microscope.

Irrigation with saline (kept at body temperature) precedes arachnoid and dural closure, and then repositioning of the laminar flap prior to muscle closure.

Solid Astrocytoma (Figs. 10.135–10.139)

These tumors, whether benign or malignant, invade diffusely the entire spinal cord. There is often a plane of demarcation between tumor and normal spinal cord. Resection, consequently, is possible through a myelotomy.

The solid astrocytoma may be malignant or benign, more or less highly vascularized, easily separable – plane of dissection – from normal spinal cord or diffusely invasive, extending tentacles from its center. If benign, it may have a succulent appearance because of microscopic cystic areas, but it may also be densely fibrotic. In either event it is very amenable to Cavitron or laser resection. It should not be necessary to use bipolar cautery to shrink these tumors or regular suction to aspirate them. If malignant, the astrocytoma of the spinal cord is a frightful adversary, most of all because complete resection is not possible and it may be vascular, edematous, not separable from "normal" tissue. Decompression is the single benefit a surgeon may offer a child with a malignant spinal cord astrocytoma, so he should limit himself in the extent of resection, performing a relaxing duraplasty closure.

For benign astrocytoma, the technique and extent of resection are very important, as are dural and lamina closure. The laminotomy technique and dural opening have already been described. One should keep the bony opening minimal (total resection has not been shown to be possible in more than 25% of patients, children and adults), for postoperative scoliolsis is a devastating reality. Lateralization of the tumor's growth may displace the median longitudinal fissue considerably from the midline, devascularizing the medullary surface, rendering selection of the myelotomy site...even random. If one cannot orient oneself, incising at the palest, most bulging point is a good rule of thumb, just as one does for cerebral tumors. Once in the tumor, the dissection,

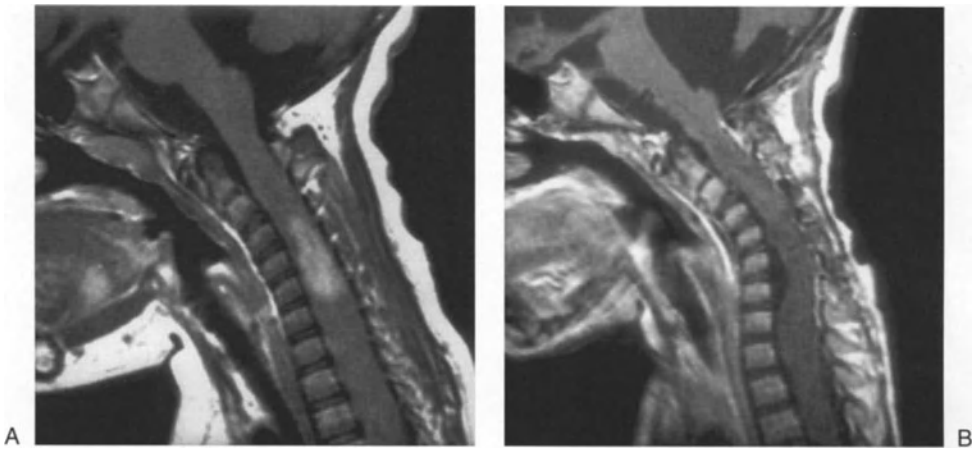


Figure 10.135. In (A) the solid astrocytoma (which we had considered to be an ependymoma) is well delimited; and, in (B) one sees an apparent total resection. However, there is more: the surgeon performed a laminectomy, and one sees the resultant gibbus. A laminotomy would have been preferable.

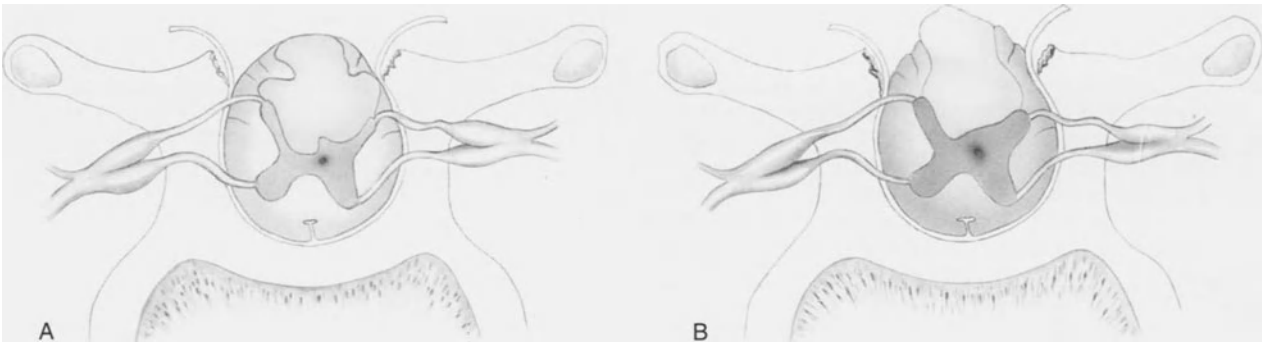


Figure 10.136. The astrocytoma of the posterior columns, the dorsal variety in our experience, expands irregularly the dorsal columns on either side of the medial longitudinal fasciculus (A) and flattens ventrally the central gray as it spreads the dorsal horns. The tumor does not invariably remain dorsal to the gray matter, for it may expand across the mid-coronal plane of the cord, or so flatten the gray matter and displace it to either side as to give one the impression it has done this. After a myelotomy has been performed (B), a portion of the astrocytoma may deliver itself through the opening, though this is not nearly so common as in ependymoma. As this happens, the gray matter gradually returns to its normal position if the tumor is small. A clear line of demarcation is not the rule in astrocytoma.

whether performed with Cavitron (preferable) or laser, should proceed very slowly. If microdissectors are used it may be possible to find a plane of cleavage but, even when this is identified, it is soon lost. Therefore, one proceeds with the "bit-by-bit" technique, performing more of an internal decompression than a tumor resection. It is not at all unusual to bivalve longitudinally the spinal cord.

There is no word-by-word manner to take one through the removal of a benign, solid spinal cord astrocytoma. However, there are a few maxims: perform the myelotomy at the most widened, palest part of the cord, and then proceed millimeter at a time to the tumor. Stay within neoplastic tissue throughout the dissection.

Ependymoma (Fig. 10.140)

The intramedullary ependymoma may occasionally be separated from the surrounding spinal cord. A midline myelotomy permits access to the centrally located ependymoma, which is vaporized (using laser). Because of the benign nature of the spinal ependymoma, one is encouraged to remove as much tumor as possible.

Very probably, the singlemost heterogeneously active CNS tumor with regard to biological aggressiveness is the ependymoma: *relatively benign in the cerebral hemisphere, relatively malignant along the lateral and midline ventricles, very benign in the spinal cord, and nonresectable in the cauda equina.*

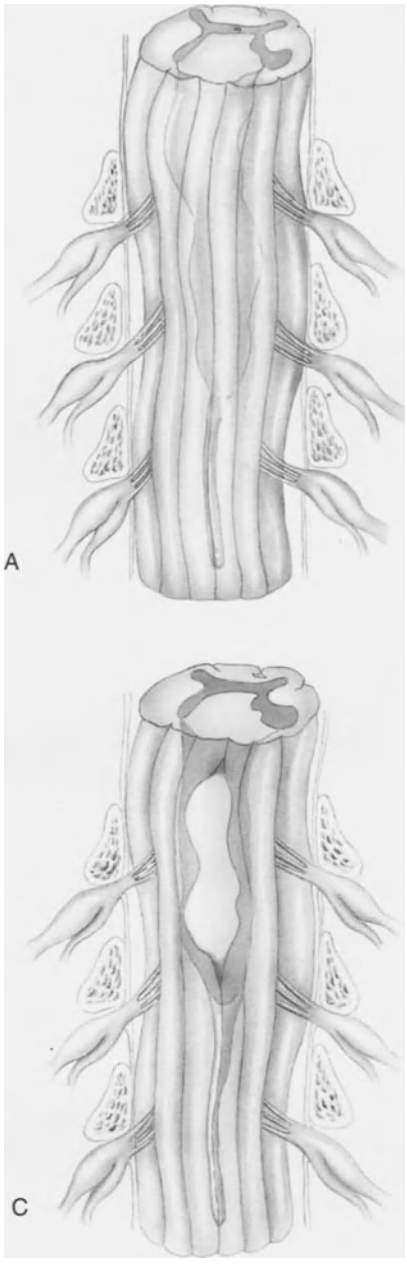


Figure 10.137. Ventral column astrocytomas are not nearly so common in our experience, maybe because before MRI we were not able to identify them. They distort the cord (A) as much as the dorsal or lateral column tumors, but if the surgeon approaches the tumor posteriorly he is not aware of these topographical changes. In (B) one has a visual explanation of why at surgery for even a relatively large intramedullary astrocytoma, the myelotomy, performed in the median longitudinal sulcus, may not bring the surgeon immediately upon the tumor: it is ventral in location. Exophytic components of medullary tumors (C), even astrocytomas, occur. In (D) the surgical approach was ventrolateral, and possible to plan only because of the horizontal MRI images.

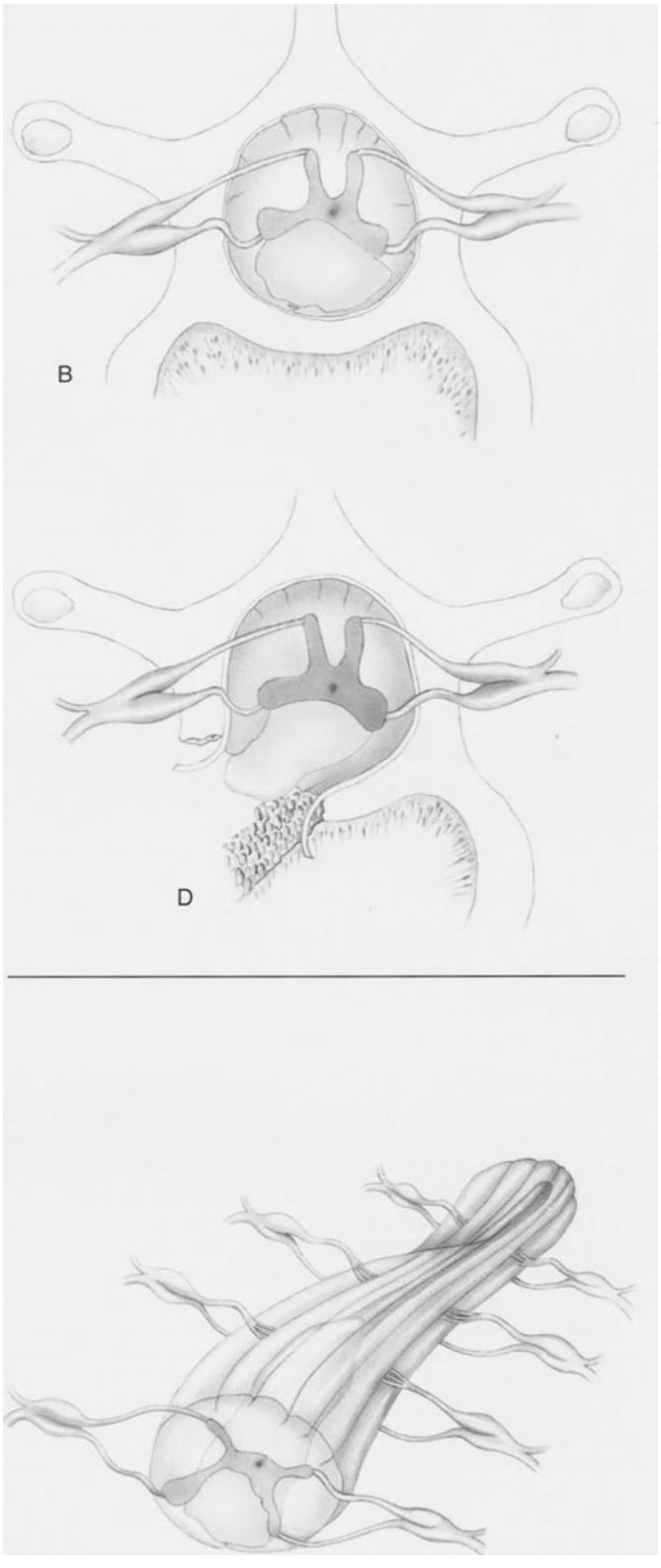


Figure 10.138. This is a schematic representation of the operative findings for a ventral astrocytoma which had been approached posteriorly: the dorsal aspect of the cord was twisted, turned upon itself, and expanded in a fusiform manner.

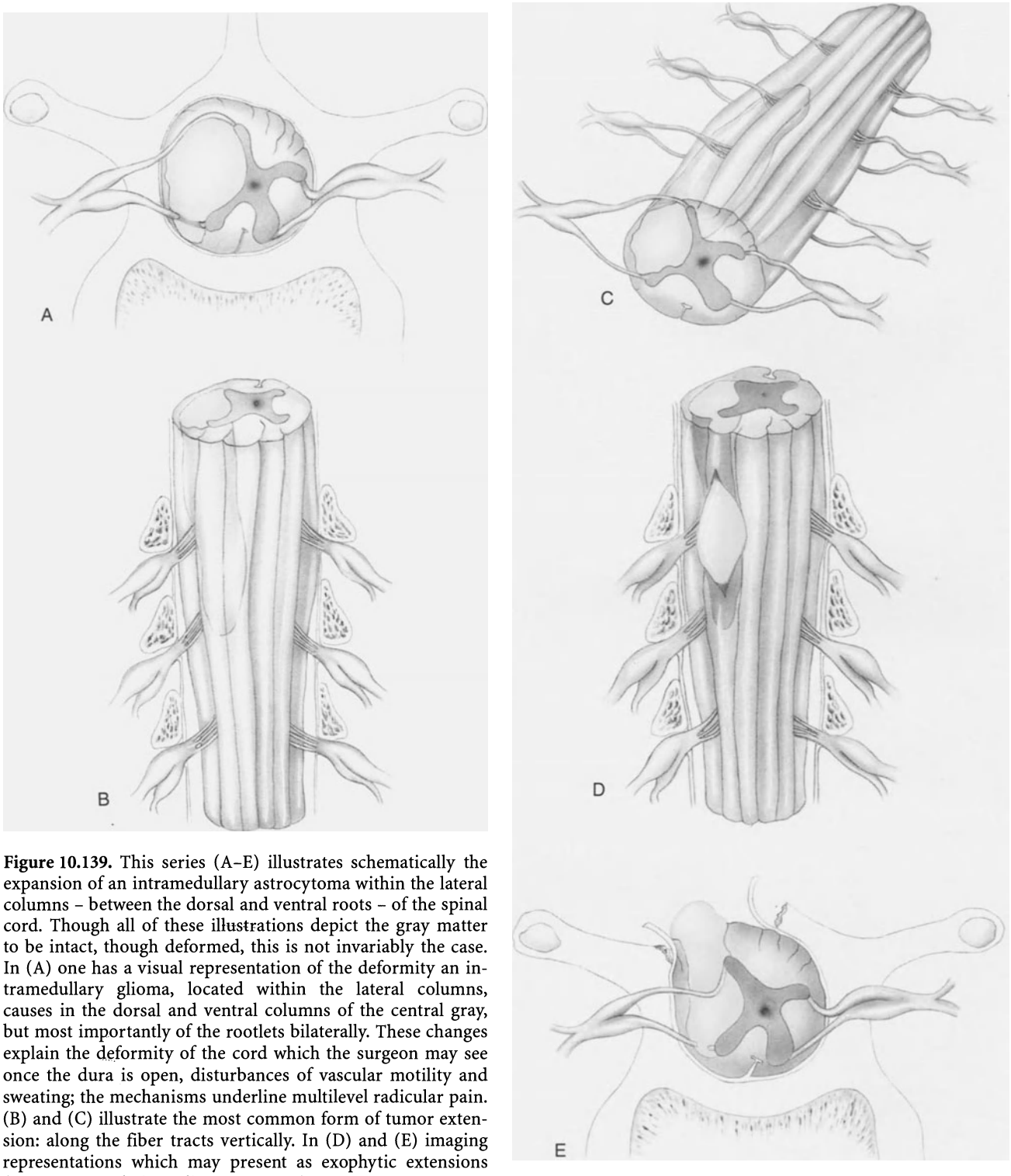


Figure 10.139. This series (A–E) illustrates schematically the expansion of an intramedullary astrocytoma within the lateral columns – between the dorsal and ventral roots – of the spinal cord. Though all of these illustrations depict the gray matter to be intact, though deformed, this is not invariably the case. In (A) one has a visual representation of the deformity an intramedullary glioma, located within the lateral columns, causes in the dorsal and ventral columns of the central gray, but most importantly of the rootlets bilaterally. These changes explain the deformity of the cord which the surgeon may see once the dura is open, disturbances of vascular motility and sweating; the mechanisms underline multilevel radicular pain. (B) and (C) illustrate the most common form of tumor extension: along the fiber tracts vertically. In (D) and (E) imaging representations which may present as exophytic extensions (most commonly in malignant tumors) or as tumor extrusions after the cord has been incised. It is emphasized that the surgeon should not attempt invariably to incise the cord along the median longitudinal fasciculus, but rather where he sees maximal fasciculus widening.

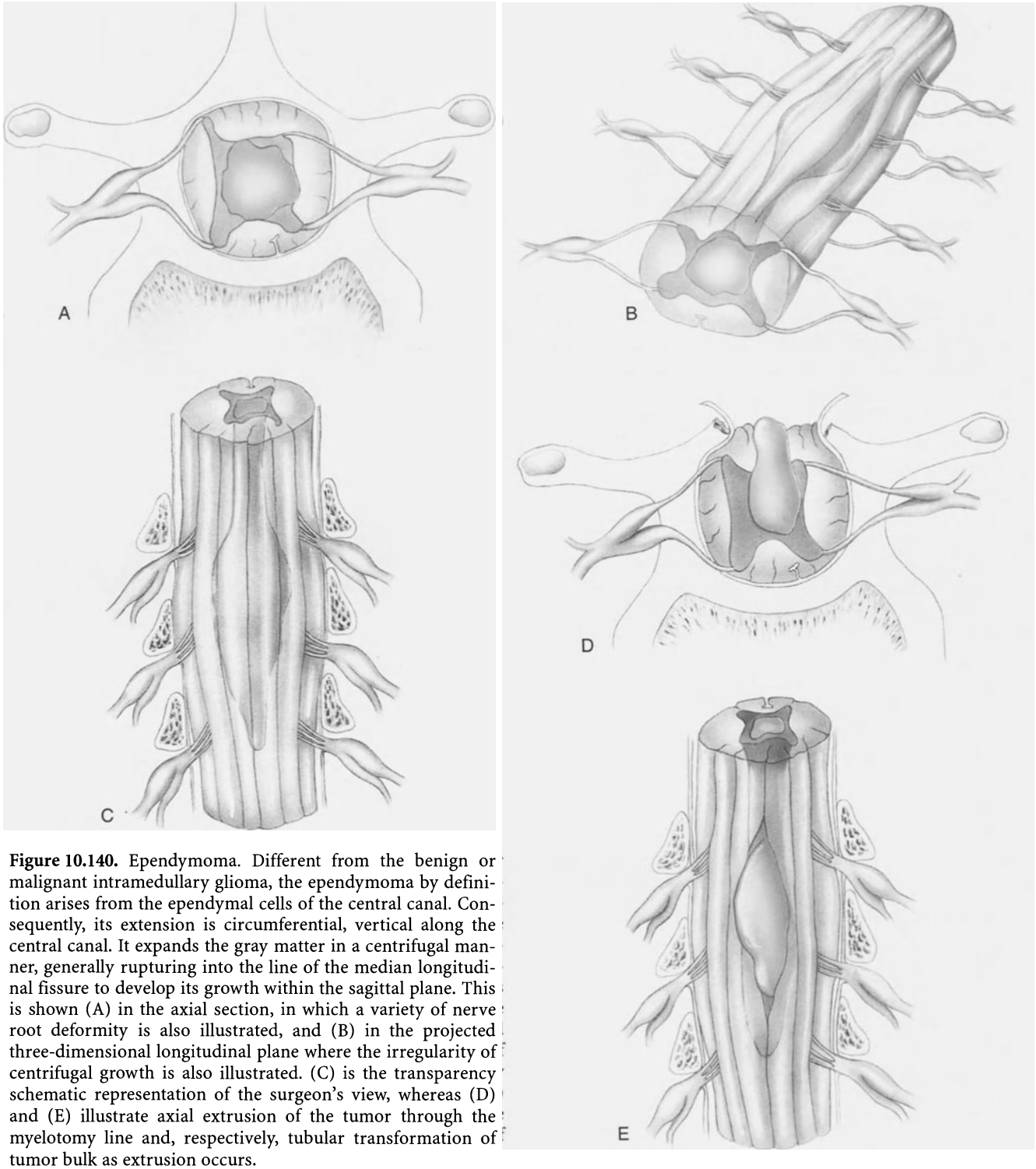


Figure 10.140. Ependymoma. Different from the benign or malignant intramedullary glioma, the ependymoma by definition arises from the ependymal cells of the central canal. Consequently, its extension is circumferential, vertical along the central canal. It expands the gray matter in a centrifugal manner, generally rupturing into the line of the median longitudinal fissure to develop its growth within the sagittal plane. This is shown (A) in the axial section, in which a variety of nerve root deformity is also illustrated, and (B) in the projected three-dimensional longitudinal plane where the irregularity of centrifugal growth is also illustrated. (C) is the transparency schematic representation of the surgeon's view, whereas (D) and (E) illustrate axial extrusion of the tumor through the myelotomy line and, respectively, tubular transformation of tumor bulk as extrusion occurs.

It is a curious neoplasm, indeed, with little or no correlation between histological appearance and biological activity. We must look further for cytological or histological parameters which permit us some degree of predictability. The spinal cord and cauda equina ependymomas put this stark reality into relief.

Ependymoma of the spinal cord in childhood is certainly not a common tumor. In fact, it is an unusual situation in which one pediatric, or general, neurosurgical center treats ten such tumors in 35 years – the practice lifetime of a neurosurgeon. Consequently, the database upon which clinical conclusions are structured is narrow. Nevertheless, we have not seen any significant differences between the adult and pediatric populations which would limit direct transfer of conclusions and tecniques – with the very significant exception of the laminotomy in children – from adults to children.

This tumor expands rather uniformly, along the longitudinal axis of the medulla spinalis, first within the central canal, in which it originates, and then the parenchyma surrounding it, which the ependymoma generally compresses and displaces. It rarely invades and destroys spinal cord nuclei or tracts. Consequently, the cord surrounding ependymoma is more or less uniformly expanded, fusiform with tapering superior and inferior extremities, so that the median longitudinal sulcus may be identified and incised. The myelotomy technique is the same as for astrocytoma. At less than three millimeters the tumor is encountered, generally immediately beneath the surface.

Once the tumor is exposed, the myelotomy is extended longitudinally, superiorly and inferiorly, slowly enough to allow the ependymoma to deliver itself partially from the intramedullary compartment without compressing the cord. The remainder of the resection is performed by working within the tumor.

Cauda Equina Ependymoma (Fig. 10.141)

When ependymoma involves the cauda equina, it layers all nerve roots as icing on a cake, rendering it almost impossible to remove tumor and preserve neural elements. Surgery should be limited to decompression. However, very occasionally the tumor is globular, attached to the film terminate...in which case resection is possible and not difficult.

Arteriovenous Malformations of the Spinal Cord

These are so rare in childhood that no attention is given to the technique for their removal in this text.

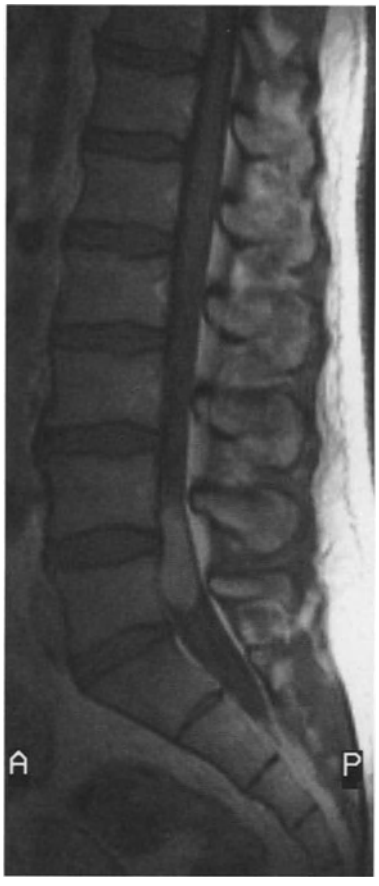


Figure 10.141. This T1 weighted post-gadolinium image reveals the filum terminale ependymoma extending from the center of L4 to the inferior surface of that of L5. At surgery, the lesion was pedunculated from the filum terminale, only moderately adherent to nerve root of the cauda equina. We erroneously diagnosed this as a neurinoma of a lumbar root since the patient presented with the clinical picture of lumbar radiculopathy. It is different from the cauda equina ependymoma, which classically expands around the nerve roots of the cauda equina coating them into one mass of ependymoma penetrated by lumbar and sacral roots. The cauda equina ependymoma by and large is not operable; the filum terminale ependymoma may be resected with relative ease.

Dermoid and Epidermoid Tumors

Since these are congenital tumors, they have been described in Chap. 14, "Congenital Anomalies."

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11 Vascular Disorders:
Surgical Approaches and Operative Technique

"Nature her selfe must be our adviser; the path she chalks must be our walk: for so while we confer with our own eirs, and take our rise from meaner things to higher, we shall at length be received into her Closet-secrets."

WILLIAM HARVEY, Anatomical Exertations Concerning the Generation of Living Creatures

The technique of femoral-cerebral catheterization in infants and children was described by us in 1980, reporting [1] a series of 1869 procedures. Clinical complications occurred in 12 of 678 or 1.8% of examinations while radiographic complications were discovered in 38 of 1869 or 1.1% of procedures. The results were compared to a similar retrospective study of a large series (2184 procedures) of direct puncture/retrograde brachial studies in a similar patient population at the same institution, in which there was a 2.6% clinical complication rate and a 4.3% radiographic complication rate. Those factors which predispose to complications, clinical or radiographic, are more pronounced in the younger age groups but affect the direct puncture/retrograde brachial technique to a greater degree. The lower morbidity, greater selectivity, and improved comfort make the femoral-cerebral route preferable for neuroangiographic studies in infants and children.

Introduction

The first cerebral angiogram in a child was reported by Moniz [2] in 1928, and the first series of percutaneous carotid angiograms in children by Torkildsen and Koppang [3] in 1950. Seldinger [4] described a percutaneous technique of catheter insertion into vessels in 1953, and in 1960 Gensini and Ecker [5] adapted the Seldinger technique to retrograde femoral catheterization of the cerebral arteries. As late as 1968, however, the method of femoral catheterization, in general, was not well accepted for use in infants and small children [6], with the exception of the pioneering work of Harwood-Nash [7], who adopted the routine use of catheter studies in 1967. Appreciation of the femoral catheter technique for cerebral angiography in children lagged behind its acceptance in the adult population because of assumed technical difficulties, necessary improvements in catheter technology, and the already low morbidity of the direct puncture approach in experienced hands [8, 9].

In recent years, several centers have reported experience with catheter angiography in the pediatric age group [10–13]. Obenchain et al. [11] were the first to analyze their group for complications of pediatric femoral-cerebral angiography, and their rate was 5% (in 100 patients). Certainly, the literature notes differences in angiographic morbidity between infants, children, and adults [11, 14], but there is a dearth of material available concerning the relative complication rates of the femoral catheter versus direct puncture/retrograde brachial techniques among infants and children [15].

Because of the low complication rate of direct puncture and retrograde injection studies [3, 9, 11, 14, 16–23] and because of the greater technical expertise necessary in femoral catheter studies [10, 12, 13, 15, 24], there has been reluctance on the part of many centers to adopt the latter route for routine neuroangiographic studies in children.

It is our observation that femoral cerebral angiography is a safe and valuable method to study and treat vascular disease in the pediatric age group. In addition to a lower morbidity and mortality, it assures a greater degree of patient comfort in terms of positioning, time of study, number of arterial punctures, and lack of irritative effects of contrast material. Further, the quality of imaging is greatly enhanced by the selective and superselective capability of the technique. Local anesthesia supported by adequate sedation are adequate for the performance of high-quality studies.

The above said and documented, one must now recognize that computed tomography angiography (CTA) and magnetic resonance angiography (MRA), both noninvasive imaging technologies, are a reality which are becoming more precise every year. They are now at a level of reliability such as to supplant catheter angio-

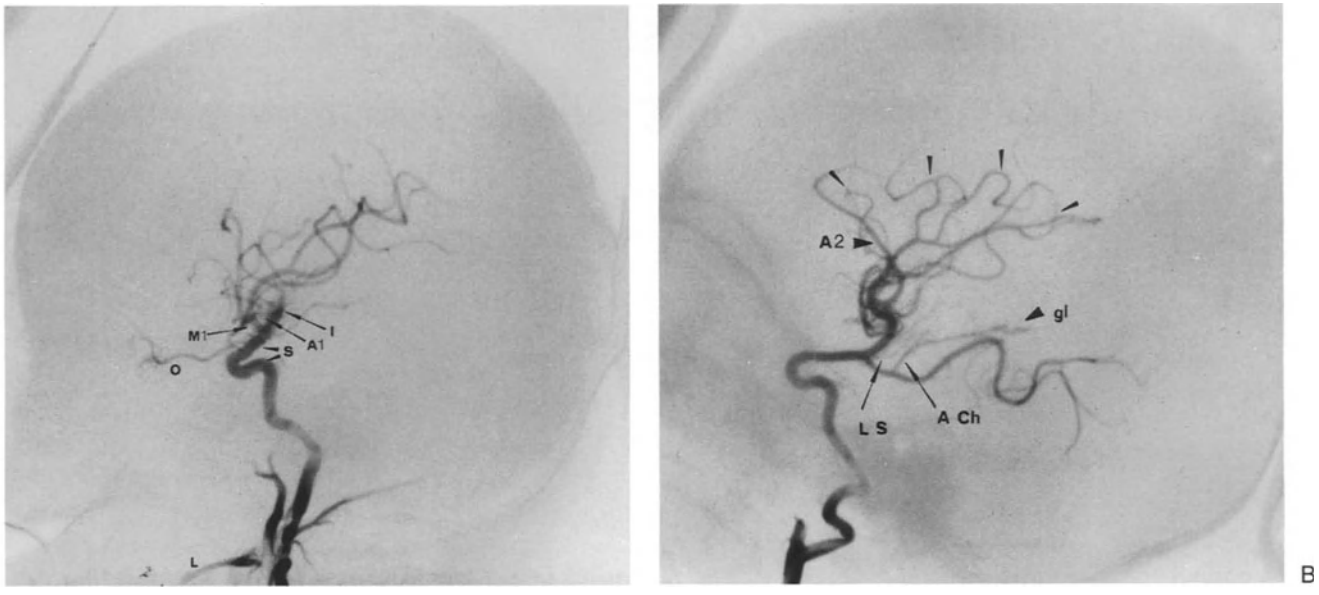


Figure 11.1. (A) The siphon (*S*) is tight in appearance; one should note the high and posteriorly located bifurcation of the internal carotid artery *(1)*, with the A-I segment of the anterior cerebral artery coursing directly anteriorly and the M-1 segment of the middle cerebral artery looping inferiorly and entering the sylvian fissure. The redundancy of these vessels is obvious. The ophthalmic *(0)* takes its origin from the internal carotid artery immediately beneath the origin of the optic nerve, and the bifurcation of the internal carotid allows the reader to evaluate the size of the supraclinoid cistern in which the internal carotid is located. Of particular interest is the size of the lingual artery *(L)* in the newborn. Indeed, judging from cerebral angiography, the majority of blood flow to the head in the newborn is to the tongue and thalamus. The sylvian complex stands out clearly in this illustration, since there is no filling of the anterior or posterior cerebral systems. (B) The lenticulostriate *(LS)* vessels stand out well between the intracisternal portion of the internal carotid artery and the anomalous origin of the anterior choroidal artery *(A Ch)*, which comes from the midportion of the posterior communicating artery. The early blush of the glomus (*gl*) of the choroid plexus within the trigone is well seen. The A-2 portion of the anterior cerebral artery is located posterior to the middle cerebral trunk, a not uncommon observation in the newborn. The sylvian vessels are looping gently back onto themselves as they exit from the surface of the insular cortex through the sulci, indicating the marginal sulcus (*arrowheads*).

graphy in all but a *very limited* number of pathological entities.

Saccular Aneurysms

Saccular aneurysms in juveniles and adolescents are dealt with technically in the same way as in adults, though the surgeon may be confident that he will not encounter arteriosclerosis or arterosclerotic plaques. *Giant aneurysms, irrespective of their location, must be resected "in toto" because of their tendency to act as tumors.* Posterior inferior cerebellar artery (PICA) aneurysms are more common, and generally larger in size. The standard techniques for exposing the suprachiasmatic, parasellar, interpeduncular, and lateral medullary regions for access to anterior or posterior circle aneurysms, or aneurysms of the vertebral basilar junction, are used.

The newborn, infant, and toddler present remarkably different anatomical and technical implications. Recognizing the fact that one is speaking of a very rare clinical entity when discussing saccular aneurysms in these age categories, it is safe to state that, by and large, they occur in three locations: bifurcation of the internal carotid artery, trifurcation of the middle cerebral artery, and origin of PICA at the vertebral artery.

Specific vascular anatomical characteristics of newborns and infants are as follows: The anterior communicating artery does not exist as a radiologically identifiable anatomical structure, the internal carotid artery is large and continues directly into the middle cerebral artery, and the PICA is located entirely within the posterior fossa.

The flow of blood from the internal carotid is almost directly into the middle cerebral artery, which appears to be the direct continuation of the internal carotid, coursing superolaterally rather than horizontally (Fig. 11.1A). Similarly, the branches of the middle cerebral artery are linear extensions of this vessel, at the limen of the insula (Fig. 11.1B). Hence, one may understand why aneurysms are more common at the bifurcation of the internal carotid artery and trifurcation of the middle cerebral artery.

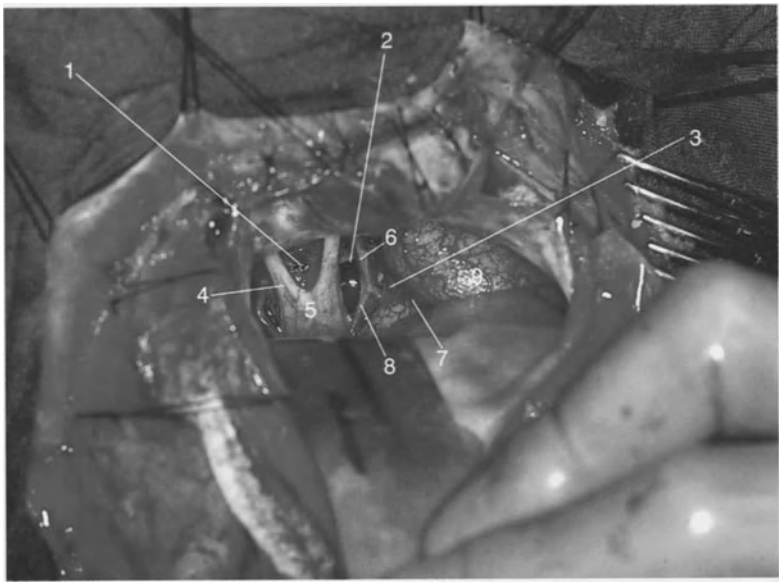


Figure 11.2. This is a photograph of a right medial frontal craniotomy in an infant, illustrating the enormity of basal cisterns and relative anatomy of optic pathways and carotid arteries. The prechiasmatic *(1),* interopticocarotid *(2),* and sylvian *(3)* cisterns have been opened, permitting complete exposure of the optic nerves (*4*) and chiasm (*5*), the internal carotid (*6*), middle cerebral (*7*), and anterior cerebral *(8)* arteries. The temporal lobe (*9*) is immediately lateral to the internal carotid artery. One may, from this photograph, appreciate how the relative stage of undevelopment of the frontal lobes and the very large basal cisterns render easy the exposure of the entirety of the anterior circle in infancy.

The shallow anterior fossa, as yet undeveloped frontal lobes, minimally developed temporal lobes, and extraordinarily large basal cisterns render access to the internal carotid and middle cerebral arteries easy and rapid (Fig. 11.2). The very large cisterna magna and relatively large lateral medullary cisterns, along with the as yet poorly developed cerebellar hemispheres, render access to the PICA and the vertebral artery (Fig. 11.3) equally easy and rapid! Consequently, aneurysms of the newborn, infant, and toddler may be approached surgically through one of two craniotomies: (1) a unilateral frontal bone flap for any anterior circle or middle cerebral aneurysm and (2) an inferior cerebellar triangle craniotomy for the giant aneurysms that occur at the origin of PICA.

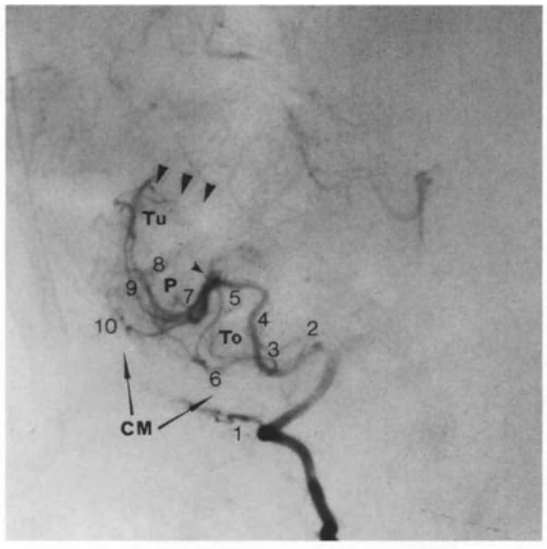
Internal Carotid Bifurcation Aneurysms

With the child in the supine position, a lateral frontal craniotomy is performed, and the dura is opened in a double trapdoor fashion. The frontal lobe is retracted superomedially, exposing the olfactory nerve anteromedially and the sylvian fissure posterolaterally. This permits the surgeon to follow the lesser wing of the sphenoid from the pterion to the anterior clinoid process. After the olfactory nerve has been coagulated and sectioned, immediately posterior to the olfactory bulb (to avoid traction, which could pull the olfactory rootlets from the cribriform plate and result in cerebrospinal fluid rhinorrhea), the optic nerve is identified within its recess at the base of the anterior clinoid. The arachnoid from around the optic nerve is opened, and the optic nerve is separated from the internal carotid artery. It is then sectioned longitudinally along the superior surface of the internal carotid artery, working from inferomedial to posterolateral, and then passing over the anterior surface of the middle cerebral artery. The dissection proceeds to the limen of the insula, opening the sylvian fissure to avoid the arachnoid exerting traction on the aneurysm.

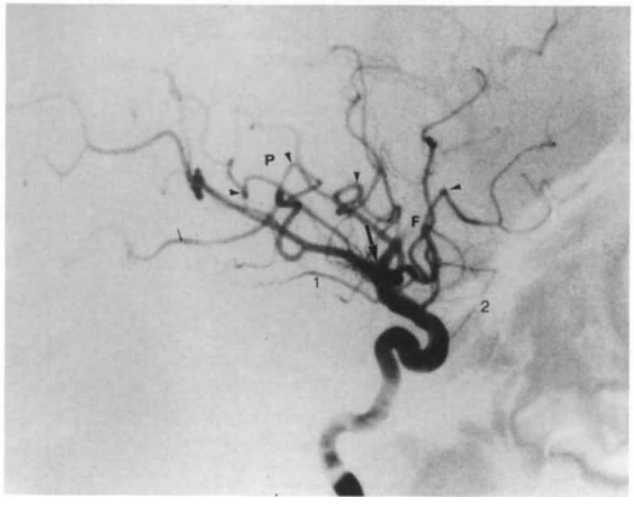
The lateral surface of the homolateral optic nerve is then separated from the medial surface of the internal carotid artery. At this time, the olfactory nerve should be identified in the olfactory sulcus along the base of the frontal lobe, and followed posteriorly to the olfactory trigone. This is the only safe and sure way of identifying the bifurcation of the internal carotid artery in this age category because of the already described apparent continuity of internal carotid and middle cerebral arteries. No effort is made to dissect out the fundus of the aneurysm. Rather, one should simply identify the neck, separate any perforating branches that may be adherent to it, and then apply the smallest clip possible. There are no plaques within these aneurysms, so the surgeon need not fear tearing the internal carotid artery or not attaining an immediate and complete occlusion of the aneurysm's neck.

Aneurysms of the Trifurcation of the Middle Cerebral Artery (Figs. 11.4, 11.5)

These aneurysms are approached in the same manner as internal carotid bifurcation aneurysms. The dissection is also the same up to and including separating the arachnoid from the middle cerebral artery and separating the optic nerve from the internal carotid artery. The sylvian fissure is opened from medial to lateral, quite an easy undertaking because of its enormous size and the thinness of the arachnoid. This separates the temporal and frontal lobes from one another, exposing the

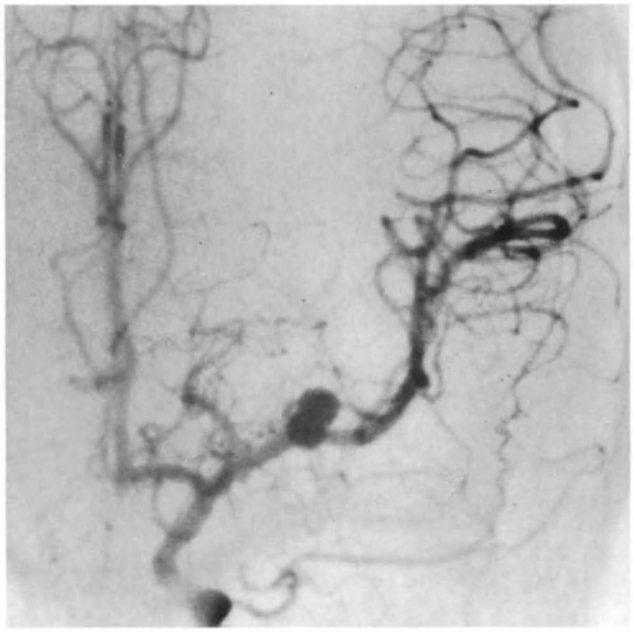


A



B

◄ **Figure 11.3.** (A) Muscular branches *(I)* are quite prominent and establish anastomotic routes between the vertebral and occipital arteries. The posterior inferior cerebellar artery (PICA) most often originates from the vertebral and courses around the medulla oblongata, where it may be divided into ventral *(2)*, lateral (*3*), and dorsal or retromedullary (*4*) segments. The retromedullary segment follows a superomedial course along the inferior rim of the IV ventricle to the obex and then, within the vallecula, courses posteriorly as the supratonsillar segment (*5*) to the posterior tonsillar sulcus where the choroidal point (*small arrowhead*) may be seen, as may the prominent choroidal blush. The apex of the tonsil (*6*) rests within the cisterna magna. The retrotonsillar segment (*7*) curves inferiorly around the undersurface of the pyramis *(P)*, at which point the vermian (*9*) and hemispherical *(10)* branches become identifiable. The artery of the superior pyramis *(8)* separates the tuber (*tu*) from the pyramis *(P)*. The great horizontal fissure (*large arrowheads*) separates the tuber below from the folium above, and is the line of demarcation between vascular supply to the inferior portion of the cerebellum by the posterior inferior cerebellar artery and to the superior portion of the cerebellum by the superior cerebellar arteries. The tonsil (*to*) may be identified clearly as may the cisterna magna *(CM)*. (B) The anterior choroidal artery *(1)* is quite large, as are the perforating vessels going to the basal ganglia and thalami. Note the prominence of the medial anterior inferior temporal artery *(2)*, which courses within the sylvian fissure and over the medial surface of the temporal lobe to the corpus amygdaloideum. This vessel is the route of anastomosis for collateral flow between the anterior and middle cerebral systems, since it may anastomose with Heubner's artery with which it occasionally establishes branches. The posterior bifurcation of the internal carotid artery (*arrow*) may be well appreciated as may the distribution of the primary sylvian vessels over the insular cortex, curving outward at the circular sulcus of the insula (*arrowheads*) and running around the frontal (*F*) and parietal *(P)* operculae to gain access to the surface of the hemisphere.



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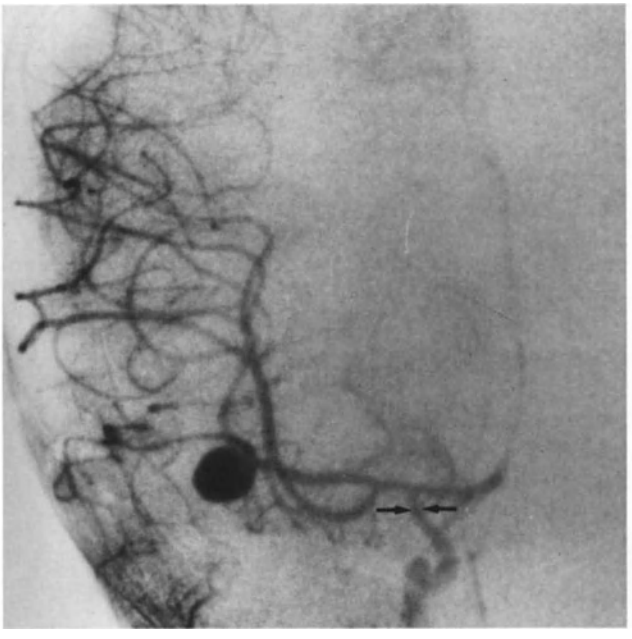

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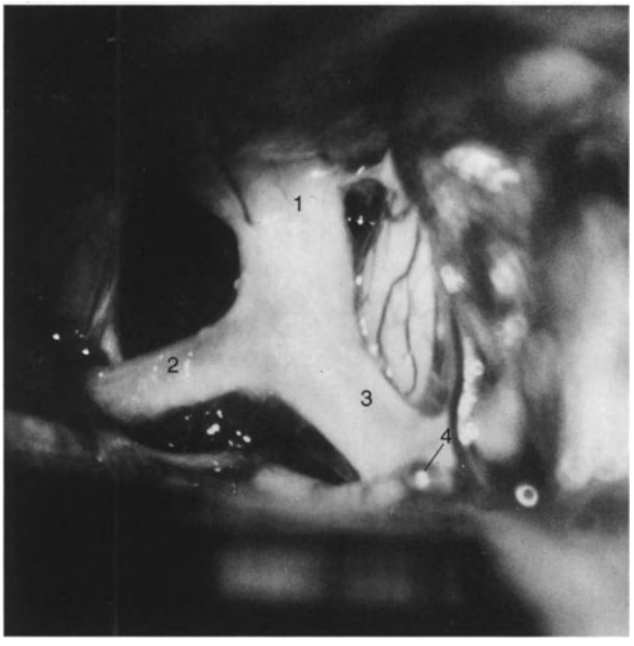


Figure 11.5. The internal carotid artery *(1)*, anterior cerebral artery *(2)*, middle cerebral artery (3), and aneurysm (*4*) at the trifurcation of the middle cerebral artery within the sylvian fissure.

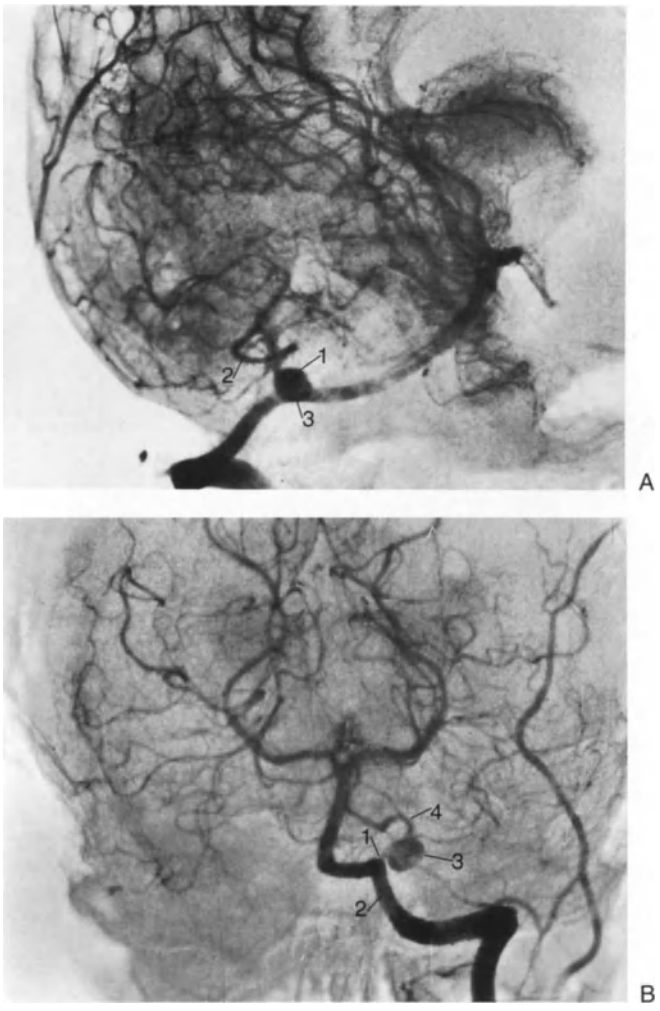


Figure 11.6. (A) The aneurysm *(1)* appears to be located at the origin of the posterior inferior cerebellar artery (PICA) *(2)* from the vertebral artery (3). (B) The AP projection, however, shows clearly that the PICA *(1)* originates from the vertebral *(2)*, and that the aneurysm (*3*) is of the PICA between its origin and lateral medullary segment (*4*).

limen of the insula and the triangular, olfactory, and temporal operculae. It may be necessary to transect the deep middle cerebral vein as it passes into the sphenoparietal sinus, rather than run the risk of tearing it from the sinus as the temporal lobe falls inferiorly. The very large insular branches of the middle cerebral artery are identified, one at a time, at their points of origin, and then followed over the short gyri of the insula as far posteriorly as the longitudinal sulcus. This permits the separation of these vessels from the body and neck of the middle cerebral trifurcation aneurysm, prior to application of an appropriately sized clip.

Figure 11.4. (A) This trifurcation aneurysm is in no way different from those observed in adults. (B) Another trifurcation aneurysm in a different child, complicated by spasm of the terminal portion of the internal carotid artery (*opposing arrowheads*).

Posterior Inferior Cerebellar Artery (PICA) Aneurysms (Figs. 11.6–11.8)

With the child in the sitting position an inferior cerebellar triangle craniotomy is performed, and the dura is opened, as illustrated in Fig. 5.12. If the PICA aneurysm is, as so often occurs, an aneurysmal dilation of the vertebral artery, extending from the foramen transversarium of the atlas to the region of the origin of PICA or the basilar artery, the arch of C-1 should be removed as far laterally as the posterior strut of the foramen transversarium. Because of the smallness of size and incomplete ossification of C-1, it is best to use a very delicate, sharp-tipped, rongeur to take away the bony spicules, taking care to remain within the periosteum. This avoids damaging the vertebral artery or rupturing the aneurysm.

At this time, one may evaluate the anatomy of the region and the extent of the aneurysm, in order to determine whether the craniotomy should be extended farther laterally, superior to the occipital condyle, or

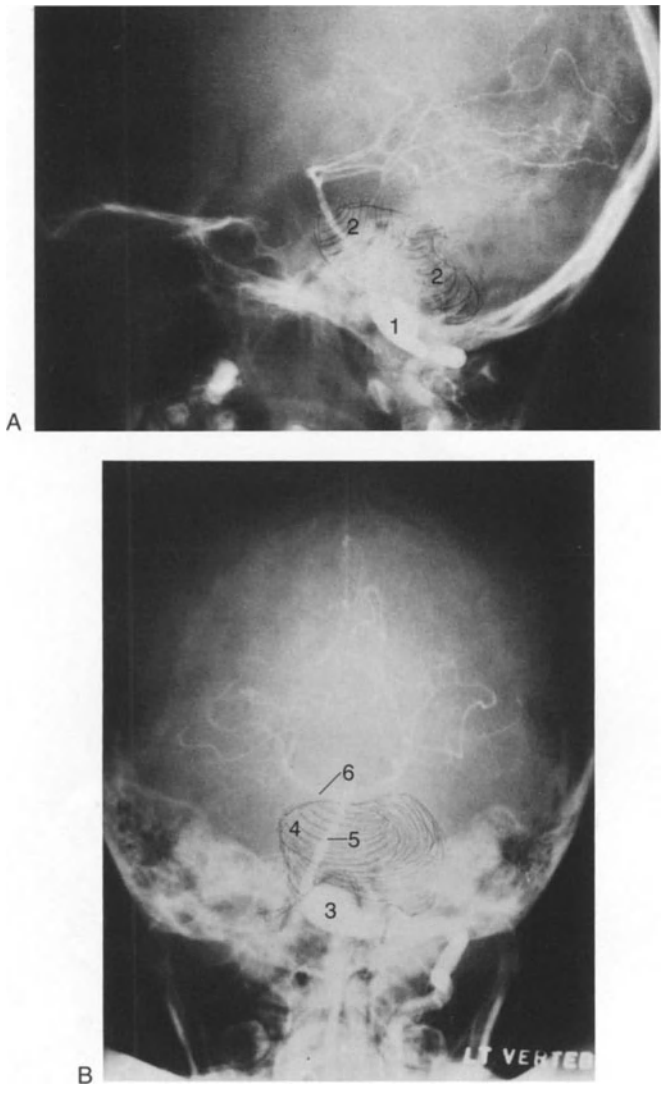


Figure 11.7. Posterior inferior cerebellar artery (PICA) aneurysms are well known to develop into giant aneuryms in infancy. Why this is so remains doubtful. Because of the giant size, and the presence of clot laminated in various stages of aging, one may invariably consider PICA aneuryms to be post-traumatic in the newborn, infant, and child, but variably so in the juvenile or adolescent. This is an angiogram of an enormous false aneurysm developing at the origin of the PICA from the vertebral artery, following a head injury. The vertebral artery *(1)* is ectatic, and then expands into an aneurysm. (A) A lateral projection, the aneurysm *(2)* has been drawn in immediately above the ectatic portion of the vertebral. (B) The half-axial projection; one may appreciate the ectasia of the vertebral artery, the base of the aneurysm (*3*), the mass effect of the false aneurysm (*4*), and the displaced basilar (*5*) and posterior cerebral (*6*) arteries.

whether the exposure is adequate. The enormous size of the cisterna magna and the lateral medullary cistern at this age generally render such lateral extension of the craniotomy excessive. However, since PICA aneurysms in the newborn and infant, as well as the toddler, may measure 3 × 2 × 3 cm in volume, it is best to evaluate the exposure before proceeding.

The vertebral artery is identified at its exit from the foramen transversarium and at its entrance into the subarachnoid space. One should take care not to attempt to dissect his way around the vertebral artery at the level of the foramen transversarium, since this could damage the vessel. It is impossible to control bleeding when the vertebral artery is torn at this point. Quite the converse is true when the vertebral artery is isolated within the cisterna magna and lateral medullary cistern. One may choose at this time to apply a temporary clip to the vertebral artery in this location, though proper *exposure* affords adequate protection in the event of damage.

►

Figure 11.8. These photographs were taken during the repair of the aneurysm illustrated in Fig. 11.7. (A) The arch of C-l has been exposed *(1),* allowing one to identify a fracture line *(2)* at the region of the foramen transversarium and (*3*) just lateral to the spine Cl. The aneurysm (*4*) bulges just above the arch of C-l. (B) The arch of C-l has been resected, and the dura sewn to the right *(1)* and to the left *(2),* exposing the posterior aspect of the enormous false aneurysm (*3*). (C) The tonsil of the left cerebellar hemisphere *(1)* was resected from the dome of the aneurysm, the vertebral artery occluded with hemoclips at its exit from the foramen transversarium of Cl, and the false aneurysm resected. The funnellike entrance of the false aneurysm of the vertebral and posterior inferior cerebellar artery (PICA) into the basilar artery was so large that it was impossible to apply any of the commercially available aneurysm clips. Consequently, a bulldog clamp *(2)* was applied, without occluding either the right vertebral or basilar arteries. (D) Operative photograph of another child, with a PICA aneurysm. When PICA aneurysms involve the PICA itself rather than a point of its origin from the vertebral artery, it is much preferable to resect the tonsil of the cerebellar hemisphere *(1).* Therefore, it is not necessary to elevate the tonsil and cerebellar hemisphere, something which would put stretch on the arachnoid and risk rupture of the aneurysm. This resection allows a direct view of the lateral surface of the medulla oblongata *(2),* and full exposure of the aneurysm (*3*). (E) After the tonsil has been resected, one need only place a self-retaining retractor *(1)* along the inferior surface of the cerebellar hemisphere, to hold it in place. This permits direct visualization of the aneurysm *(2),* the distal segment of the PICA coming out of its fundus (*3*), the IX and X cranial nerves crossing the meridian of the aneurysm (*4*), and the proximal portion of the PICA entering the aneurysm (*5*). (F) After the IX and X cranial nerves have been dissected from the surface of the aneurysm *(1)* and the XII cranial nerve *(2)* slid inferiorly, one may see the VII and VIII cranial nerves (*3*).

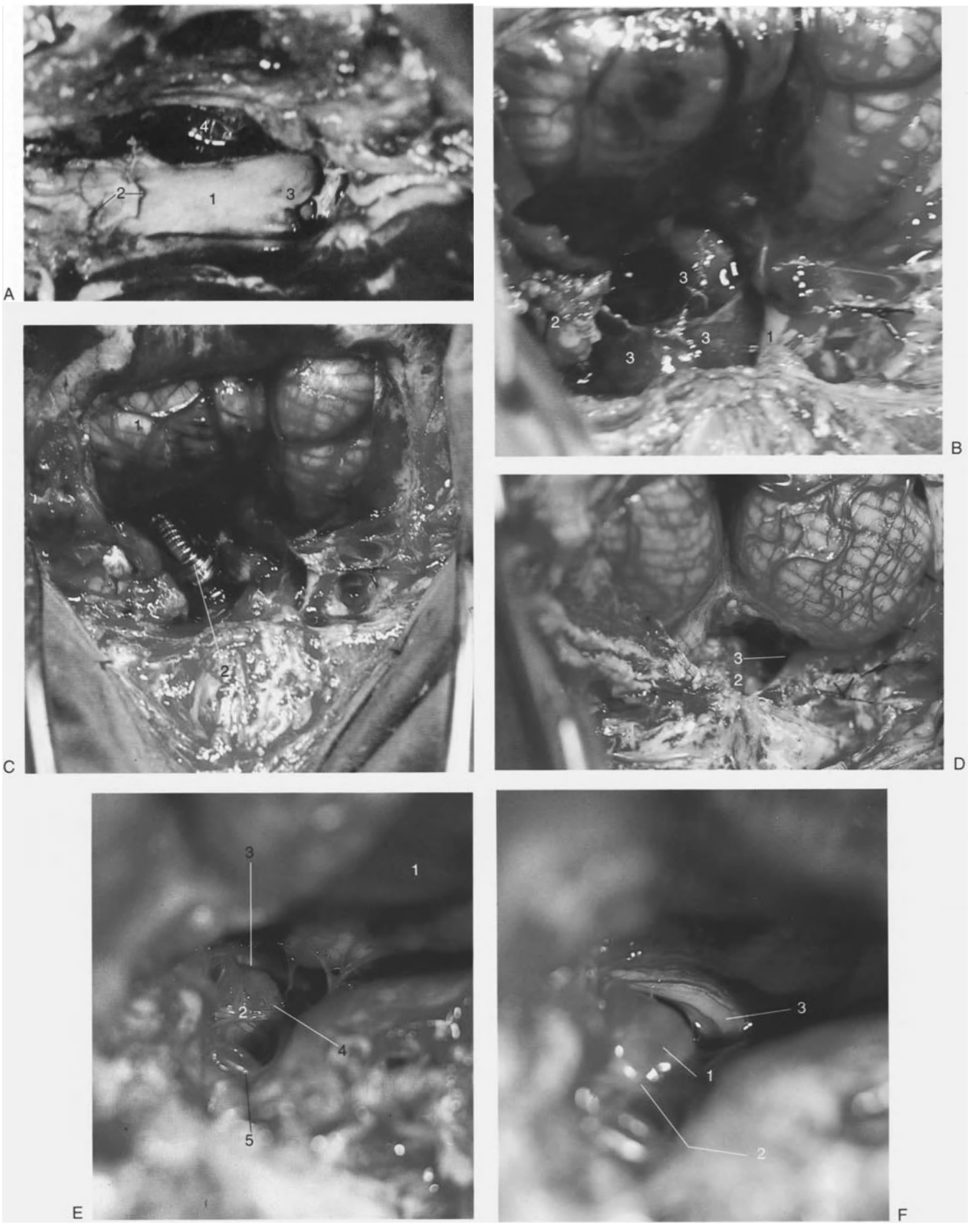


Figure 11.8. Legend see p. 360.

The tonsil should be resected! If it is elevated, the arachnoidal stretching may tear the aneurysm and the traction permanently damage the IX, X, XI, and XII cranial nerves. *Also, elevating the tonsil and cerebellar hemisphere may force these very large aneurysms into the lateral medullary surface, causing cardiac arrest.* The spinal accessory portion of the XI cranial nerve should now be inspected, and transected, but not dissected from the surface of the aneurysm. This allows the aneurysm to bulge into the operative field, over the posterior and inferior aspects of the olive. If the IX and X cranial nerves are excessively stretched over the posterior aspect of a giant aneurysm, leaving the surgeon to conclude that it will not be possible to deliver it without compressing the lateral surfaces of the medulla oblongata and pons, rootlets of one or the other – not both – should be sectioned with microscissors before proceeding to deliver the aneurysm and identify its neck. *The technique of dissection should not be such as to push the aneurysm in the direction of the medulla oblongata.*

If the aneurysm is small, it may be separated from the olive so as to view that portion of the vertebral artery located between aneurysm neck and the basilar artery. If its size is such as to preclude doing this safely, a temporary clip should be placed on the vertebral artery proximal to the origin of the aneurysm. One then opens into the aneurysm so as to collapse it and expose the most distal portion of the vertebral artery. The placement of a clip along the neck of the aneurysm, or trapping the aneurysm between two clips, one proximal and one distal, are decisions that must be made at the time of surgery. PICA aneurysms may be of the PICA itself, with the main trunk entering into and exiting from the aneurysm. In such instances, if it is not possible to clip the neck, the aneurysms must be trapped with two clips. An occipital artery/PICA bypass may be performed after the aneurysm is removed. Presently, stents are available, but we have had no experience with their use.

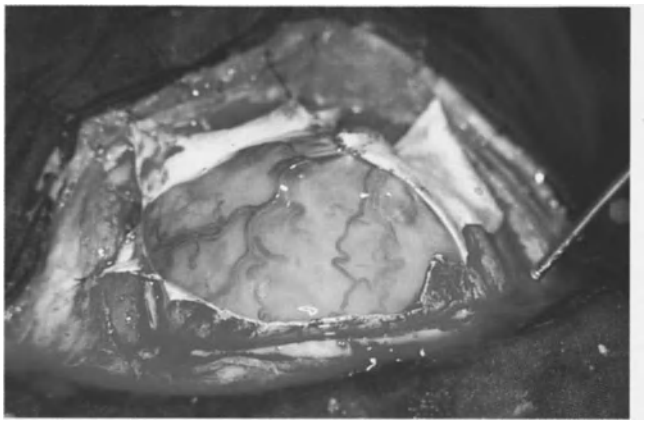


Figure 11.9. Opalescence of the arachnoid, which one almost invariably observes in children with arteriovenous malformations. Whether this is secondary to the shunting of arterial blood into the venous system, the increase in intracranial pressure, and repeated microscopic subarachnoid bleeds is purely speculative.

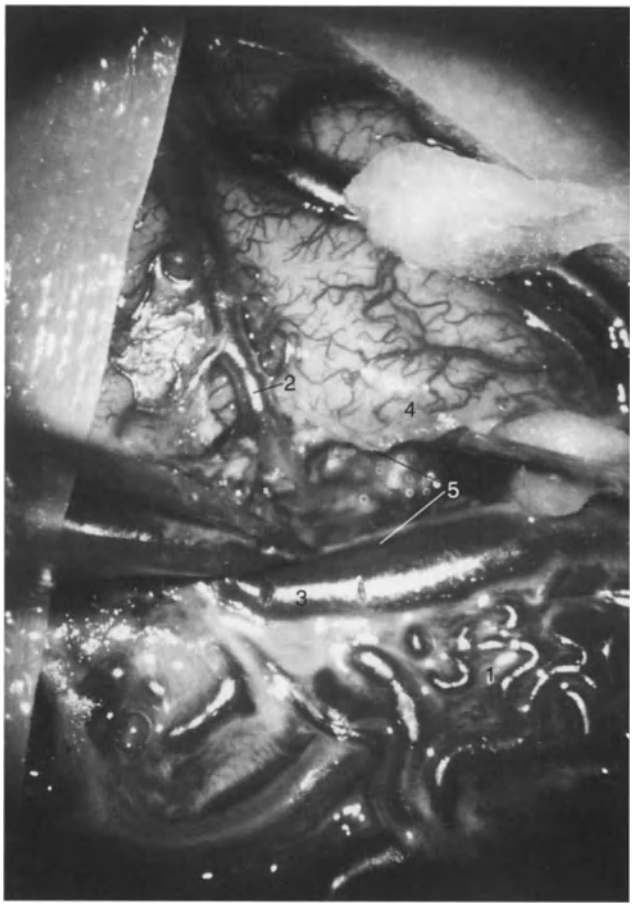


Figure 11.10. The ideal technique for resecting arteriovenous malformation consists of separating the malformation – racemose *(1);* efferent *(2);* afferent (3) – components from the surrounding brain (*4*) by performing a cerebrotomy along the border between normal brain and vascular anomaly (*5*).

Vascular Malformations (Figs. 11.9–11.12)

Arteriovenous malformations, with the exception of fistulae involving the galenic system, are almost nonexistent in children less than 2 years of age. Vascular malformations may be venous or arteriovenous. They may be transcranial, dural, leptomeningeal, or limited entirely to the extracranial area. The leptomeningeal malformations, in turn, may be meningopial or choroidal. Therefore, we may classify craniocerebral vascular (arterial and/or venous) pathology anatomically as scalp, transcranial venovenous or arteriovenous, dural, parenchymal, leptomeningeal, and choroidal arteriove-

nous, *with all except the venovenous consisting of dynamic, high-pressure, fistulae.*

The transcranial venovenous variety offers no threat of rupture, does not usually result in progressive neurological deficits or seizures, and may be treated surgically without exposure of the cerebral cortex. They are potentially dangerous in the event of scalp lacerations, but only to the extent that they may form the anatomical basis for air emboli, at most a very theoretical chain of events. They are, however, unsightly, and often frighten the parents.

Dural arteriovenous fistulae may cause hydrocephalus and high output heart failure, just as those involving the galenic system; the parenchymal, leptomeningeal, and choroidal may hemorrhage. Chronic arachnoiditis is a long-term complication, which appears morphologically as thickening or opalescence. Macrocephaly and intellectual or neurologic deficits may result from any of the vascular abnormalities other than those limited to the scalp.

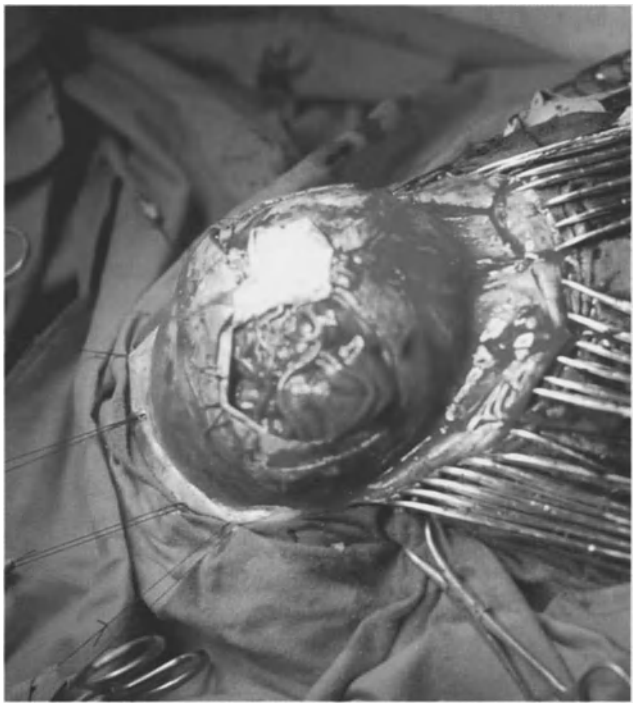
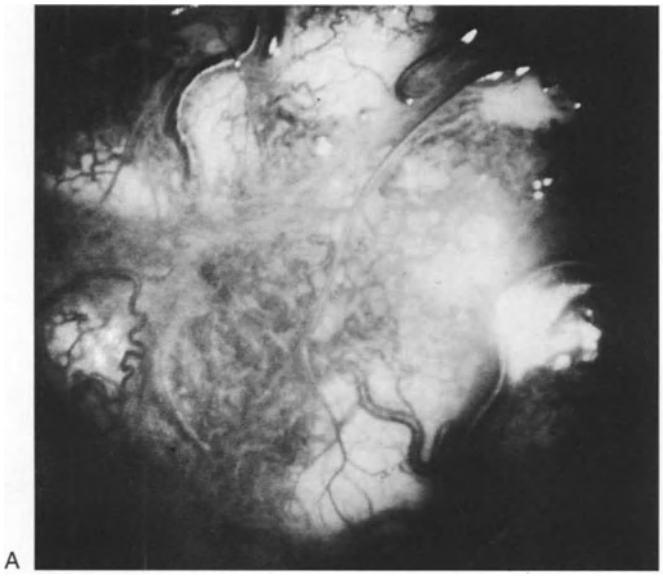


Figure 11.11. This is the appearance of the cerebral cortex in a child with an arteriovenous malformation of the galenic system. Note the granular appearance of the arachnoid, the "bag of worms" appearance of the cortical draining veins, and the arterialization of some cortical vessels.



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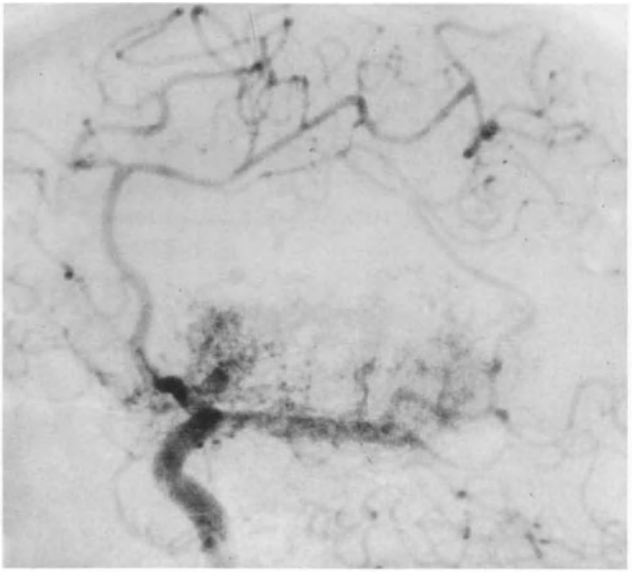

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Figure 11.12. (A) Cerebral cortex of a child with an arteriovenous malformation of the galenic system, illustrating the *Moya-Moya* effect within the cortical vessels. This becomes obvious only after the child has reached 5 or 10 years of age, when major draining sinuses such as the transverse, jugular, sigmoid, or cavernous have become obstructed from intraluminal thrombosis. Collateralization of flow increases, with retrograde flow (venous to arterial!) developing first, and then *Moya-Moya* appearance on the surface. (B) It is not common for arteriovenous malformations of the galenic system to have a predominantly racemose component, nourished by perforating branches of the anterior, middle, and posterior cerebral system.

Transcranial Venovenous Shunts

In order to treat these surgically one must identify the precise point in the skull where there is a macroscopic, diploic, venous communication. Very often, these are located along the superior sagittal or transverse sinuses, although they may also occur anywhere over the calvarium. The subgaleal and intradermal portion of the venovenous shunt presents clinically as a serpiginous, racemous malformation, which may be obliterated by digital scalp compression, and which fills in a retrograde fashion. One may not rely on angiography to illustrate all shunting sites. Rather, clinical inspection of the scalp is essential. MRA, and more recently, CT reconstruct angiography are very effective, noninvasive imaging studies.

Surgical treatment consists of reflection of a scalp flap, exposure of the cranium and identification of the transcranial venovenous fistulae. As these are identified, one at a time, they are coagulated, transected, and each bony diploic channel is obliterated with bone wax. Care should be taken to reflect the scalp flap without dissecting the pericranium from the underlying skull, since this greatly increases the amount of bony bleeding and subjects the child to undue risk of air emboli.

Transcranial Arteriovenous Fistulae

Though these may exist as isolated clinical entities, they almost invariably occur in association with dural arteriovenous fistulae. We have never seen an association between either transcranial or dural arteriovenous fistulae and leptomeningeal or choroidal malformations. The arteries afferent to the fistulae are most commonly branches of the superficial temporal artery, and second most commonly branches of the occipital artery. Their identification by selective external carotid angiography is not difficult and their isolation, ligature, and transection is a simple surgical procedure. However, when there is a dural component to the transcranial arteriovenous fistula, one may not expect that simple ligature of the extracranial vessels afferent to the fistula will suffice.

Dural Arteriovenous Fistulae

(Figs. 11.13, 11.14)

These are invariably high-flow arteriovenous malformations, involving arterial branches of the external carotid system and the dural venous sinuses. The superior sagittal sinus is rarely involved, the tentorium cerebelli and transverse dural sinuses are most often involved, and the cavernous sinus is involved only after a sphenoid sinusitis or an injury.

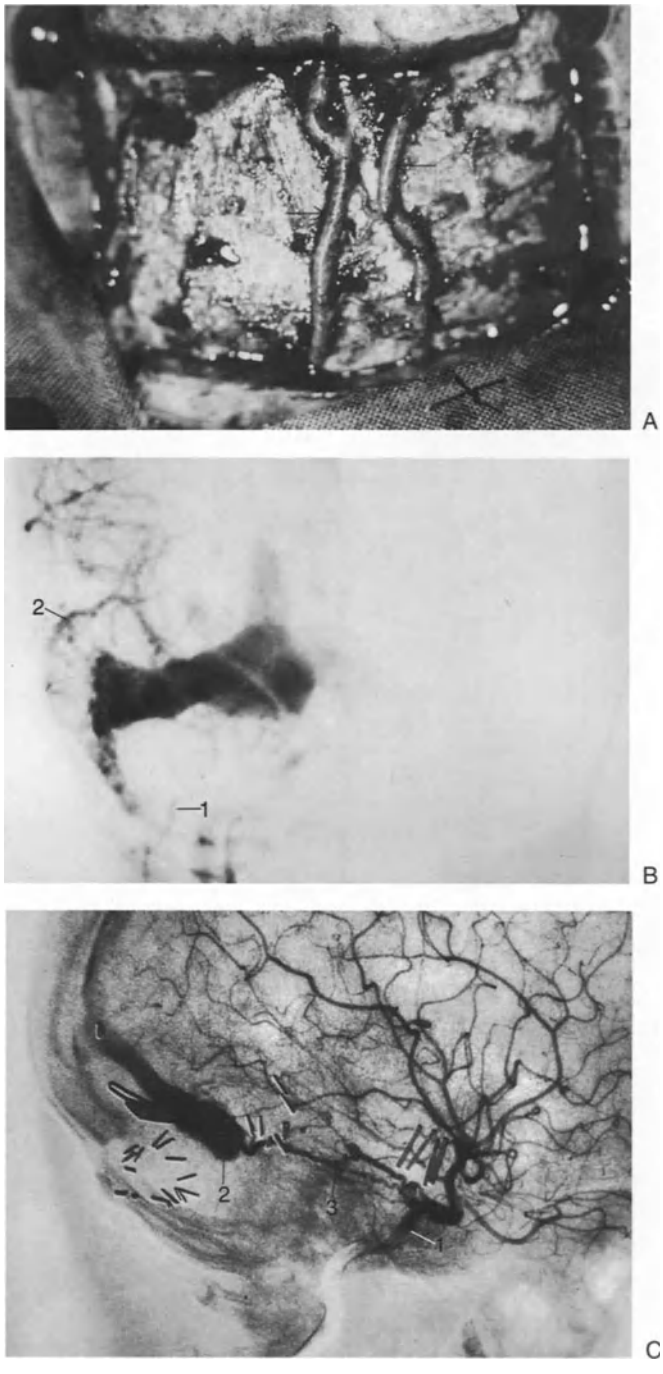
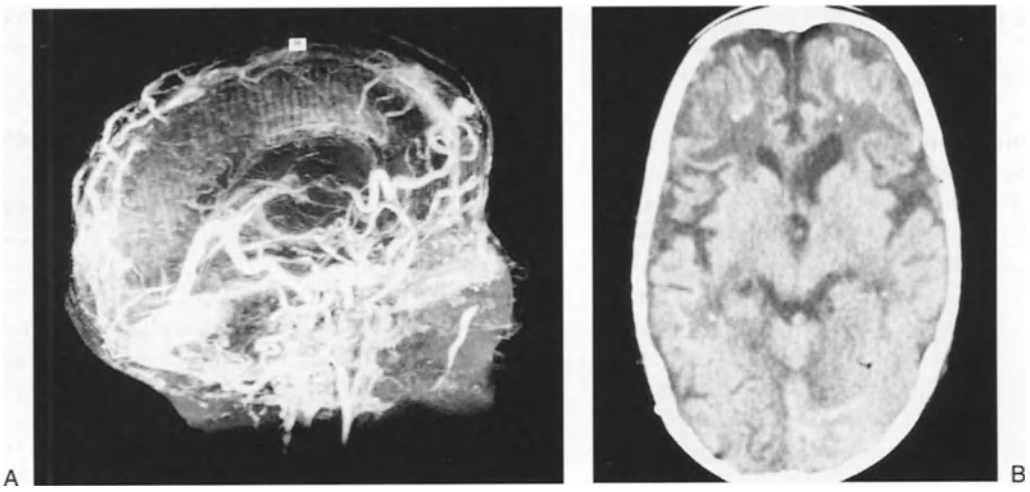


Figure 11.13. (A) Hypertrophied meningeal arteries (*arrows*) feeding into the transverse sinus. (B) Preoperative arteriogram illustrating the middle meningeal (*1*) and occipital (*2*) feeders to the tranverse sinus. (C) Postoperative arteriogram illustrating complete occlusion of meningeal and occipital feeders. There is, however, shunting from the internal carotid, (*1*) to the transverse sinus (*2*) through the tentorial arteries, Bernasconi-Cassinari (*3*). Note that this child has hypertensive hydrocephalus, something I have commonly observed in children with high-flow arteriovenous shunts into the centrencephalic venous system and the dural sinus.



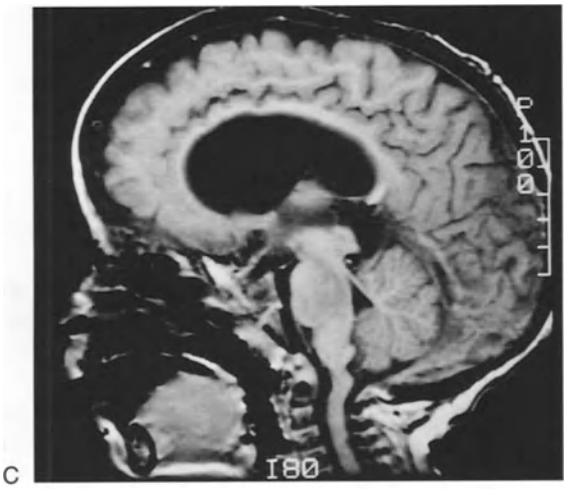


Figure 11.14. This figure shows a venogram (A), an axial T1 weighted MRI scan (B), and a sagittal T1 weighted MRI scan (C). In the venogram one notes the complete occlusion of the superior sagittal sinus with the shunting of all of the venous drainage through the medullary system of the hemispheres and into the deep venous system, the straight and transverse sinuses, and the cavernous sinus. Anteriorly, the structure which appears to be the anterior third of the superior sagittal sinus is a remarkably dilated cortical vein. In the axial MRI study one sees cortical atrophy to the point of redundant gyri, severe dysplasia of the white matter, dilation of the sylvian fissures and the subarachnoid spaces over the insula bilaterally, and posteriorly venous drainage from the occipital lobes into the straight sinus. In the sagittal T1 weighted image one notes to advantage the advanced cortical atrophy and redundant gyri, the remarkable dilation of the sulci, progressive dilation of the lateral ventricle, a Chiari I alteration secondary to the active and progressive ventriculomegaly, and buckling at the bulbomedullary junction as a complication of tonsillar herniation: tetraventricular hydrocephalus of the communicating variety superimposed upon progressive cerebral atrophy secondary to complete occlusion of the superior sagittal sinus.

The transcranial arterial feeders to the venous sinuses are identified angiographically and/or with the use of a Doppler at the operating table, isolated, ligated, and transected. One may not expect these vessels to remain closed if they are simply coagulated or ligated. They must be transected.

The dural arteriovenous shunt consists of branches of the anterior, middle, or posterior meningeal arteries, afferent to a dural sinus. When this occurs, there is arterial blood (under arterial pressure) within the dural sinuses. Consequently, these sinuses do not expand and collapse with inspiration and expiration. Very often, the arterial pressure within the dural sinuses results in either spontaneous thrombosis of the dural sinuses or of the bridging cortical veins to the dural sinuses. Infarction of cerebral parenchyma, secondary to retrograde flow from the dural sinuses through the draining parenchymal veins and into the cerebral substance, causes convulsions and neurological deficit. When the arteriovenous fistula consists of meningeal and meningohypophyseal branches from the internal carotid artery to the petrosal or transverse sinuses, the tentorium cerebelli becomes a virtual labyrinth of sinusoidal, venous, and arterial structures. Consequently, surgical correction of such dural arteriovenous fistulae entails transection of the feeding vessels at their point of entry into the tentorium. If this is not done, *if the dural sinus* (whether it be petrosal or transverse) *is simply ligated, the tentorium is converted into a dural aneurysm* as the arterial blood that enters it from the meningohypophyseal branches is trapped.

Following identification of the individual dural tributaries to the arteriovenous fistulae involving the sinuses, they are exposed surgically, ligated, and transected. One uses the same technique for occluding these vessels afferent to a dural arteriovenous malformation

as for occlusion of the meningeal arteries when opening the dura. The only difference is in size. Craniotomy for exposure of the afferent dural branches is preferable to bur hole opening.

In those cases in which the distal portion of the dural sinus has thrombosed, as not uncommonly occurs in the region of the petrosal or sigmoid sinus, and when there is retrograde arterial flow into the torcular Herophili or straight sinus, one may safely occlude the proximal portion of the dural sinus, providing the tentorium cerebelli has not been converted into an arteriovenous labyrinth by tentorial and meningohypophyseal branches of the internal carotid artery. This eliminates the retrograde flow of arterial blood into the cerebral parenchyma. If the tentorium cerebelli is, in fact, a labyrinth of venoarterial structures, and if the surgeon chooses to occlude the most proximal portion of the transverse sinus so as to eliminate retrograde flow of arterial blood into the cerebral parenchyma, he must then apply hemoclips to the tentorium from the transverse sinus to the tentorial opening laterally, along its insertion onto the petrous apex, and medially along its transition into the falx cerebri. This entails a craniotomy that extends above and below the transverse sinus, elevation of the occipital lobe and depression of the superior cerebellar hemisphere, and exposure of the tentorial edge along the line of the ambient cistern: a formidable undertaking.

Superior Sagittal Sinus Thrombosis

Thrombosis of the sinuses, especially the transverse and superior sagittal, has been a known clinical entity since before the time of cerebral angiography: otitic hydrocephalus was long considered the result of postinfectious thrombosis of the transverse and sigmoid sinuses. However, idiopathic thrombosis of the superior sagittal sinus was not a known clinical entity until the time of cerebral angiography in children (l960s) when it was first diagnosed and then considered a complication of disturbances in hydration.

Superior sagittal sinus thrombosis occurring in the perinatal period, independent of trauma or infection, has been observed, and the patient illustrated in Fig. 11.14 is such a case. The complications of this pathology are devastating: uncontrollable seizures, first delays and then arrest in psychomotor development, progressive cerebral atrophy, and then either severe pan cerebral pathology or death.

Parenchymal Arteriovenous Malformations

Hemispherical Arteriovenous Malformations
(Figs. 11.15–11.17)

In its simplest terms surgical management of leptomeningeal arteriovenous malformations consists of isolating the racemous component from its afferent vessels prior to resection. Some surgeons choose to do this by occluding first the venous (draining) structures, whereas others choose to occlude first the arterial (feeding) vessels. In fact, with the rarest of exceptions, one works along the surface of the malformation, in the plane between normal parenchyma and leptomeningeal arteriovenous malformation, coagulating and transecting feeding and draining vessels, as one separates the racemous and aneurysmal components of the arteriovenous malformation from the surrounding cerebrum. The identifiable afferent structures to the racemous and aneurysmal components, carrying arterial blood, are exposed and isolated, whenever possible. Similarly, the

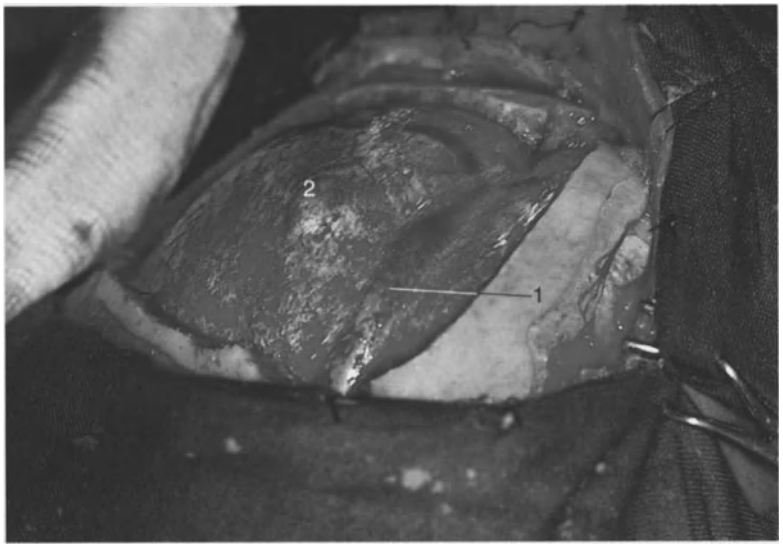


Figure 11.15. For hemispherical arteriovenous malformations feeding extensively into the superior sagittal sinus (SSS) (*1*), with aneurysmal dilation of some elements of the malformation (*2*), it is best to reflect a craniotomy that extends on both sides of the SSS.

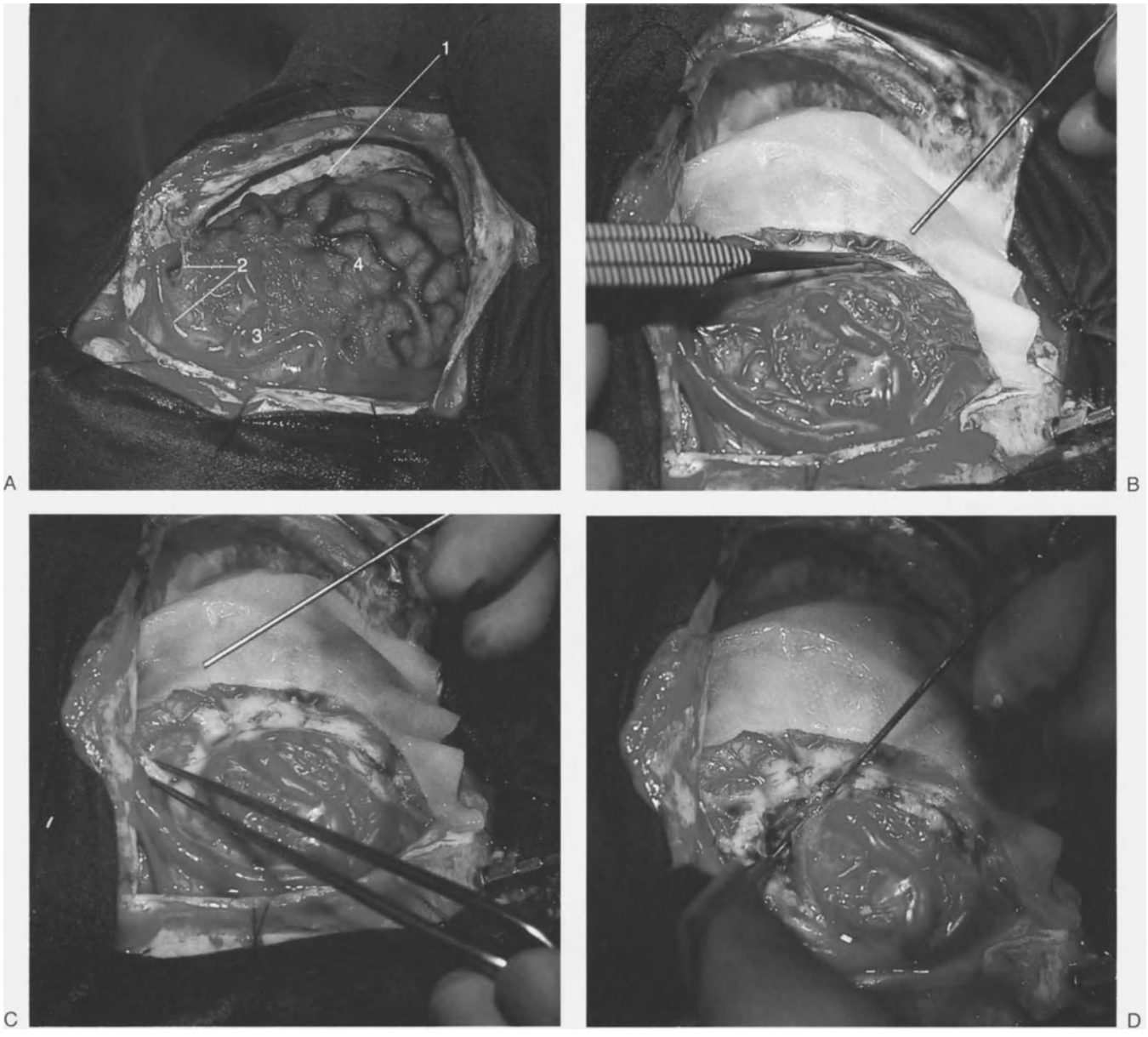


Figure 11.16. This large arteriovenous malformation of the right frontal lobe drained into the anterior two-thirds of the SSS. (A) Three of these draining veins were coagulated and transected, allowing the frontal lobe to fall from the SSS, exposing the falx cerebri (*1*). The two largest draining veins (*2*) were left intact because of the sharp border between the arteriovenous malformation (*3*) and normal brain (*4*). (B) After bipolar coagulation of the arachnoid along the superior limbus of the malformation, the arachnoid is transected with microscissors. (C) Once the bipolar coagulation and transection of arachnoid have been completed around the circumference of the arteriovenous malformation, the bordering brain is covered with Telfa and one of the last two draining veins is coagulated, preparatory to transection. (D) At this time the minuscule bridging vessels between malformation and surrounding brain are coagulated and transected, freeing the distended arteriovenous mass (which has swollen because of coagulation of almost all of the draining vessels). Fluffy cottons are insinuated into the interval between brain and malformation.

identifiable efferent vascular structures, draining the racemous and aneurysmal components, are also isolated. Both are protected with fluffy cotton and/or Telfa. The major efferent vessels are then coagulated and transected.

If the racemous component appears to collapse, one is advised to coagulate and transect one or more of the major afferent (draining) structures, so as to facilitate separation of the malformation from the surrounding parenchyma. Identification of the limbus between mal-

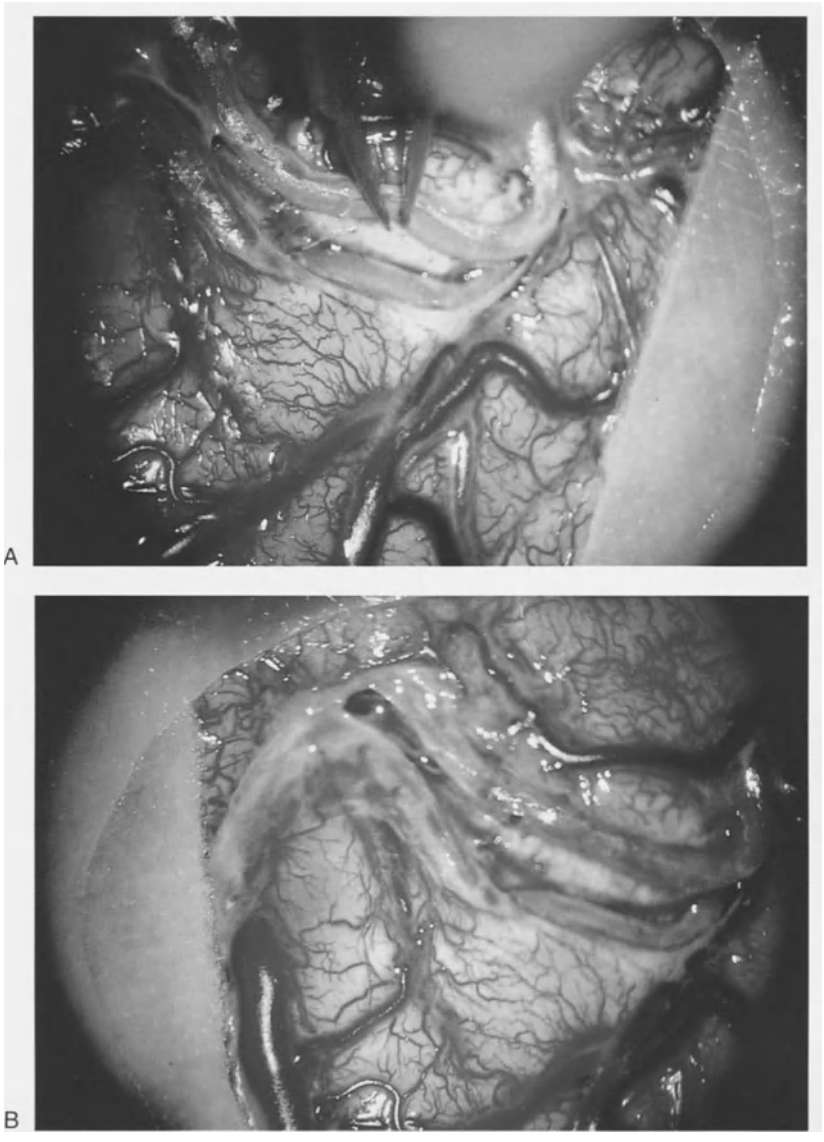


Figure 11.17. Smaller arteriovenous malformations, such as the microarteriovenous fistula (which would be called cryptic if it had bled, since surely it would not be identifiable) are resected by separating the anomalous vessels from the surrounding brain in stages, as shown in (A) and (B).

formation and normal brain is followed by coagulation of the opalescent arachnoid along this line. The coagulated arachnoid is cut with microscissors, and then the multitude of microscopic bridging vessels are coagulated and transected prior to laying fluffy cotton into the created gutter. This allows the pulsating "tumor" portion of the malformation (racemous and aneurysmal components) to be delivered from the surrounding cerebral parenchyma as ever larger fluffy cottons are lain into the interval between malformation and brain. This latter process permits the surgeon to collapse gradually the racemous component of the malformation, diminishing both its volume and flow, and thereby eliminating any surface bleeding. The aneurysmal portion is also collapsed, taking care to do this by laying the fluffy down from distal to proximal, so as to force the arterial blood from it and into the draining, afferent, vessels. Remaining efferent and afferent vessels are coagulated and transected, one at a time, as the racemous and aneurysmal components are unfolded and lifted from the parenchymal bed.

Venous Angiomas (Fig. 11.18)

The venous angioma is another vascular pathology which has only recently become commonly known, though certainly even during the time of cerebral angiography this pathology was occasionally identified. Very probably, it was responsible for the "spontaneous" intracerebral bleeds in both children and adults. Today, with nuclear magnetic resonance one is able to identify these areas of pathology and then document precisely their nature with classical cerebral angiography.

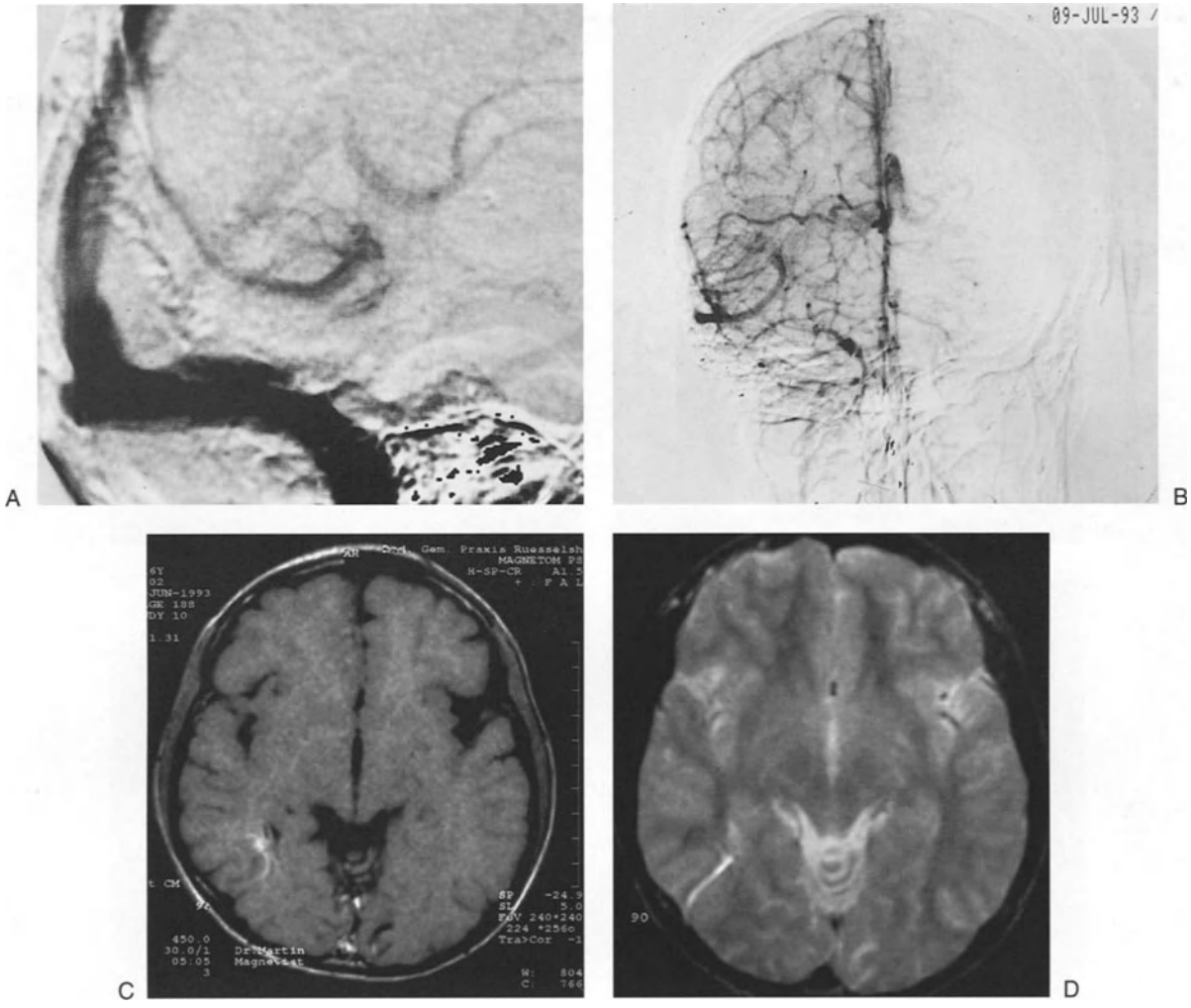


Figure 11.18. In these illustrations, (A) and (B) are angiographic venograms, whereas (C) and (D) are axial MRI studies. In (A) the venous anomaly is clearly seen nestling inferior to the great vein of Galen and draining posterosuperiorly into the superior sagittal sinus. (B) The venous anomaly is now seen to come from the center of the occipital lobe in this half axial venogram. (C) and (D) are, respectively T1 and T2 weighted axial MRI studies revealing the malformation to take origin apparently at the trigone of the right lateral ventricle and then to pass through the white matter and across the sulcus to reach the surface of the junction between the supra marginal and angular gyri. This origin at the trigone, most probably the glomus of the choroid plexus, is not unusual for vascular pathology in children (see the operative findings illustrated in Fig. 11.26B,C).

Cerebellar Hemisphere

Arteriovenous Malformations

The very rare cerebellar hemisphere arteriovenous malformations are managed just as the cerebral arteriovenous malformations. However, one must take note of the fact that the draining veins from cerebellar hemisphere vascular malformations course either superiorly into the transverse sinus or anteriorly into the petrosal and jugular sinuses. Consequently, one does not have access to these structures until the majority of the racemous and aneurysmal components have been dissected from the surrounding cerebellar hemisphere *and compressed.* Of more significance still is the fact that the arterial structures afferent to the malformation are invariably located within the interval separating the cerebellar peduncles and brainstem (medially) from the floculonodular lobes (laterally). The posterior inferior cerebellar artery (PICA), anterior inferior cerebellar artery (AICA), and the main branches of the superior cerebellar artery are located in the parasagittal plane separating the brainstem from the cerebellar hemisphere inferiorly, and the vermis from the cerebellar hemisphere superiorly. *With these anatomical considerations in mind, it becomes clear that cerebellar hemisphere arte-*

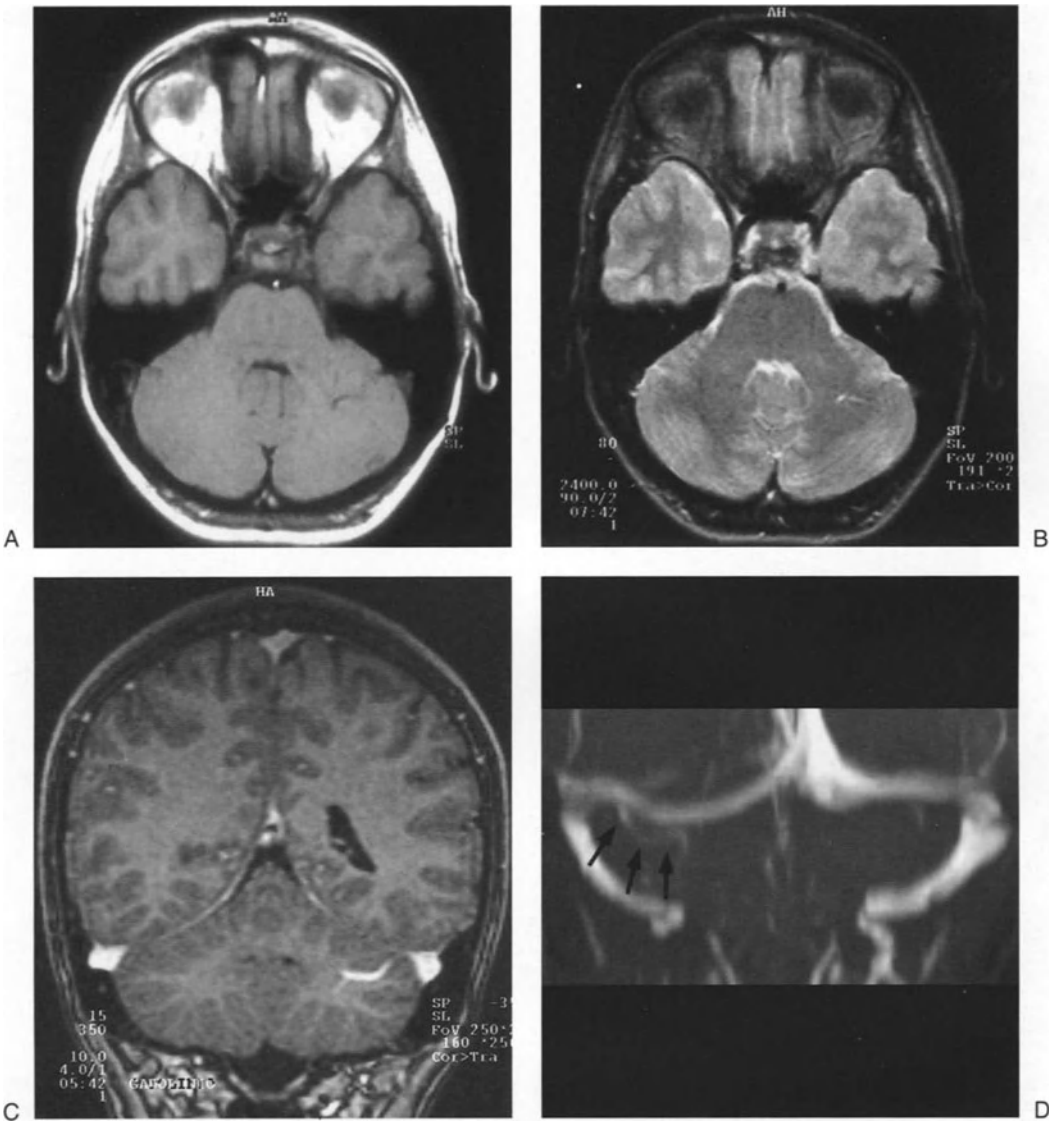


Figure 11.19. This is a series of magnetic resonance studies, cerebral and vascular, of a small venous angioma. (A) is an axial SE600/15, (B) is an axial SE2400–90, (C) is a coronal MPR 3D 10/4 pot-gadolinium, and (D) is an angio TOF 2D 35–39/10 half axial study. On the axial images there is a linear structure in the left cerebellar hemisphere which shows a low signal on T1 and a high signal on T2. The post-gadolinium coronal three-dimensional image of the lesion has a high signal image of the peripheral vessel. In the half axial reconstruction of the venous angiogram TOF two-dimensional sequence, one sees connections of the pathological vessel with the transverse sinus.

riovenous malformations are dissected from the surrounding parenchyma before either the afferent arterial or efferent venous components may be identified, coagulated, and transected.

As the efferent tributaries to the malformation are identified, they should be lifted from the underlying parenchyma, so as to ascertain that there are no recurrent branches to the brainstem, *before* they are coagulated and transected. The afferent venous structures should be coagulated as far from the transverse, tentorial, petrous, and jugular sinuses as possible. After this is done, Avitene or Surgicel strips are placed over the stumps, plastering them to the dural structures so as to minimize risks of reopening and subsequent massive venous bleeding or air embolism. The laying of Telfa strips over the Avitene or Surgicel affords additional protection throughout the remainder of the operative procedure and assists the establishment of a seal between Avitene, Surgicel, and dura.

Medial cerebellar hemisphere or vermian malformations may receive tributaries from the internal occipital branches of the posterior cerebral arteries, the long cir-

cumferential branches of the basilar artery, or the lateral posterior choroidal artery. Their afferent drainage may feed into the supraculminate system, the great vein of Galen, or the basal vein of Rosenthal. If this is identified at angiography, one may choose to reflect both suboccipital and medial occipital craniotomy flaps, so as to have access to the entirety of the transverse sinus, the tentorium, and the three major falcotentorial cisterns (ambient, quadrigeminal, superior cerebellar) (Fig. 11.19).

Orbital Cavernoma (Fig. 11.20)

The cavernoma, cavernous hemangioma, may occur anywhere. Since at the present time it is considered to be a pathological alteration of the vascular system, and not a neoplasm, it is included in this chapter. However, the name cavernoma implies directly a space occupying lesion.

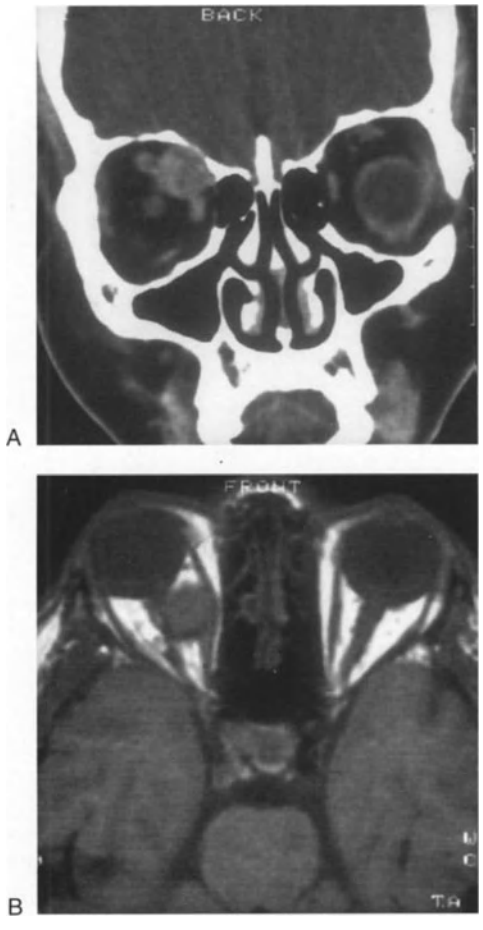


Figure 11.20. Cavernous hemangioma. In (A) a coronal SE 2400/90, and (B) an axial SE 600/22, one is able to identify the cavernoma in the intraconal space, medial to the optic nerve, *on the right*. The T2 weighted image shows a high homogeneous signal intensity. The well-circumscribed nature of this lesion makes it quite feasible for the surgeon to remove it with a very high probability of not damaging the intraorbital structures.

Brainstem Arteriovenous Malformations (Figs. 11.21, 11.22)

The decisive factor concerning operability of a brainstem arteriovenous malformation is whether the afferent and efferent vessels are located on the *surface* of the stem or *traverse* it. Surface malformations may be dissected from the lateral aspects of the pons varolii or the medulla oblongata. Malformations adherent to the peduncles, the collicular plate, the dorsal aspect of the pons, or the floor of the IV ventricle are inoperable. Similarly, those malformations that have the racemous and/or aneurysmal components in the center of the brainstem are inoperable because the afferent and efferent vessels traverse the stem, almost invariably from ventral to dorsal, with the afferent vessels being branches of the vertebral basilar system and the efferent vessels draining into the petrosal sinuses, the basal veins of Rosenthal, the great vein of Galen, and the supraculminate system.

It is often necessary to operate on resectable arteriovenous malformations of the brainstem in stages, since they may be nourished by branches of PICA, the vertebral artery, AICA, the superior cerebellar artery, and long circumferential branches of the basilar artery. Also, the drainage may be into the inferior vermian veins, the inferior petrosal sinus, and the superior petrosal vein. Such combinations of efferent and afferent vasculature necessitate at least two stages, one through an inferior cerebellar triangle approach, and the other through a posterior inferior temporal craniotomy and subtemporal approach to the ambient cistern. Because of the anatomical characteristics already described, one does not need to consider transecting the tentorium to gain access to the inferior and lateral surfaces of the pons. However, an occipital craniotomy may be necessary for a transtentorial approach to the vascular pathology with the region of the three major dorsal cisterns (superior cerebellar, quadrigeminal, ambient).

Before beginning dissection of the malformation, with the intent to separate it from the surface of the stem, one must identify individually the efferent and afferent vessels, and then coagulate the efferent vessels over a 3- or 4-mm distance before their entrance into the racemous or aneurysmal components. It is best to avoid clipping and transecting these vessels as long as possible, so as to minimize risk of dislodging the clip from a transected vessel during subsequent dissection. As a group of afferent vessels are dissected, coagulated, clipped, and transected, they should be covered with moistened Telfa strips.

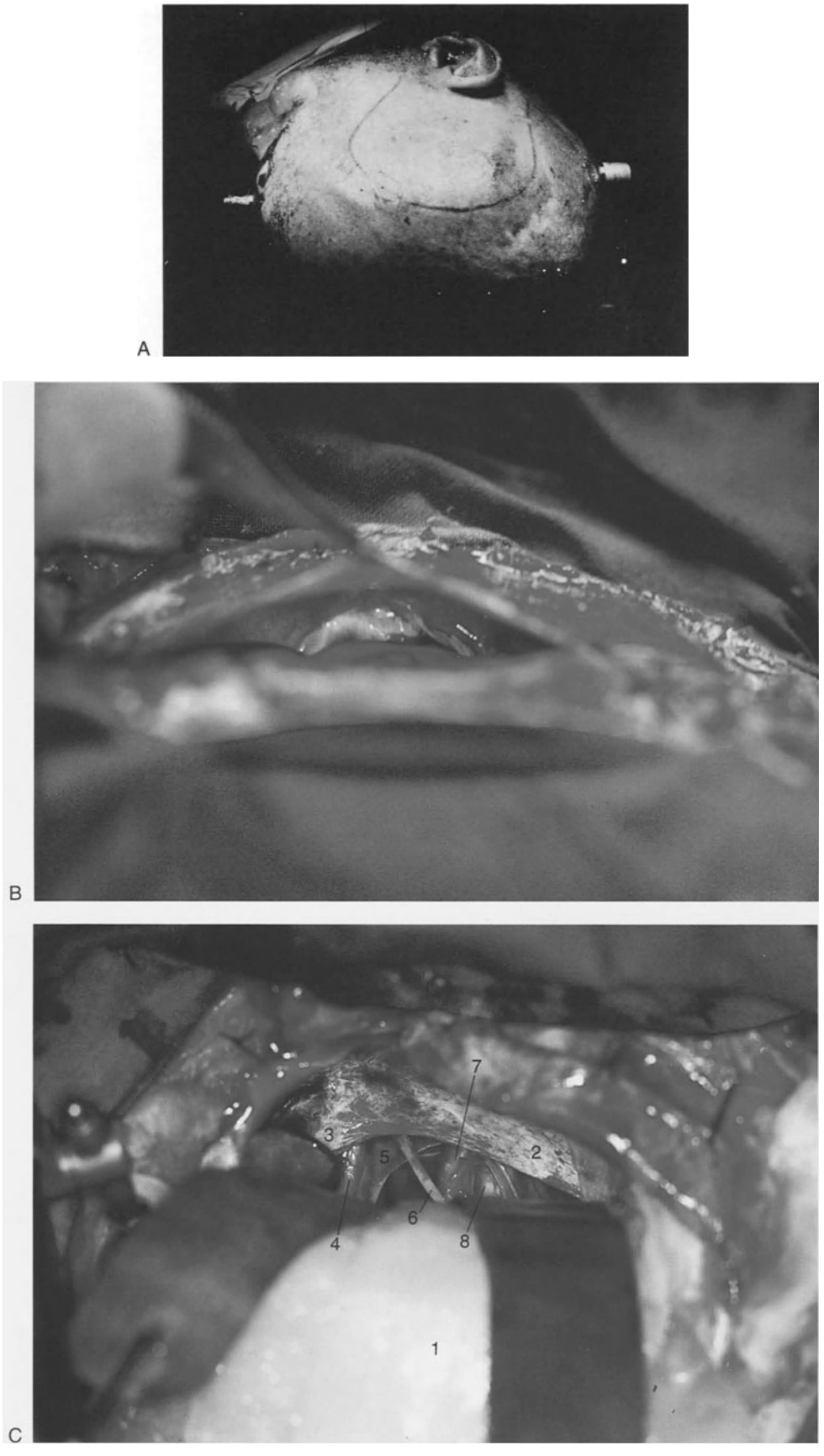
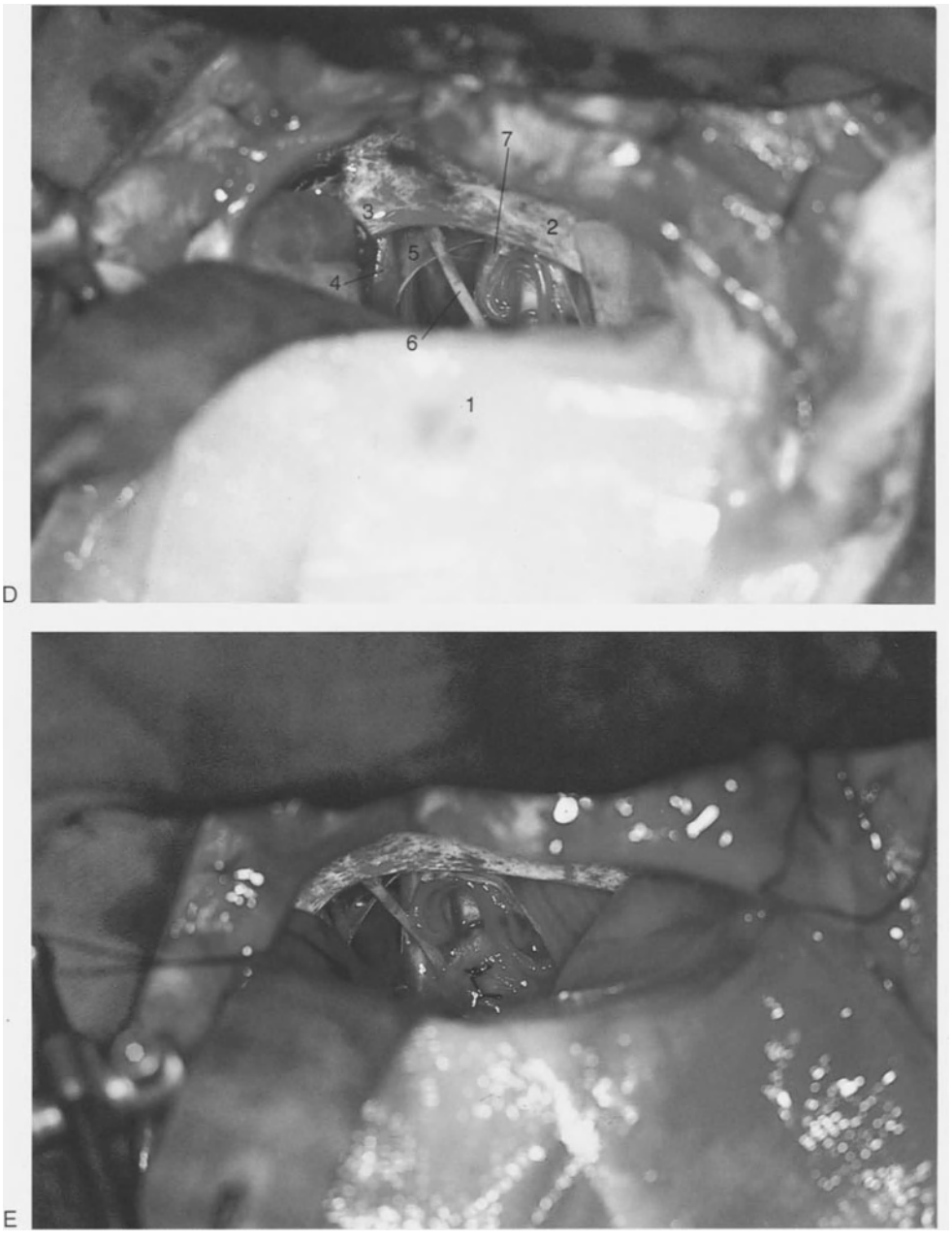


Figure 11.21. Legend see p. 373.



◀ **Figure 11.21.** Brainstem arteriovenous malformations are approached through a temporal flap, under the temporal lobe, along the tentorium to the ambient cistern. (A) Temporal flap. (B) The osteoplastic bone flap has been reflected and the dura opened and slightly elevated, exposing the sigmoid sinus. (C) The temporal lobe has been covered by Telfa and retracted (*1*), exposing the tentorial edge (*2*), posterior clinoid (*3*), internal carotid artery *(4)* arachnoid membrane of the lateral surface of the interpeduncular cistern (*5*), III cranial nerve (*6*), basilar artery (*7*), and pons (*8*). (D) Further retraction, using gravity and removing one of the two spatulas, exposes the anomalous vessels along the surface of the pons. (E) Two of the anomalous vessels have been ligated and two remain to be ligated. (A1–E1) see pp. 374–376.

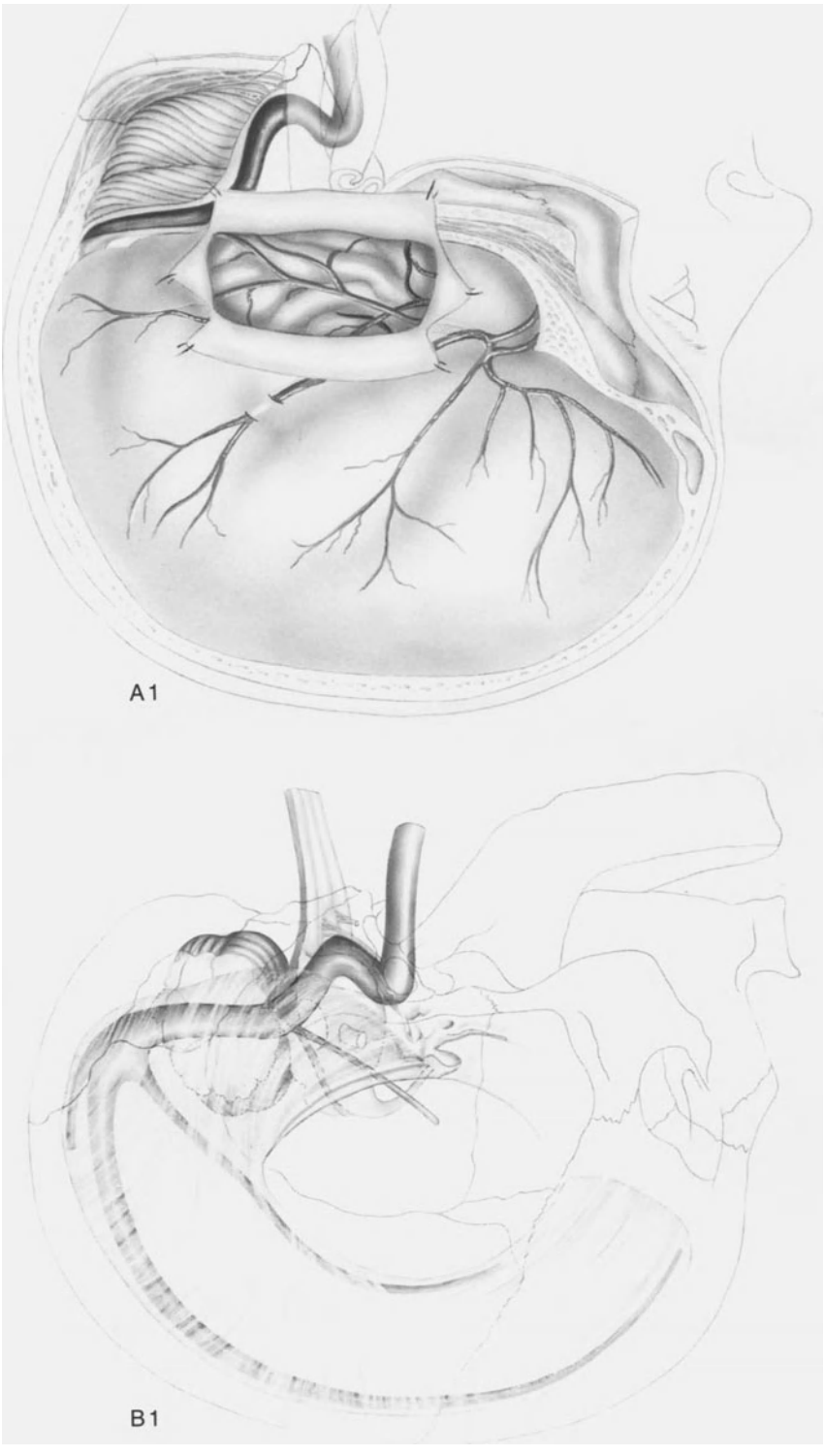


Figure 11.21. (A1) The dural opening need only expose the middle and posterior thirds of the temporal lobe, but it must be brought down to the floor of the middle fossa. It is preferable not to expose the vein of Labbé. This postition of the child's head facilitates retraction and minimizes compression damage to the temporal lobe...but one must take great care not to permit stretching of the vein of Labbé. (B1) The brainstem (from III ventricle floor to the cervical roots of XI) is seen through "the transparent" tentorium, permitting the reader to realize that mid-brain may be worked on effectively without opening the tentorium, but that the upper portion of the pons wants tentorial opening... and that lower pons wants sectioning posteriorly to, and across, the superior petrosal vein: The tentorial flap may then be reflected, exposing the stem as shown in (C1). (C1–E1) see pp. 375, 376.

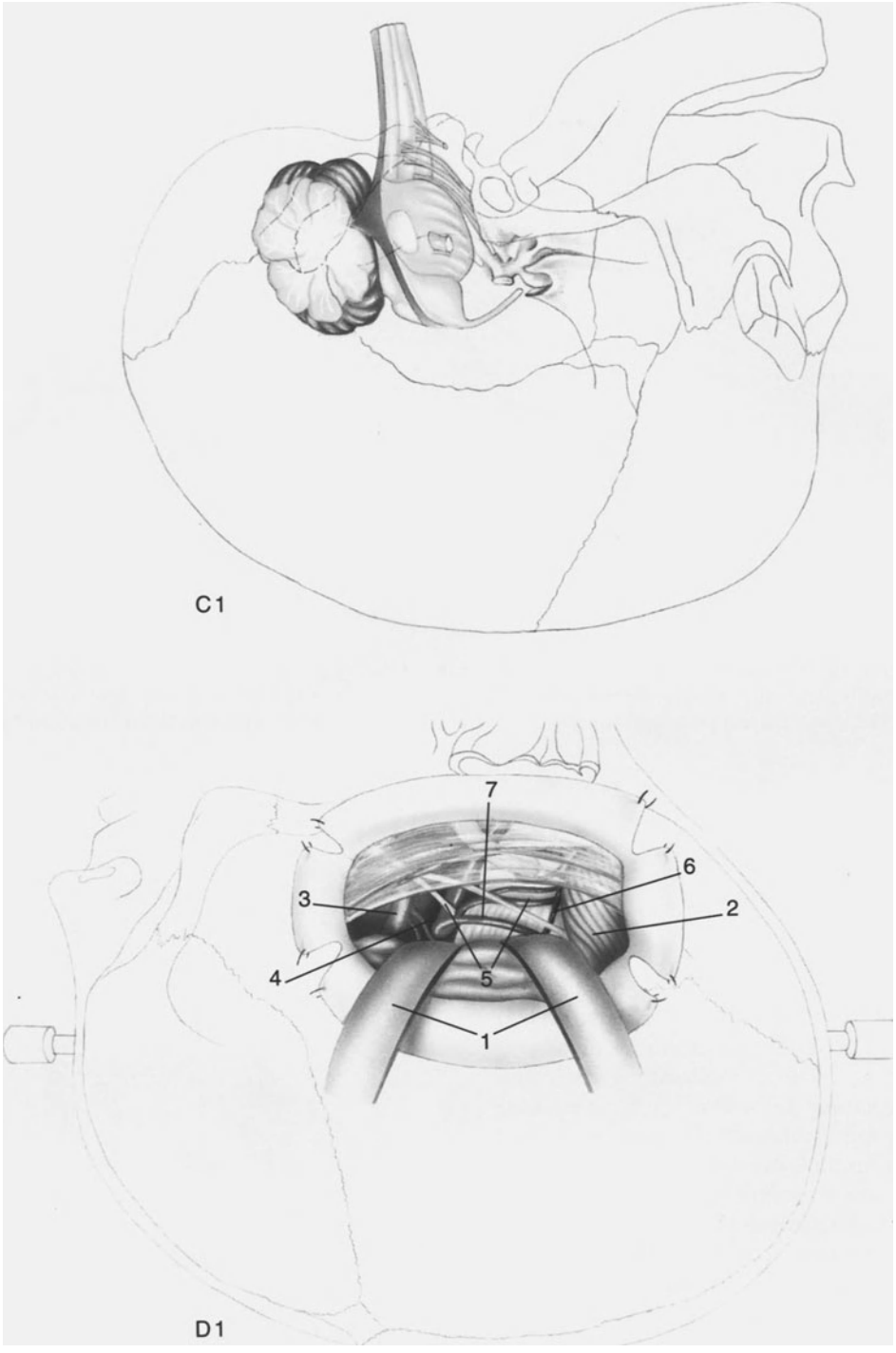


Figure 11.21. (C1) With the child supine, head dropped minimally below the horizontal axis of the trunk, the brainstem is fully visible after the tentorium has been opened and its lateral flap brought to anchor either to the periosteum or dura at the edge of the temporal flap. The sigmoid sinus is now posterior to the area of access: out of the way. (D1) This drawing was taken directly from an intraoperative procedure on the pons. It reveals (1) the ease of retraction of the temporal lobe; (2) the lateral surface of the cerebellar hemisphere protruding above the tentorial edge; (3) the carotid artery with its anterior choroidal and posterior communicating artery branches; (4) the basilar fundus with its posterior cerebral and superior cerebellar branches; (5) the III and IV cranial nerves; (6) the transverse pontine vein; and (7) the superior portion of the pons... all above the line of the tentorial edge.

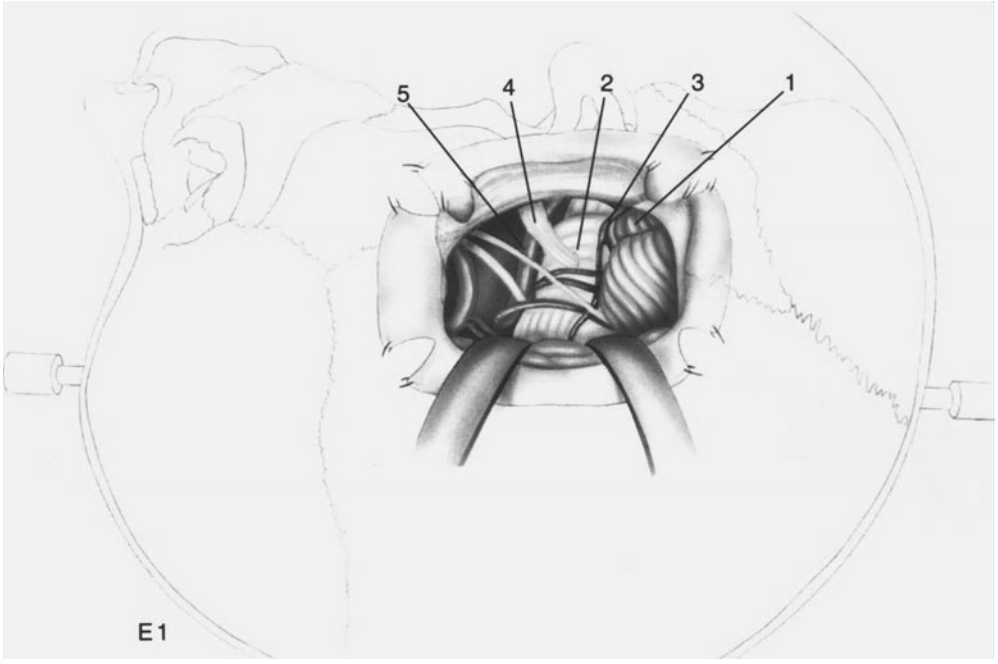


Figure 11.21. (E1) Once the tentorium has been cut and sewn out of the direct operative field, one sees (1) the flocculus and lateral recess of the IV; (2) the inferior pons and superior portion of the bulb (olive); (3) the inferior extension of the transverse pontine vein; (4) the sensory root of V at the entry zone; and (5) the inferior portion of the trunk of the basilar artery.

One then separates the malformation from the surface of the brainstem by using a fluffy cotton to tease them from one another, and then rolling the cotton over the surface of the racemous component, compressing and separating it evermore from the brainstem. Bipolar forceps may be used to shrink the racemous and aneurysmal components, steadily diminishing the flow through the malformation. Because of the fact that these malformations are fed by the same branches of AICA that nourish the choroid plexus within the lateral recess of the IV ventricle, one may expect to encounter a cluster of pathological vessels around the exit of the vestibular and auditory components of the VIII nerve (along the medial surface of the lateral recess). This is most effectively dealt with by using bipolar forceps, least effectively by attempting to use hemoclips: coagulation shrinks choroid plexus and racemous arteriovenous malformations, so that one may identify and preserve the VIII cranial nerve, clips tending to stick to the applicator or tear open choroidal tissue. Similarly, those portions of the brainstem malformation being fed by branches of PICA may extend into the inferior medullary velum and involve the IV ventricle choroid plexus within the roof. They are treated the same way technically. Varices, whether holding venous or arterial blood, extending into or abutting upon the floor of the IV ventricle, should not be coagulated or dissected. One may hope that they will subsequently coagulate spontaneously, and should not risk opening into them.

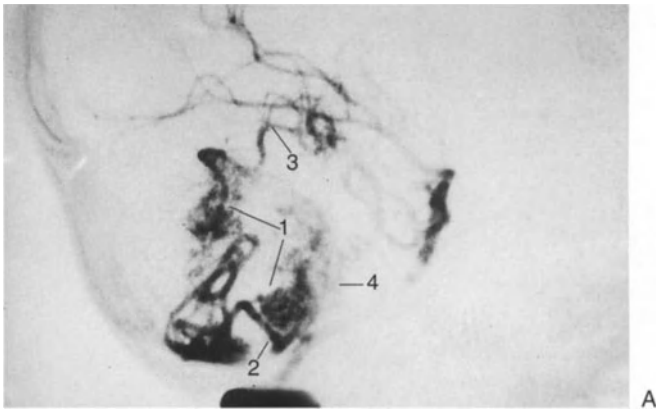


Figure 11.22. (A) This is an arteriogram of a child with an arteriovenous malformation of the brainstem, with anomalous vessels (*1*) coming from the posterior inferior cerebellar artery (PICA) (*2*) and the superior cerebellar (*3*) and anterior inferior cerebellar arteries (*4*). (B–D) see p. 377.

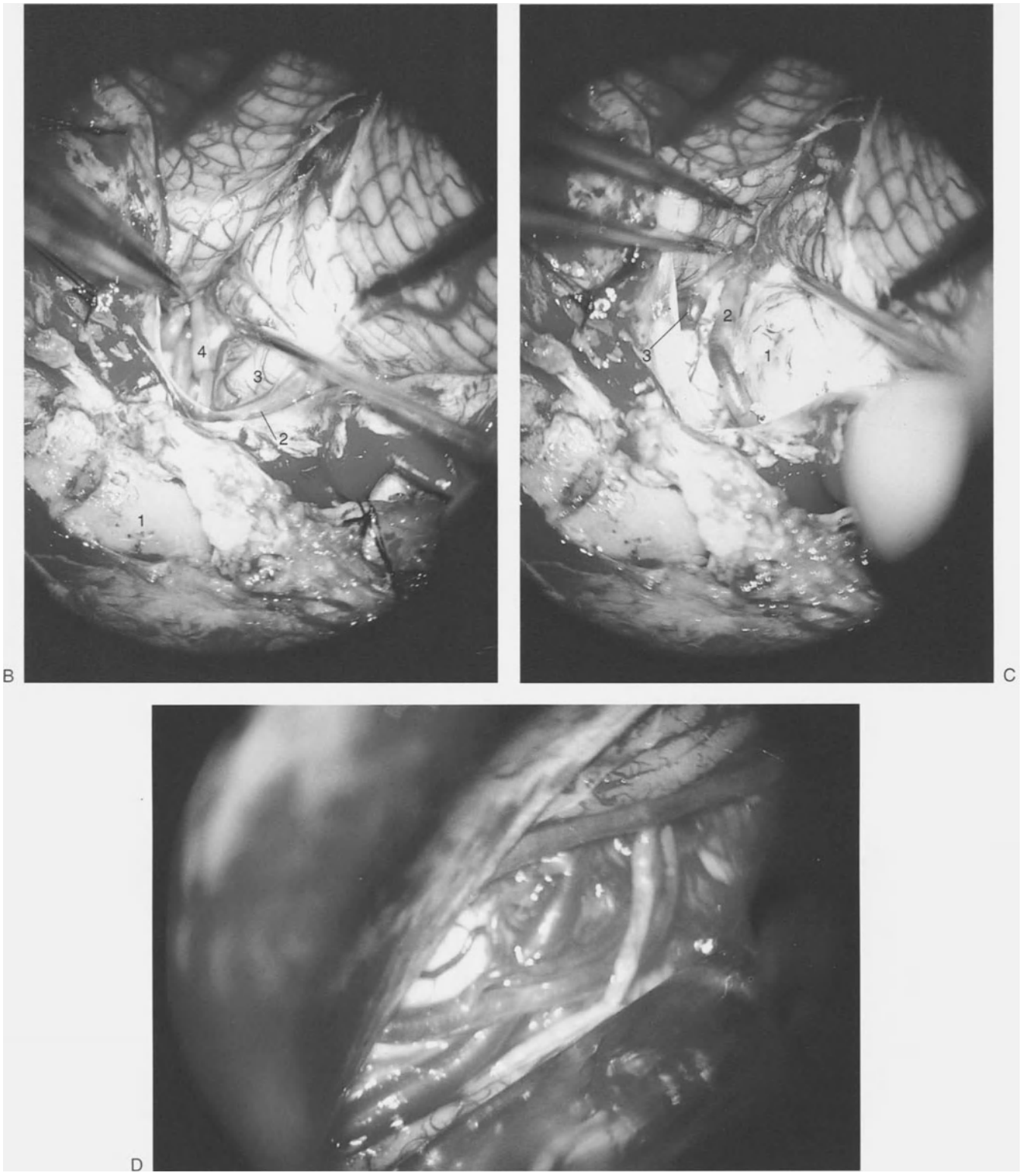


Figure 11.22. (B) A suboccipital craniotomy, with preservation of the arch of C1 (*1*), permits exposure of the PICA (*2*) in its course around the tonsil (*3*) and into the region of the vallecula (*4*). (C) The medial surface of the tonsil (*1*) has been coagulated and covered with Telfa, in order to facilitate exposure of the main trunk of the PICA entering the IV ventricle through vallecula (*2*) to nourish the portion of the arteriovenous malformation within the IV ventricle (*3*). (D)After the feeders from the PICA and the malformation within the IV ventricle have been occluded and removed, at another sitting, those branches from the suprior cerebellar and anterior inferior cerebelar arteries are exposed, coagulated and transected.

Spontaneous Intraparenchymal Hemorrhage
(Figs. 11.23, 11.24)

Intraparenchymal hemorrhage has long been associated with the vasculopathies of arteriosclerotic origin, considering them outside of pediatric age ranges. However, spontaneous occlusive and hemorrhagic vascular accidents occur in children of all ages and may be associated with coagulopathies or nonarteriosclerotic vasculopathies. They present clinically in identically the same manner as all intracerebral hemorrhages.

Intraventricular Arteriovenous Malformations
(Fig. 11.25)

Intraventricular arteriovenous malformations may be subclassified into those occupying the lateral ventricle, the III ventricle, or the IV ventricle. For all intents and purposes, the IV ventricle arteriovenous malformation does not exist independent of involvement of either the brainstem or vermis. Consequently, no particular tech-

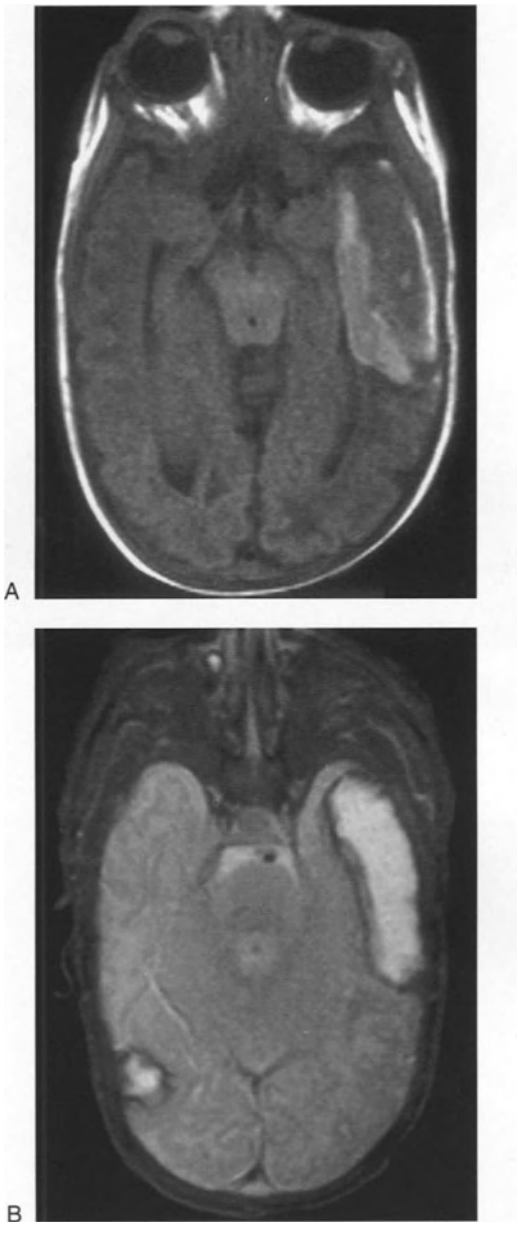


Figure 11.23. Spontaneous intracerebral hemorrhage. (A) and (B) of this illustration are, respectively, coronal and sagittal T1 MRI studies which illustrate the intracerebral hemorrhage in the area immediately superomedial to the left temporal horn. Whether the hemorrhage comes from an occult venous anomaly, a venous anomaly associated with the choroid plexus of the temporal horn, a coagulopathy, or a vasculopathy could not be asserted

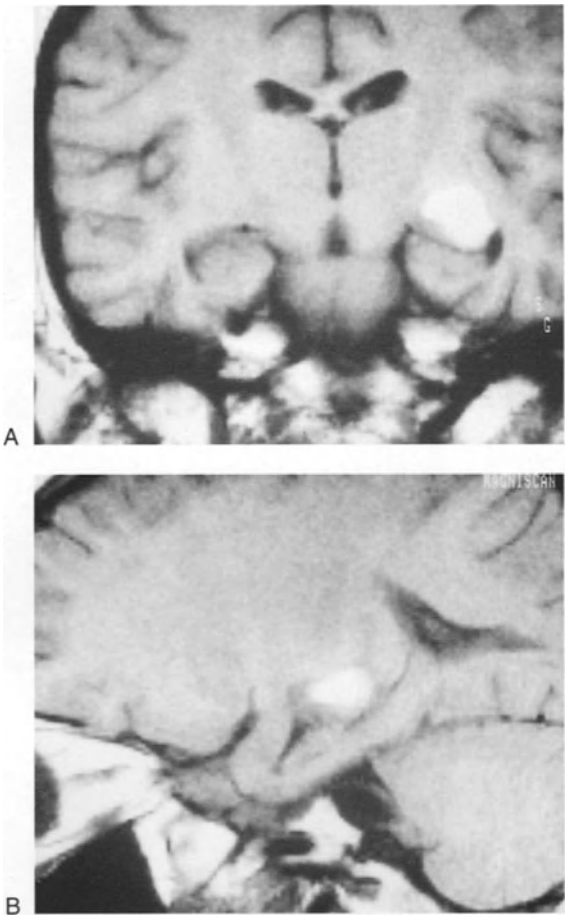


Figure 11.24. Spontaneous intraparenchymal hemorrhage in a newborn with platelet deficiency. This child, whose gestational age was 36 weeks, had a low birth weight. (A) is an axial SE 600/15 and (B) is an axial SE2400/90. The images reveal a left temporal lobe hemorrhagic area with a high signal on the T2 weighted image (B), and a low signal on the T1 weighted image which represents methoglobin. At the periphery of the T2 image the low signal represents hemosiderin. There is also a lesion in the contralateral temporo-occipital area.

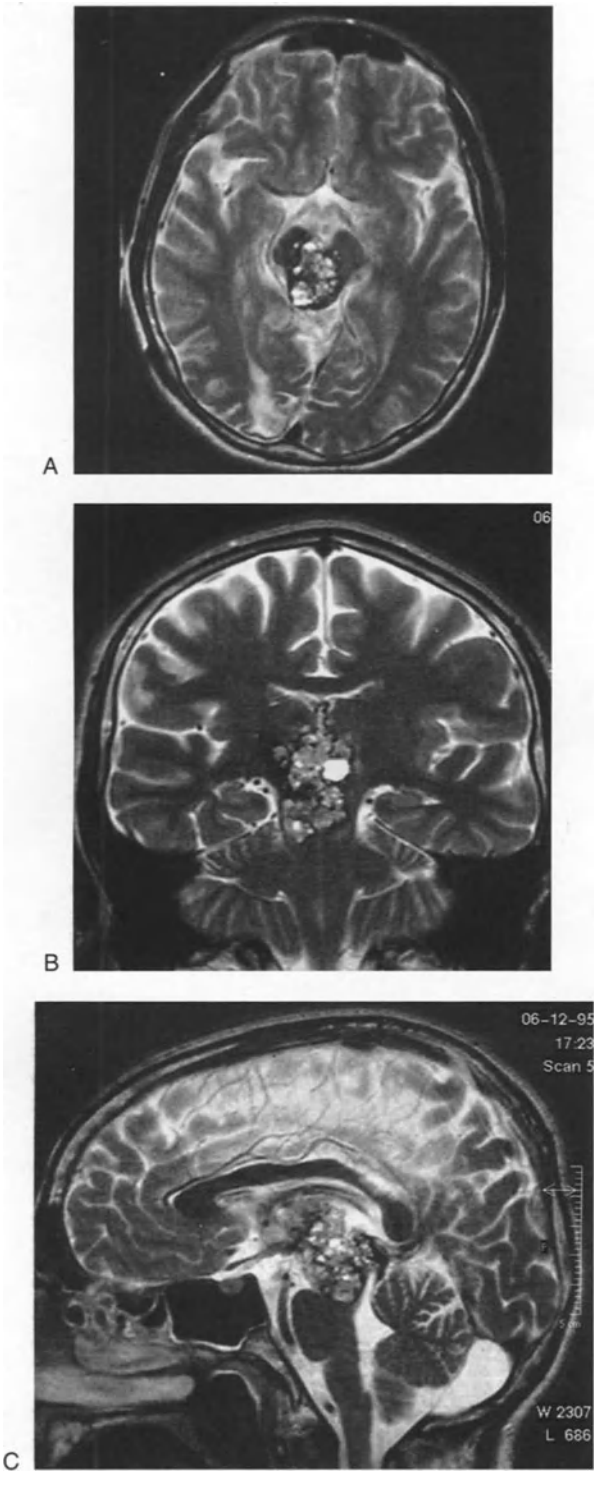


Figure 11.25. Cavernoma (cavernous angioma) of the midbrain. (A) axial, (B) coronal, and (C) sagittal are turbo SE 5000/130 images. They show *dishomogeneous hypo/high signal intensity mass* in the midbrain. The entirety of the mid brain, portions of the peduncles along with substantia nigra and tegmentun and tectum, is involved. The cavernous angioma has a lobular appearance, it extends beyond the collicular plate as it bulges into the collicular cistern, and fungates anterosuperiorly into the III ventricle.

nical considerations (different from those already described for cerebellar hemisphere, vermian and brainstem lesions) apply to their management. The lateral ventricle arteriovenous malformation is a relatively common lesion in childhood, whereas the III ventricle malformation is very rare in this age group.

Lateral Ventricle Malformation (Fig. 11.26)

These involve the glomus of the choroid plexus, being fed by medial and lateral posterior choroidal arteries, seldom extending as far anteriorly as the region of the pes hippocampus and amygdala. They tend to expand on both sides of the choroidal fissure, within the glomus of the choroid plexus intraventricularly, and within the region of the ambient cistern medial to the choroidal fissure. It is most unusual for them to embed themselves within the calcar avis or collateral eminence, and seldom do they extend along the terminal sulcus to the foramen of Monro. However, one must be prepared to have access to the temporal pole, the region of the trigone, and the terminal sulcus as far anteriorly as the foramen of Monro, when approaching arteriovenous malformations of the lateral ventricle. This is easily accomplished, and presents little or no risk of morbidity, when the right side is involved. However, for malformations of the choroid plexus in the dominant hemisphere, the surgical considerations are quite different and the risks of morbidity are awesome. Therefore, *it is impossible to exaggerate the amount of care and deliberation involved in deciding whether to operate a malformation within the ventricular system of the dominant hemisphere!*

For the nondominant hemisphere, an osteoplastic temporal flap is reflected and the dura is opened in a double trapdoor fashion, prior to performing a cerebrotomy and entering the right lateral ventricle at the trigone. Self-retaining retractors are then placed into the temporal horn and body of the right lateral ventricle, elevating them so as to hold the cerebrum up and to keep the ventricle distended. The malformation will be identified immediately within the glomus. At this time one may determine whether it is extending into the body and temporal horn of the lateral ventricle.

It is best to begin resection by coagulating the choroid plexus, whether normal or prey to arteriovenous malformation, at the tip of the temporal horn, shrinking it as one proceeds to use the bipolar forceps, thus opening the choroidal fissure and identifying the anterior choroidal artery immediately. This vessel should be coagulated and transected. The dissection then proceeds posteriorly in the direction of the glomus, coagulating the malformation, or choroid plexus, as one proceeds, opening completely the choroidal fissure and entering into the ambient cistern. One will identify the IV cranial nerve and the vein of Rosenthal, which generally

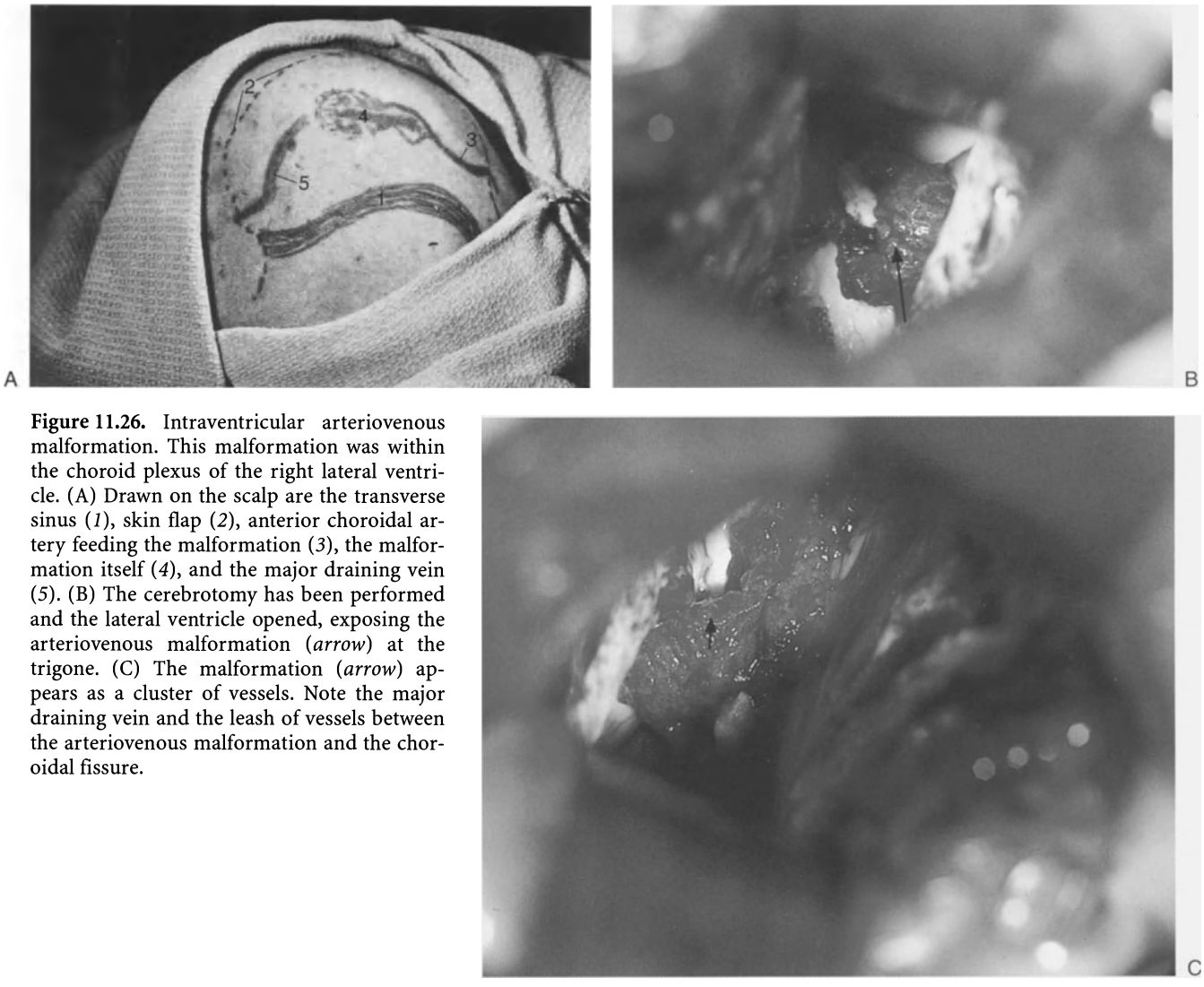


Figure 11.26. Intraventricular arteriovenous malformation. This malformation was within the choroid plexus of the right lateral ventricle. (A) Drawn on the scalp are the transverse sinus (*1*), skin flap (*2*), anterior choroidal artery feeding the malformation (*3*), the malformation itself (*4*), and the major draining vein (*5*). (B) The cerebrotomy has been performed and the lateral ventricle opened, exposing the arteriovenous malformation (*arrow*) at the trigone. (C) The malformation (*arrow*) appears as a cluster of vessels. Note the major draining vein and the leash of vessels between the arteriovenous malformation and the choroidal fissure.

is draining arterial blood, since the efferent vessels from the malformation drain into this vessel, the terminal vein, and, indirectly, the galenic system. As the glomus is approached, the malformation may be seen to consist of small patches of normal choroid plexus, extensive racemous areas, and very large aneurysmal components, consisting of the terminal vein and efferent vessels to the vein of Rosenthal. These may be safely coagulated, their rootlets cut from the surrounding ependyma over the calcar avis and collateral eminence, and then, superiorly, from the terminal sulcus. At this transition point, one leaves the choroidal fissure, so that there is no longer risk of damaging the vein of Rosenthal or the IV cranial nerve. The malformation may now be lifted from its bed if it had been limited to the glomus, or the dissection may be continued to the foramen of Monro if it extends that far anteriorly. There is no need to resect normal choroid plexus from the terminal sulcus or the foramen of Monro. Those portions of malformation extending through the choroidal fissure into the ambient cistern are dissected and coagulated as one opens the choroidal fissure, extending posteriorly from the tip of the temporal horn, then superiorly and medially. If it is necessary to operate an arteriovenous malformation of the choroid plexus in the dominant hemisphere, a frontoparietotemporal bone flap should be reflected and the dura opened.

Access to the glomus is attained through a cerebrotomy placed posterior and superior to the supramarginal and angular gyri, entering behind Wernicke's area in the cortex and superior to the radiations of Gratiolet along the lateral surface of the trigone. This minimizes risks of speech impairment or hemianopsia. Once the ventricle is entered, the same technique is used for resection as in the nondominant hemisphere, unless the malformation extends along the terminal sulcus to the region of the foramen of Monro. If it does, one must move anteriorly to perform a cerebrotomy within the

superior frontal convolution, anterior to the motor strip, so as to enter portion 2 of the frontal horn. Placement of the self-retaining retractor within the frontal horn elevates it and exposes the foramen of Monro. This permits the same access to the most anterior portion of the lateral ventricle choroid plexus, without undue risk of damage from retraction.

Third Ventricle Arteriovenous Malformations

Third ventricular arteriovenous malformations, when they occur, are generally so small in size as to put the neuroradiologist in difficulty concerning the diagnosis. They are easily confused with normal choroid plexus of the roof of the III ventricle.

The surgical approach and technique for obliteration of arteriovenous malformations within the roof of the III ventricle is the same as for resection of papillomas of the roof of the III ventricle. The arterial feeders and the venous drainage are the same. The fundamental difference between the two is that the arteriovenous malformation is very small and easily confused with normal choroid plexus, whereas the papilloma is large. In fact, at the time of surgery one may not be able to identify with certainty the presence or location of an arteriovenous malformation because of the presence and appearance of normal choroid plexus. Consequently, it is recommended that the surgical procedure, if undertaken, since generally it is not indicated, be limited to coagulating the choroid plexus within the roof of the III ventricle and that no attempt be made to identify and obliterate afferent and efferent vessels.

Arteriovenous Fistulae Involving Galenic System and/or Perimesencephalic Leptomeninges

Introduction

Vein of Galen aneurysms are rare vascular malformations which develop during intrauterine life. Their clinical presentation depends on the patient's age at onset: severe symptoms such as congestive heart failure and hydrocephalus are common in newborns and younger children, whereas in older children and adults headache and hemorrhage are not unusual findings.

Aneurysmal dilatation of the vein of Galen may be divided into two different entities, according to the embryogenic period in which the malformation develops. It may be defined either as a direct arteriovenous fistula ("true vein of Galen aneurysm") between the choroidal and/or quadrigeminal arteries and overlying a single median venous sac (persistence of the embryonic median prosencephalic vein of Markowski), or it may be a true arteriovenous malformation (AVM) with secondary enlargement of the vein of Galen. The first entity develops at between 3 and 11 weeks of intrauterine life, while the second one has a later embryogenic development. The vein of Markowski is not the proven precursor of the vein of Galen. Several vessels may contribute to the arterial supply, among them the terminal portion of the pericallosal, the anterior and posterior choroidal, the lenticulostriate, the superior cerebellar, the posterior cerebral, and the middle cerebral arteries.

On computerized tomography (CT), the vein of Galen is demonstrated as a homogeneous mass enhancing brightly after contrast administration. Magnetic resonance imaging (MRI) usually shows a signal void because of the turbulent flow in the malformation.

These malformations have been treated differently in the past. For a long time, surgery was the only treatment. Recently, endovascular techniques, particularly those using isobutylcyanoacrylate, especially for newborns, have significantly reduced morbidity and mortality. Nonetheless the results are still unsatisfactory in terms of hydrocephalus and mental retardation. Therefore we reviewed the current literature [25–29] and our personal surgical (pre-interventional neuroradiology) experiences relating to this subject, to compare the different approaches and to ascertain whether surgery, with the refinements introduced by microvascular techniques, is a sound alternative to the currently almost universally used endovascular therapy.

During the past 10 years diagnostics have, once more, set the pace for and established the basis upon which treatment of what has heretofore been considered an ominous, if not incurable clinical entity, has been structured. The detailed angiographic (arteriographic) analyses of the vascular alterations (malformative, ectatic, fistulous) first reported by Raybaud and subsequently elaborated upon and put into clinical perspective by Lasjaunias, have brought us to a point where we may now understand and treat properly both the vascular alterations and their secondary complications.

As so very often occurs here, too, progress proceeding from diagnosis to treatment has been the direct result of ever-increasing specialization. Without the identification of neuroradiology as a distinct specialty, and the subsequent evolution of pediatric neuroradiology, the obliteration or exclusion of arteriovenous malformations and aneurysmal dilation of the galenic system would have made no progress. Following Schechter's suggestion that the term "aneurysm of the vein of Galen" either be abolished or qualified [30] since it is not an aneurysm, Raimondi classified his series of cases into *arteriovenous fistulae of the galenic (lesser and greater) system* and identified specifically the entity of *aneurysmal dilation of the vein of Galen* as a consequence of arteriovenous fistulization from anterior (pericallosal) and posterior (choroidal, retrosplenial, perforating, etc.) [30–32] circulation arteries. The aneurysmal dilation of the lesser, greater, or both galenic

systems was described by him to be the direct result of arteriovenous fistulization. In fact the primary fistulizing arterial system (pericallosal, retrosplenial, choroidal) served as the basis upon which the specific surgical approach (parietal, infratemporal, suboccipital) was based. Though this facilitated greatly identification of the tributary arteries to the aneurysmal dilation of the galenic system, it represented no qualitative basis upon which entirely new technical procedures could be structured. Indeed, it was not until the already mentioned works by the two French neuroradiologists, first Charles Raybaud and subsequently Pierre Lasjaunias, were reported that arteriovenous malformations and aneurysmal dilation involving the embryonic and fetal deep venous circulations could be effectively managed.

Presently, we have the theoretical and technical wherewithal to diagnose and treat, effectively in the majority of cases, arteriovenous malformations and aneurysmal dilation of the galenic system in children. We also have full clinical knowledge of the complicating problems: cardiac failure, hydrocephalus, mental retardation. It remains now for the clinicians diagnosing and managing these childhood anomalies to select which entities should be managed by endovascular interventional neuroradiology, which by classical neurosurgical techniques. It also remains for us to decide whether children with complicating hydrocephalus should be shunted and, if so, whether this should be performed before or after exclusion of the fistula and/or aneurysmal dilation.

The mėdian prosencephalic vein (medial vein of the prosencephalon), a single midline structure, drains the choroid plexuses (of both the lateral and third ventricles) from the 7th through the 12th weeks of intrauterine life. Subsequent to this, the medial vein of the prosencephalon disappears and the two internal cerebral veins may be identified. It is the persistence of the medial vein of the prosencephalon, an embryonic structure, and the occasionally abnormal arterial pattern located over the cerebral hemispheres, which permit one to distinguish arteriovenous fistulae ("vein of Galen aneurysm") from the arteriovenous malformation of the cerebral hemispheres, the basal ganglia, and the brainstem [33].

The medial vein of the prosencephalon, when it persists, has been considered by Raybaud [34] to be the aneurysmal sac of the arteriovenous malformation of the galenic system, identifying the time of occurrence as the 3rd month of intrauterine life: some time between the disappearance of this (embryonic) prosencephalic vein and the appearance of the choroidal vessels.

This fundamental anatomical concept separates arteriovenous fistulae of the galenic system from the arteriovenous malformations, chronologically, in that the latter pathologic entity is characterized by the presence of normal arterial and venous structures [35]. Consequently, the arteriovenous malformation develops after the normal vasculature has formed; the arteriovenous fistula of the galenic system is a true pathological alteration of vascular development [33]. An additional morphologic characteristic of arteriovenous fistulae of the galenic system is the retention of the embryonic venous pattern, most commonly represented by a plexus or sinus of the falx. The straight sinus may or may not be patent.

Of extreme interest to the clinician is the fact that the dural sinus and the deep venous system, "draining" the aneurysmal dilation of the medial prosencephalic vein, are often subject to thrombophlebitis; consequently, the combination of extensive deep venous anomalies, persistence of such embryonic venous structures as the medial prosencephalic vein, single or plexiform sinuses in the falx, nonpatent straight sinus, very significant changes in blood flow and turbulence through the area of the arteriovenous fistula, all contribute to thrombophlebitis [32, 36]. The result: thrombotic occlusion of the major tentorial sinuses (transverse) and draining veins [31, 32].

The progressive occlusion of the tentorial sinuses (straight, transverse, sigmoid) results from the high flow of arterial blood producing a mechanical thrombophlebitis first and thrombotic occlusions subsequently. When, as in some cases of arteriovenous malformations of the galenic system, the straight sinus is congenitally absent, the thrombophlebitis is all the more marked and deep venous drainage through tentorial sinuses impaired steadily, as collateral flow develops into and through the cavernous sinuses and ophthalmic vein. In these cases, with absence or occlusion of the sigmoid sinus and jugular bulb, the flow through the superior and inferior petrosal veins is obstructed, accentuating collateral flow through the orbital venous system. The result is quite obvious clinically: low-grade hemorrhagic conjunctivitis, varying degrees of venous distention of the palpebrae, and serpiginous venous ectasia of the scalp over the frontal and temporal areas. One may palpate pulsatile flow through these venous systems, which emit an audible bruit. The impairment of visual acuity is not rare in children with this marked degree of collateral flow, and very often mental retardation also results.

One, consequently, immediately becomes aware of the need to distinguish between arteriovenous malformations (the classic AVMs which are so common in adults, so very rare in children under 15 years of age, and extremely rare in children under 10 years of age) on one hand, and the arteriovenous fistulae of the galenic system on the other. The next distinction to be made is between true malformations of the galenic system and "false" ones. Lasjaunias et al. [37] identified arteriovenous shunts draining into a venous structure ("venous pouch"), but without reflux into hemispheri-

cal veins (which Raimondi and Cerullo called "aneurysmal dilation of the great vein of Galen" [32]) from those characterized by drainage into the pouch and then retrograde flow into hemispherical and midline veins, which generally drain into the great vein of Galen. It was the latter group that they designated "cerebral AVM with vein of Galen ectasia" (and which Raimondi and Cerullo had already called "arteriovenous malformation of the galenic system" [32]). Another name Lesjaunias et al. gave to this anomaly was "vein of Galen aneurysmal dilatation." The former group, arteriovenous shunting into a venous pouch without reflux, was called "vein of Galen aneurysmal malformation." Hence, the "vein of Galen aneurysmal dilation" consists of just that: dilation of the great vein of Galen to such an extent that the adjective aneurysmal could be applied. The second form is considered by them to be a true malformation: "vein of Galen aneurysmal malformation."

The above classification is not to be considered an escape into intellectual gymnastics, since it provides very precise and complete criteria upon which the clinician may base decisions regarding occlusion or exclusion of the pathology from the arteriovenous circulatory system. Consequently, what Raimondi and Cerullo called *aneurysmal dilation of the great vein of Galen is currently best considered an arteriovenous shunt draining into a venous pouch*, but not having reflux drainage into hemispherical veins. This venous pouch may easily be resected surgically, since the procedure consists of nothing more than isolating the pouch and then occluding the os at its point of entry into the pouch, a simple and safe surgical procedure. However, the very factors that make it a simple procedure from an operative point of view apply to endovascular occlusion. What Raimondi had referred to as arteriovenous malformation of the galenic system, on the other hand, is presently best termed "vein of Galen aneurysmal dilation," since the arteriovenous malformation is not necessarily limited to the galenic system but, rather, *drains* into the galenic system. In fact, any arteriovenous malformation draining exclusively or predominantly into the galenic system causes dilation of the lesser, the greater, or both galenic systems; therefore, the term "vein of Galen aneurysmal dilation" is most appropriate, and most directly indicates the procedure to be performed. In these cases, the endovascular approach is preferable.

Probably one of the most significant contributions to the current status of knowledge on this subject, which is far from clear and certainly about which there is no unanimity, was made by Raybaud et al. [34]. These authors made the point that the venous structure that becomes ectatic (aneurysmally dilated) is the medial vein of the prosencephalon, and that it, in fact, is the structure that involutes as the great vein of Galen is formed.

The vein of Galen aneurysmal dilation may occur in association with any arteriovenous malformation of the brain the draining structures of which are tributaries to the great vein of Galen. The vein of Galen aneurysmal malformation, on the other hand, *has no* vein of Galen. It is the medial vein of the prosencephalon that persists and becomes dilated secondary to the presence of the arteriovenous fistula feeding into it. Hence, the *true* malformation is the blind pouch of the medial prosencephalic vein, from which blood is not shunted into other arteries or veins: the medial prosencephalic vein is malformed. The *false* malformation is ectasia, dilation of the normally formed vein of Galen, which is enlarged because it serves as a conduit for the passage of blood from any cortical or centrencephalic vascular malformation into the other arteries or veins.

In a more recent work, Lasjaunias reviewed his total series of vein of Galen aneurysmal malformations and vein of Galen aneurysmal dilations [33]. Excluding the vein of Galen aneurysmal dilations from the study, he was able to document that the very first anatomical anomaly of the arterial and venous systems to occur embryologically is the vein of Galen aneurysmal malformation. He did this on the basis of the presence of alternate patterns of vessels draining the deep cerebral structures, corresponding to the 80-mm stage. In this work, he was able to confirm the hypothesis put forth by Raybaud, adding support to the theory that the vein of Galen aneurysmal malformation develops before the vein of Galen aneurysmal dilation. He also observed that other arteriovenous malformations of the brain occur after the cerebrovascular structure is completely formed. Furthermore, it was suggested that vein of Galen aneurysmal malformations, as already stated, consist of persistence and dilation of the medial vein of the prosencephalon and arteriovenous shunts involving either the choroidal arteries or mural shunts [33].

With regard to children, one of the most common fistulae found is the arteriovenous fistula between branches of the posterior circle (and, occasionally, the pericallosal artery) and the galenic system, commonly referred to as "aneurysms of the vein of Galen," but which should for simplicity of expression be called vein of Galen aneurysmal dilation. Although, most certainly, aneurysmal dilation of one of the galenic veins invariably occurs when such a fistula develops, it is not truly an aneurysm. This distinction is important, since its surgical treatment is directed toward excluding the arterial flow into the galenic system, and not occlusion or resection of the "aneurysmal" dilation or malformation of the vein of Galen.

The galenic system of veins consists of the paired, paramedian, and internal cerebral veins located within the tela choroidea of the roof of the III ventricle (the lesser veins of Galen), and the single large anastomotic vein (the great vein of Galen), which receives the inter-

nal cerebral veins and the veins of Rosenthal. Fistulae between the choroidal arteries in the roof of the III ventricle (medial posterior choroidal branches of the posterior cerebral artery) and the internal cerebral veins (lesser veins of Galen) may occur as independent entities. Similarly, inferior retrosplenial, quadrigeminal, and superior cerebellar arteries may establish fistulous communications directly with the great vein of Galen. Unnamed branches of the internal occipital and superior cerebellar arteries may also penetrate the great vein of Galen directly. The terminal (splenial) branches of the pericallosal artery often enter the superior surface of the aneurysmal dilation or malformation (of the vein of Galen) when such an arteriovenous fistula is present.

One may, then, observe an arteriovenous fistula with aneurysmal dilation of (1) only the internal cerebral veins (lesser galenic system), (2) only the great vein of Galen, or (3) the entire galenic system (lesser and greater veins of Galen).

Heart Failure, Arteriovenous Shunting, Thrombophlebitis, and Hydrocephalus

One may observe progressive, intractable, high-output failure without hydrocephalus in the newborn, and communicating or obstructive hydrocephalus with or without high-output failure in the infant and toddler. When the great vein of Galen is either simply dilated or an integral part of the malformation, hydrocephalus may occur as an associated condition secondary to compressive occlusion of the aqueduct of Sylvius by the dilated vein of Galen; as the result of a dramatic increase in venous flow and pressure; and/or as the consequence of progressive thrombosis secondary to thrombophlebitis of the major venous draining structures.

Despite the absence of signs of collicular plate compression in any of the heretofore reported cases of hydrocephalus in children with dilation or malformation of the galenic system, most authors, erroneously in our opinion, attributed the hydrocephalus to compressive occlusion of the aqueduct by the dilated vein of Galen. Nevertheless, it is, in the absence of definite evidence to the contrary, best to consider that this may be a pathogenetic factor. More common, and very much more probable as causes, are the coexistence of dramatic increases in cerebral intravascular flow and pressure along with such clear evidence of progressive venous thrombophlebitis of the dural sinuses and major draining veins. These latter pathological events are associated with (we think pathogenic of) the communicating hydrocephalus so very commonly complicating aneurysmal dilation or malformation of the galenic system.

Confronted with the child, with either a vein of Galen arteriovenous malformation or a vein of Galen aneurysmal dilation, who has ventriculomegaly, one must determine whether the hydrocephalus is pathogenic or simply anatomical. In the event it is pathogenic, the next decision that must be made concerns shunting. Unless there is megacephaly and/or a bulging anterior fontanelle, in an infant, we may not a priori diagnose pathogenic hydrocephalus. Of course, grade III ventriculomegaly [30, 38, 39], the setting-sun sign, a shrill cry, paraparesis, delayed development of milestones, etc., are all clear-cut clinical indications of pathogenic hydrocephalus. On the other hand, distention of the subarachnoid spaces and the fissures, grade I or grade II ventriculomegaly, in the absence of clinical signs of cerebral embarassment, are not indicative of pathogenic hydrocephalus.

Either the arteriovenous malformation or the arteriovenous dilation of the galenic system may give ventriculomegaly. Arteriovenous shunts, especially those involving the centrencephalic system, may cause ventriculomegaly (hydrocephalus) because of shunting of arterial blood, under arterial pressure, into the venous system, thereby increasing the venous outflow pressure. Aneurysmal dilation of the great vein of Galen, associated with these centrencephalic arteriovenous fistulae, deceptively indicates a compressive factor, suggesting to the clinician that the aqueduct of Sylvius is either compressed or occluded, thereby causing obstructive hydrocephalus. This is not the case. Aneurysmal dilation of the great vein of Galen may compress the superior portion of the cerebellum, shifting it downward, thus causing dislocation of the cerebellar tonsils across the line of the foramen magnum (Chiari I malformation), resulting in constrictive hydrocephalus [30, 31]. It is doubtful that simple transforaminal herniation of the cerebellar tonsils in children with anomalies of the galenic system is the sole cause of hydrocephalus. *Rather, it is more likely that the centrencephalic arteriovenous shunting, increased venous outflow pressure, and resultant occlusion of tentorial sinuses represent the true pathogenesis of hydrocephalus in these children.*

In light of the fact that the hydrocephalic process in these children is most likely hemodynamic, there is little reason to suggest that the most desirable approach to managing the hydrocephalus is a ventriculoperitoneal shunt prior to correction of the vascular pathology. Rather, the most rational approach seems to be exclusion of the arteriovenous shunt first. If, thereafter, hydrocephalus persists and is symptomatic, one may safely proceed to a ventriculoperitoneal shunting procedure.

The thrombophlebitic changes are the most insidious, potentially the most damaging, and those which make it mandatory for some of these arteriovenous fistulae to be treated rapidly and surgically: to eliminate the high-flow/high-pressure dynamics, the causes for both the potential cardiac failure *and* the even more damaging cerebral infarction secondary to thromboph-

lebitic occlusion of the major venous drainage pathways. Waiting 6–18 months may be acceptable, and even preferable, in children without evidence of hydrocephalus and some degree (need for digitalization) of cardiac failure. However, either of these pictures of clinical decompensation, and especially the presence of both simultaneously, indicates an urgent situation requiring relatively immediate exclusion of the arteriovenous shunt, whether into a dilated or malformed galenic system.

It is essential to determine whether the lesser or greater galenic system is involved in the fistula and to identify specifically the arterial flow into the fistula. Aneurysmal dilation of the internal cerebral vein, secondary to arteriovenous fistula, is characterized by hypertrophy and increased number of both the anterior choroidal and the middle posterior choroidal arteries. The latter vessels penetrate the tela choroidea of the roof of the III ventricle and shunt into the internal cerebral vein at that point. Branches of the anterior choroidal artery shunt into the choroid plexus along the choroidal fissure. The aneurysmal dilation of the entire internal cerebral vein and retrograde dilation of the terminal and subependymal veins may occur in the absence of involvement of the greater vein of Galen. The retrograde filling may also extend into the terminal and subependymal veins.

The choroid plexuses and subependymal veins are almost invariably distended and both the anterior and posterolateral choroidal arteries are hypertrophied as a result of the increased flow into the centrencephalic venous system. It is generally difficult to identify the posteromedial choroidal arteries. The hypertrophy of the choroidal vessels and the choroid plexuses of the lateral ventricles along with the distention of the subependymal veins (which contain arterial blood) are additional factors that contraindicate insertion of a shunting system prior to exclusion of the vascular anomaly.

The fistulous communication may exist exclusively between arterial branches of the posterior circle and the great vein of Galen. In this instance, only the great vein of Galen dilates, without retrograde filling of the lesser vein, and the posterior communicating and posterior cerebral arteries may be remarkably dilated along with the posterior choroidal arteries. Similarly, the superior cerebellar arteries may shunt directly into the great vein of Galen. In these conditions, there is a complete aneurysmal malformation of the vein of Galen or of the galenic system.

There is no general rule concerning degree of shunt, aneurysmal dilation of the lesser galenic system, aneurysmal dilation of the greater galenic system, aneurysmal malformation of the entire galenic system, presence of hydrocephalus, and age. In fact, the newborn may have progressive, intractable, high-output failure or aneurysmal dilation of the great vein of Galen in association with arterial shunting into it. Therefore, in considering the congenital anomaly "arteriovenous malformation of the galenic system"/"aneurysmal dilation of the great vein of Galen," it is advantageous to distinguish between the onset of signs and symptoms in the newborn and those at a later age, since the cardiac changes, caliber of the caroticovertebral systems, arterial tributaries to the galenic system, and size of the venous outflow structures are quite different.

In children diagnosed when newborn, heart failure is invariably present, and intractable high-output failure is the most common type, so that the arteriovenous fistulae of the galenic system are generally diagnosed by the cardiologist who is evaluating the child for the cause of the heart failure at birth. The caroticovertebral systems are widely dilated, and the major tributary arteries to both the lesser and greater galenic veins are generally perforating branches of the carotid and basilar arteries, although the pericallosal artery may occasionally shunt directly into the galenic system at the splenium. The middle cerebral system distal to its perforators does not shunt into the malformation. The lesser vein of Galen (internal cerebral vein) very often is widely dilated and opens directly into the great vein of Galen. The great unknowns are: do the branches coming from the posteromedial and posterolateral perforating complexes pass through the midbrain and posterior floor of the III ventricle? around it? are they vessels "dedicated" to the fistula or hypertrophied perforators?

Endovascular Occlusion or Open Surgery

Though there are as yet no hard and fast rules that permit one to determine whether an individual patient should be treated neurosurgically or with endovascular interventional neuroradiology, it appears at present that the large majority of children with arteriovenous anomalies involving the galenic system are being treated with endovascular techniques. Until approximately 1985, however, open surgical procedures or no operative treatment were the alternatives. Since that time, the truly dramatic progress in interventional neuroradiology has permitted some centers to accumulate truly remarkable experience [33, 40]! The only reports of extensive operative neurosurgery experience hold material treated prior to 1980, and group arteriovenous malformations and aneurysmal dilations of the galenic system together [32, 41].

Consequently, it is not presently possible to compare the two approaches, but it appears that the pediatric neurosurgical world has, with definite exceptions, chosen to recommend endovascular management. Those infants and children with either malformation or dilation involving the galenic system should initially be evaluated for endovascular treatment, though children

with the pouch-like dilation of the medial prosencephalic vein may successfully and with extraordinarily low morbidity be treated with operative neurosurgery.

Treatment of vein of Galen aneurysm remains a controversial subject. First of all, it is a rare malformation and apart from some large series in the literature the vast majority of the published material is case reports.

Timing of treatment is still a controversial issue. In one of the articles reviewed, the authors were able to collect 19 [28] cases from the current literature, plus 2 of their own, in which spontaneous thrombosis of the malformation had taken place.

Furthermore, it must still be decided just which is the best treatment for these lesions, if a single treatment of choice may be identified. Hydrocephalus in this pathological entity is an important issue. It may be triventricular hydrocephalus, which appears early in the course of the disease and is caused by the mass effect of the lesion and blockage of the CSF circulation through the aqueduct of Sylvius. However, one may also see hydrocephalus secondary to increased venous pressure and retrograde flow of arterial blood into the venous system. Embolization alone may solve the problem, it may cause it, or it may complicate it. Later in the course of the disease it is possible to observe the appearance of tetraventricular communicating hydrocephalus. These are serious problems which represent a major drawback to embolization, one which is underestimated in the current literature and understated by authors using embolization techniques.

The pathophysiology of tetraventricular communicating hydrocephalus is to be sought in the complete occlusion or failure to develop of the vein of Galen, which causes an inverted flow, venous engorgement, and subsequently venous hypertension. Furthermore, in newborn the venous compartment plays a fundamental role in CSF drainage because of the immaturity of the arachnoidal villi. This hydrocephalus is responsible for the severe intellectual impairment and psychomotor retardation which affect most of the subjects treated by embolization. From a theoretical point of view, better results may be achieved by trying to restore an anatomically correct circulation, leaving a channel in the vein of Galen aneurysm, to preserve venous drainage. Hernesmiani [26] tried this using an encircling clip, but eventually he was unable to demonstrate flow in control angiography. This may be due either to the technical difficulties of the procedure or to the tendency of large sinuses and veins to thrombose spontaneously.

Using microvascular technique, and with the refinements introduced in instrumentation, surgery in deep fields such as the falcotentorial junction is no longer prohibitively hazardous. It is associated with no more intra- or postoperative complications than embolization – a fact which warrants emphasis. The surgical risks of such a procedure may be lessened by detailed angiography. This has to show clearly all the afferent vessels of the malformation, so that the correct surgical approach may be chosen. An approach which is correctly planned reduces excessive manipulation of the malformation and thus also the risk of bleeding, and the retraction one has to use on the brain to reach the pathological vessels.

Surgical risk is related to age: the younger the patient, the higher the risk. Cardiomegaly is another factor significantly influencing the surgical and embolization risks. There is unanimity that newborns with intractable high-output failure are untreatable.

The risk of hemorrhage either during the surgical or embolization procedure, or in the postoperative period, is high; furthermore, these patients do not tolerate even small amounts of loss of blood, and it is not possible to work at a low systemic pressure due to cardiac decompensation.

The close vicinity of the malformation to important structures such as the thalami, the mesenchephalon, and the deep venous drainage add another challenge to this difficult surgery. However, as yet there has been no "real-time comparison" between endovascular embolization and surgical isolation of the shunt: during the past 10 years, when the most recent and dramatic advances in microsurgical technique have occurred, the treatment method used has been almost exclusively embolization by interventionists. This is unfortunate, since only the vascular component of this complex cerebrovascular, anatomophysiological lesion has been focused upon. The parenchymal and intraventricular hemorrhages during treatment, the varying stages (degrees) of hydrocephalus, and the creation of a solid mass at the location of the aneurysmal dilation of the vein of Galen are all negative aspects of embolization which must be factored into results that are being classified as "good." It is to the advantage of the children suffering from these malformations that a fresh, surgical, approach to their treatment be undertaken, *especially in light of the fact that the best results being obtained today by endovascular treatment are identical to those reported for direct surgical treatment 15 years ago.* Until now, surgeons have generally deferred to the interventionists. However, if one reviews the literature carefully [42–53], it becomes clear that the results reported by interventionists during the past 5 years are not clinically better that those reported by operating surgeons previous to 1982. Considering that the major advances in microsurgical instrumentation and techniques have been made since 1982, one must reconsider the advantages of surgery.

Anatomic Classification and Surgical Anatomy

In infants and children, arteriovenous malformations of the galenic system may be subdivided *anatomically* with regard to surgical strategy and technique into three categories: superior, inferior, and posterior. Such

a classification permits the surgeon to plan appropriately the operative approach. The category is "superior," when the tributary arteries enter the superior surface of the anomalous galenic system; "inferior" when they enter its ventral surface; and "posterior" when they course around the aneurysmal dilation to enter at the posterolateral tentorial surface. This classification permits the surgeon to plan a biparietal craniotomy approach along the falx cerebri for the *superior* group; a two-stage, bilateral temporal craniotomy and supratentorial approach to the tributaries on either side in the *inferior* category; and a suboccipital craniotomy with infratentorial/supracerebellar approach in the *posterior* category.

In either event, the efferent (tributary) arteries are identified, clipped, and transected, as far from their entry into the aneurysmal dilation as possible. After all tributaries have been occluded and cut, the anomalous galenic system is left as is, without any attempt to diminish its size (either by inserting imbricating sutures or opening and reconstructing it).

Superior Category (Figs. 11.27, 11.28)

In the superior category, the pericallosal and posterior cerebral arteries enter the aneurysmally dilated anomalous lesser and/or greater vein(s) of Galen along median and paramedian planes. Careful study of the lateral and half-axial arteriographic projections permit identification of the point of entry of the tributaries into the aneurysm and, most importantly, the planning of the operative procedure. If all afferent vessels enter the aneurysm in the median (sagittal) plane, a unilateral parietal flap suffices for exposure and surgical access to tributaries from both the right and left pericallosal and posterior cerebral systems. If, however, the tributaries enter in a parasagittal or superior lateral plane, bilateral parietal flaps are necessary: it is not possible to work safely and effectively along the opposite superolateral surface of the aneurysm. Tributaries must be dissected over a distance of at least 6 mm. Consequently, those coming from the posterior cerebral system and the medial posterior chorioidal artery are coursing inferosuperiorly, nestled within the wall of the aneurysmal dilation. One cannot expose an adequate length by working over the dome of the aneurysm, so that one is obliged to be in a position to expose the lateral surface without compressing or excessively retracting the dome. If a biparietal flap is indicated, then it is best to do this in one stage. Access to the corpus callosum and anomalous galenic system is facilitated somewhat by the fact that children with arteriovenous fistulae involving the galenic system generally have a paucity of bridging cortical veins: most of the arterial blood is being shunted into the fistulae.

If a biparietal flap is reflected in an infant, one should leave the superior sagittal suture intact, cutting the parietal bone approximately 3–4 mm parallel and lateral to the suture. This affords protection to the SSS and permits the surgeon to prevent kinking (and, consequently, diminished flow) of the sinus during the operative procedure. If a toddler or juvenile is being operated on, the biparietal flap should be such as to leave a strip of bone over the superior sagittal sinus.

After reflection of the flap(s) and opening of the dura, one may observe a cerebrum which has the appearance of a ball of vessels containing oxygenated blood (in the newborn who is in high output failure, as illustrated in Figs. 11.12, 11.23 C), or a normal-appearing cerebrum with few cortical veins and arteries (the toddler and juvenile). The parasagittal surface of the parietal lobe is separated from the superior sagittal sinus and Telfa lain over it for protection as the falx is identified and then the parietal lobe retracted laterally. The pericallosal cistern is generally obliterated because of the posterior displacement and compression of the corpus callosum by the aneurysm. One notes immediately the remarkable size of the pericallosal artery(ies). These vessels are dissected from the corpus callosum and covered with fluffy cotton, which is kept moist, as the splenium and/or body of the corpus callosum is opened in the midsagittal plane. A Penfield #4 dissector is used to split the corpus callosum, since this instrument does so bloodlessly and also allows the surgeon to feel the underlying aneurysm. Once the corpus callosum has been sectioned, it is stripped from the aneurysm with the use of wet fluffy cottons, exposing the insertion of the pericallosal tributaries into the anomalous galenic aneurysm. Each tributary should be dissected over as long a distance as possible from its point of entry into the aneurysm, and then covered with Telfa. Once all pericallosal tributaries have been identified and dissected, one moves lateralward along the superior surface of the aneurysm to identify the tributaries coming from the medial posterior choroidal and posterior cerebral systems. This approach should be along the homolateral side, so that exposure of the tributaries, for example, coming from the right medial posterior choroidal artery, should be approached along the right side of the falx cerebri. This gives excellent visualization of the superior lateral surface of the aneurysm and provides the surgeon adequate access to these tributaries along a sufficient length of vessel to permit dissection, ligature or clipping, and transection. The entire superior lateral surface must be inspected for tributaries.

After all tributaries, on both sides (median and paramedian along the homolateral side, paramedian on the contralateral side), have been isolated, one proceeds to ligature or clip and transect. Depending upon the size of the tributaries, either small or medium hemoclips are applied. The recommended technique for application is to bring the jaws of the hemoclip over the surfaces of the afferent vessel, and then to close them very gradually, taking care to observe the jaws of the applica-

tor as this is done to be sure that the clip closure does not skew. This could result in cutting rather than occluding the tributary. It is preferable to apply two clips, separated from one another by 1 mm, at the aneurysmal extremity of the vessel and then two more, separated from one another by a similar distance, approximately 3 mm away. The vessel is cut at the center of this 3-mm distance, allowing approximately a 1-mm stump on either end. This clipping technique provides maximum assurance of vascular occlusion.

Inferior Category (Fig. 11.29)

The inferior category is characterized by tributary branches from the posterior cerebral and posterior choroidial arteries entering the anomalous galenic system along its inferior lateral surface. Consequently, access to these tributaries is obtained through a temporal flap, elevating the temporal lobe, and entering the ambient cistern after exposing the rim of the tentorial foramen. This permits exposure of all afferent vessels within the pia-arachnoid of the ambient cistern, so that they may be occluded proximal to entry into the aneurysm. The temporal lobe must be elevated from the tentorium, so that the hippocampal gyrus and ambient cistern may be exposed. Since the vein of Labbé is so very variable in anatomical location and size, no generalities concerning its management exist. However, if one follows the tentorium from the petrous apex to the ambient cistern, gradually elevating the temporal lobe as one proceeds, correct orientation is maintained. At the rim of the tentorial foramen, the ambient cistern bulges prominently. Once opened, there is a gush of cerebrospinal fluid and the IV cranial nerve comes immediately into view, with the tributaries to the anomalous galenic system aneurysm being located deep to this cranial nerve. These tributaries are branches of the posterior cerebral system.

Though the inferior supply to galenic system aneurysms is almost invariably bilateral, the degree of dilation and number of tributary vessels vary from one side to the other, so that no rules apply. Consequently, one may find it necessary to operate bilaterally. When this is the case, surgery should be staged, with the most extensive system of tributaries being operated on first, and with a period of 3–5 months intervening.

After the temporal flap has been reflected and the dura opened, the temporal lobe is elevated until the ambient cistern is identified and opened. This gives egress to cerebrospinal fluid and brings the superior cerebral and medial posterior choroidal arteries into the center of the operative field. One may follow them posteriorly, superiorly, and medially in order to identify their tributaries to the galenic aneurysm. It is best to orient oneself by exposing these vessels along the lateral surface of the pontomesencephalic junction and then to coagulate them along their course toward the aneurysm.

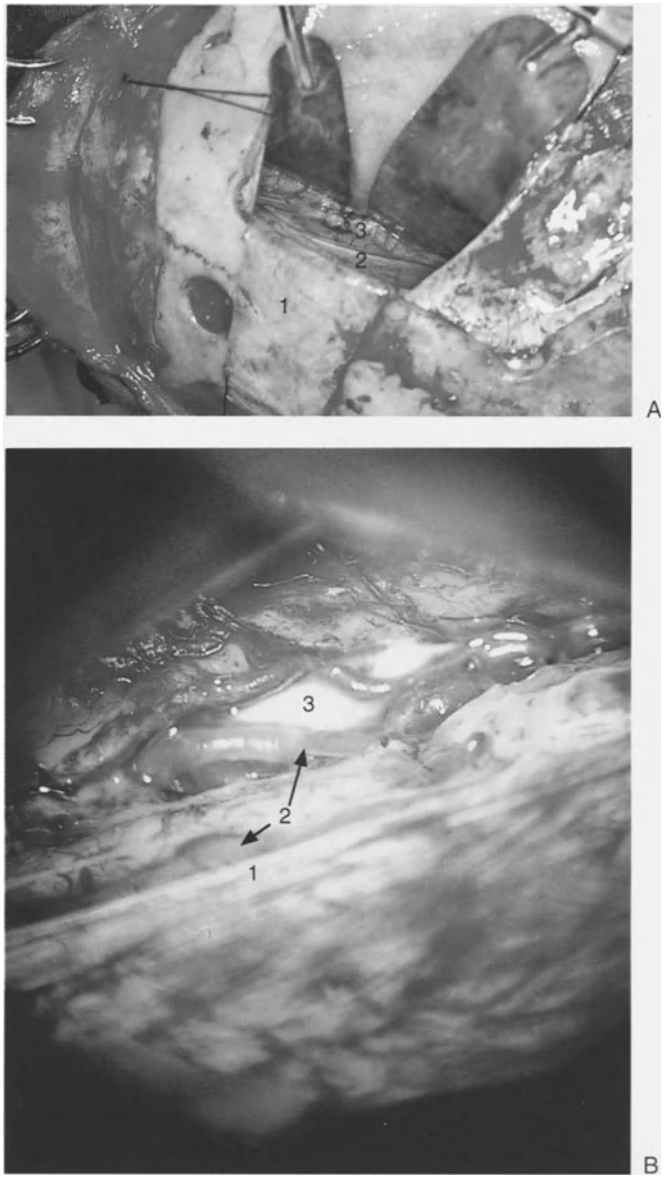


Figure 11.27. Parasagittal approach to arteriovenous malformation of the galenic system. (A) A parietal bone flap has been reflected, the dura (*1*) sewn over the sagittal plane, the falx cerebri (*2*) and pericallosal cistern (*3*) exposed. (B) The inferior sagittal sinus (*1*), pericallosal feeders to the malformation (*2*), and underlying corpus callosum (*3*) after the pericallosal cistern has been opened. (C) Dissection of all pericallosal feeders to the malformation is followed by application of hemoclips to the vessels, prior to transecting them. (D) After the pericallosal feeders have been dealt with, the corpus callosum is split, and the underlying arteriovenous malformation exposed. In this child, there was a combined aneurysmal dilation of the lesser (*1*) and greater (*2*) galenic veins. One sees branches from the posterior cerebral artery (*3*) coursing over the great vein of Galen aneurysm, to enter the lesser vein of Galen aneurysm. (E) A Sundt clip (*1*) has been applied to the redundant lesser vein of Galen, a Heifitz clip (*2*) to the last feeding vessel to the greater vein of Galen. All arterial feeders to the malformation have been occluded. The author does not recommend imbricating the aneurysm of the vein of Galen (*3*). ►

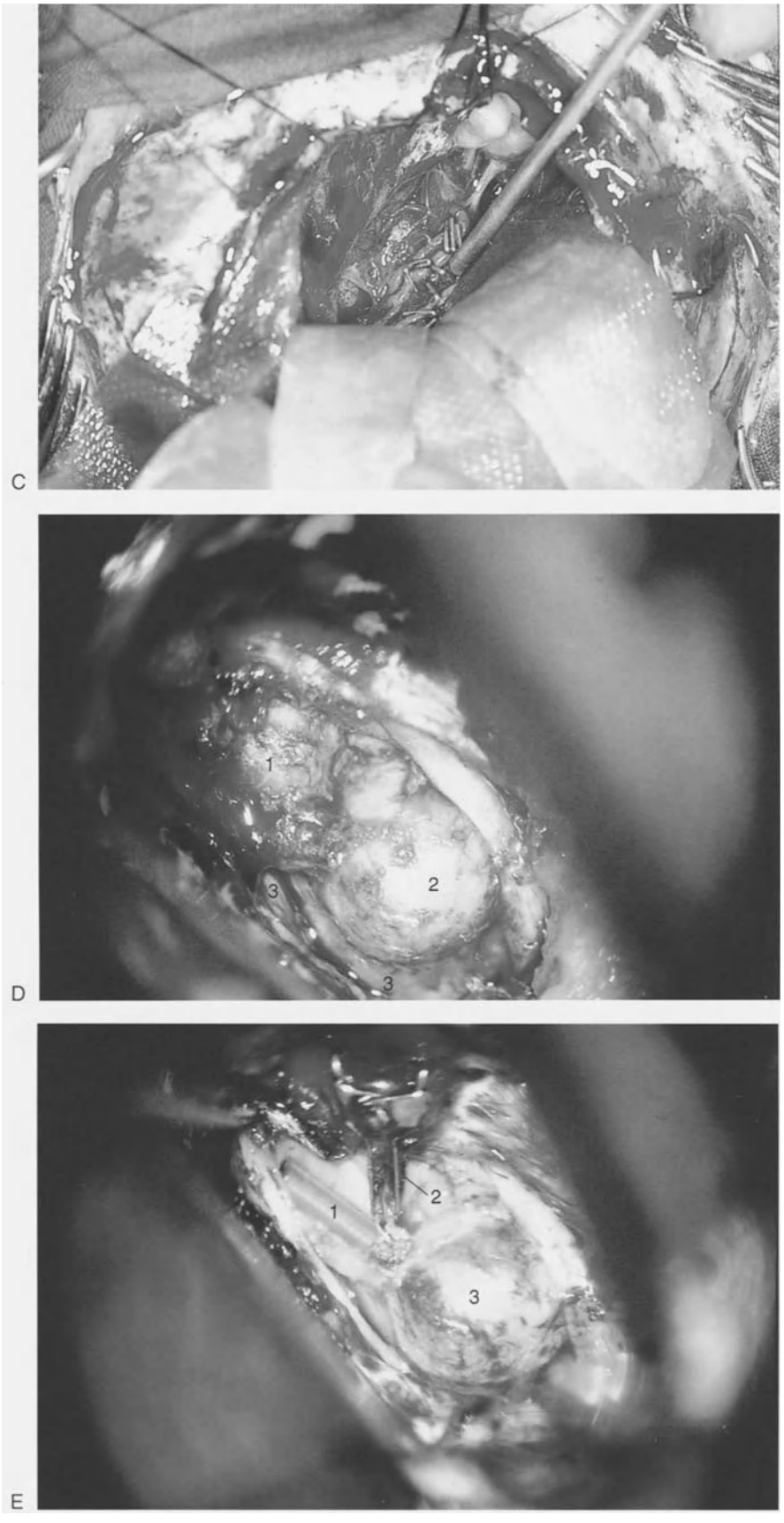


Figure 11.27. Legend see p. 388.

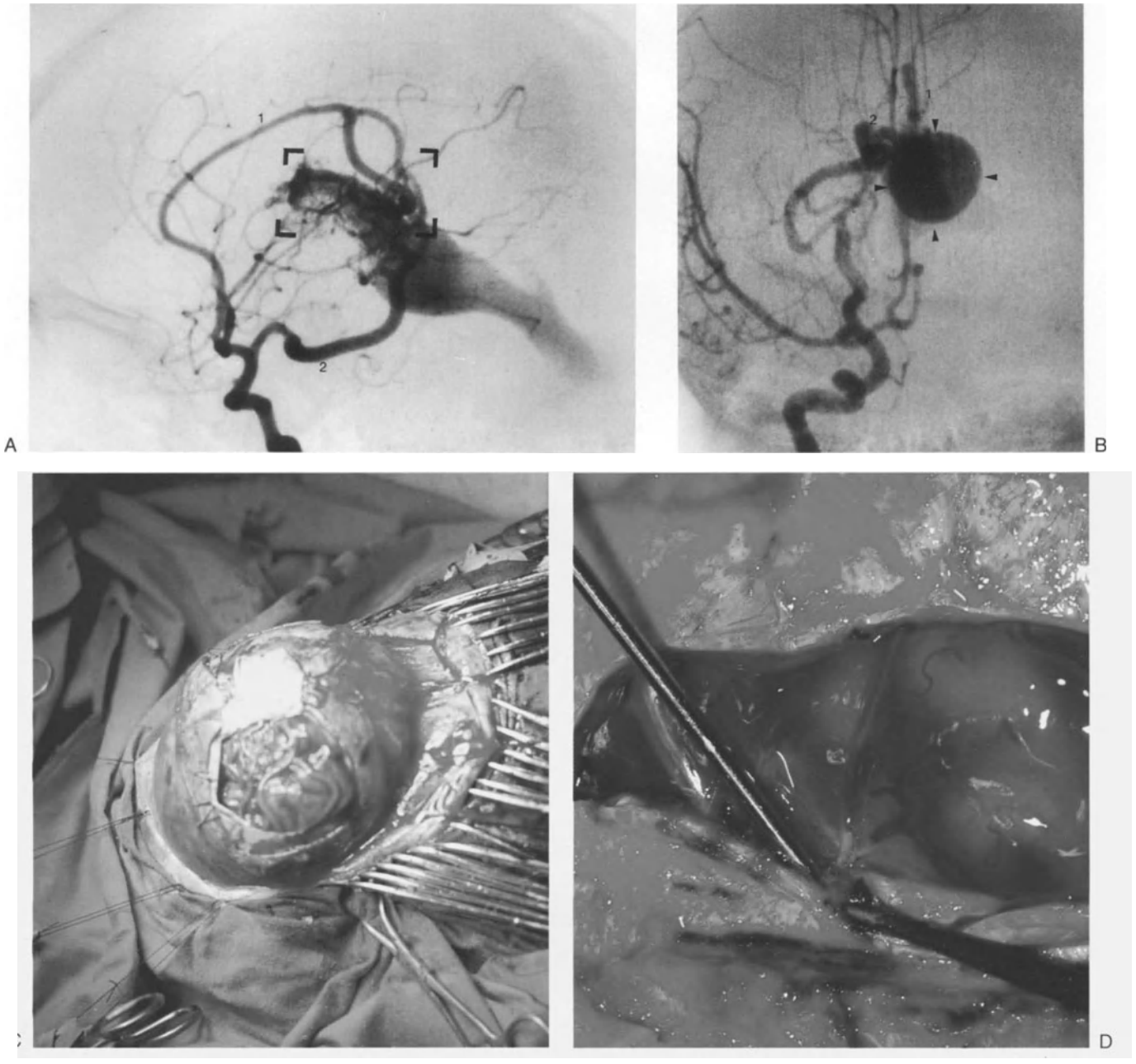


Figure 11.28. (A) Both the pericallosal and posterior cerebral systems are tributary to the lesser and greater veins of Galen in the "superior category." The entry into the galenic system is along the superior surface (*box*). (B) The balloon-shaped vein of Galen (*arrowheads*) is filling from the pericallosal artery and branches of the posterior cerebral system, both of which may be seen to enter the superior surface of the galenic system. The parasagittal approach to occlusion of the feeding vessels is recommended in this example of the "superior category." (C) In the newborn in high output failure during the first 24 h of life, the entire cerebrum consists of anomalous vessels having the appearance of a bag of worms, leading one to understand how difficult operating these children is, and to predict that the outcome offers little hope for normal cerebral function. This "bag of worms" appearance is herein illustrated. (D) One has an opportunity to appreciate how adherent the minute bridging vessels are, all of which necessitate coagulation and transection. The underlying arachnoid and subarachnoid space have an appearance of a fresh subarachnoid hemorrhage, though this has not occurred. (E) see p. 391.

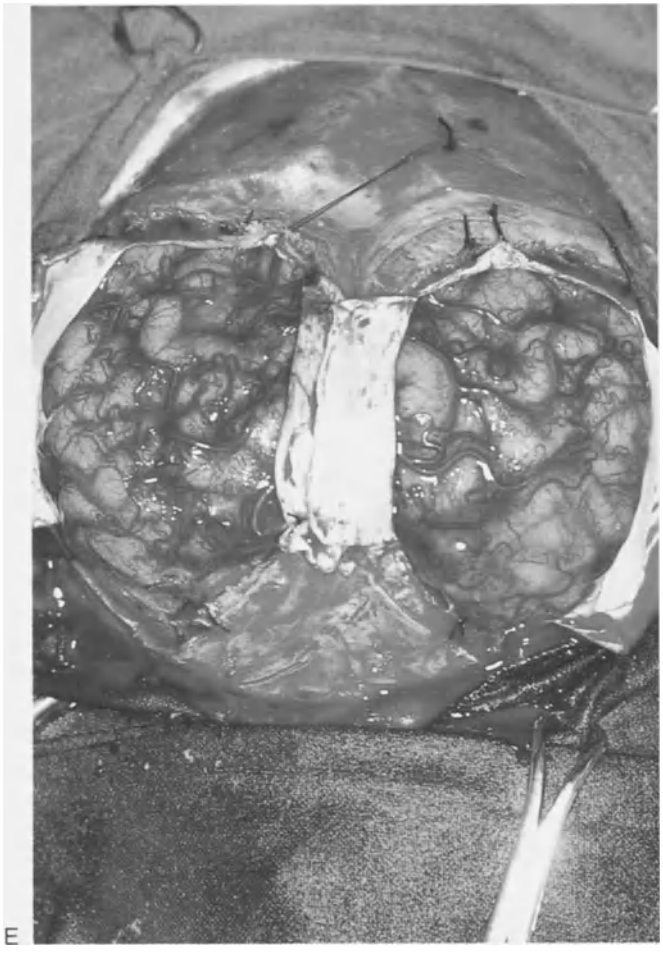


Figure 11.28. (E) A biparietal flap has been reflected. One notes the *Moya Moya* appearance of vessels of both cerebral hemispheres, something which does not occur in children under 2 years of age with the exception of newborns in progressive, high output failure.

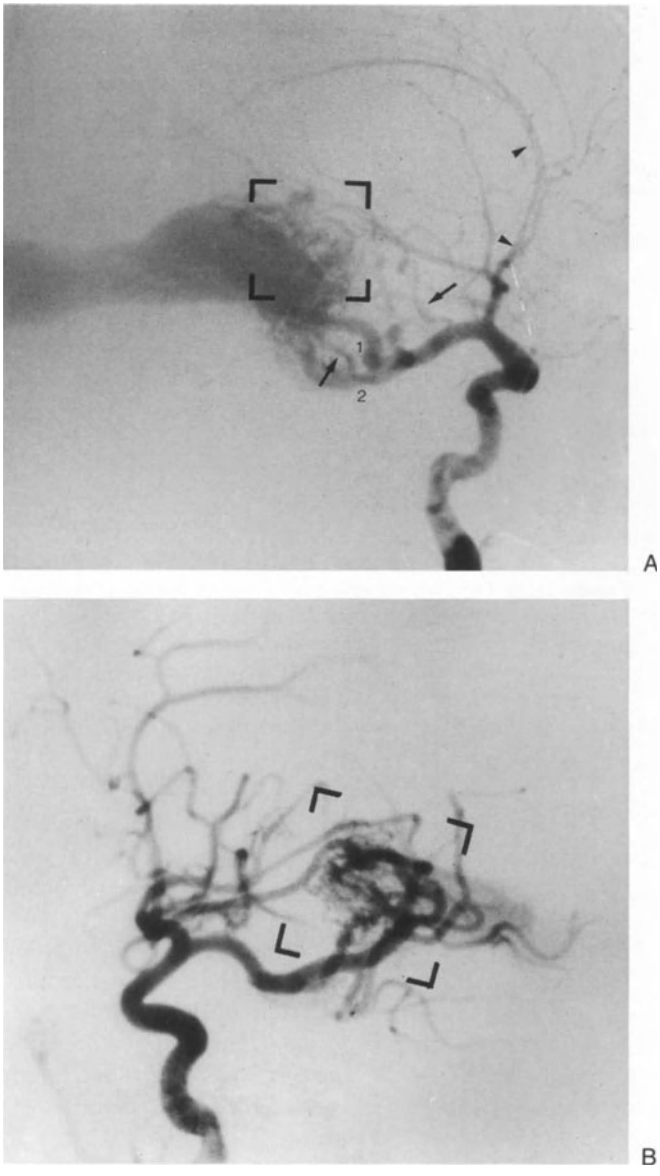


Figure 11.29. (A) The medial (*1*) and lateral (*2*) posterior choroidial arteries, as well as long and short perforating branches of the posterior cerebral system (*arrows*), course around the brainstem and enter the inferior surface of the galenic system in this example of the "inferior category." There is a rather diffuse, racemose component (*box*). The bowing of the pericallosal artery (*arrowheads*) indicates the presence of hydrocephalus. (B) Entry into the inferior surface of the galenic system and some of the racemose component are illustrated (*box*) in this second example of the "inferior category."

The larger tributaries should be dissected along their entire course from the major nourishing vessel to the aneurysm. Generally, this system of tributaries has a more fragile vascular wall than those entering the superior surface of the aneurysm: more often pia-arachnoidal malformations. Some afferent vessels, however, have a rather rigid wall. The fragile vessels need only be coagulated and transected. Those vessels with a relatively rigid wall must be dissected over an extensive (5–7 mm) area, doubly ligatured or clipped on either end, and transected.

Posterior Category (Figs. 11.30, 11.31)

When the tributary vessels to the arteriovenous fistulae involving the anomalous changes of the galenic system enter the aneurysm along its posteroinferior surface, the surgical approach is through the posterior fossa, reflecting a superior cerebellar triangle flap and approaching the feeders along the infratentorial, supracerebellar plane. This approach provides excellent exposure of the inferior medial and inferior lateral surfaces of the aneurysm, at the point of entry of the afferent vessels.

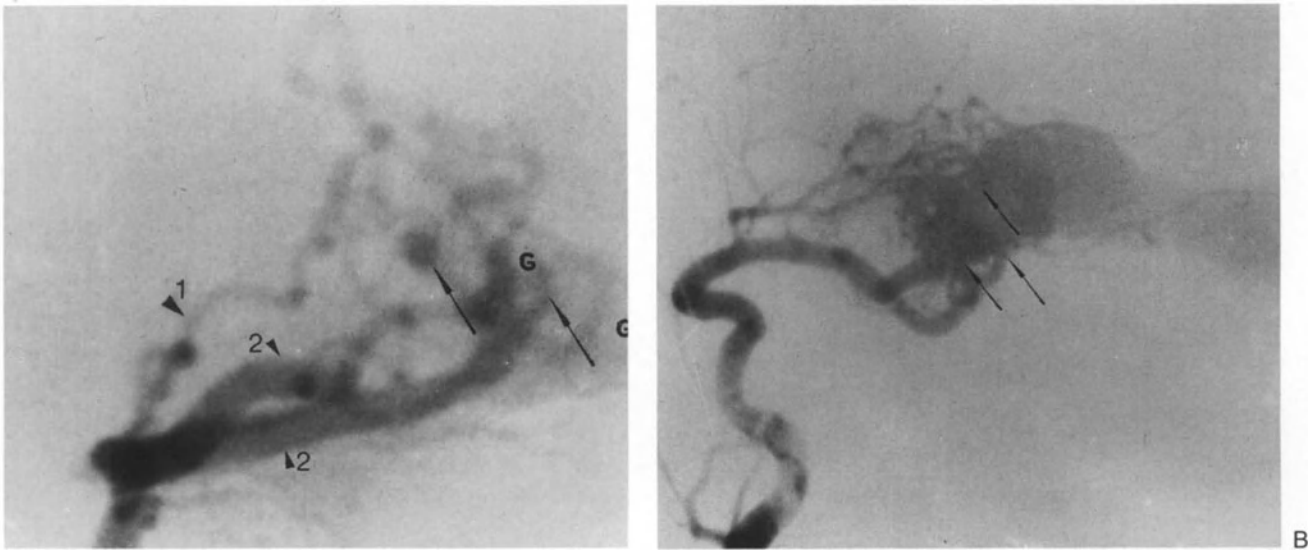


Figure 11.30. (A) The hypertrophied interpeduncular perforating branch (*1*) of the basilar fundus is passing through the brainstem to gain access to the quadrigeminal cistern. The remainder of the afferent vessels (*2*) to the aneurysmal dilation of the great vein of Galen (*G*) course almost horizontally within the ambient cistern, around the brainstem, and over the quadrigeminal plate. They are entering the great vein of Galen along the posterior surface (*arrows*). (B) Posterior entry (*arrows*) of the afferent branches to the racemose component of the arteriovenous malformation and a bulbous dilation of the great vein of Galen.

The significant anatomical characteristics are an almost complete absence of bridging tentorial veins, running from the superior surfaces of the cerebellar hemisphere and vermis to the transverse sinus and tentorium. The superior cerebellar cistern is often surrounded by thickened arachnoid, as is the quadrigeminal cistern. Both of these are densely adherent to the anomalous galenic aneurysm superiorly, the culmen monticuli of the cerebellar vermis posteroinferiorly, and the dorsal pontomesencephalic surface anteroinferiorly. There is seldom a true straight sinus!

The great vein of Galen (or an anomalous vessels in its stead), converted into an enormous aneurysm, fills the entirety of the tentorial opening at its most posterior and superior surface, so that one sees only this structure after the superior cerebellar and quadrigeminal cisterns have been opened. In fact, the aneurysmal dilation of the anomalous great vein of Galen fills the tentorial opening posteriorly, at times compressing the collicular plate as it is displaced anteriorly. The ambient cistern borders the galenic aneurysm on either side, filling the interval between this latter structure and the tentorial edge. Within the ambient cistern are located the posterior cerebral artery and its tributaries to the galenic aneurysm; superior lateral to the ambient cistern are the isthmus of the hippocampal gyrus and the hippocampal formation.

After a superior cerebellar triangle free bone flap has been reflected, the dura is exposed and a single trapdoor dural opening is made. Neither the transverse sinus nor the torcular Herophili should be exposed. The superior bridging veins are identified, coagulated, and transected. This allows the superior surfaces of the cerebellar hemisphere and vermis to fall inferiorly, so that one may expose the superior cerebellar cistern, open it, and then expose the quadrigeminal cistern. Once this is exposed, the aneurysm falls immediately into the operative field. The tentorial edges are then identified and, subsequent to this, the ambient cisterns opened, exposing the major trunks of the posterior cerebral arteries and their tributaries to the aneurysm. These tributaries nestle within the aneurysm, so that it is necessary to dissect each from the aneurysm, using blunt dissection to separate one from the other, and taking care to identify the point of entry of tributary into aneurysm. Generally, there are afferent vessels coming from the internal occipital artery and the quadrigeminal branches of the posterior cerebral artery. These vary in number and size. After the tributaries have been identified, and dissected, bilaterally, they are double-clipped and transected, using the same technique as for the superior and lateral categories. One will note gradual diminution in volume and distention of the aneurysm as each set of tributaries are occluded. The aneurysm is not opened or imbricated (Figs. 11.32–11.36).

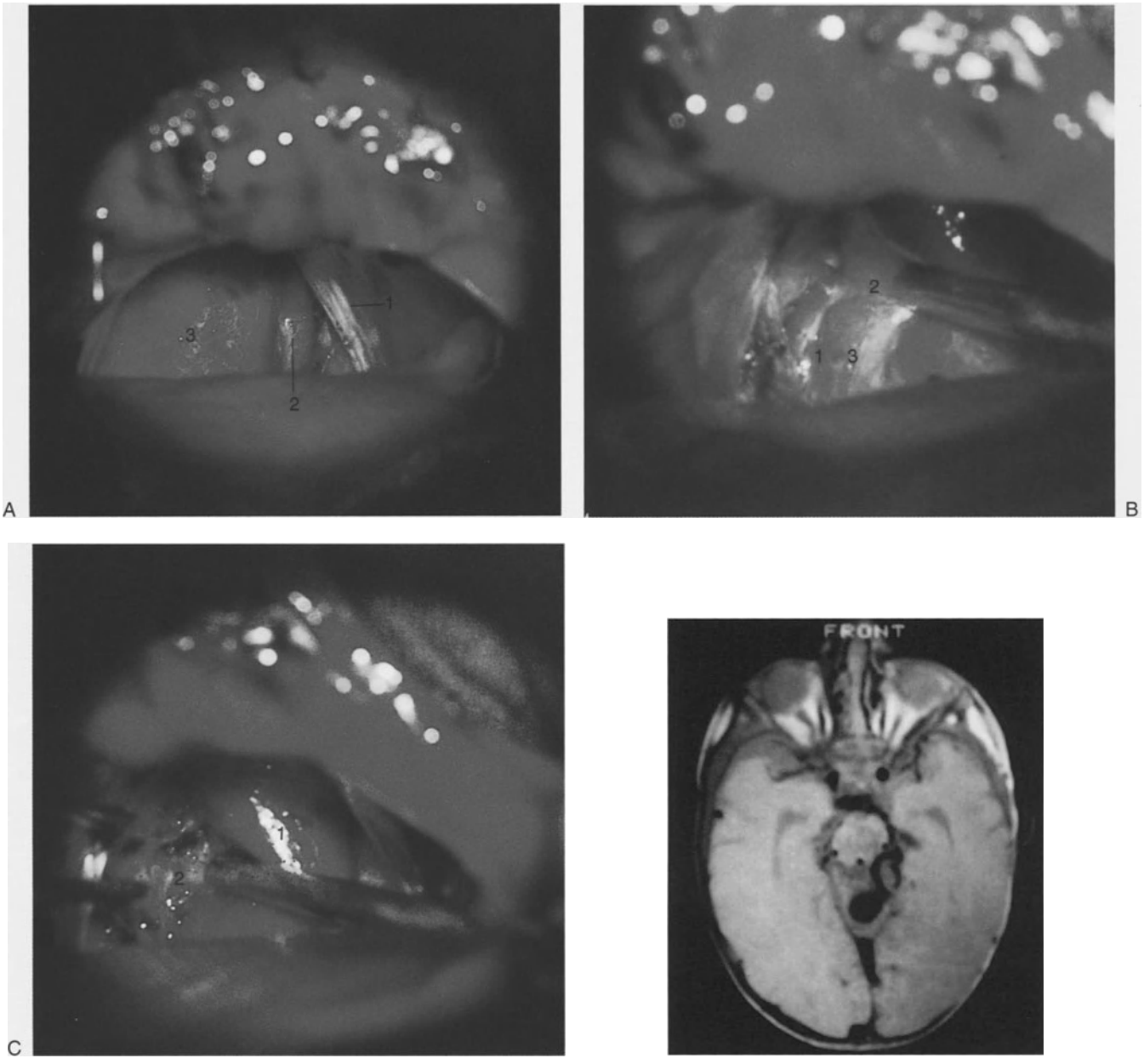


Figure 11.31. Suboccipital approach to arteriovenous malformation of the galenic system. (A) One may identify the right tentorial edge (*1*), and posterior cerebral feeder (*2*) entering the aneurysmal dilation of the great vein of Galen (*3*). (B) The posterior cerebral feeder (*1*) is being dissected, as adventitia (*2*) from over the great vein of Galen (*3*) is reflected to the contralateral side. (C) The great vein of Galen (*1*) has been freed completely from the posterior lateral feeder (*2*), so that the point of entry of this latter vessel into the former may be identified, freed, and occluded.

Figure 11.32. Arteriovenous malformations of the galenic system. This axial projection reveals the dilated vein of Galen posterior to the brainstem, and a large medial posterior choroidal feeding vessel. This patient was 34 years old at the time this MRI study was performed. He had remained asymptotic throughout his life, and this was an "incidental" finding.

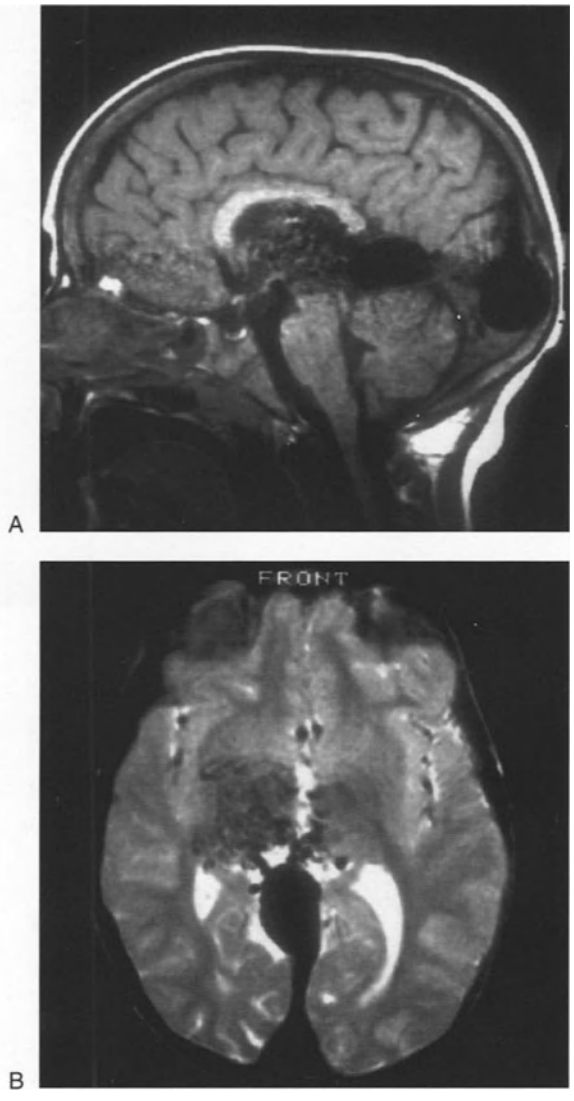


Figure 11.33. Arteriovenous malformation of the galenic system. In (A) a sagittal 600/15 image, one sees immediately the abnormal dilation of the torcular Herophili, and the pathological dilation of the vein of Galen which is draining directly into it. The culment monticuli of the cerebellum is depressed by the venous dilation, and the cerebellar tonsil is displaced across the line of the foramen magnum. In (B), an axial SE 2400/90 image, one sees flow void along the right transverse sinus and within the torcular Herophili. Note also the multitude of vessels in the right thalamus, a result of arteriovenous shunts.

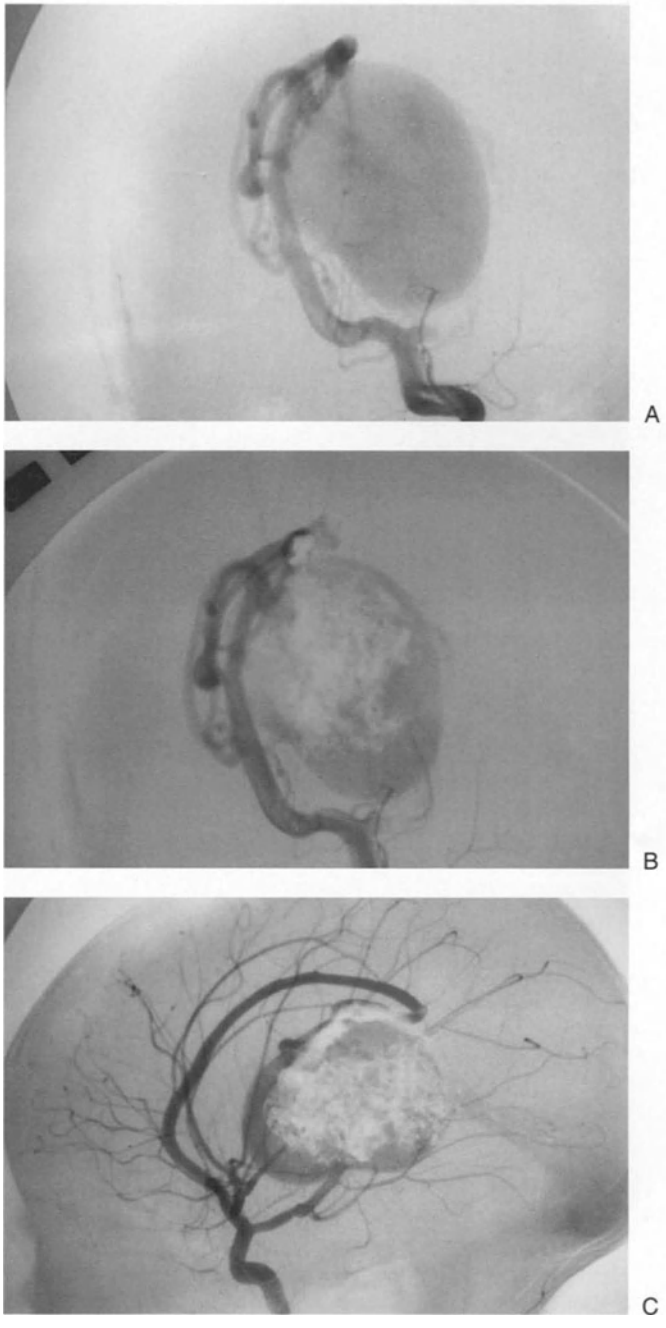


Figure 11.34. Arteriovenous malformations of the Galenic system. (A) This pretransarterial embolization reveals the enormous dilation of the arteriovenous malformation. (B) This is an arteriographic study of the same malformation, in the same projection immediately after transarterial embolization. It reveals clot formation within almost the entirety of the aneurysmal transformation of the galenic vein. (C) This is the lateral arteriogram of the same patient, immediately after transarterial embolization, revealing the clot formation and some persistent shunting into the straight sinus and torcular Herophili.

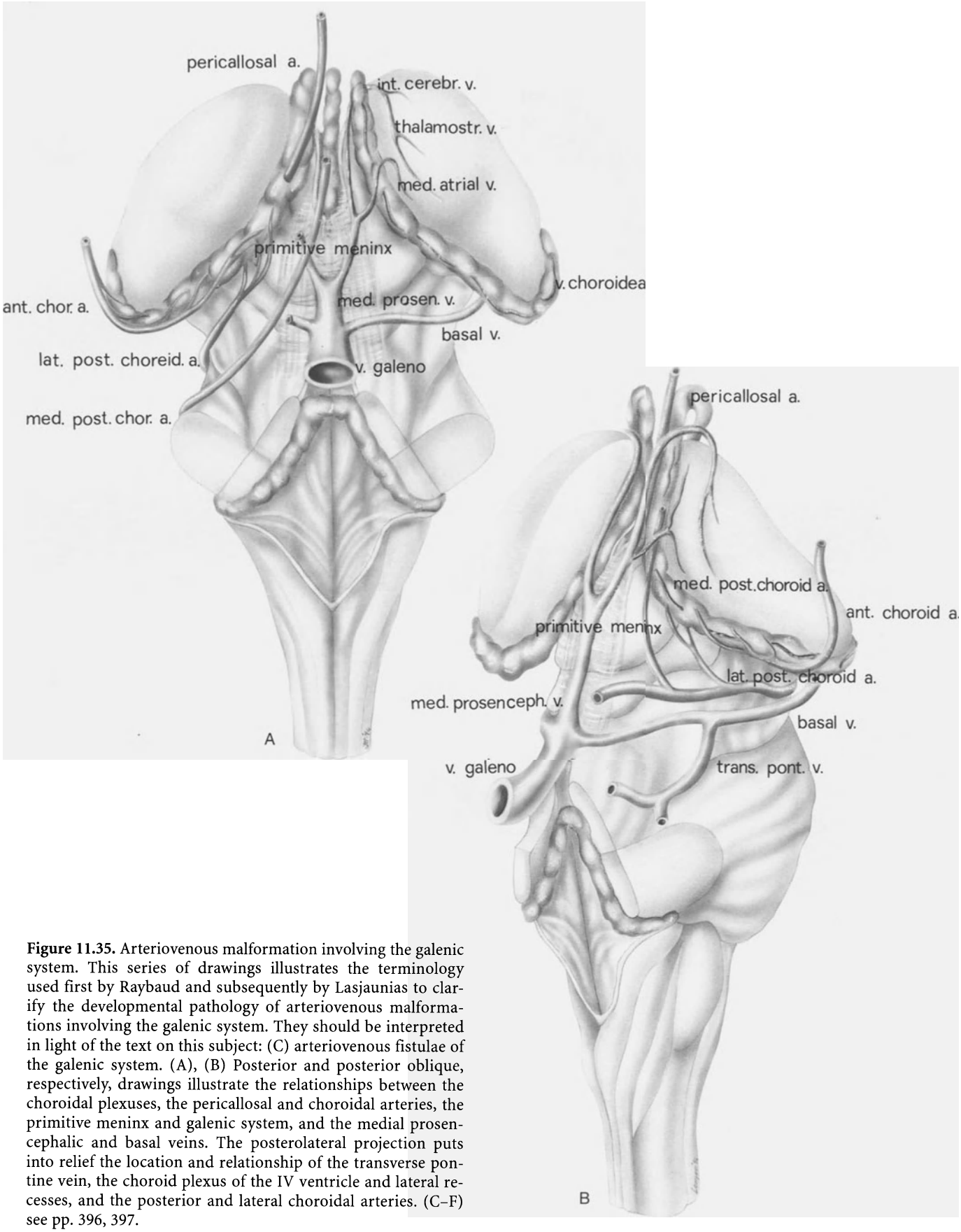
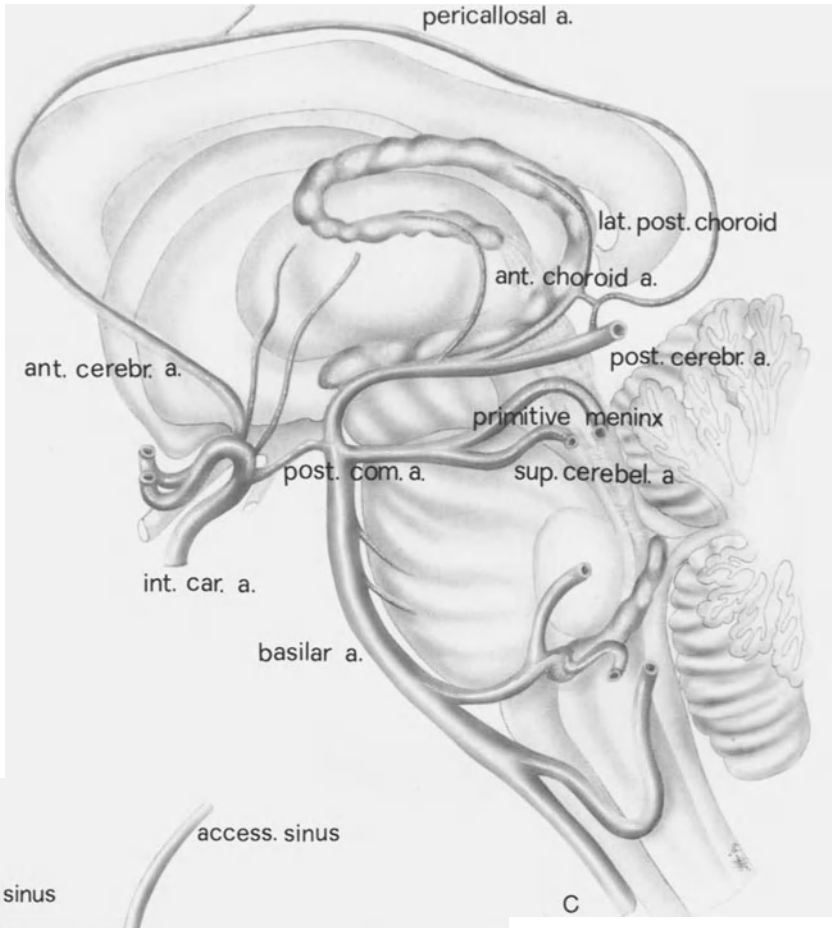
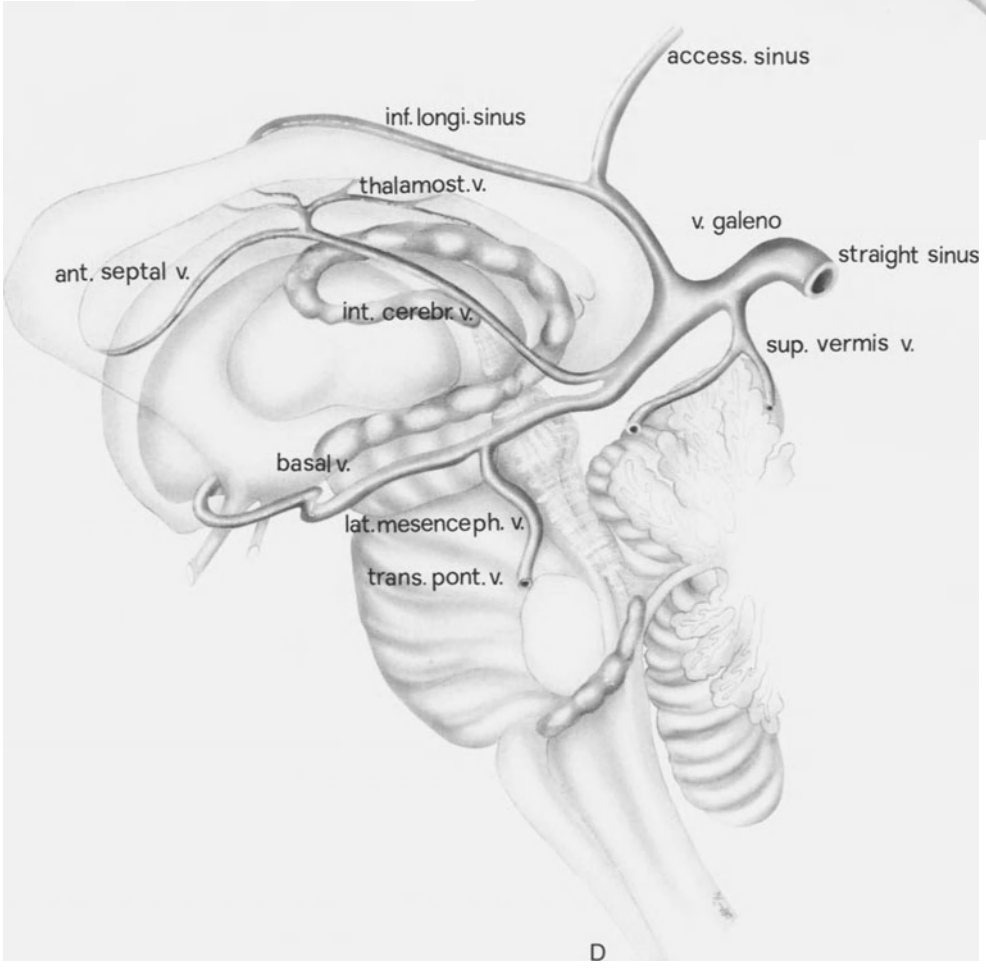


Figure 11.35. Arteriovenous malformation involving the galenic system. This series of drawings illustrates the terminology used first by Raybaud and subsequently by Lasjaunias to clarify the developmental pathology of arteriovenous malformations involving the galenic system. They should be interpreted in light of the text on this subject: (C) arteriovenous fistulae of the galenic system. (A), (B) Posterior and posterior oblique, respectively, drawings illustrate the relationships between the choroidal plexuses, the pericallosal and choroidal arteries, the primitive meninx and galenic system, and the medial prosencephalic and basal veins. The posterolateral projection puts into relief the location and relationship of the transverse pontine vein, the choroid plexus of the IV ventricle and lateral recesses, and the posterior and lateral choroidal arteries. (C–F) see pp. 396, 397.

Figure 11.35. (C) and (D) are lateral drawings which assist the reader to visualize the arterial (C) and venous (D) anatomy vis-à-vis the choroid plexuses of the lateral and IV ventricles, the corpus callosum and midbrain, the primitive meninx and brainstem, the galenic venous system and its tributaries, and the pericallosal arterial structures connecting the anterior and posterior circulations. (E, F) see pp. 397.





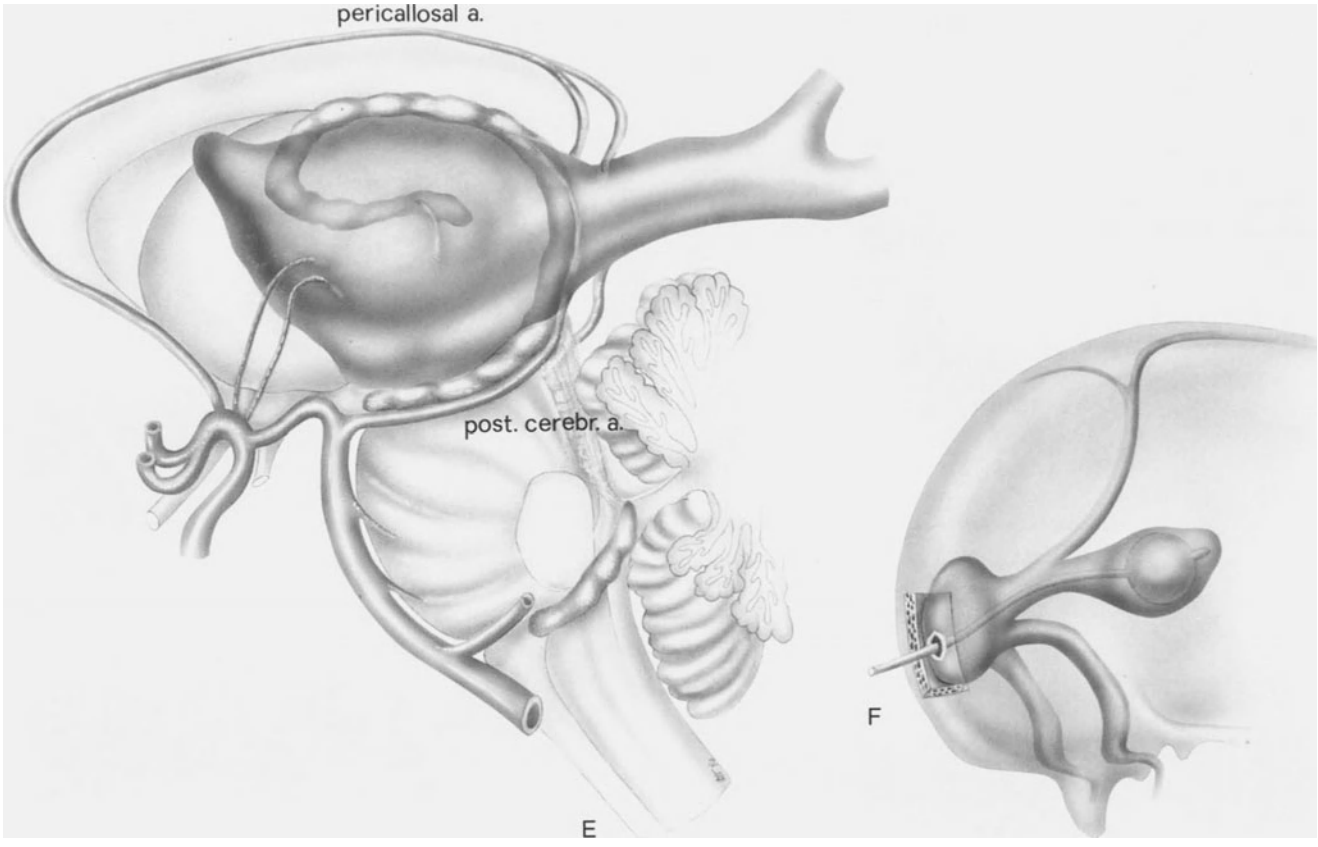


Figure 11.35. (E) In this drawing an aneurysmal venous dilation, commonly and probably equally commonly incorrectly, considered to be the great vein of Galen, is projected as it occupies the III ventricle and compresses the mid-brain. The course of the lateral ventricle choroid plexus is drawn in perspective; the entrance of the terminal portions of the pericallosal arteries into the venous structure, and of the posterior choroidal arteries with both contributing to the arteriovenous shunt are also clearly indicated. The perforating vessels are herein drawn entering the malformation, which they almost invariably do; however, they seldom perforate the mid-brain. (F) This is a drawing of the first procedure we performed in 1978, to occlude the aneurysmal dilation of the galenic system by performing a bur hole at the inion, opening the torcular herophili with a No. 11 blade, inserting a Foley catheter to the blind pouch of the malformation, and then inflating the catheter to occlude subtotally the malformation. The child's shunt disappeared completely, and immediately, and his heart rate became regular (he was a newborn in high output, intractable, failure). That night the child was apparently perfectly normal. The following morning the child suffered acute cerebral decompensation and died: autopsy revealed infarction of the centrencephalic portions of both hemispheres.

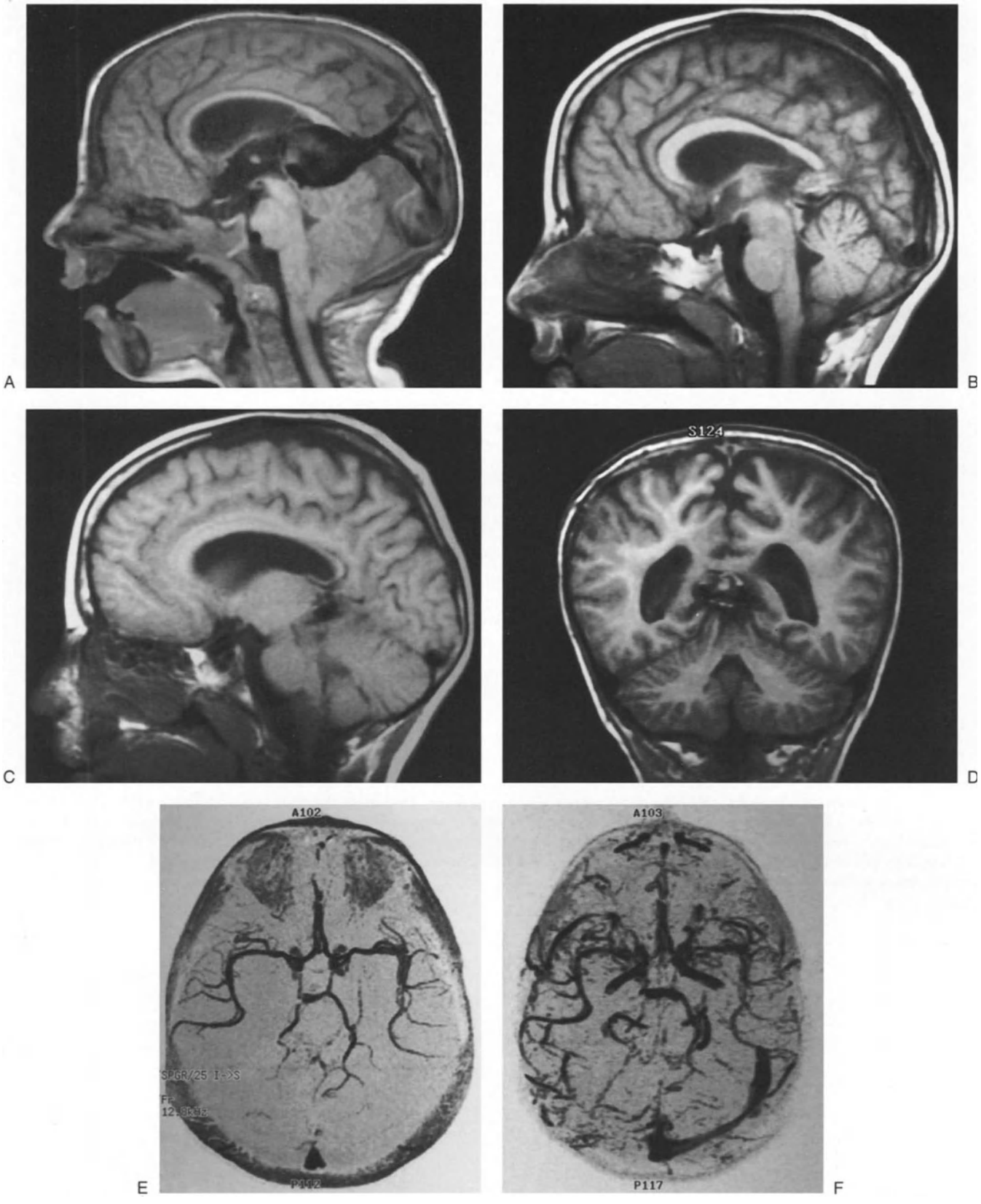


Figure 11.36. Legend see p. 399.

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◀ **Figure 11.36.** Arteriovenous malformation of the galenic system. This is a most interesting case for two reasons: it reveals complete occlusion of the arteriovenous malformation of the galenic system accomplished via transarterial embolization; it also reveals hydrocephalus secondary to venous hypertension... not compression of the collicular plate... with a clinical symptomatic Chiari I alteration secondary to compression of the culmen monticuli. (A) This sagittal T1 image reveals dilated lateral ventricles, enormous dilation of the aneurysmal transformation of the galenic system, compression of the culmen monticuli with its convexity converted into a concavity, and herniation of the tonsils across the line of the foramen magnum (the Chiari I alteration). In (B), (C), and (D) are reproduced the T1 images in the sagittal, parasagittal and coronal projections, respectively, revealing (1) postembolization occlusion of the aneurysmal dilation of the galenic system; (2) a clot in the infraspinal region of the aneurysmal dilation of the galenic system; (3) a normal culmen monticuli now that the aneurysm has been embolized; and (4) the return of the cerebellar tonsils into the posterior fossa. In (E) and (F), the early and mid arterial phases, respectively, of the control angiogram performed on this child reveal no vascular malformation.

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12 Infections

"If some things are too luxuriant, it is owing to the richness of the soil; and if others are not arrived to perfection or maturity, it is only because they are overrun and oppressed by those of a stronger nature."

ALEXANDER POPE, Essay

Seldom in the history of surgery has a dictum, in this case "ivi pus, ibi evacuo" (wherever there is pus, it must be evacuated) been so true, for so long! In the management of intracranial infections, in fact, all surgical procedures are directed simply to removing the foreign body, excising the infected tissue, or draining the pus. Surgery is not indicated for the treatment of subdural empyema because there is no pyogenic exudate in the subdural space. For shunt infections, the foreign body (shunt) must be removed, the granuloma (when present and on the surface) should be excised, and the pyogenic cerebrospinal fluid (CSF) drained (external CSF drainage).

The literature on shunt infections is endless, the approaches to treating them very variable, and the results unpredictable. However, the majority of Pediatric Neurosurgery Centers throughout the world seem to be reporting similar experiences. Therefore, we have chosen to present our thoughts and experiences through the medium of a study performed on our patients with the assistance of Dr. Mario Ammirati [2].

Infection is an ominous complication of CSF shunts: it may turn an otherwise successful neurosurgical operation into a nightmare, with its far-reaching effects on the child's survival and development, and on the length and cost of the hospitalization [12, 13, 17, 21].

Despite numerous efforts, including the use of prophylactic antibiotics and the surgical isolator [4, 11, 14, 24], the infection rate [13] in shunted hydrocephalic children remains high, ranging from 7% to 30%. Numerous series have given a figure of approximately 20% [1, 7, 15, 19, 23, 24]. Among those factors possibly related to shunt infection, age at the time of shunt placement and etiology of the hydrocephalus have received considerable attention in the literature, with different conclusions being drawn regarding their significance in the genesis of shunt infection [8, 10, 16, 18, 20–25].

A total of 431 consecutive patients who underwent their first CSF shunt insertion at Children's Memorial Hospital over a 10-year period were retrospectively studied with regard to the relationship between the etiology of the hydrocephalus, age at the time of shunt placement, and infection rate (Tables 12.1–12.4). Forty percent of the patients had constrictive hydrocephalus and meningomyelocele, 33% congenital communicating or obstructive hydrocephalus, and 18% tumors. Intraventricular hemorrhage and meningitis accounted for the remaining 8%. Eighty-three percent of the patients were less than 1 year old at the time of surgery; 18% were 1 week old or younger. A total of 1485 procedures were performed with an average of 3 procedures per patient. Ninety-six patients had infections, resulting in a 22% infection rate per patient and a 6% infection rate per procedure. No significant correlation was evident between etiology of the hydrocephalus and infection rate ($P > 0.05$), even though meningomyelocele patients seemed to be more prone to infection than congenital hydrocephalus patients ($P = 0.06$). Age at the time of shunt placement was related to infection rate, with younger patients having more than older ones ($P < 0.01$). More in-depth analysis of the relationship between age and infection rate was possible in the meningomyelocele and congenital hydrocephalus groups, owing to the significant number of these patients that fell

Table 12.1. Etiology of the hydrocephalus

Meningomyelocele	174 (40%)
Congenital hydrocephalus	144 (34%)
Intraventricular hemorrhage	23 (5%)
Meningitis	12 (3%)
Tumors	78 (18%)
Total	431

Table 12.2. Infection rate per patient and per procedure

	Patients	Procedures	Procedures per patient	Infections	Infections (% per patient)	Infections (% per procedure)
1973–1978	184	661	3.5	42	22.8	6.3
1978–1982	247	824	3.3	54	21.8	6.5
Total	431	1485	3.4	96	22.3	6.4

Table 12.3. Etiology of the hydrocephalus and infection rate

	Meningomyelocele	Congenital hydrocephalus	Intraventricular hemorrhage	Meningitis	Tumors
Infected	48 (27.5%)	25 (17.3%)	7 (30.4%)	4 (33.3%)	12 (15.3%)
Not infected	126 (72.5%)	119 (82.7%)	16 (69.6%)	8 (66.7%)	66 (84.7%)
Total	174	144	23	12	78

$P > 0.05$ (all); $P = 0.06$ (MM-CH).

Table 12.4. Etiology of the hydrocephalus and age at the time of shunt placement[a]

	1 week	2–8 weeks	9–52 weeks	52 weeks
Meningomyelocele	33	113	25	3
Congenital hydrocephalus	41	39	55	9
Intraventricular hemorrhage	2	15	6	0
Meningitis	0	2	8	2
Tumors	0	3	14	61

[a] 98.2% of the MM and 94% of the CH were shunted within 52 weeks; 78.2% of the Tumors were shunted at 52 weeks of age.

into each one of the subdivisions chosen with respect to age at the time of shunt placement.

The 431 patients underwent 1485 procedures, with an average of 3.4 procedures being performed on each patient (range 1–24). Ninety-six patients were infected, resulting in a 22% infection rate per patient and a 6.4% infection rate per procedure. Regarding the number of procedures and the infection rate for two consecutive 5-year periods, we observed that the number of procedures per patient and the infection rate remained essentially the same in the two periods. There was no strong correlation between different etiologies of the hydrocephalus and infection rate ($P > 0.05$), even though when meningomyelocele patients were compared with congenital hydrocephalus patients a trend was evident with the former having more infections than the latter ($P = 0.06$). While almost all the meningomyelocele and congenital hydrocephalus patients were shunted within the 1st year of life (98.2% and 94%, respectively), the vast majority (78% of patients) with tumors were shunted when they were older than 1 year.

There is a correlation between the age at the time of surgery and the infection rate (Table 12.5), with patients shunted within the 1st week of life being exposed to a higher risk of infection than those shunted at 9 weeks of age or older ($P < 0.05$; $P < 0.01$). Patients in the 2- to-8-week interval seem to occupy an intermediate position.

No meaningful correlation was possible between etiology and the hydrocephalus, age at the time of shunt placement, infection rate in the tumor, IVH, or meningitis patients due to the small number in each age group (IVH, meningitis) and skewed distribution (tumor, 78% of which were shunted after 1 year of age). Such a correlation was possible for the meningomyelocele (MM) and congenital hydrocephalus (CH) patients because of the significant number of patients of each group who were shunted in each of the time spans (< 1 week; 2–8 weeks; 9–52 weeks) within the year (Tables 12.6–12.8). Of the MM patients, 48% shunted in the 1st week of life got infected versus 24% shunted between 2 and 8 weeks and 16% when shunted between 9 and 52 weeks of age ($P < 0.01$). There was no significant difference in the in-

Table 12.5. Infection rate and age at the time of shunt insertion

	1 week	2–8 weeks	9–52 weeks	52 weeks
Infected	26 (34.6%)	40 (23.3%)	21 (19.2%)	9 (11.8%)
Not infected	49 (65.4%)	131 (76.7%)	88 (80.8%)	67 (88.2%)
	75	171	109	76

$P < 0.02$ (all); $P > 0.05$ ($\leq$ 1 week vs. 2–8 weeks); $P > 0.05$ (2–8 weeks vs. 9–52 weeks); $P > 0.05$ (9–52 weeks vs. 52 weeks); $P < 0.05$ ($\leq$ 1 week vs. 9–52 weeks); $P < 0.01$ ($\leq$ 1 week vs. $>$ 52 weeks); $P > 0.05$ (2–8 weeks vs. $>$ 52 weeks); $P < 0.01$ ($\leq$ 1 week vs. 2-∞ weeks); $P > 0.05$ (2–8 weeks vs. 9-∞ weeks).

Table 12.6. Meningomyelocele: infection and age at the time of shunt placement[a]

	1 week	2–8 weeks	9–52 weeks
Infected	16 (48.4%)	27 (23.8%)	4 (16%)
Not infected	17 (51.6%)	86 (76.2%)	21 (84%)
	33	113	25

$P < 0.05$ (all); $P > 0.01$ ($\leq$ 1 week vs. 2–8 weeks); $P > 0.05$ (2–8 weeks vs. 9–52 weeks); $P < 0.05$ ($\leq$ 1 week vs. 9–52 weeks); $P < 0.01$ ($\leq$ 1 week vs. 2–52 weeks). [a] Only three meningomyelocele patients were shunted at 52 weeks of age.

Table 12.7. Meningomyelocele: infection and age at the time of shunt placement

	2–3 weeks	4–5 weeks	6–7–8 weeks
Infected	18 (26%)	7 (25%)	3 (19%)
Not infected	51 (74%)	21 (75%)	13 (81%)
	69	28	16

$P > 0.05$ (all); $P > 0.05$ (2–3 weeks vs. 4–5 weeks); $P > 0.05$ (4–5 weeks vs. 6–7–8 weeks); $P > 0.05$ (2–3 weeks vs. 6–7–8 weeks); $P > 0.05$ (2–3 weeks vs. 4–8 weeks).

Table 12.8. Congenital hydrocephalus: infection rate and age at the time of shunt placement[a]

	1 week	2–8 weeks	9–52 weeks
Infected	9 (21.9%)	9 (23%)	7 (12.7%)
Not infected	32 (78.1%)	30 (77%)	48 (87.3%)
	41	39	55

$P < 0.05$ (all); $P > 0.05$ ($\leq$ 1 week vs. 2–8 weeks); $P > 0.05$ (2–8 weeks vs. 9–52 weeks); $P > 0.05$ ($\leq$ 1 week vs. 9–52 weeks); $P > 0.5$ ($\leq$ 1 week vs. 2–52 weeks). [a] Only 9 children with congenital hydrocephalus were shunted at 52 weeks of age.

fection rate in the MM patients shunted within the 2-to-8-week period ($P > 0.05$). The infection rate in CH patients shunted at 1 week, 2–8 weeks, or 9–52 weeks of age was not significantly different ($P > 0.05$).

It is sufficient to mention the ill effects of shunt infection on the child's survival and development to highlight the importance of preventing its occurrence [12, 13, 17, 21]. It is generally agreed that infection takes place due to operative contamination and subsequent colonization of the shunt apparatus [3, 5]. Attempts to reduce the incidence of this colonization by using perioperative antibiotics or a surgical isolator have not been successful [8, 10, 18, 20, 21, 24]. In fact, recent series still give an infection rate of about 15%–20%/patient [1, 23]. A better understanding of the factors related to shunt infections and their subsequent manipulation, whenever feasible, might prove valuable in decreasing the incidence of shunt infections in children.

Our overall infection rate of 22%/patient life-time is similar to that reported in the literature [1, 21, 23]; and

our patient population, with respect to age and etiology of the hydrocephalus, is fairly well representative of a neurosurgical ward in a major pediatric hospital [1, 21, 22]. The four time subdivisions were chosen based on clinical experience to identify: very early, early, late, and very late shunt insertion with regard to patient age. Indeed, the first two subdivisions correspond to the newborn and very early infant period, and the fourth to the childhood stage [6].

Regarding the relationships between etiology (limited to a comparison between congenital hydrocephalus and MM) of the hydrocephalus and infection rate, we demonstrate that the P value ($P = 0.06$) approaches significance. Whether this indicates a trend cannot be stated. Nevertheless, comparison of the infection rate in the MM and CH groups, which are the only two groups that have a significant number of patients shunted in each of the three time periods within the year, certainly suggests a very gray area. In fact, our finding of an increased infection rate in the MM patients is in agreement with previous reports [16, 22]. We did not do a bilateral comparison with the other groups of patients because of the small number of patients in the IVH and meningitis groups and the skewed distribution of the tumor patients, 78% of whom were shunted after 52 weeks of age.

The relationship between age at the time of shunt placement and the infection rate, with younger patients having a higher infection rate, must be carefully interpreted because it is different in the different etiological groups, remaining valid in the MM group but questionable in the CH groups. There are some reports in the literature linking infection rate and age in shunted hydrocephalic children [8, 10, 14, 16–18, 23, 25], but none looks at how this relationship behaves in the different etiological groups. We were able to study this because of the significant number of MM and CH patients who were shunted in each of the three time subdivisions.

Our finding of MM patients having a very high infection rate when shunted within the 1st week of life is not surprising. Indeed, a meningomyelocele may be regarded as a clean-contaminated or contaminated wound [9], and the insertion of a foreign body (shunt) in close temporal relationship to the closure of the contaminated wound (meningomyelocele repair) may increase the incidence of shunt infections.

CSF shunt infection is a complex phenomenon. Among other factors, the different etiologies of the hydrocephalus and the age at the time of the shunt insertion probably have a bearing on the infection rate. When comparing the infection rate in different series, it is important that the patient population be similar with regard to the age at the time of shunt insertion and etiology of the hydrocephalus. Our review of the literature reveals that in those series in which at least 50% of the patients were shunted at less than 1 year of age, the infection rate was never below 10% and was around 20% in most major series [7, 13, 18, 21].

We consider in detail the MM and CH patients because these two groups of patients are fairly well stratified within the year regarding age at the time of shunt placement, enabling us to draw some meaningful conclusions from our observations. MM and CH, by definition, are shunted early in life; tumors are shunted later. Therefore, it is not reasonable to compare these as two groups (MM and CH, and tumor) regarding infections, postshunt, occurring in relation to early age:

1. Etiology of the hydrocephalus is not related to the infection rate in our series, but there is a trend suggesting that that MM may have a higher infection rate than CH patients ($P = 0.06$). This finding needs further confirmation.
2. MM patients shunted within the 1st week of life have a significantly higher infection rate than MM patients shunted at 2 weeks of age or older ($P < 0.01$). The same does apply to CH patients ($P = 0.05$).

As the correlation of meningomyelocele and shunt placement in the 1st week of life is associated with a very high infection rate (48%), a waiting course may be rewarding in this group of patients, delaying shunt placement until 2 weeks of age or older unless the increase in intracranial pressure becomes life-threatening.

Osteomyelitis

The osteomyelitic bone is removed with a rongeur and discarded, taking the resection peripherally to the locus of infection, and being careful to extend it into areas where there is spontaneous diploic bleeding and no evidence of cortical (bone) digestion by the septic process. If the infected bone is either a free or osteoplastic flap, it should be removed and discarded. The temptation to conserve portions of the bone is to be resisted! Once the infected flap has been discarded one should inspect the edges which border upon the craniotomy site to be sure they are healthy. In fact, it is good practice to curette these edges until diploic bleeding is encountered.

One should avoid opening the dura when operating on a child for osteomyelitis, whether it be secondary to a systemic infection, an infection of the nasal passages, or craniotomy, lest a pathway be opened for extension of the infection into the subdural or intraparenchymal tissue. Computed tomography (CT) scan and/or cerebral angiography should be used preoperatively to learn whether the infection is limited to the skull. Even when the osteomyelitis is post-traumatic, and there is an overlying infection of the periosteum and scalp, one should not open the dura to inspect for an underlying infection. In fact, subdural empyema is treated exclusively with antibiotics, as is leptomeningitis. Intracere-

bral abscess is readily diagnosed with current neuroradiological techniques.

After the infected bone has been removed, the surrounding bone edges curetted and periosteum debrided, the field is irrigated copiously with topical antibiotic solution such as bacitracin, Penrose drains are inserted through stab wounds in the surrounding scalp, and the scalp flap is closed with a single layer of mattress sutures. Wire offers no advantages, is difficult to remove, and is not as efficient for the proper placement of inverted mattress sutures (which are so essential to providing full thickness apposition of the scalp edges).

Epidural Empyema

The epidural empyema represents what is presently a truly unusual clinical entity, but also one which may invariably be successfully treated. The clinical diagnosis is definitive only when angiography shows separation of the dura mater from the inner surface of the skull – as indicated either by visualization of a displaced meningeal artery, or uniform "staining" of the inflamed outer layer of the dura mater. Otherwise, one may suspect the presence of an epidural empyema, but not distinguish it from an intraparenchymal abscess. A CT or MRI scan does not permit distinction between epidural empyema and brain abscess. When the differential diagnosis between brain abscess and epidural empyema is entertained, the surgeon is advised to place his first bur hole over the suspected area of loculated pus within the epidural space.

Treatment of the epidural empyema consists of placement of a single bur hole over the collection of pus, drainage of the pyogenic exudate, irrigation of the cavity with copious amounts of antibiotic solution, insertion of two or more Penrose drains through stab wounds, and closure of the scalp incision with a single layer of inverted mattress sutures. The drains are removed gradually, over a period of 3–10 days, depending upon reexpansion of the brain and obliteration of the cavity. A CT scan suffices to monitor the healing process.

Subdural Abscess and Subdural Empyema

(Fig. 12.1)

The difference between subdural abscess and subdural empyema is that the former is a well-encapsulated collection of liquefied exudate (pus), and the latter is a diffuse, fibrinous, pyogenic exudate adherent to adjacent pachy- and leptomeninges, and subjacent cortex.

The subdural *empyema* is not a surgically treatable entity. It is included here only because of the temptation which one may have to operate, considering it comparable to an epidural empyema or confusing it with a subdural abscess. Quite the contrary is true!

Anatomopathologically speaking the subdural empyema consists of a collection of fibrinous exudate within the subdural space, varying in thickness from 0.5 to 2 mm, intimately adherent to the inner surface of the dura externally and the arachnoid membrane internally. At times, it may be so thick and globular in form as to resemble a meningioma. Attempts to resect this fibrinous exudate result in the stripping of the arachnoid from the surface of the brain, damaging the cortex and causing diffuse petechial hemorrhages. The insertion of drains into the subdural space does not facilitate egress of the consolidated exudate. Once the diagnosis of subdural emypema is made (either neuroradiologically or after a bur hole reveals that there is no epidural collection of pus and dural opening reveals the presence of pyogenic exudate within the subdural space), the treatment consists of a period of 4–6 weeks of intravenous antibiotic therapy. The subdural *abscess,* however, is treated surgically with craniotomy, or bur holes, and evacuation of the pus, followed by antibiotic therapy. Anticonvulsant medication is essential because of the high incidence of seizures in these patients.

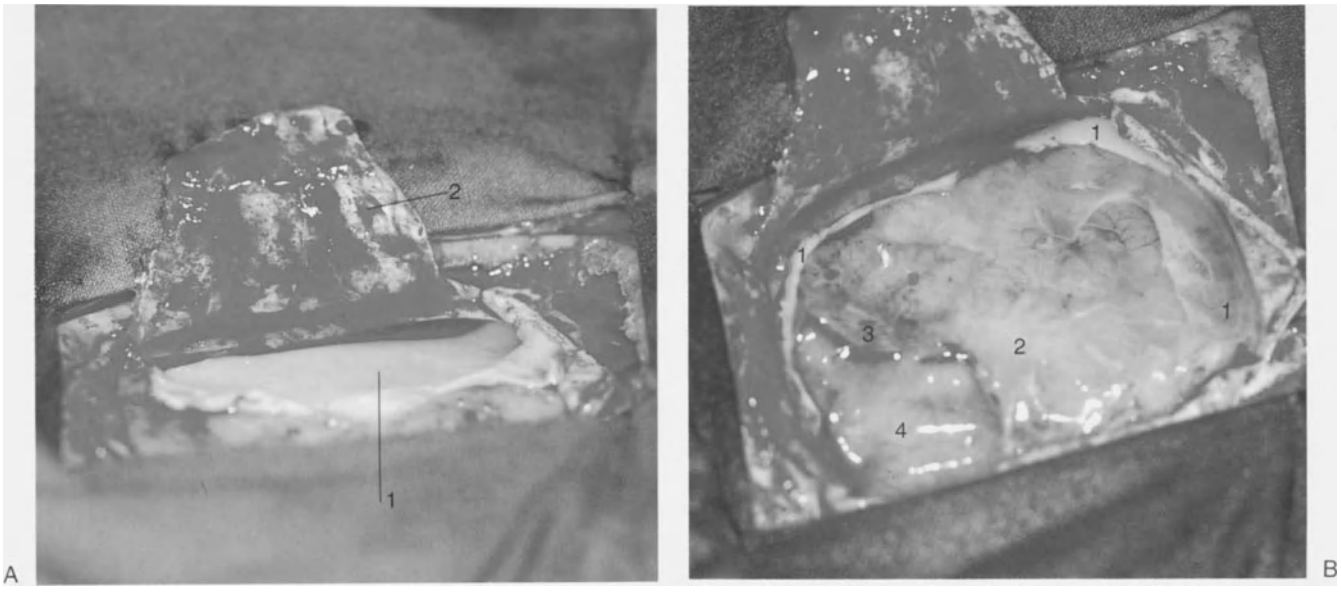
Acute Meningitis with Hydrocephalus

(Figs. 12.2–12.4)

Acute leptomeningitis causes inflammation of the pia-arachnoid and the cortical surface, may be suspected clinically and confirmed angiographically, and results either in cerebral edema or, less commonly, acute communicating hydrocephalus. The leptomeningitis and cerebral edema are managed medically. Acute communicating hydrocephalus represents a neurosurgical emergency, and must be treated with external ventricular drainage lest it result in secondary cerebral edema, porencephaly, and both transtentorial and transforaminal herniation. The external ventricular drainage should be maintained until the intraventricular cerebrospinal fluid cultures and chemistries are normal (see specific reference to external ventricular drainage in Chap. 15) and the leptomeningitis has been cured medically.

Brain Abscess

Abscess formations are not invariably uniform accumulations of liquid pus. In fact, they generally consist of multilobulated areas of liquefied parenchymal digestion, with varying densities of liquefaction and fibrinous exudate collected within a number of septated cavities within the abscess. *Therefore, one may not assume that simple puncture of the abscess cavity will give*



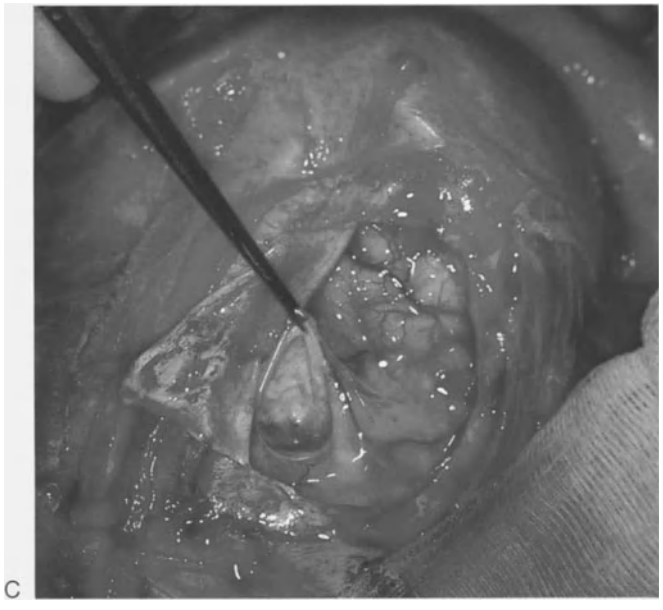


Figure 12.1. (A) Here one notes the pearl white subdural empyema membrane (*1*) beneath the dural (*2*) opening. (B) This child suffered a subdural empyema. One notes the dense outer membrane at the corners of the opening (*1*), and an inner membrane (*2*) adherent to the underlying arachnoid, pia, and cortex (*3*). Extensive bleeding and decortication occur when attempts are made to separate the pyogenic exudate from the underlying parenchyma. Some locules of empyema are visible (*4*). (C) The subdural empyema is adherent to a necrotic portion of brain.

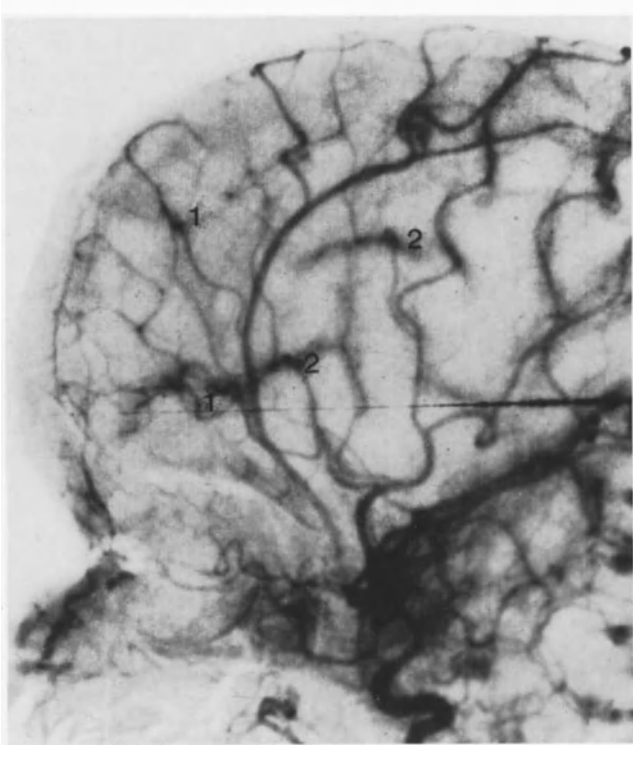


Figure 12.2. The arterial phases are characterized by "halo" formation around the sulcal branches of the anterior (*1*) and middle (*2*) cerebral system.

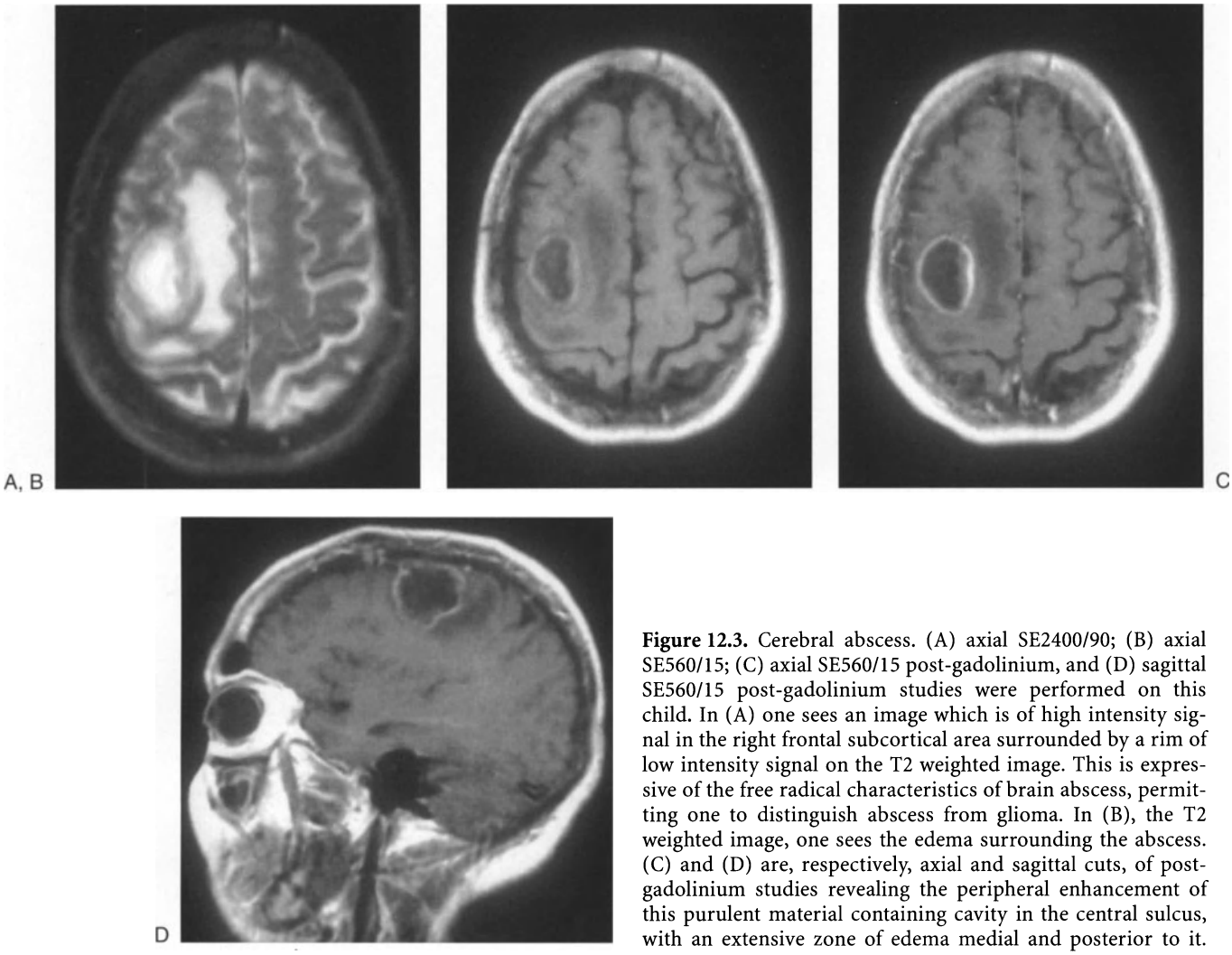


Figure 12.3. Cerebral abscess. (A) axial SE2400/90; (B) axial SE560/15; (C) axial SE560/15 post-gadolinium, and (D) sagittal SE560/15 post-gadolinium studies were performed on this child. In (A) one sees an image which is of high intensity signal in the right frontal subcortical area surrounded by a rim of low intensity signal on the T2 weighted image. This is expressive of the free radical characteristics of brain abscess, permitting one to distinguish abscess from glioma. In (B), the T2 weighted image, one sees the edema surrounding the abscess. (C) and (D) are, respectively, axial and sagittal cuts, of post-gadolinium studies revealing the peripheral enhancement of this purulent material containing cavity in the central sulcus, with an extensive zone of edema medial and posterior to it.

egress to all of its contents, nor that drainage of its liquefied exudate will result either in collapse of the wall or cure. To be sure, some abscesses may be successfully treated with single puncture and drainage. This is not, however, invariably true. Intravenous antibiotics should be begun immediately the diagnosis of intracerebral abscess is suspected.

In light of the fact that patients with brain abscesses resected before they lose consciousness, either from brain shift or following a full major seizure, may be cured and that the mortality rate in those who are not surgically treated until after a full major seizure or loss of consciousness approximates 50%, *treatment of brain abscess is to be considered an absolute neurosurgical emergency.*

Currently, as since the dawn of surgical management of intraparenchymal abscesses, two distinctly different forms of operative treatment exist: (1) simple bur hole, abscess puncture, and drainage of pus and (2) craniotomy and in toto resection of the abscess. Those abscesses located within the region of the basal ganglia or the frontotemporoparietal area of the dominant hemisphere are best treated with simple puncture and drainage of whatever amounts of pyogenic exudate may be removed. Abscesses in all other locations are best treated with craniotomy and total resection. The reason for the initial attempt at simple drainage of basal ganglia and speech area abscesses is obvious. The reason for total resection of those abscesses located in surgically accessible areas is that one may not be certain that there is only one cavity within the abscess: in fact, there are often multiple "daughter-abscesses."

Bur Hole and Cannula Drainage

A single bur hole is placed over the most desirable access line to the abscess cavity. A cruciate incision, just large enough to permit entrance of a brain cannula, is made in the dura. One should use a blunt cannula,

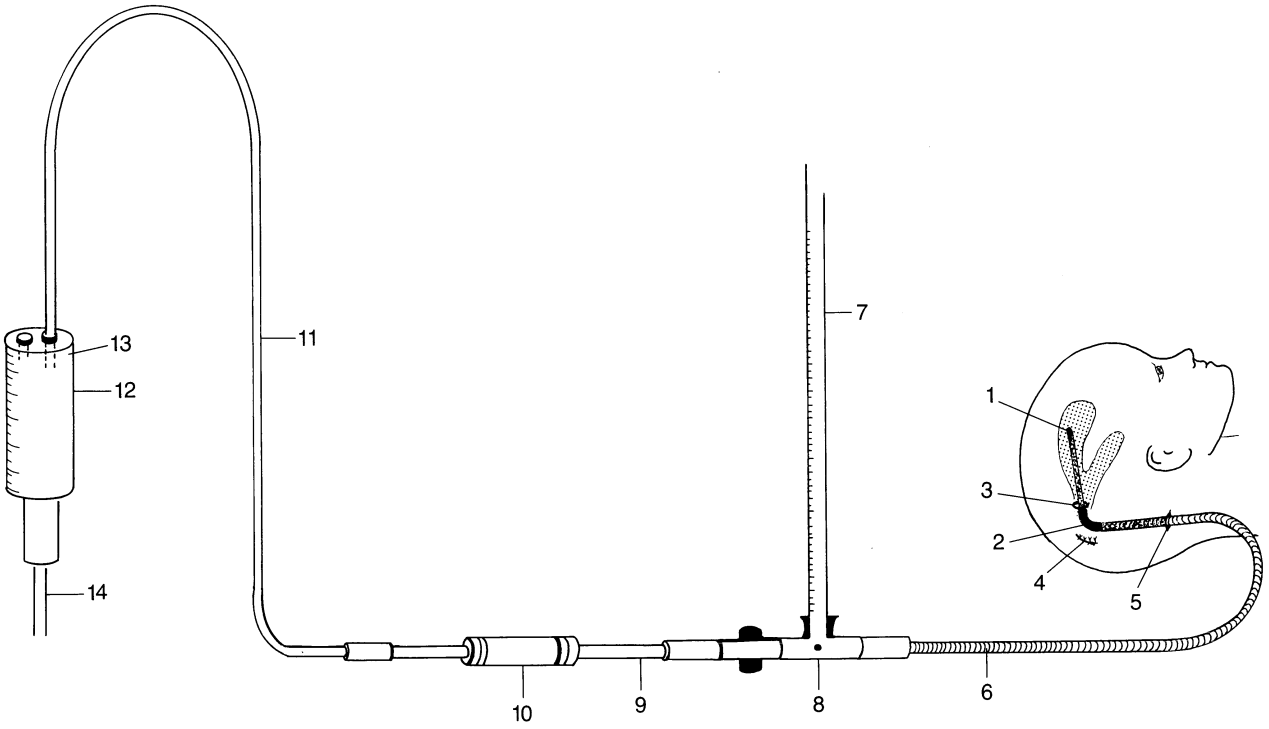


Figure 12.4. External ventricular drainage. *1,* ventricular catheter; *2,* right-angle connector, optional, since the ventricular catheter may be run under the skin and then through the stab wound; *3,* drill hole; *4,* skin incision; *5,* stab incision; *6,* spring tubing; *7,* manometer; *8,* three-way stopcock; *9, 10, 11,* distal connection system with or without one-way valve included; *12,* burette set so that dropper (*13*) is at the same levels as head and heart; *14,* cerebrospinal fluid collection tubing.

avoiding either the very sharp or flat-tipped cannulas. Care must be taken to hold the cannula in a direct line as it is inserted through the abscess wall, since the variability of thickness of the wall is such that on occasion it may be very dense and difficult to penetrate. If this is the case, the cannula will be deflected over the surface of the abscess and into surrounding edematous brain. Because of the fact that the inflammatory reaction around the abscess cavity consists of neovascularization and glial tissue, this deflection may result in brain damage and/or intracerebral hematoma. Once the abscess cavity is punctured, the obturator is removed. If there is not a spontaneous egress of pus, one should apply a 5-ml syringe to the cannula so as to aspirate the pyogenic exudate.

Take care to collect material for both aerobic and anaerobic culture, and to measure carefully the volume of pus withdrawn from the cavity. Once no more purulent material may be withdrawn, the cavity should be irrigated with a volume of antibiotic solution which measures no more than 75% of the total volume of pus withdrawn. The irrigating solution should be injected and withdrawn, using fresh antibiotic solution in each aliquot. When there is no longer fibrinous exudate within the returning solution, one considers the procedure ended. Some surgeons known to the author choose to insert a drain into the abscess cavity; most do not. The skin is closed, as in all procedures for intracranial infections, with a single layer of inverted mattress sutures. Follow-up CT or MRI scans should be performed the evening of surgery, and then at 3-day intervals until the abscess cavity has collapsed completely. If there is CT or MRI evidence of increase in volume of the abscess, one should proceed straightway to repeat puncture and drainage of the abscess if it is located either in the basal ganglia or speech area, or with a definitive operative procedure: craniotomy and removal of the entire abscess, if it is located elsewhere.

Craniotomy and Resection of the Abscess

Following craniotomy and double trapdoor dural opening to expose the entirety of cerebrum over the abscess, a cerebrotomy is performed, in either a sulcus or a gyrus, over the most flattened portion of cortical tissue. If the abscess has leaked into the subarachnoid space, one will immediately note opalescent arachnoid and/or injection of the cortical surface, suggestive of meningitis. These visible signs are most indicative of the location of the abscess. They are also suggestive of the point at which the abscess ruptures into the subarachnoid

spaces. Consequently, working within this portion of inflamed and flattened cortex may result in rupture of the abscess with outpouring of pus. Though every effort should be made to avoid this, there is no reason to think that such an event will result in diffuse leptomeningitis or increase the risk of postoperative subdural or epidural empyema, osteomyelitis, or wound infection.

After the cerebrotomy and exposure of the "pointing" surface of the abscess, one should extend the cerebral incision from one end of the abscess to the other, so as to permit delivering it through the parenchymal opening. This will be greatly facilitated if the pus is not drained, and if the surgeon takes a great deal of time in "delivering" it. Fluffy cottons, no wider than 2 or 3 mm and varying in length from 2 to 4 cm, should be inserted over the abscess capsule, laying one over the other as they are positioned circumferentially, to separate the neovascular and gliotic surfaces of the brain from the capsule. This allows the abscess to bulge through the cerebrotomy and out of its parenchymal bed. One should avoid the use of spatulae or dissectors.

If the abscess ruptures during dissection, the pus should be aspirated completely from the surface of the brain and from within the abscess cavity. One may then pick the capsule up with pituitary forceps, separating it subsequently from the surrounding brain with the use either of a Penfield #1 dissector or a small tuft of fluffy cotton held within Cushing forceps. As this is done, rather abundant venous bleeding may occur, a result of tears in the neovascular structures surrounding the abscess wall. Attempts to coagulate these vessels are futile. It is best to remain with the task at hand, removal of the abscess, and then to irrigate the parenchymal bed with saline after the capsule has been removed. Fluffy cottons may also be placed within the cavity and aspirated until all bleeding has stopped.

Drains should not be placed within the abscess cavity. The dura is closed with 4–0 sutures, the bone flap is reapproximated and anchored into position, and the skin is closed with the usual Cloward sutures.

One need not treat the patient with 4–6 weeks of intravenous antibiotics. A single follow-up MRI or CT scan suffices to provide information concerning the postoperative status of the brain.

Pyocephalus

It is well known that brain abscess does not, with the rarest of exceptions, occur in children under 2 years of age. However, it is not unusual to encounter a case of pyocephalus in the newborn or infant. The term "pyocephalus" indicates intraventricular pus present in such quantities as to replace completely the cerebrospinal fluid, dilate the ventricles, and give the surgeon the impression that the cerebral mantle is little more than an abscess cavity. Its diagnosis is always serendipitous, and its treatment consists of placement of bilateral external ventricular drains, irrigating alternatively one ventricle and draining the other, under very low pressure. This should be done over a prolonged period of time. The external ventricular drains are managed just as when treating a child with ventriculitis.

Though hydrocephalus may remain as a permanent complication of pyocephalus, this is not invariably true. Therefore, one is justified in removing the external ventricular drains after the infection has cleared. If, subsequently, hydrocephalus becomes apparent, a shunt may be inserted.

Ventriculitis

Infection is the single most common, and dangerous, complication of shunting systems. It also represents the most common cause for shunt failure. Control of intracranial pressure in hydrocephalic children, consequently, becomes a serious problem when a shunt gets infected!

There are three commonly accepted means of treatment for ventriculitis complicating hydrocephalus: (1) removal of the infected shunt and immediate reinsertion of a new shunt, combined with antibiotic therapy; (2) removal of the infected shunt and initiation of antibiotic therapy, with insertion of a new shunting system after the infection has cleared (both of these procedures have serious limitations, subjecting the child to unnecessary risks of reinfection in the former and uncontrollable increase in intraventricular pressure in the latter); and (3) *external ventricular drainage, with appropriate antibiotic treatment, which is, in fact, the treatment of choice.*

The external ventricular drain consists of a single Spring Catheter (Codman, Randolph, Massachusetts) shunting system inserted into the lateral ventricle, through a split trochar (see the section on shunt revisions in Chap. 15). A stab wound should be made approximately 2 cm from the skin incision (used for the twist drill opening and insertion of the split trochar). The spring catheter is brought through the stab wound, passed beneath the skin, and then inserted into a lateral ventricle. A "lock-clip" is used to anchor the spring catheter to the skin, and then the external ventricular system is connected.

Though the most common reason for inserting an external ventricular drain is ventriculitis, it is also used in the treatment of acute hydrocephalus secondary to intraventricular or subarachnoid hemorrhage, and in those instances where the ventricular fluid has a protein content in excess of 500 mg%.

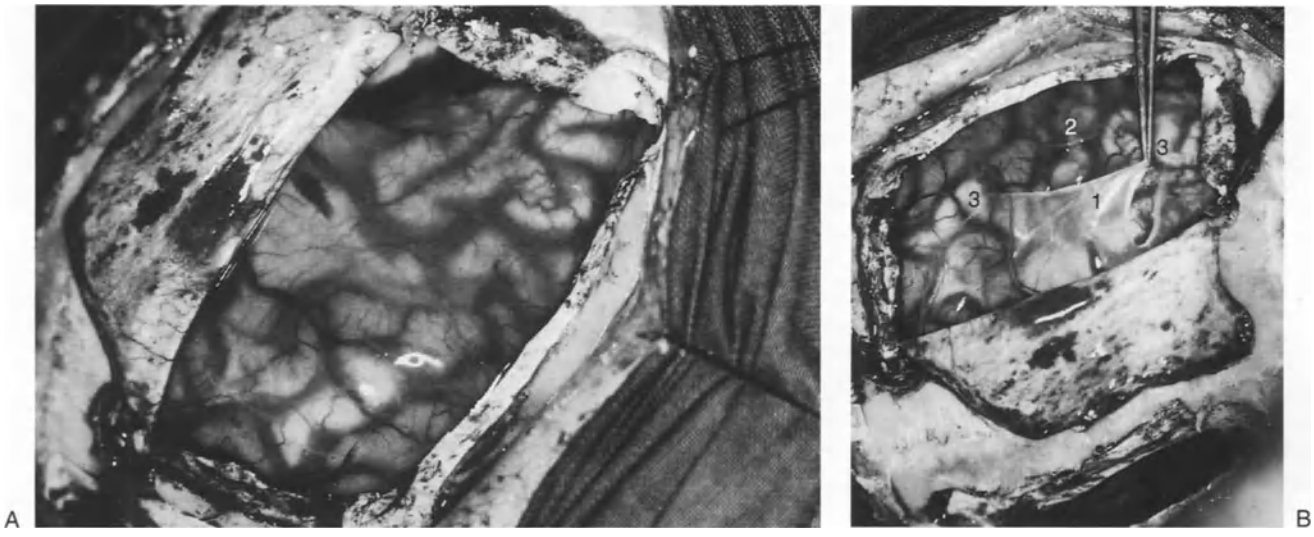


Figure 12.5. (A) The remarkable cerebral atrophy that occurs in association with long-standing subdural effusion, in a child who had suffered meningitis. One notes the opalescent membranes over the surface of the hemisphere, the enormous sulci, and the shunken gyri. (B) The outer leaf (*1*) of the subdural effusion membrane is elevated, exposing underlying effusion, inner leaf (*2*), and extensive areas of devascularization of cerebral gyri (*3*).

Subdural Effusions (Fig. 12.5)

Inflammatory processes either of the meninges or cerebral parenchyma, much more commonly the former than the latter, may result in accumulation of fluid which is either actively infected or presently sterile but secondary to an active infection, within subdural spaces. This results in the formation of an accumulation of fluid contained within two neoformed membranes (outer and inner) bordering, respectively, the inner surface of the dura and the arachnoid. At times, especially in the particularly young, the accumulation of fluid may occur so rapidly, accompanied by such severe cerebral edema, and then become so extensive in volume as to cause an acute increase in intracranial pressure, with splitting of sutures and bulging of the anterior fontanelle. Other times, the accumulation of this fluid within the subdural space is insidious in onset and gradually progressive, developing at the expense of the underlying brain without causing a clinically apparent increase in intracranial pressure. These effusions are extremely difficult to treat. Resection of their membranes, repeated subdural taps, and shunting procedures have all failed. The treatment of subdural effusions consists of bur hole and drainage of the fluid in the acute cases, when there is an increase in intracranial pressure; and lowering of the superior sagittal sinus in the chronic cases (see the section on chronic subdural hematoma in Chap. 13) when there is progressive diminution in cerebral volume and "hanging veins."

Cerebritis and Cerebellitis (Figs. 12.6, 12.7)

With the exception of secondary porencephaly, which may become loculated, isolated, and expansile so as to act as a space-occupying lesion, there are no surgical indications for cerebritis. If a porencephalic cavity, which is expansile in nature, does develop, craniotomy with marsupialization of the cavity and the underlying lateral ventricle or shunting are the treatments of choice.

Inflammatory processes of the cerebellum (cerebellitis) may also result in the formation of intraparenchymal cavities that may become expansile. These are best treated with resection of the cyst wall. Another form of

►

Figure 12.7. Cerebellitis. (A) The degenerative necrotic changes within the cortex of both cerebellar hemispheres (*1*) has resulted in adhesions of the hemispheres to one another. The membrane of the cisterna magna has been removed, exposing the tonsils (*2*) and the dorsal aspect of the medulla spinalis (*3*). (B) The right cerebellar hemisphere (*1*) is being separated from the one on the left, in an attempt to open the cerebrospinal fluid pathways. The tonsils (*2*) have been freed from the underlying medulla oblongata, exposing the floor of the IV ventricle (*3*). One sees the remnant of the dense, fibrotic arachnoid membrane of the cisterna magna (*4*). This surgery was not followed by improvement in the child's clinical course. It was necessary to insert a cystoperitoneal shunt. This is, in fact, the postinfectious form of Dandy-Walker cyst (first described by Walter Dandy).

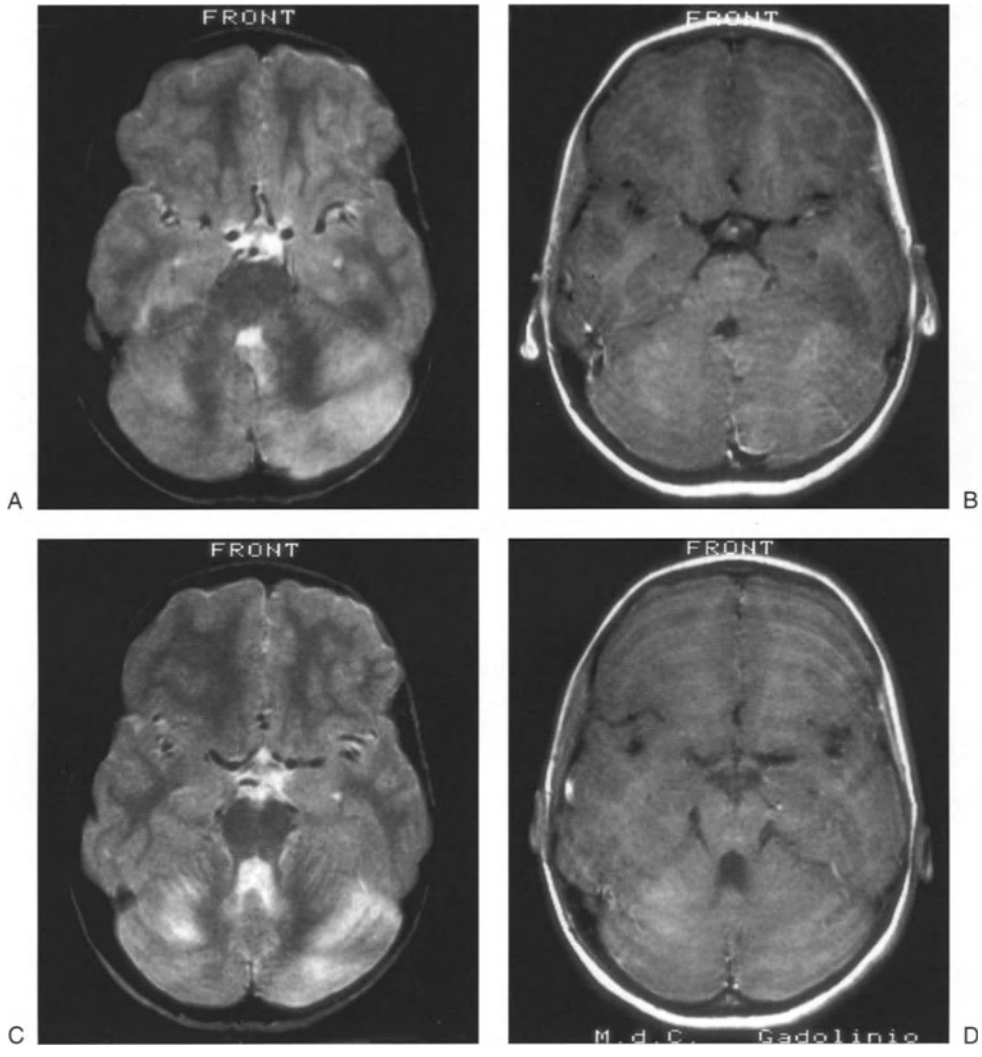


Figure 12.6. Bilateral cortical cerebellitis, pre- and post-therapy. In (A), an axial SE2400/90 image, there are high signal intensity zones in both cerebellar hemispheres, more evident *on the left*, representing edema and inflammation. The ventricle is compressed by edema. In (B) an axial SE560/15 post-gadolinium study, there is postenhancement of the image. The diagnosis, cerebellitis, was made, and the child was followed with MR studies after specific treatment. (C) Axial SE2400/90 2 weeks post-therapy and (D) axial SE560/15 post-gadolinium, also 2 weeks post-therapy, reveal partial resolution of both the edema and the enhancement, and the IV ventricle has expanded into its normal position.

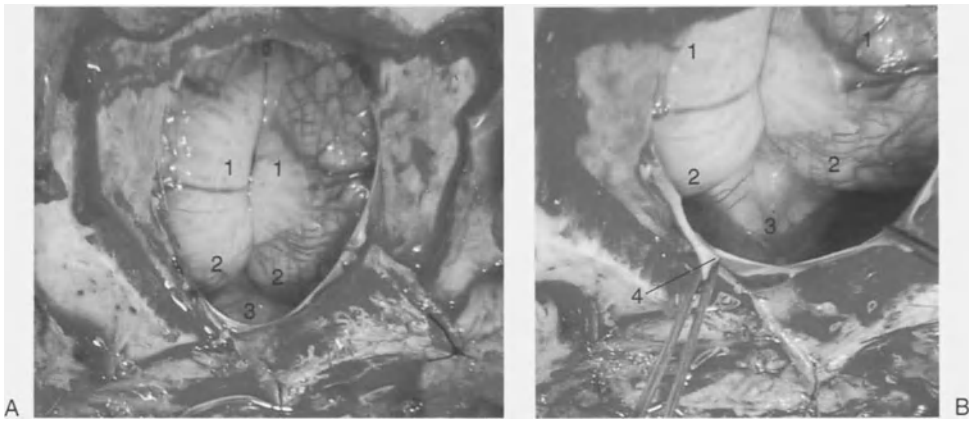


Figure 12.7. Legend see p. 410.

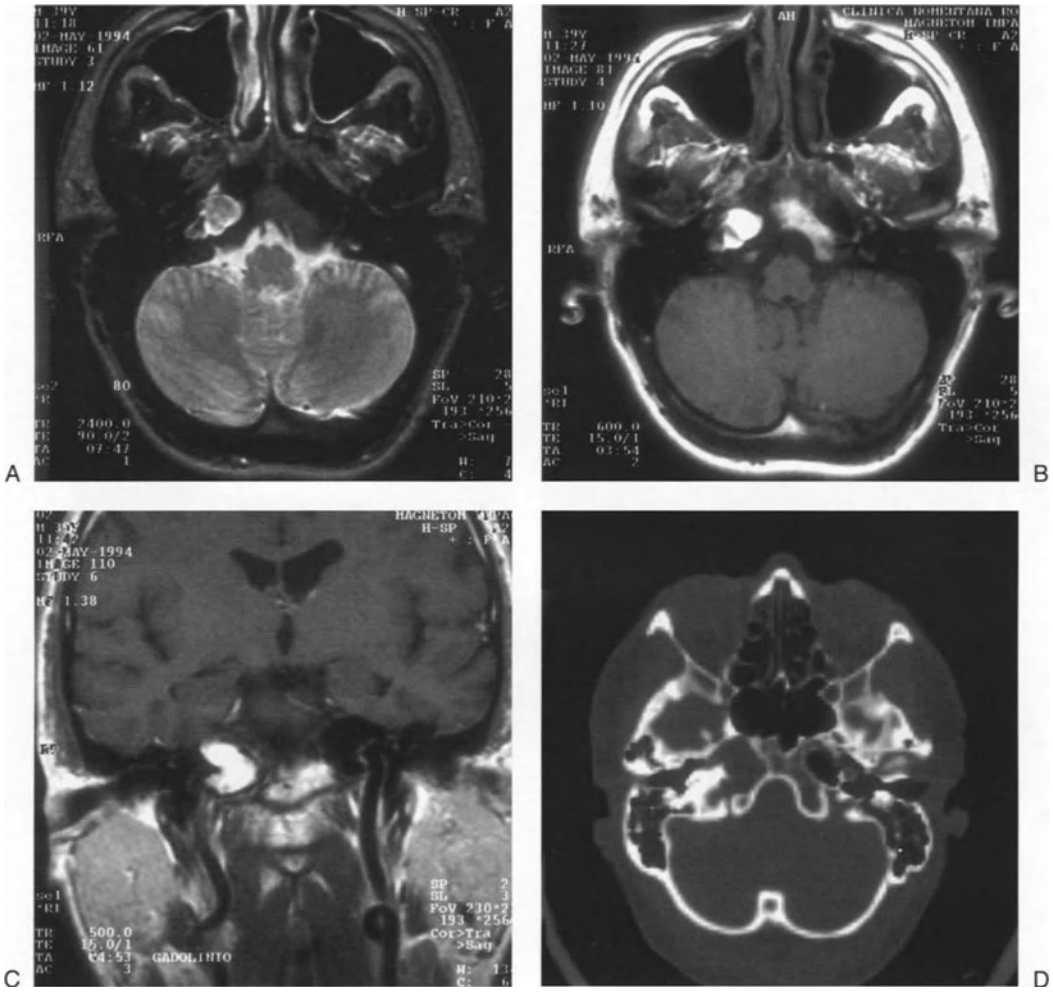


Figure 12.8. Cholesterinic granuloma of the petrous apex. (A) is an axial SE2400/90, (B) an axial SE600/15, (C) a coronal SE500/15, and (D) an axial CT scan bone window. In the right petrous apex there is a soft tissue mass showing the same signal intensity as fat. The hyposignal intensity on the T2 weighted image (A) and the high signal intensity on the T1 weighted image are well seen. The CT scan reveals alterations in the bony structure of the right petrous apex.

cerebellitis, that involving the cerebellar cortex and overlying pia-arachnoid at the insertion of the arachnoid membrane of the cisterna magna, may result in a postinfectious Dandy-Walker cyst. In essence, the inflammatory process provokes intense adhesions between arachnoid of the cisterna magna, pia-arachnoid, and cortex, obstructing completely the outlet foramina of the IV ventricle and sealing the cerebellar hemispheres and tonsils to one another and the brainstem.

Surgical removal of the adhesions is not followed by a cure. It is preferable to perform a cystoperitoneal shunt.

Wound Infections (Figs. 12.8, 12.9)

All wound infections are neurosurgical emergencies and, consequently, should be treated immediately.

Stitch Abscess

Stitch abscesses are treated by removal of the infected stitches. The wound is opened and inspected for any necrotic or foreign material, which is, in turn, removed before the wound is scrubbed with surgical soap for approximately 10 min. The tissue is then allowed to granulate in by secondary intention, with medical personnel scrubbing it with surgical soap for 10 min, 3 times a day, until healing has occurred.

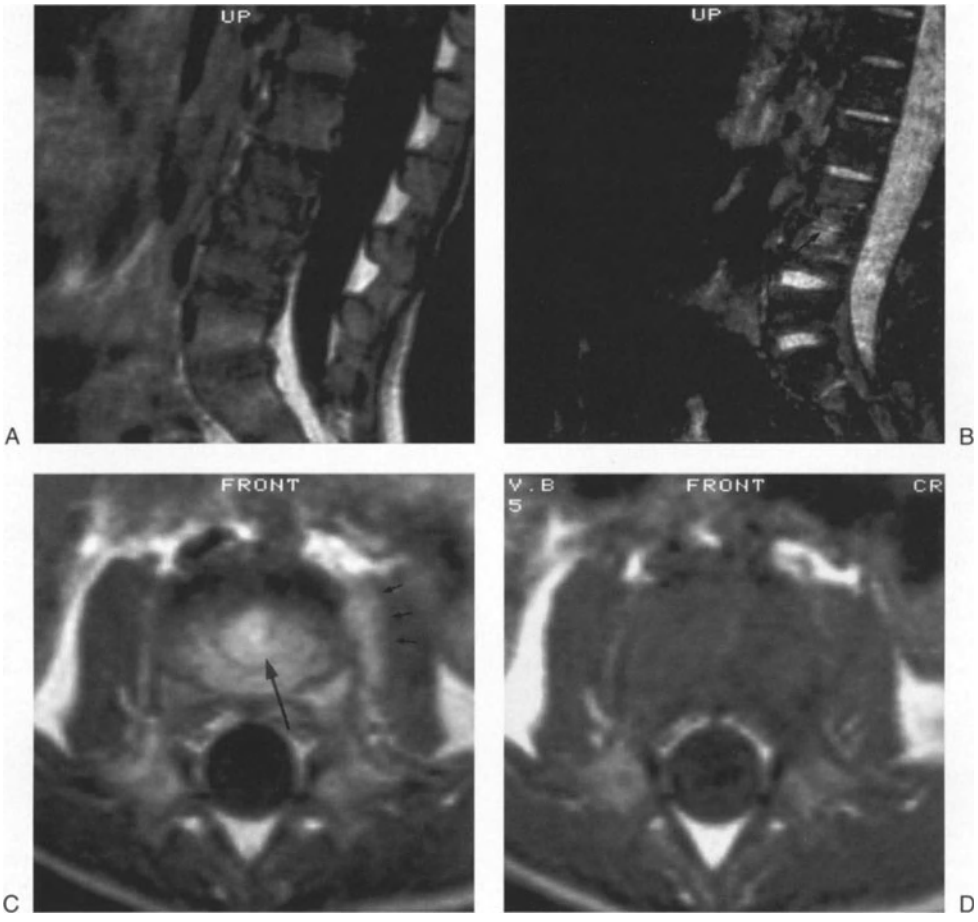


Figure 12.9. Spondylodiscitis, L3–4. We have no information concerning the cause of this clinical entity, so it is termed "idiopathic." There is a hyposignal intensity on the T1 weighted image of the vertebral body of L3 and L4 (A), with a hypersignal of the intravertebral disk (*arrows*) best seen in (B). (C, D) The post-gadolinium injection reveals enhancement of the disk with inflammatory tissue in the left paravertebral space, shown, respectively, with the *large arrow* and the *small arrows* in (C).

Superficial Infection

Surface infections such as excoriations are treated with a single surgical scrub and the application of antiseptic material. Clean dry dressings are applied.

Deep Infection

Whether secondary to trauma or a complication of an operative procedure, these are treated with opening of the wound; removal of foreign substances, granuloma, suture material, and so on; and debridement and removal of any bone fragments or flaps. The bone is discarded. The entire area is then scrubbed with surgical soap and irrigated with copious amounts of antibiotic solution. Such deep infections are best left open, so that they may be washed with surgical soap for 10 min, three times a day, allowing them to granulate in. One should not perform a secondary surgical closure on such wounds. If the wound is extensive, one may elect, at a later date, after there is a healthy base of granulation tissue, to request that the plastic surgeons assume its management.

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13 Trauma

"And yet a mysterious light seemed to show me his boyish head, as if in that moment the youth within him had, for a moment, gleamed and expired."

JOSEPH CONRAD, Lord Jim

The management of craniocerebral trauma in the child is, in contrast to the adult, more dependent upon clinical neurological evaluation, fastidious observation and monitoring of neurological and vital signs, and the timely use of diagnostic procedures. The significance of the neurological evaluation, comparison of the Children's Coma Score (CCS) to the Glasgow Coma Score (GCS), and the discussion of the Children's Outcome Scale (COS) are presented in this text. The parameters to be observed during the stay on the pediatric neurosurgical floor, or in the pediatric neurointensive care units, are also presented. The same applies for the diagnostic studies, although it is essential at this time for one to review and to analyze critically the diagnostic studies that should be ordered and the value to be attributed to them in the decision-making process: when to operate and what operation to perform.

The single, most abused diagnostic study in emergency rooms, whether in adult or in children's hospitals, is the "routine" skull X-ray. They are generally of poor quality because they are performed on an emergency basis and the child is frightened and unmanageable. Rarely do they contribute significantly to the management of the child. In fact, whether a skull fracture is present or absent, if present whether it is depressed or elevated, and if it is depressed whether it is simple or compound, are of no significance in and of themselves at the time of initial evaluation of the child. In essence, one must first determine whether the child has suffered a loss of consciousness, what its present state of neurological function is, whether there are associated internal or osseous injuries, and what the immediate course of the clinical events is. Certainly, no child will be carefully observed while in the waiting area of the X-ray department or on the X-ray table. Consequently, critical moments and, alas! hours, are often lost between the time the child is sent to the X-ray department from the emergency room for routine skull X-rays and the time it reaches a well-staffed and fully equipped nursing station.

The proper time to request skull X-rays, or computed tomography (CT) scans, for that matter, is after the child has been examined by a qualified pediatric neurosurgeon, who will then determine, on the basis of a careful history regarding the mechanics of the injury, seizure occurrence, loss of consciousness, external signs of injury, clinical and laboratory evaluations for cerebral or internal injuries, and observation of the clinical course, which diagnostic studies are indicated and, more important, when these studies should be performed.

For example, if there is a full-thickness scalp laceration or protruding brain, the surgeon will not need skull X-rays, since immediate surgery is indicated and the wound will be adequately inspected at that time. He may desire a CT scan to learn whether there is an associated intracerebral hematoma, which would change his operative plans completely. The knowledge that a linear skull fracture may be present, on the other hand, does not contribute in any way to the immediate decisions for patient management. Of course, deterioration in neurological condition may result from expanding intracranial clots (epidural or subdural), which may result from the same sequence of traumatic events that would cause a linear skull fracture: diploic hemorrhage from a diastatic fracture and epidural hemorrhage over the superior sagittal sinus (SSS) from a linear or depressed fracture across the sinus.

The current state of the art of CT scanning is such that one may obtain much more information from a carefully performed CT scan than a combination of plain skull X-rays and a hastily performed CT scan: information concerning presence and nature of a skull fracture, intracranial hemorrhage or cerebral contusion, diffuse subarachnoid hemorrhage or cerebral edema. There is no place for magnetic resonance imaging

(MRI) in the management of *acute* head injuries, but there is for subsequent evaluation.

Younger children suffer much greater damage from such deceleration impact injuries as falls, and acceleration burst injuries such as blows to the head. Approximately 50% of children whose fontanelles have not closed suffer skull fractures from either acceleration or deceleration injuries, whereas only 29% of children with closed fontanelles and sutures suffer fractures. Similarly, children less than 6 months of age suffer the highest death rate among motor vehicle occupants (9/100,000), 1-year-old children suffer the second highest rate (4–5/100,000), and children between the ages of 6 and 12 years suffer the third highest death rate (3/100,000).

The age demarcation between good and poor outcomes in morbidity and mortality resulting from head injuries in childhood is sharp, occurring at the time of fontanelle closure. If one studies age distribution histograms of children with open and closed fontanelles, it becomes apparent that fontanelle closure occurs at approximately 1 year of age, permitting the clinician to superimpose the closed fontanelle population upon children older than 1 year of age. These observations (a higher incidence of skull fracture in children with open sutures and fontanelles, greater morbidity and a higher mortality in the same age category) suggest that infants are more vulnerable to blunt trauma than toddlers in that they suffer more craniocerebral damage and a much higher incidence of neurological deficit and death. Whether the primary impact itself or a delayed cerebral response to this primary impact, one resulting from very different cerebral anatomy and physiology, is the causative factor is not possible to ascertain at this time. The facts are that *infants have less chance of a good recovery after blunt head injury than do children older than 1 year of age*, 13.4% poor recovery in children less than 1 year of age and 4.9% in those between 1 and 3 years of age. Similarly, the highest incidence of admission to hospital for head injuries in childhood occurs during the first 3 months of life (18%). It then falls off in an almost linear fashion to reach the lowest incidence between the 18th and 21st months of life (3%), before leveling at a plateau of 7% during each trimester between the 21st and 36th months of life.

One may not, with certainty, attribute these very significant differences in age-incidence/hospital-admission epidemiology exclusively to noxae characteristic of individual trimesters during newborn, infancy, and toddler ages of life: passage through the birth canal, falls in the delivery room, slipping from the parents' arms during feeding, falls from a high chair or bassinet, crawling over the siderails of the crib, running and playing, and so on. The corollary of these observations – one which may explain why the rule *the younger the child the greater the damage to the brain from blunt trauma* – applies: it may be that open fontanelle and sutures predispose a child to a higher incidence of subdural hematoma. It has been observed that 32% of the open fontanelle population suffer subdural hematoma as a consequence of blunt head injury, whereas only 5% of the closed fontanelle population suffer this complication. In fact, in a review of our own work, we observed that 79% of all post-traumatic intracranial mass lesions were in infants and that 93% of these were subdural hematomas! Of these patients 7.5% (4 of 53) were toddlers, most suffering a CCS admission of 11 and none suffering a CCS below 8.

The newborns and infants suffering subdural hematomas are generally admitted late, at a time when they are symptomatic (failure to thrive, seizures, vomiting, lethargy) with clinical evidence of increasing intracranial pressure (split sutures, bulging fontanelle, sunset phenomenon, etc.). Toddlers and older children, however, most often present in hospital immediately after a head injury significant enough to cause a subdural hematoma. Their long-term symptoms and signs are expressive of either a postconcussion syndrome (irritability, diminution in academic performance, headache, behavior change, etc.), or of severe cerebral damage. One may only hypothesize concerning the protection which closed sutures and fontanelles provide against the formation of subdural hematomas. Among the possibilities are that the clot formation may more easily assume clinically significant volume within an expansile skull, that the absence of pacchionian granules and arachnoidal adhesions to the dura both predispose to tearing of bridging cortical veins and the accumulation of blood along the vertex, and that the malleability of the skull permits more severe compression and distortion injury of the brain.

The occurrence of the subdural hematoma (whether acute or chronic) is responsible for the extreme difference in outcome between open and closed fontanelle populations. This is expressed by the fact that excluding all subdural hematoma patients (83 of 462 in those studied by us) results in a drop of poor outcome from 4.9% to 2.8% in the closed fontanelle population and from 13.9% to 4.6% in the open suture population. Still, however, children under 1 year of age suffer a higher incidence of poor outcome.

One is confronted, consequently, with a series of dilemmas when discussing craniocerebral trauma in *children.* In the literature, it is almost invariably assumed that children are everybody under 18 years of age. The anatomical, physiological, developmental, and sociological differences among the individual age categories of childhood are enormous, so that the clinician must separate one group from another when studying, or treating, head-injured children: the child evolves through entirely different structural and functional conditions as it ages. During the first 3 years of life, the central ner-

vous system continues to mature at almost the same rate as during the intrauterine period. Progression from the newborn existence (approximately 22 h of sleep, with the waking intervals being limited almost entirely to feeding) to the 30-month-old child (who speaks, obeys commands, engages in meaningful play activities, and solves problems) is an infinitely greater change than those that occur over any other period in a lifetime.

At birth the brain neuronal population is the same as in adult life, but the glial cell proliferation, synaptic connections, and dendritic arborization have only begun. They progress almost logarithmically throughout the first 2 years of life, whereas myelinization of the central nervous system begins with the 1st year of life, but then continues progressively through the 10th year. By 4 years of age, the child's brain weighs approximately 75% of the adult, whereas it is only 25% of that weight at birth: the result of glial growth, increased volume of neurons and axons, and myelinization. In fact, cerebral volume is not expressive of greater amounts of brain water, since there is a rapid decline in brain water during the first 2 years of life, at which time the child's brain is hydrated to the same extent as the adult. These structural factors are the basis for the characteristic pathoanatomical brain injuries suffered by infants less than 3 months of age – tears of the corpus callosum, the midbrain at its junction with the cerebral hemispheres, subcortical white matter, and the temporal and orbitofrontal lobes. The anatomical factors causative for these lacerations are malleability of the skull, quasi-gelatinous consistency of a brain composed almost entirely of cells and without myelinated axons, small subarachnoid spaces, and large basal cisterns.

On the other hand, older infants suffer tears at the pontomedullary junction, diffuse petechial hemorrhages, and cellular necrosis. *Pari-passu* with the changes in cerebral structure, there are significant alterations in mesenchymal (skull, periosteum, sutures, dura) anatomy. The floating squamous bones of the calvarium approach and then join one another, the fontanelles become obliterated as membranoous bone forms between the two layers of the periosteum, the sutures lose their anatomical definition as a mesenchymal bridge between the endosteum (outer layer of the dura) and the periosteum, the tables of the skull thicken, and the diploë converts from sinusoidal chambers to an irregular mass of channels running perpendicular to the surface of the skull. Thus, by 1 year of age, the clinically evident cutoff point between a high and low incidence of subdural hematoma, the sutures have closed and the skull has become solid.

It is precisely this anatomical maturation, progressing from a mass of developing cells covered by pliable and isolated plates of membranous bone tethered at the sutures, into solidly protected, highly specialized nuclear aggregates with myelinated axons, which makes it impossible to use the same clinical criteria to evaluate the nature, severity, and course of craniocerebral injury in the different ages of childhood; and dangerous to grade childhood head injury along the lines of criteria used in adults!

These anatomical, physiological, and developmental facts have been uniformly ignored by authors writing on the subject of pediatric head trauma. In fact, neonates, infants, toddlers, juveniles, and adolescents (and, alas, young adults) have invariably been considered a homogeneous group. Of especial significance is the fact that, with only one exception (the Raimondi Hirschauer paper) this subject has never been studied – reported – chronologically, analyzing clinical observations in monthly, or yearly age-time frames in the critical first 3 years of life.

The efficacy of surgical or medical (e.g., the use of Decadron), as well as intensive care and management with the institution of barbiturate coma, has been measured using the GCS (a grading system derived exclusively from the study of adults!) and then evaluated from a cohort of *all* ages of childhood. This has resulted in imprecise reporting of the incidence of subdural hematoma, intracerebral hematoma, cerebral laceration or contusion, brain swelling, cerebral vascular dilation, and alterations in circulating cerebral blood volume.

The GCS is a standardized, and generally accepted, convention for evaluating the severity of head injury in adults, *but it is not applicable to infants or toddlers!* It is based upon interpreting varying degrees of higher integrative functions (obeying command, being oriented, spontaneous eye opening). Such a system permits disarticulation and resolution of individual integrative functions, *but does not allow one to interpret functional alterations at subcortical and brainstem levels,* which are normal performance levels for newborn and infants. In fact, "a normal infant would not score better than 4 (of 6) on the motor exam (flexor withdrawal) or 2 (of 5) of the verbal exam (incomprehensible sounds)" on the GCS. Also, the response to eye opening other than spontaneous would be misleading, since newborn and infants generally close their eyes when feeling pain.

In addition to these obvious inadequacies of the GCS in evaluating craniocerebral injury in childhood, one must consider the fact that we do not know how much time is necessary to evaluate the effects of cerebral damage. Some forms of damage become less noticeable over the years and others remain obscure until a later age when more demands (motor and intellectual) are put upon the brain.

"Moderately" brain-injured infants and toddlers very often do not show signs of sensory, motor, cognitive, or behavior impairment for many years, despite "apparent" complete recovery soon after the injury. (This is as true for cerebral injury as it is for hydrocephalus, me-

ningitis, porencephaly, vasospasm, and the dysraphic state). Very severe neurological deficits are the rule when porencephaly or meningitis occur in the neonatal period, and moderate deficits are much more common when meningitis and encephalitis occur during the first 2 years of life.

One cannot accept the anecdotal reports that less damaging long-term effects result from injuries suffered in the early years of life, that young children "outgrow" or are more resistant to cerebral damage. Approximately 20% of head-injured children suffer hyperkinesis, difficulty in anger control, impaired attention, and headache: elements of the "post-traumatic syndrome." Also, studies of delinquent children with psychiatric problems reveal that a statistically significant number of them had suffered head or face injuries in childhood. Therefore, normal neurological or IQ examinations in children who have suffered head injuries are no assurance that permanent and significant brain damage has not occurred.

In a publication coauthored by Dr. Jeffrey Hirschauer [1], we reported on a study of 462 head-injured children (between 1 and 36 months of age) in whom the injuries ranged from trivial to deep coma. Most of this introductory text is transcribed from that work. Penetrating head injuries were not considered in that study, nor were birth injuries. The results permitted the categorization of a Children's Coma Score (CCS) and the preparation of a Children's Outcome Scale (COS), both predicated entirely upon direct observations of injured infants and toddlers.

The three elements of the neurological examination that form the basis of the Children's Coma Score are motor, ocular, and verbal, with the total score being the sum of the three subscores:

Ocular response: maximum score = 4
- 4 Pursuit
- 3 Extraocular muscles (EOM) intact, reactive pupils
- 2 Fixed pupils or EOM impaired
- 1 Fixed pupils and EOM paralyzed

Verbal response: maximum score = 3
- 3 Cries
- 2 Spontaneous respirations
- 1 Apneic

Motor response: maximum score = 4
- 4 Flexes and extends
- 3 Withdraws from painful stimuli
- 2 Hypertonic
- 1 Flaccid

- Total maximum score assignable = 11
- Minimum score = 3

Since infants are unable to speak and injured toddlers generally neither obey commands nor respond appropriately to verbal stimuli, we allocated no points to responses requiring such complex behavior as speech or stimulus localization.

Five COS categories were designated:

I Excellent recovery
II Moderate, but nondisabling deficit
III Either a severe motor or cognitive deficit
IV Vegetative
V Death

Categories I and II are considered "good" outcome, III–V "poor" outcome.

Comparison of the GCS with the CCS permits one immediately to recognize the fact that the maximum score on the GCS is 15, whereas it is only 11 on the CCS. In addition to this, one notes that the former permits good discrimination of higher integrative functions, whereas the latter provides for evaluating subcortical and brainstem functions. In fact, comparing the GCS with the CCS for higher integrative functions reveals that in the GCS a score of 12–15 is awarded when the patient is either "normal" or at the very least able to open his eyes to command, and is at its worst when he is not responding (motorwise or cognitively) either to verbal command or pain.

None of these higher integrative functions are testable in infants and toddlers. Cortical functions are expressed by a score of 9–11 on the GCS, but infants and toddlers must get full marks, 11, on the CCS, the score of a normal child in this age range. Subcortical functions score out at 5–8 on the GCS, and 8–10 on the CCS, illustrating that the CCS is more sensitive for appraising subcortical function. Adults who are functioning at the brainstem level score from 3 to 4 on the GCS, whereas infants and toddlers score 3–7 on the CCS.

Despite the fact that seizures, split sutures, a bulging fontanelle, and a high-pitched cry are the common presenting signs of increases in intracranial pressure in infants, thus resulting in a much higher percentage of head-injured infants being brought to the hospital early and treated early, infants suffer greater cerebral damage and do not do as well as toddlers. There is no greater incidence of seizures, lateralizing signs, or skull fractures in either age category. Neurologically intact children with open sutures at the time of admission to hospital are more likely to have a "poor" outcome, an observation that applies across the full range of admission CCS right down to a score of 3.

The single most reliable examination for evaluating the outcome in children under 3 years of age is the *ocular examination*, being most consistent with regard both to "good" and "poor" outcome. About 95% of children with an ocular examination of 4 do well, whereas no child with an ocular examination of 1 survives. This degree of reliability does not occur in either the motor or verbal evaluations, since one may predict that approximately 20% of the children in the lowest motor ca-

tegory and 40% of those in the lowest verbal category may enjoy a good outcome. It may well be that the ocular examination is most reliable, and that there is greater consistency between the ocular examination and outcome over the first 2 years of life than, for example, motor evaluation, because all oculomotor functions (including near reflex and ocular pursuit) are present by the time a child is 2 months of age. On the other hand, only the most basic motor functions – sucking, reflex grasp, startle reflex – are present in a 2 month old, whereas a 2 year old has a full range of fine motor movement. Cortical spinal myelinization occurs much later than optic pathway myelinization. The same maturation processes for respiration explain the relative unreliability of the verbal categories. Apnea is very common in the non-brain-damaged newborn, whereas it is nonexistent in toddlers.

In comparing the three aspects of the Children's Coma Score (ocular, verbal, motor), and applying the expected observations to the clinical condition, one may expect that 100% of the children with closed fontanelles and 94% of those with open fontanelles will have a "good" outcome with an ocular score of 4, 79% and 64% with an ocular score of 3, and 55% and 22% with an ocular score of 2. The motor examination permits one to conclude that 100% of the closed fontanelle and 94% of the open fontanelle children with a motor score of 4 have a "good" outcome, whereas the percentages change to 81 and 42 with a score of 3, and 0 and 100 for those with a score of 2. In the verbal examination, 98% of the closed fontanelle and 92% of the open fontanelle children with a score of 3 enjoyed a "good" outcome, whereas the percentages were 56 and 8, respectively, for those with a score of 2.

Radiographic evidence of *post-traumatic splitting* of the sutures is indicative both of a "poor" outcome and a higher incidence of post-traumatic seizures. Palpation of the open fontanelle provides good evidence regarding outcome, since only 5% of the children with a soft fontanelle suffer a "poor" outcome, 16% of those with a full fontanelle suffer a "poor" outcome, and 50% of those with a tense fontanelle suffer a "poor" outcome. Only 12% of the children with a soft fontanelle have subdural hematomas, whereas the percentages rise to 71 and 83, respectively, for those with full and tense fontanelles.

The presence of a *linear skull fracture* may not be correlated with outcome, unless it is bilateral! In 7% of the children without, and 4% of those with, linear fractures the outcome is "poor," whereas 26% of the children with bilateral fractures suffer a "poor" outcome. One must also distinguish between a linear fracture and a diastatic fracture, since 33% of the children with diastatic fractures suffer "poor" results.

Post-traumatic *seizures* occur in 10% of the children with depressed fractures and 7% of those without fracture. Irrespective of whether a child's sutures and fontanelle are still open or closed, ocular deviation and hemiparesis may not be correlated with a "poor" outcome, whereas simple hemiparesis (in the absence of ocular deviation) is significantly associated – in 26% of the cases – with a "poor" outcome. The presence of an extensor plantar response does not alter these relationships. In fact, a unilateral Babinski reflex, along with hemiparesis and ocular deviation, are indicative of a benign course in 97% of head-injured children under 3 years of age. Whether the coexistence of this triad is expressive of a seizure, rather than discrete neurological destruction, is not known, though approximately 62% of the children with this triad may be expected to suffer focal motor seizures sometime during the hospital stay, and 23% of them remain permanently epileptic. When these numbers are compared to the incidence of post-traumatic seizures in the general population (only 3%!) this correlation becomes most meaningful. It is also of value to realize that such hard evidence of neurological deficit as an extensor plantar response, ocular deviation, and hemiparesis actually are indicative of a benign course. Developing upon the possibility that the triad – extensor plantar response, ocular deviation, hemiparesis – may be a seizure equivalent, and that it is correlated with a benign course, one must immediately separate this clinical picture from tonic-clonic post-traumatic seizures, whether focal or generalized. Of the children with focal seizures, 14% suffered poor outcome and another 14% had recurrent seizures, with 69% of the post-traumatic seizure patients being less than 1 year of age. About 27% of children with generalized seizures suffered a "poor" outcome and 10% have recurrent seizures, with 76% of the patients being less than 12 months of age. Late seizures occur more commonly in children who suffer early focal seizures, and 66% of the patients who develop late seizures may be expected to suffer a poor outcome. The incidence of post-traumatic seizures is higher in infants than in toddlers (26% vs. 10%), with evidence that the toddler has the same incidence of post-traumatic epilepsy as does the adult. In comparing the age categories, 26% of the newborn, 18% of the children between 1 and 12 months of age, and approximately 10% in patients older than 1 year of age, one realizes that the seizure rate drops after the 1st month of life, remaining high during the 1st year, and then leveling off thereafter.

Bilateral retinal hemorrhages correlate very directly with poor outcome, whereas a unilateral retinal hemorrhage is of no significance at all. Split sutures and bilateral retinal hemorrhages coexist very often, both indicating a sudden and severe rise in intracranial pressure. In fact, 65% of the children with bilateral retinal hemorrhages have extra-axial hematomas. These signs indicate to the clinician that immediate surgery is essential, especially in light of the fact that timely removal of the

clot is associated with a good outcome in exactly the same percentage of cases as subdural hematoma alone, without split sutures or retinal hemorrhages: 69% "good" recovery in both groups. One may conclude that unilateral retinal hemorrhages are qualitatively and quantitatively different from bilateral hemorrhages – less severe, occur in older children, less often associated with extra-axial hematoma – and *that bilateral hemorrhages are a reliable sign indicating the need for emergency treatment.* They are a result of an acute increase in intracranial pressure, as are split sutures and diastatic fractures. In fact, however, hemorrhages are more reliable as an indicator of an increase in intracranial pressure than a bulging and tense anterior fontanelle, since an expanding extra-axial hematoma does not invariably cause a tense or bulging fontanelle.

Approximately 79% of the children suffering intracranial mass lesions are less than 1 year of age and subdural hematoma is the pathological lesion in 93% of the cases, with the chronic variety occurring in two-thirds of the children. There is no difference in the outcome of children with subdural hematoma treated with bur holes, subdural puncture, or subdural peritoneal shunts. In fact, these children may have all three forms of treatment, and the subdural fluid continues to reaccumulate. Those treated with craniotomy and evacuation of the hematoma enjoy a much better outcome, 73% "good" results in open fontanelle and 62% "good" results in closed fontanelle children. If "hanging veins" are present on cerebral angiography, lowering of the superior sagittal sinus is effective treatment in 90% of the children. Epidural hematoma and intraparenchymal edema are most unusual in open fontanelle children, though they represent 25% and 12%, respectively, of the intracranial clots in toddlers. An epidural hematoma in a toddler should invariably be associated with good outcome, if treated before pupil changes occur, with a "poor" outcome if treated after pupil changes have occurred, and with either the vegetative state or death if the pupils are fixed and dilated. On the other hand, irrespective of the status of the pupils, intraparenchymal mass lesions are associated with severe edema and poor outcome.

Injuries of the Scalp

Scalp injuries range from abrasions through subgaleal or subperiosteal hematoma to lacerations of various depth. The scalp abrasion is best treated with a simple scrub with surgical soap and application of antiseptic, without shaving the area, since this only adds to the scalp injury and presents the risk of bringing contaminants into the depth of the dense connective tissue. Subgaleal or subperiosteal hematomas are not to be treated under any circumstances. Puncturing the subgaleal or subperiosteal space to remove hematoma is to be discouraged because removal of the freshly formed clot is difficult; the disadvantages rest in the risks of introducing bacteria into the hematoma (a superb culture media) with complicating subdural or subperiosteal empyema resulting. The subgaleal empyema is easy to manage in that one need only incise the scalp, give egress to the pus, and insert Penrose drains. The subperiosteal empyema, on the other hand, is extraordinarily painful, presents grave risks of osteomyelitis, and may necessitate removing varying amounts of the calvarium.

Scalp lacerations are closed in the emergency room, using the same technique for hemostasis and scalp closure as described in Chap. 2, unless the child is to be taken to the operating room because of a surgical head injury.

Fractures

The determining factors in fracture management are whether the fracture is compound or depressed, since, generally speaking, compound fractures necessitate immediate surgery, and depressed fractures, when not compound, may necessitate (elective) surgery.

Linear Fractures

The linear skull fracture is, in and of itself, of no surgical significance. It assumes "warning" value when it traverses the course of a meningeal artery or dural sinus. In essence, a change in clinical signs, expressive of increasing intracranial pressure or a focally expanding mass lesion, in a child who has a linear fracture that crosses one of the above-mentioned vascular structures, obliges the surgeon to conclude that a hematoma is accumulating until proven otherwise. One should be alert to the possibility that a linear fracture across the transverse sinus may result in an epidural hematoma expanding within the supratentorial compartment, the infratentorial compartment, or both. Similarly, a linear fracture across the SSS may indicate an epidural hematoma expanding to the right or the left of the sagittal plane, or, more ominous still, one expanding directly over the midline and compressing the SSS.

Arterial epidural hematomas, resulting from torn meningeal vessels, are common in infants, but they do not occur (almost) exclusively within the distribution of the middle meningeal artery as they do in the adult. In fact, epidural hematomas from tears in posterior meningeal vessels are as common as those resulting from middle meningeal injury. The most common cause of epidural hematoma in the infant is diploic bleeding, in the toddler bleeding from a meningeal artery.

Diastatic Fractures

Diastatic fractures may be classified either as *linear, comminuted*, or *stellate*, depending upon the number and form of fragments one may identify. In its simplest terms, however, it is best to consider a diastatic linear fracture as one consisting of a break in skull continuity, with the bone edges separated by more than 5 mm over a distance of greater than 2 cm.

These fractures are all too often harbingers of either epidural hematoma or meningocele spuria. It is recommended that they not be operated on, that the child be followed closely, and that there be a high index of suspicion for any of the already described complicating clinical conditions. One may, however, be justified in choosing to explore diastatic linear fractures, so as to inspect for the presence of an epidural hematoma or a tear in the dura. Given the potentially lethal complications of the former and neurological damaging complications of the latter, this course of action is advisable. If any epidural hematoma is found, the diastatic fracture is simply extended into a craniotomy adequate for evacuating the hematoma, and stopping dural bleeding and oozing from the inner surface of the skull. If a dural tear is found, the dura is opened and the subdural space inspected. If a subdural hematoma is encountered, it is dealt with surgically. If not, the dura is closed and one proceeds with scalp closure. This prevents the formation of a meningocele spuria.

Basal Linear Fractures

Linear fractures of the base of the skull are not managed surgically. It is almost impossible to diagnose them neuroradiologically in the newborn and infant, and extraordinarily difficult to do so in the toddler and adolescent. One may, categorically, make the diagnosis of basal linear fracture if one finds either hemotympanum in the absence of a ruptured drum or battle sign (or raccoon eyes) in the absence of direct injury to the temporal or naso-orbital areas.

There has been much discussion concerning the advisability of treating linear fractures of the base of the skull prophylactically with antibiotics. Some have sustained that antibiotics should be used only when there is cerebrospinal fluid rhinorrhea; others feel they should be used immediately after the diagnosis of a linear fracture of the base of the skull is made. Still others think there is no place for prophylactic antibiotics in the management of these injuries.

Depressed Fractures (Fig. 13.1)

Closed depressed fractures may be elevated or not, depending upon the orientation and experiences of the individual neurosurgeon. There are no hard and fast data concerning the association of a persistent, untreated, depressed fracture with permanent neurological deficit or seizures. However, most neurosurgeons accept the arbitrary determination that a depression of 5 mm or greater indicates surgery. This seems reasonable, especially in light of the fact that an operative procedure for elevating a depressed fracture presents minimal risks,

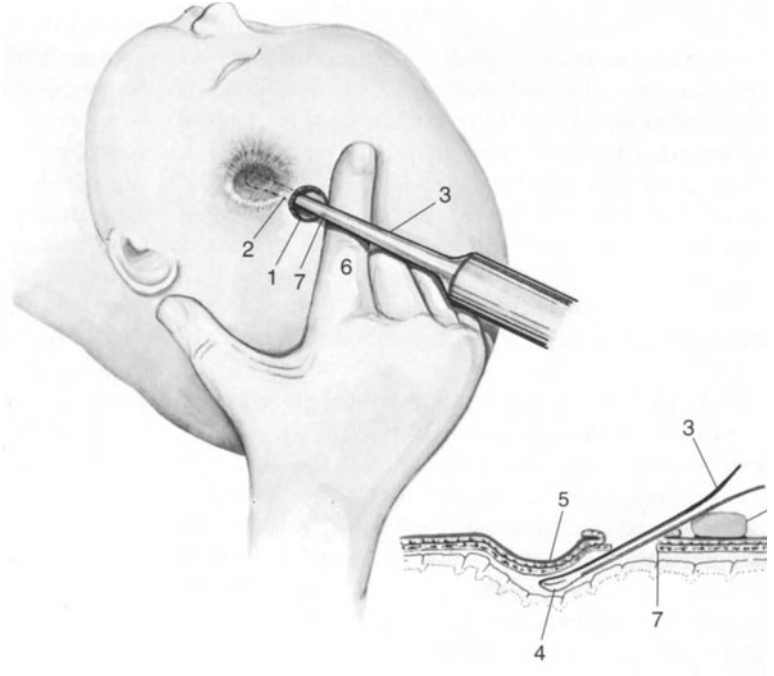


Figure 13.1. The "ping-pong" fracture is elevated by placing a bur hole opening (*1*) immediately peripheral to the point where the normal skull ends and the depression begins (*2*). A periosteal elevator (*3*) is then inserted through the bur hole, bringing its tip (*4*) to the very nadir of the "ping-pong" fracture (*5*). The surgeon should take care to use his own finger (*6*) as the fulcrum, not the normal skull at the edge of the bur hole opening (*7*), lest he inadvertently cause another depressed fracture.

and that it affords the patient the added protection of having the depressed fracture elevated and the dura inspected for tears which, if present, may be repaired before a meningocele spuria develops.

In the newborn or infant, depressed fractures may be either "ping-pong" fractures, consisting of nothing more than a conversion of the post-traumatic concavity of the skull into a return to the normal convexity, with the depth of depression indicating the potential cerebral concussion. Comminuted fractures, with impaction of spicules into the dura or brain, are most unusual in the newborn and infant, as 'ping-pong' fractures are most unusual in the toddler and juvenile.

The ping-pong fracture is treated by making a perforator opening in the skull just peripheral to the limbus of the depressed area, and then slipping a Penfield #4, or #1, dissector, whichever is most appropriate, between the skull and the dura. The depression is then elevated, with the surgeon taking care to use his finger, *not* the bone edge, as the fulcrum for the dissector, since this latter could result in further depression of the calvarium.

Surgical management of a comminuted fracture does not necessitate placement of bur hole, since one simply removes the spicules, one at a time, with a fine-tipped rongeur, inspects the underlying dura, and then replaces the spicules and proceeds to closure of the scalp, if the dura is not damaged. If the dura is damaged, it should be opened to inspect the subdural space to ascertain that there is no clot present. The surgeon then proceeds to dural closure, replacement of the bony spicules, and scalp closure. Of course, this is proper management of closed, not compound, comminuted fractures.

Compound Skull Fractures (Figs. 13.2–13.4)

The compound depressed skull fracture represents an acute neurosurgical emergency, since one is obliged to debride the wound, inspect the fracture area for persistent bleeders, and remove bone spicules and foreign substances from the epidural space and surface of the brain. One should not explore the subcortical area of the brain for foreign substances nor should one enter this area in search of spicules identified either on CT scan or skull X-ray. Debridement is best limited to the surface, whether removing damaged brain, bone fragments, or foreign bodies.

The compound depressed fracture of the convexity of the calvarium is managed surgically, after surface debridement of the scalp and periosteum, in the same way as a closed comminuted depressed fracture, with one exception: in the compound depressed fracture the bony spicules and fragments are discarded, not reapproximated into their previous position and left in place! These fragments must be considered contaminated, possible foci for infection.

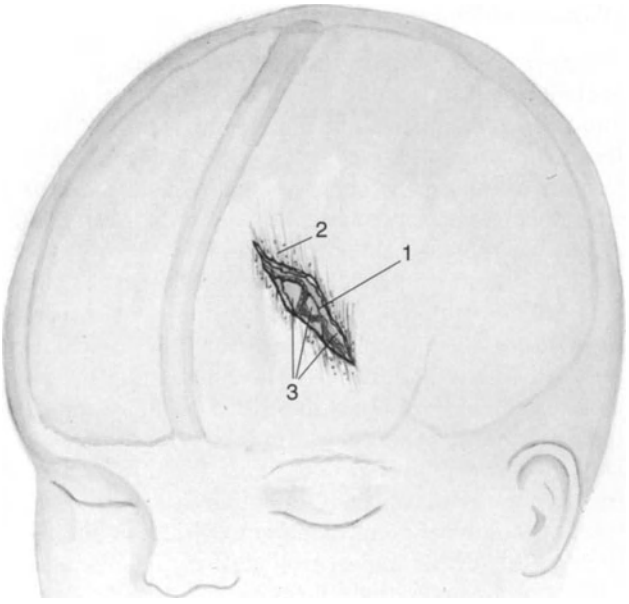
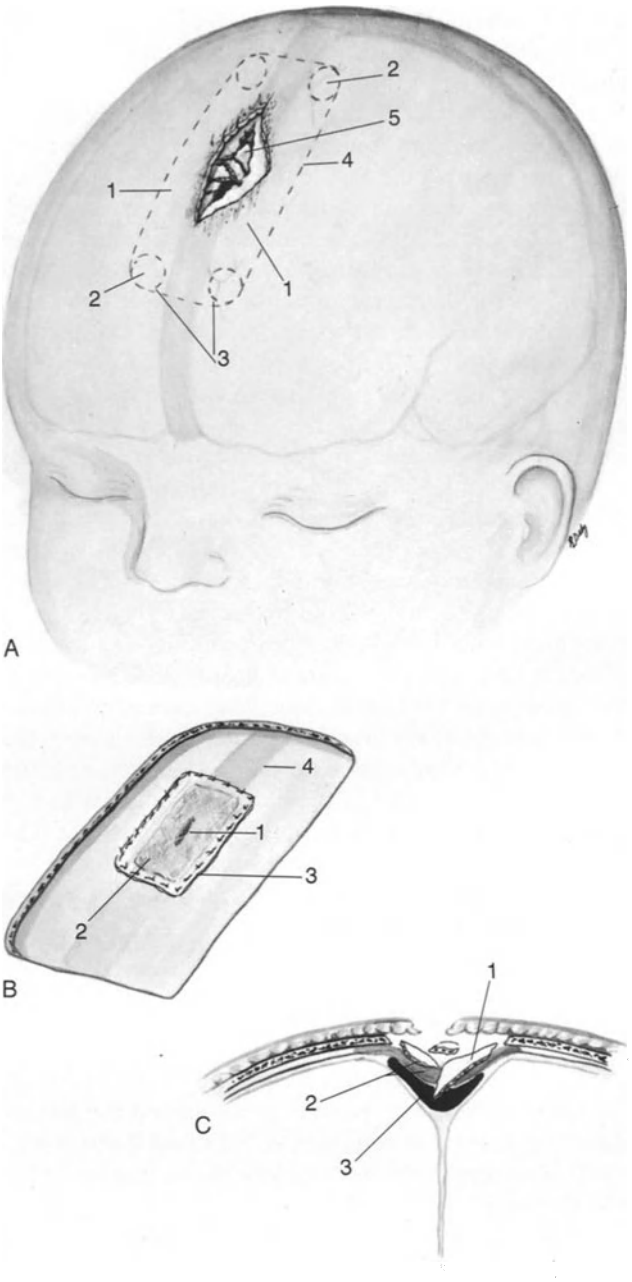


Figure 13.2. A compound depressed fracture is characterized by a full-thickness scalp laceration almost invariably accompanied by extensive tissue destruction from the compressive force, and single or multiple bony spicules, fragmented and, at times, impacted into the underlying dura and brain. The laceration (*1*) and surrounding areas of tissue damage (*2*) may overlay the bony spicules (*3*). This does not invariably occur.

The compound depressed skull fracture over the superior sagittal or transverse sinuses presents entirely different surgical implications, operative risks, and technique. Once one diagnoses a compound depressed fracture over one of these sinuses, either clinically or with skull X-rays, one should proceed immediately to CT scan, with coronal and sagittal reconstruction, in order to evaluate as precisely as possible the depth of penetration or compression of the sinus by impacted bones. Before operating, or making a decision to operate, one should perform MRI angiography, so as to evaluate the caliber and patency of the involved sinus beneath the impacted bone. If the sinus is almost completely occluded, and the clinical condition of the child is expressive of this occlusion, then the surgeon has no alternative but to proceed to elevate the depressed fragments and repair the lacerated sinus. If, on the other hand, there is only minimal compression of the sinus, or the child's neurosurgical condition is excellent, one should limit oneself to debridement and repair of the scalp, leaving the fracture fragments untouched. *The risks of opening a spontaneously closed rent in a major venous sinus are of such potentially lethal danger that the safest form of treatment entails the judgment that a potentially infected wound is much less dangerous to the child than a torn venous sinus.*



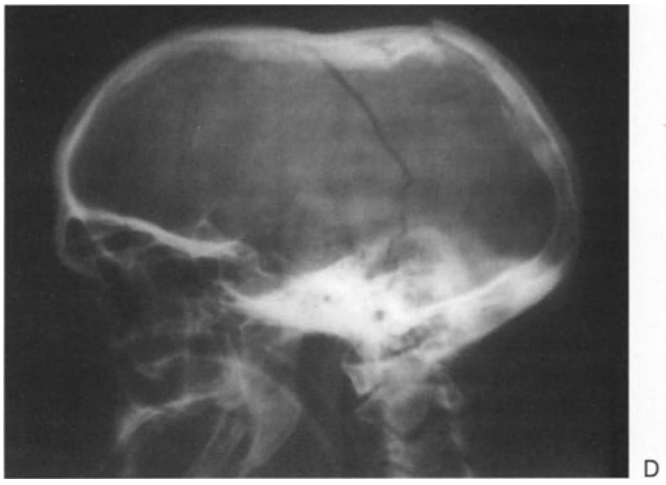


Figure 13.3. (A) In the event of a depressed fracture, compound or closed, over the superior sagittal sinus (SSS), one should reflect a quadrilateral bone flap (*1*), taking care to position the bur holes (*2*) so that their medial convexity borders upon the lateral surface of the SSS (*3*). Once the bur holes have been placed, and the osteotomy incisions (*4*) made, one may elevate the free bone flap, taking extreme care to identify and remove, when possible, the impacted spicules (*5*) of the depressed fracture. (B) After the bone flap has been reflected and the spicules removed, the SSS may bleed. Consequently, one should be prepared to repair it. Avitene is used to cover the rent in the sinus (*1*) and then this is compressed with a Surgicel strip (*2*), prior to bringing a periosteal graft (*3*) into the field and sewing it over the Surgicel strip and the SSS (*4*). (C) This is a cross-section representation of a depressed fracture over the SSS, illustrating the bony spicules (*1*) and small epidural hematoma (*2*) which is invariably present, causing the depression, deformity, and at times occlusion of the SSS (*3*). (D) This is a plain X-ray of the first coconut fracture we had ever seen and diagnosed as such: at a weekly conference at the Cook County Hospital in 1964, the Visiting Professor from India, Dr. Jacob Chandy, informed us that the fracture we were studying at that time (this illustration) occurred very commonly among Indian children...who sat under the palm trees to enjoy the shade. He coined the term "coconut fracture." We have adapted this term. Our case, however, resulted from a Coca Cola bottle being thrown from the roof of a high-rise building.

In the event one is obliged to remove the depressed fracture fragments from the superior sagittal sinus, one should be prepared to stop immediately the outpouring of venous blood and to proceed with repair of the sinus so as to guarantee patency. This entails extension of the scalp laceration into an S-shaped incision after the scalp has been shaved, the laceration has been washed and debrided, and the head draped for a biparietal (in the event of a depressed fracture over the SSS) or an occipital/suboccipital (in the event that the transverse sinus has been compressed) approach. Bur holes are placed 4 cm to either side of the depression, and 2.5 cm from the center of the involved sinus, thereby presenting the surgeon potential access to the entire sinus. The bur holes should be connected to one another by use of Gigli saw, freeing the bone flap from over the involved sinus. This permits the surgeon to lift simultaneously the bone flap and depressed fragments from the surface of the sinus, assuring complete access, and control, in the event the torn sinus reopens and bleeding occurs, as compressing fragments are lifted away. Discard the depressed fragments, irrigate the surface of the compressed sinus, and inspect it carefully. Avitene should be placed over the involved sinus and the freed bone flap placed back into position and anchored into place prior to proceeding with closure of the scalp. One may,

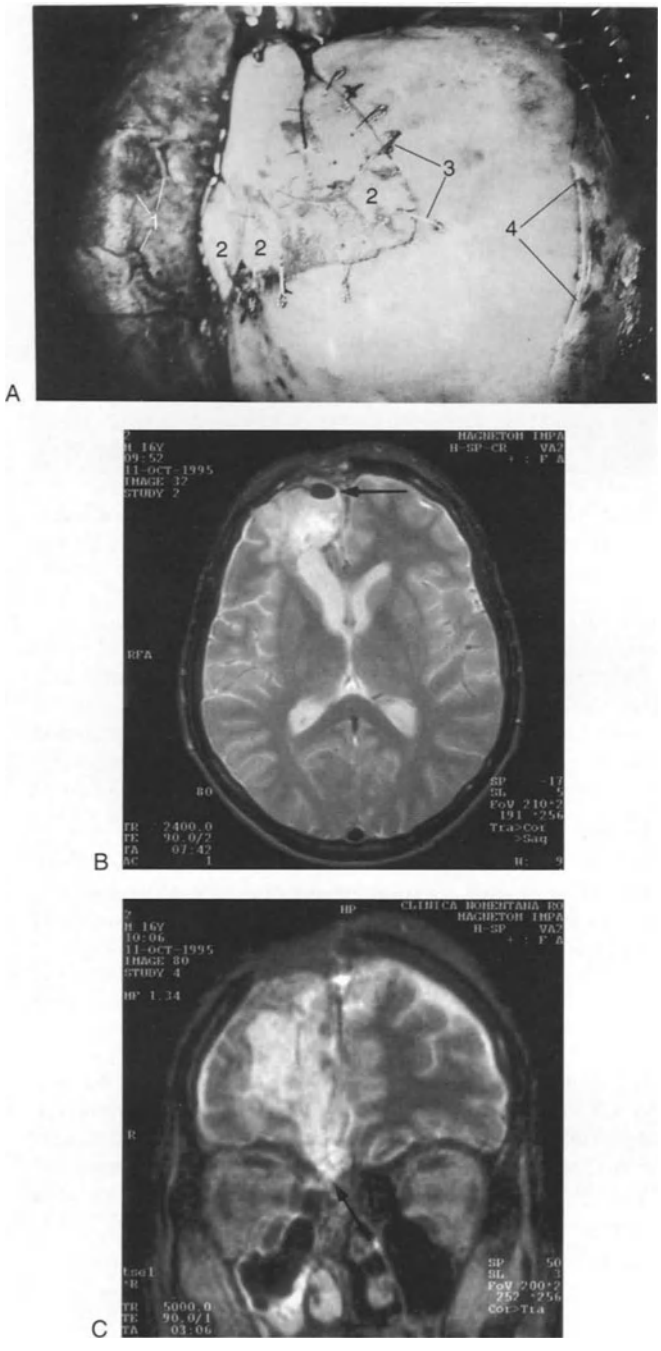


Figure 13.4. (A) Improper technique for management of compound depressed skull fractures. One notes the scalp laceration (*1*), multiple bone fragments (*2*), and wiring of these back into position at the time of closure (*3*). The compound fracture, by definition, *contaminates* the wound. The bone fragments are free and may readily be infected. In addition to this, the stripping of periosteum (*4*) devitalizes the remaining bone, subjecting it even further to the risk of osteomyelitis. (B) This is an axial SE2400/90 MR study of a post-traumatic craniocerebral injury which reveals malacia of the right frontal lobe with dilation of the right frontal horn and air bubbles indicating pneumocephalus (*arrow*). In (C) a coronal SE5000/90, the T2 weighted image permits easy identification of the frontoethmoidal bone defect with the cerebrospinal fluid leakage (*arrow*).

at a subsequent date, choose to perform a cranioplasty if the skull defect is so large as to present risks to the child.

In the event that the involved sinus bleeds profusely at the time the depressed fracture is elevated, one must immediately cover the rent in the sinus with a large fluffy cotton, and continue to irrigate the cotton as one applies the sucker to the bleeding site, thus both keeping the field dry and diminishing considerably the risk of air embolism. Gentle compression is applied to the rent in the sinus by both the tissue forceps and the suction tip, as the fluffy is gradually rolled toward the site of the tear, progressively applying smaller fluffies so that one has a more manageable working area. Prepared sandwiches of Avitene strips and Surgicel are applied to the surface of the sinus, bridging the rent, and fluffies placed over them immediately, as the sucker is brought to the site of maximum bleeding. This seals the Avitene to the sinus and closes the breech. If this suffices, one should leave the fluffy cotton over the sandwiches for a period of 10–15 min. At this time, another sandwich is placed over the tamponading one, and sealed to it, utilizing the same fluffy technique. One then removes the periosteum from the free bone flap, places it over the tamponading sandwiches, after having removed the fluffies, and anchors it to the underlying dura with 4–0 suture, attaining as tight an apposition as possible. The fragments of depressed fracture are discarded, and the freed bone flap is repositioned and anchored into place prior to closure of the scalp.

Cerebral Contusion and Edema

Cerebral contusion, if lobar and the focus of an expanding mass, should be resected as an emergency procedure. No attempt at resection is indicated if the contusion is centered in the parietal lobe or the insular cortex of either side.

In the past, many different operative procedures have been used, in vain, to treat cerebral edema: lobectomy of nonedematous brain, hoping to provide internal decompression; leaving the dura open and not anchoring the bone flap in place, hoping to allow some room for expansion of the swelling brain; performing a relaxing duraplasty and discarding the bone flap or performing radical craniectomy. There is no place in the management of acute head injuries, or chronic head injuries for that matter, for discarding a bone flap or extensive craniectomy with the hope that this will alleviate intracranial pressure and "give room for cerebral expansion." This procedure is without foundation, though it has been repeated over the years by neurosurgeons, with hope of minimizing cerebral edema. It results only in worsening the cerebral edema and further complicating the neurological damage.

Epidural Hematoma (Figs. 13.5, 13.6)

The arterial epidural hematoma may be located over the supratentorial convexity or the cerebellum. The venous epidural is located over the superior sagittal or transverse sinuses. Though, in point of fact, both the superior sagittal and transverse sinus epidural hematomas are collections of clot between the dura and the inner table of the skull, they are not conventionally considered to be the same as epidural clots secondary to tears in the meningeal arteries. Similarly, epidural clots between the squamous occipital bone and the dura of the posterior fossa have such different clinical presentations as to be commonly, and erroneously, considered other than "epidural hematomas." The fact of the matter is that in the child there is no common clinical picture that may be considered typical of an epidural hematoma. Rather, epidural hematoma is, in the child, an anatomical location of a rapidly collecting blood clot, no more, no less.

The technical implications are, indeed, quite different when considering a convexity (supratentorial) epidural hematoma on one hand, and a posterior fossa epidural on the other.

Convexity Epidural Hematoma

Convexity epidural hematomas may be midline, in which case they are located either over the superior sagittal sinus and are secondary to tears in this structure, or laterally (beneath the pterion or the parietal bone), in which case they are secondary to a tear in the middle meningeal artery or a diastatic fracture. The lateral epidural may result from a compressive/decompressive injury which causes an "explosive" force. This pushes the convexity of the calvarium into the brain and is followed, instantaneously, by an immediate rebound of the convexity of the calvarium, creating a vacuum between the skull and the dura, resulting in a stripping of the latter from the former. Consequently, hemorrhage may occur in the epidural space. This is the most life-threatening of epidural hematomas in the newborn and infant. Because of the rigidity of the calvarium in the toddler and juvenile, this pathogenetic sequence of events does not occur.

One must not expect invariably to find a torn meningeal vessel when lateral epidural hematoma is surgically evacuated. Rather, in the newborn and infant this is an exception. In the toddler and juvenile, it is a rarity. In the adolescent, it is exactly the same as in the adult. Following evacuation of the hematoma, one should make a diligent search for the bleeding source. If a rent is not found in a meningeal vessel, it may safely be assumed that the bleeding occurred from the inner surface of the skull. Generally, reexpansion of the brain will reapproximate the dura to the surface of the inner table of the skull so that recurrent bleeding will not occur. One should apply very soft bone wax to the inner surface of the skull, or, preferably, reflect a free bone flap along the limbus of the epidural hematoma. This separates that portion of the calvarium with inner table bleeding from the surrounding cranium and, therefore, eliminates the source of bleeding. Recurrent epidural hematomas generally result from continued bleeding from the inner table of the skull, generally not from rebleeding meningeal arteries.

Posterior Fossa Epidural Hematoma

The posterior fossa epidural hematoma results either from a diastatic fracture of the squamous portion of the occipital bone or a tear in the transverse sinus. They expand rapidly, result in a massive collection of blood in the epidural space and obliteration of the cisterna magna, and cause (1) compression of the hindbrain and (2) obstructive hydrocephalus. For these reasons, the clinical picture is one of rapid, if not clinically instantaneous, increase in supratentorial pressure secondary to triventricular hydrocephalus.

The surgical implications of such a catastrophic clinical entity as a posterior fossa epidural hematoma are, consequently, directed toward compensating the triventricular hydrocephalus, and evacuating the epidural hematoma. A single lateral ventricular drain compensates the triventricular hydrocephalus and a suboccipical craniotomy (inferior cerebellar triangle) permits one to evacuate the epidural.

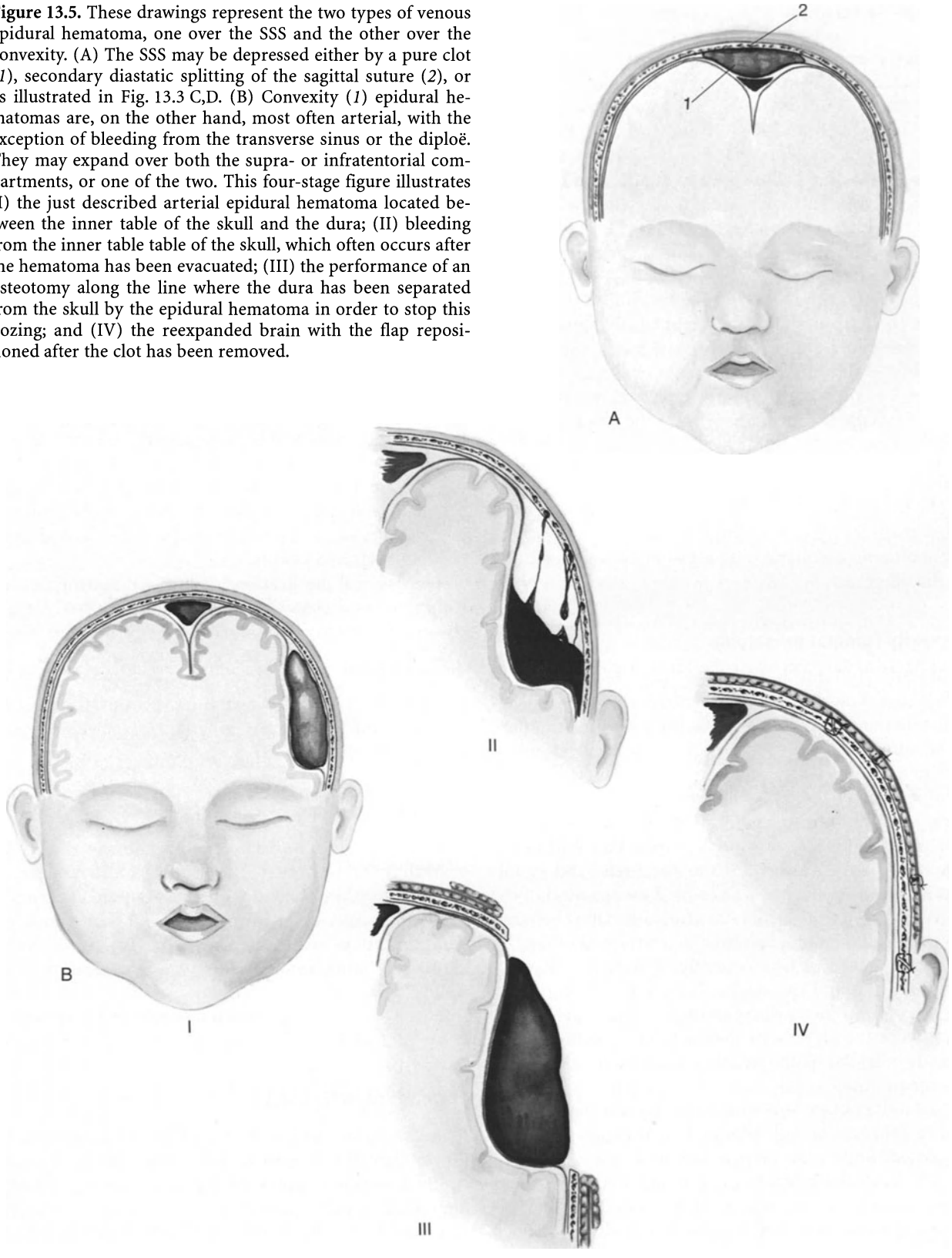
Subdural Hematoma

The subdural hematoma is a subdural collection either of fresh blood or liquefied derivatives of hematogenous breakdown. Consequently, one may speak in terms of acute, subacute, and chronic subdural hematoma, depending upon whether the blood is fresh, freshly clotted, or liquefied. Another, more commonly used though less commonly defined, classification of acute, subacute, and chronic subdural hematoma is presented in the following sections.

Acute Subdural Hematoma

Acute subdural hematoma is diffuse craniocerebral injury with varying degrees of scalp, skull, and cerebral contusion. This is characterized clinically by a severely depressed conscious level and anatomopathologically by cerebral contusion and fresh blood over the cerebral convexities, ranging in thickness from 1 to 3 mm. Surgical removal of this blood does not affect the clinical outcome.

Figure 13.5. These drawings represent the two types of venous epidural hematoma, one over the SSS and the other over the convexity. (A) The SSS may be depressed either by a pure clot (*1*), secondary diastatic splitting of the sagittal suture (*2*), or as illustrated in Fig. 13.3 C,D. (B) Convexity (*1*) epidural hematomas are, on the other hand, most often arterial, with the exception of bleeding from the transverse sinus or the diploë. They may expand over both the supra- or infratentorial compartments, or one of the two. This four-stage figure illustrates (I) the just described arterial epidural hematoma located between the inner table of the skull and the dura; (II) bleeding from the inner table table of the skull, which often occurs after the hematoma has been evacuated; (III) the performance of an osteotomy along the line where the dura has been separated from the skull by the epidural hematoma in order to stop this oozing; and (IV) the reexpanded brain with the flap repositioned after the clot has been removed.



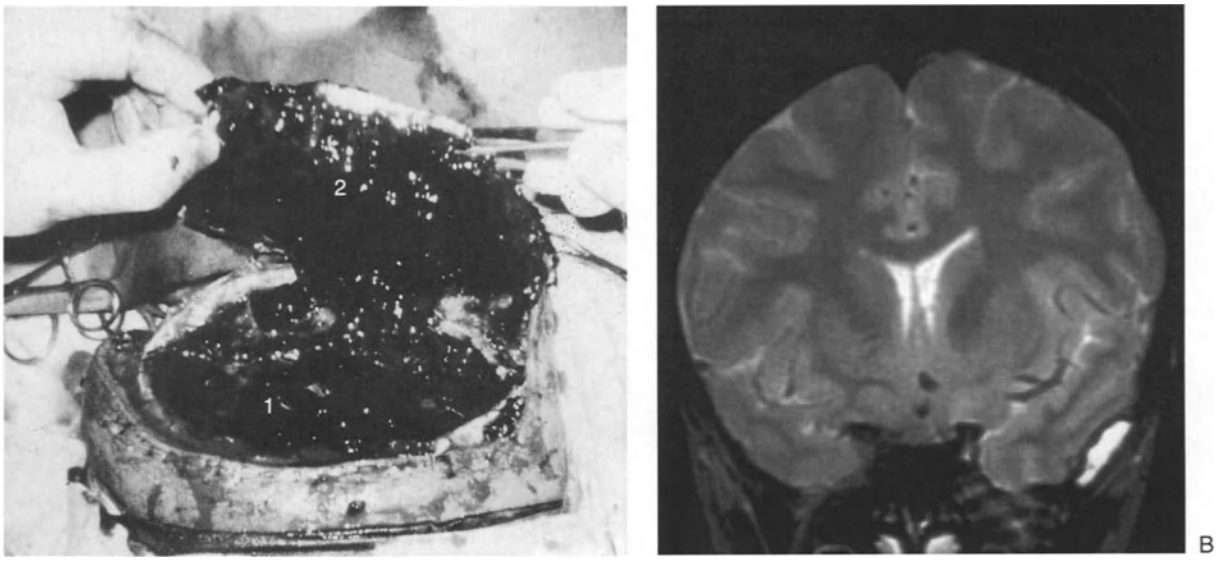


Figure 13.6. (A) Epidural hematoma, illustrating a clot over the dura (*1*) and along the inner table of the skull (*2*), so as to demonstrate that the craniotomy line may not always border upon the most peripheral portion of the blood clot. Consequently, epidural hematoma may extend beyond the line of craniotomy, separating the dura from the inner table of the skull, providing the pathogenesis for continuous oozing from the inner table of the skull. When this bleeding is persistent, one should perform an osteotomy as illustrated in the previous figure to the point where the dura is separated from the inner table of the skull. (B) This is a coronal SE2400/90 image of a small left epidural hematoma located over the temporal pole. The epidural is in the chronic stage, something we see more often now that we have magnetic resonance to monitor hematomas and no longer find it necessary to operate immediately all diagnosed epidural hematomas. This image shows an extraparenchymal area with high signal intensity and the rim of low signal intensity representing hemosiderin. The area of high signal intensity is an expression of methemoglobin. The shape of the lesion is typical for epidural hematoma. It should be noted that this hematoma was not operated on, but followed conservatively to its natural resolution.

Subacute Subdural Hematoma (Fig. 13.7)

Subacute subdural hematoma is characterized anatomopathologically by a freshly formed clot within the subdural space, one which is large enough to be pathogenetic for progressive deterioration in conscious level and neurological function. Timely removal of the clot reverses the clinical deterioration. CT and MRI studies suffice to make the diagnosis. The operative procedure, craniotomy and removal of the freshly formed clot, should be performed immediately the diagnosis is made. When the dura is opened, the clot is visible, though it generally does not deliver itself into the field, very likely since the degree of cerebral edema caused by subacute subdural hematomas in the early phases of their formation is not severe. The bleeding source is either a torn bridging cortical vein or a rent in a venous sinus. Cortical arteries may occasionally be damaged from blunt injury to the head, causing the subacute subdural. Similarly, cerebral contusion may be accompanied by a damaged cortical vein. Consequently, after the subacute subdural hematoma has been evacuated one must make a diligent search for the bleeding site, aware of the fact that *a single hematoma may be caused by more than one damaged vascular stucture.* No drain is left in the subdural space, the dura is closed, and the bone flap is anchored into position.

Chronic Subdural Hematoma (Figs. 13.8–13.11)

Chronic subdural hematoma is a complicated clinical condition, one which is not understood, and, most unfortunate of all, a waste basket diagnosis used to describe end-stage cerebral damage secondary to injury or infection.

The post-traumatic, chronic subdural hematoma in the infant represents one of the most common entities in pediatric traumatology. It is also the least understood and most controversial. There are many assumptions concerning this clinical entity. The first is that this type of subdural collection of blood is the result of acute bleeding into the subdural space. The increase in size is thought by some to be the result of its oncotic pressure and by others of repeated bleedings from the vascularized subdural membranes. It is also stated that the chronic subdural hematoma may be diagnosed simply by inserting a needle into the subdural space and obtaining egress of the liquefied hematoma. The last assumption is that the chronic subdural hematoma is generally cured by daily subdural taps and drainage of the

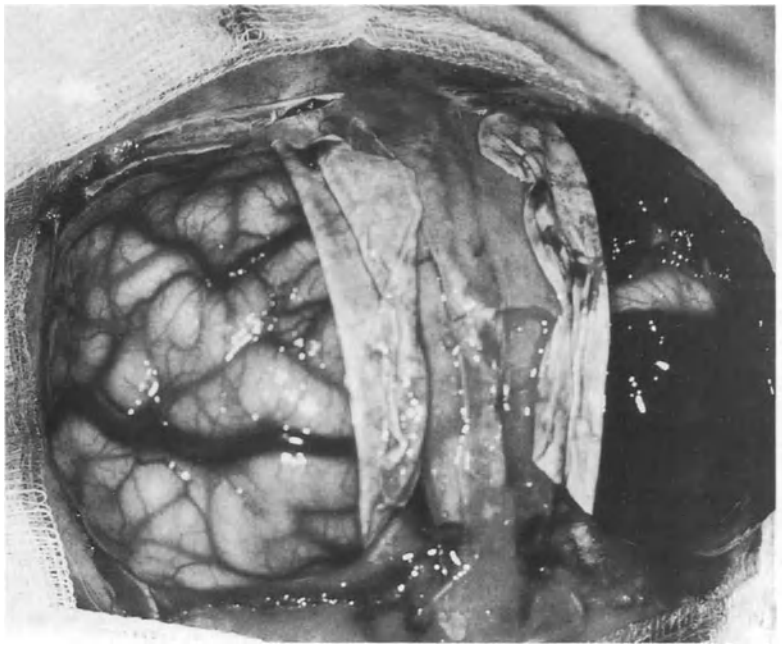


Figure 13.7. This is an operative photograph of a child with bilateral subacute subdural hematomas. The hematoma from the left has been removed and the one on the right exposed. In this child, the biparietal craniotomy was followed by simultaneous openings of the dura, so as to minimize chances of transfalcical herniation. This latter could result from removing the clot on one side and then proceeding to perform a craniotomy and durotomy on the other side.

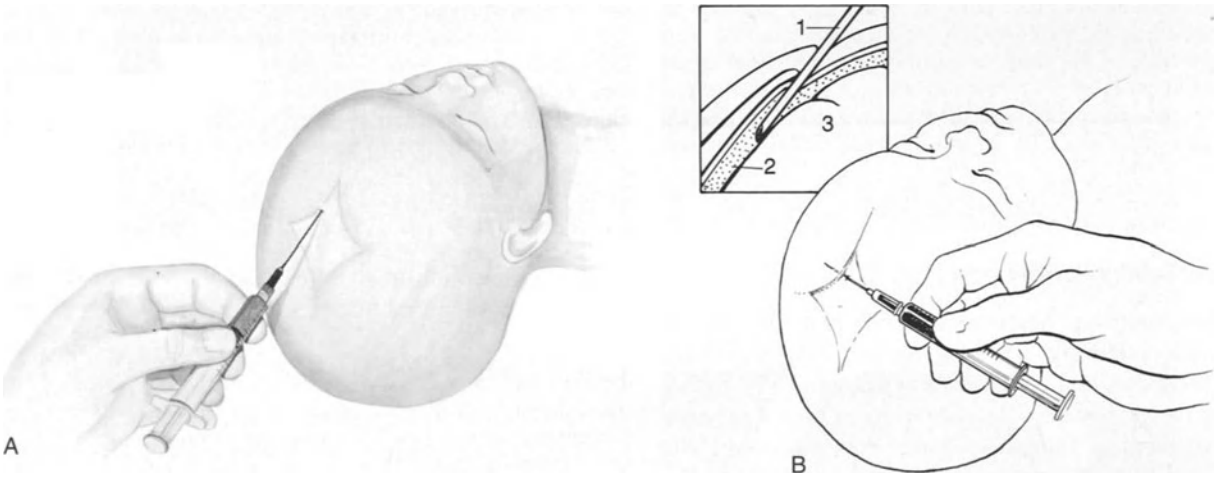


Figure 13.8. (A) Incorrect technique for performing subdural taps: the needle is directed perpendicular to the fontanelle, a direction which increases risks of penetrating the brain or damaging a cortical bridging vein. (B) The correct technique: the needle is inserted through the skin and beneath the dura, at an oblique angle, so as to minimize the risks of penetrating the brain. It is directed away from the cortical bridging veins. *The inset* shows the desired position of the needle tip (*1*) in the subdural space (*2*), not entering brain (*3*). This procedure has a role only where current imaging techniques are unavailable.

fluid. In the event that this needle drainage of the subdural fluid is not adequate, bilateral craniotomy and resection of the subdural membrane have been considered the definitive cure.

Unfortunately, the chronic subdural hematoma is not invariably the result of venous bleeding into the subdural space, nor is the progression in its size due primarily to the high oncotic pressure. Although insertion of a needle into the subdural space often permits one to diagnose a chronic subdural hematoma, this is not invariably the case; repeated needle drainage of the subdural collection of fluid is neither therapeutic nor safe, since fluid continues to reaccumulate and the needle may serve as a means for introducing bacteria into this ideal culture medium. Resection of the subdural membrane is not generally followed by a disappearance of the subdural hematoma and reexpansion of the cerebral hemispheres.

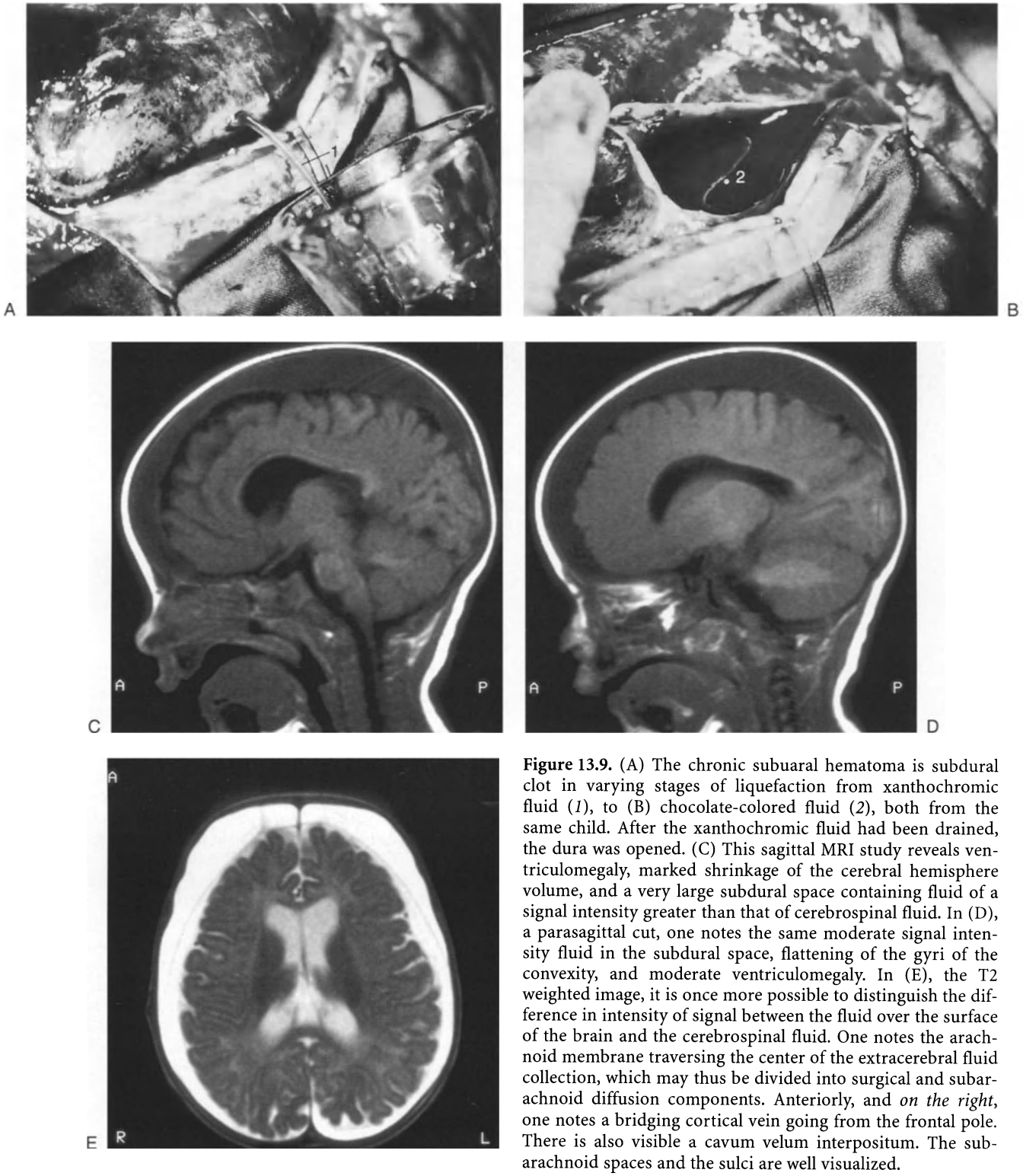
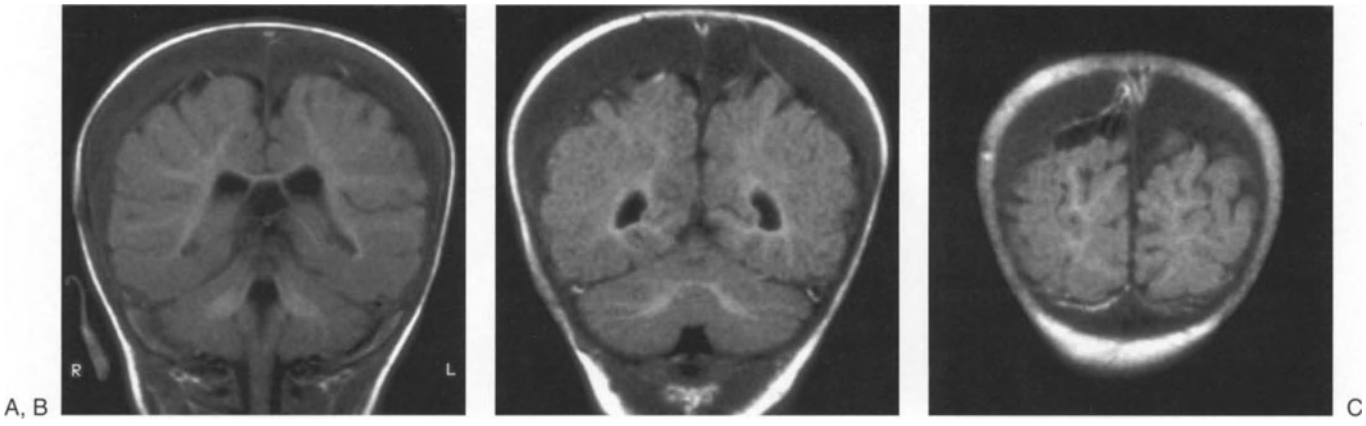


Figure 13.9. (A) The chronic subuaral hematoma is subdural clot in varying stages of liquefaction from xanthochromic fluid (*1*), to (B) chocolate-colored fluid (*2*), both from the same child. After the xanthochromic fluid had been drained, the dura was opened. (C) This sagittal MRI study reveals ventriculomegaly, marked shrinkage of the cerebral hemisphere volume, and a very large subdural space containing fluid of a signal intensity greater than that of cerebrospinal fluid. In (D), a parasagittal cut, one notes the same moderate signal intensity fluid in the subdural space, flattening of the gyri of the convexity, and moderate ventriculomegaly. In (E), the T2 weighted image, it is once more possible to distinguish the difference in intensity of signal between the fluid over the surface of the brain and the cerebrospinal fluid. One notes the arachnoid membrane traversing the center of the extracerebral fluid collection, which may thus be divided into surgical and subarachnoid diffusion components. Anteriorly, and *on the right*, one notes a bridging cortical vein going from the frontal pole. There is also visible a cavum velum interpositum. The subarachnoid spaces and the sulci are well visualized.



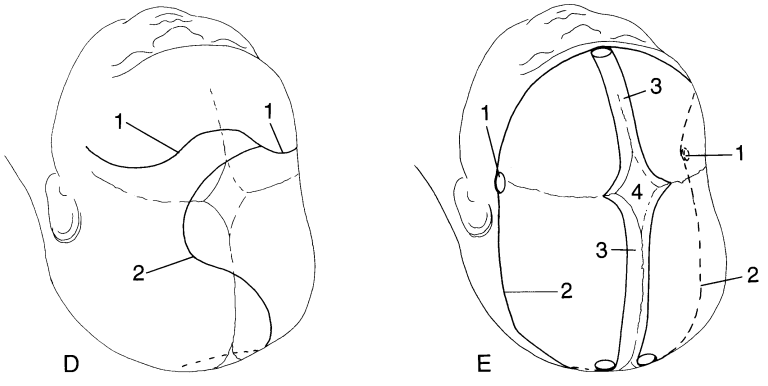


Figure 13.10. Chronic subdural hematoma progressing over time, resulting in massive accumulation of liquid within the subdural space and, to a certain extent the subarachnoid spaces. In (A–C) of this figure, coronal MRI scans are illustrated; in (D—E), a schematic representation of the pre- and post-superior sagittal sinus lowering procedure is presented. (A) This coronal section through the line of the posterior third of the body of the corpus callosum, pineal gland, and IV ventricle illustrates remarkable shrinking of the cerebral hemispheres and subdural fluid within both the subdural and subarachnoid spaces, though for the most part and especially over the vertex one may identify cerebrospinal fluid. Bridging veins may be seen to pass from both cerebral hemispheres across the accumulation of subdural fluid in the direction of the superior sagittal sinus. In fact, *on the left*, the vein may be followed from the cerebral cortex to the superior sagittal sinus. There is also an opening up of the velum interpositum. (B) This coronal section of the same study in the same child, cut through the occipital horns, reveals the very remarkable collection of subdural fluid and the bridging cortical vein *on the left*, passing from the superior parietal gyri to the superior sagittal sinus... across the subdural collection of fluid. (C) Again, the same child, and the same study but a cut through the occipital lobes illustrating the very large accumulation of subdural fluid and, most significant of all, the bridging cortical veins, going from the right occipital lobe to the superior sagittal sinus. *It is the preservation of the bridging cortical veins which permits one to prevent severe and devastating brain damage, to return the child to its normal cerebral function.* This cannot be accomplished simply by draining the subdural fluid. It is done by bilateral craniotomy, draining the subdural fluid, lowering and advancing the superior sagittal sinus, performing a duroplasty, and reconstructing the skull in a form of a *reduction cranioplasty.* These are schematically represented in (D) and (E) of this illustration. Technique for *lowering the superior sagittal sinus* and performing *reduction cranioplasty.* (D) The bifrontal (*1*) parasagittal S-shaped (*2*) incisions are herein illustrated. (E) The bur holes (*1*) and craniotomy margins (*2*) are indicated. Note that a sagittal strip of bone (*3*) is left on either side of the sagittal suture and that the craniotomy margin skirts along the borders of the anterior fontanelle (*4*).

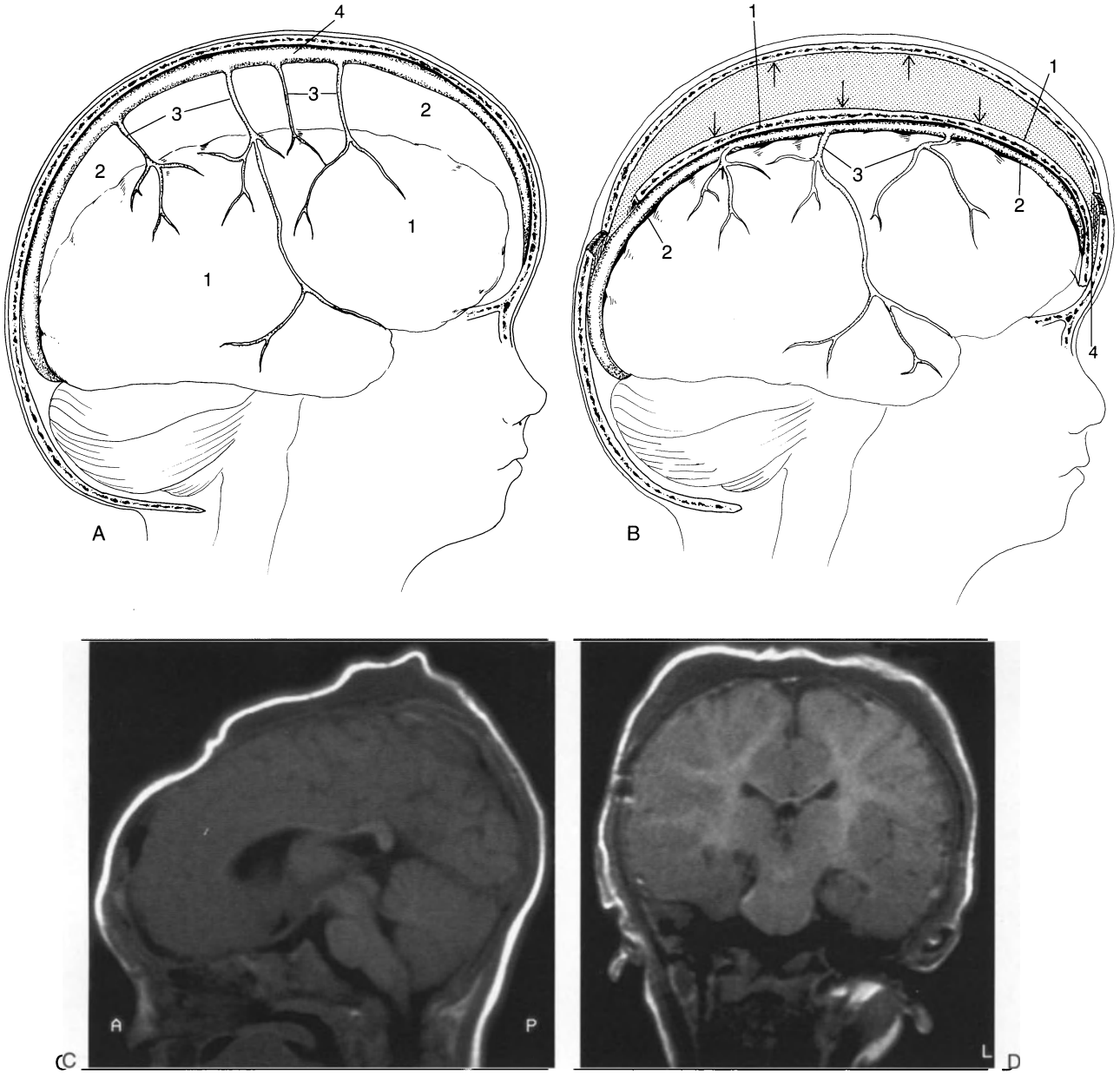


Figure 13.11. (A) This drawing represents schematically the anatomical changes of the bridging cortical vessels which result in diminished cerebral venous drainage first and then occlusion of all cortical bridging veins, and how lowering the SSS restores the normal venous anatomy and venous drainage. It illustrates the diminution in volume of the cerebral hemisphere (*1*) and the large subdural collection of fluid (*2*) within which one may identify the "hanging veins" (*3*) as they bridge across the subdural space to enter the superior sagittal sinus (*4*). The lowering of the SSS was designed to bring surgically the bridging veins into normal anatomical position, thereby converting the "hanging veins" into normal veins. (B) The sagittal suture and cranial vault 1.5 cm on either side of it (*1*) have been dropped onto the convex surface of the hemispheres, bringing the superior sagittal sinus (*2*) into normal position, converting "hanging veins" into anatomically normal bridging veins (*3*). The subdural space has been eliminated. The sagittal suture and surrounding bone have been advanced (*4*). This drawing allows one to appreciate the extent to which the sagittal suture and superior sagittal sinus have been lowered from the preoperative position of the expanded cranial vault (*opposing arrows*). (C) This sagittal T1 image was registered 10 days postlowering of the superior sagittal sinus and reduction craniotomy. One sees that there is no longer a subdural space, the ventricular system has returned to normal, and that there is a redundancy of scalp, especially at the vertex. (D) This is the coronal T1 weighted MRI study done on the 10th postoperative day. Here, the return to normal of the ventricles is clearly seen (to be compared with Fig. 13.10A) and one may identify the superior sagittal sinus, falx cerebri, and dura mater resting snugly upon the surface of the fully expanded hemispheres. The scalp is redundant.

In fact, the etiology, pathology, and pathogenesis of the chronic subdural hematoma are still poorly understood. The treatment of this clinical entity, consequently, remains uncertain.

The neuroradiologic diagnosis of the chronic subdural hematoma is best made with CT or MRI scans.The imaging studies permit:

1. The diagnosis of a chronic subdural hematoma
2. An evaluation of the location and extent of the subdural hematoma
3. Information concerning the degree of secondary cerebral atrophy and/or ventricular enlargement
4. Information concerning the bridging cortical veins

Subdural Taps and Resection of Membranes

Chronic collections of fluid in the subdural space may result from trauma, or may complicate meningitis. Several types of surgical treatment have been advocated. In 1912 Henschen [1a] and then Gilles [2], independently, recommended subdural taps as a useful method of diagnosis and treatment. In 1933 Peet and Kahn [3] performed subdural taps to distinguish "external" hydrocephalus from subdural hematomas. Since that time, subdural taps have been performed for diagnosis and treatment by many, with Salmon [4] recommending them as the procedure of choice for both diagnosis and treatment. Ingraham and Matson [5], in 1944, recommended a combination of consecutive treatments by aspiration through subdural taps, bur holes, craniotomy, and stripping of membranes. In 1957, Ransohoff [6] suggested subdural peritoneal shunt, and in 1961 Shulman and Ransohoff [7] demonstrated that the removal of the membranes did not alter the course of the disease. Time and experience have proven them to be correct. In the same year Collins and Pucci [8] found that subdural membranes regress and disappear after shunting of the fluid to the peritoneal cavity. This, unfortunately, is at best a rare occurrence.

In 1971 McLaurin and coworkers [9] again recommended subdural taps as the treatment of choice for chronic subdural hematoma, but this practice is not accepted by the majority of pediatric neurosurgeons and is, in our opinion, both ineffectual and dangerous.

The original concept that chronic subdural hematoma fluid results from venous hemorrhage and that progressive expansion of the clot is the consequence of osmotic and oncotic pressure forces, shifting fluid into the clot, has little support at this time, though there is some support for repeated bleeds from the subdural membranes.

The lack of understanding of the physiopathology of persistent collections of fluid in the subdural spaces has been the reason for failure to find a successful mode of treatment.

Careful study of the changes in course, calibre, and direction of entrance of the cortical bridging veins into the superior sagittal sinus permits one to theorize that the persistent recurrences of liquid and its steady increase in volume as the brain atrophies ... result from venous and flow obstruction.

Pathogenesis of Chronic Subdural Hematoma

The etiological factors that contribute to chronicity of a subdural fluid in children remain unproven. We postulate a two-stage pathogenesis secondary to a single causative factor. However, before moving on to discuss the causative factor and the pathogenetic mechanisms it is essential to identify the terms we are discussing: (1) chronic, (2) subdural, and (3) hematoma.

1. By *chronic*, with regard to this particular entity, we are obliged to recognize that we mean two things, long-standing and recurrent. In fact, we could safely, in the majority of cases, add...progressive.
2. Though we refer to these collections of liquid as *subdural* in location, we would be just as correct or just as incorrect if we said they were epiarachnoidal. They are located between the inner layer of the dura and the arachnoid, but they are also, in fact, not free collections within this space, for they invariably are completely contained within membranes. Often, there are many more than one fluid collection, each with its own limiting membranes.
3. Though most often these collections begin as *hematomas*, this is not invariably the case, nor is trauma invariably the cause. Infections, dehydration, and acute episodes of electrolyte imbalance have all been reported. What all these components have in common is residual hypertonic liquid, which because of its elevated osmotic and oncotic partial pressures draws more liquid from adjacent structures – dura or vessels or brain – into its mass. The physicochemical sequence across the semipermeable membrane which forms along its surface is known to all who have studied chemistry.

In light of the above we can understand why, referring to this entity as external hydrocephalus, it is so very often progressive and recurrent. The membranes are not the cause, for if they were removing them would solve the problem. Similarly, the liquid contained within the membranes is not the cause.

The electrolyte and/or protein partial pressures within the liquid is the cause. Regarding the two-stage pathogenesis, the first is the increasing accumulation of liquid contained within the membranes and the second is the progressive stretching and narrowing of the cortical veins that bridge the subarachnoid and subdural spaces to enter the sagittal sinus.

This latter process causes diminished drainage of venous blood from the cerebral hemispheres because of remarkable diminution in caliber and verticalization of the bridging veins, as they pass directly superiorly from the cortex, across the fluid (or clot) filled subdural space, to enter the superior sagittal sinus at a right angle, and ultimately leading to thrombosis of the "hanging veins." Narrowing and angulation of these veins results in elevated back pressure, favoring the formation of a transudate which passes across the cerebral parenchyma or vascular walls into the subdural collection. The "hanging veins" likely diminish venous drainage into the superior sagittal sinus, triggering vasogenic brain edema. This would explain the deterioration of brain function in these children. "Hanging veins" are pathogenetic for both the transudate which accumulates progressively in the subdural spaces and the cerebral atrophy which results. The psychomotor retardation is the clinical expression of the resultant cerebral damage. Accordingly, *lowering of the superior sagittal sinus* onto the cerebral hemispheres, bringing it to rest in its normal anatomical position along the superomedial surfaces of the hemispheres, restores the cortical veins to their normal positions and reestablishes physiological venous blood flow from the cerebral hemispheres into the draining sinuses.

Persistent reaccumulation of subdural fluid with evidence of increasing intracranial pressure, characterized either by progressive enlargement of the head, bulging fontanelle, splitting sutures, and/or progressive psychomotor retardation, with imaging evidence of progressive craniocerebral disproportion and characteristic cortical "hanging veins," are indications for surgery. The recommended surgery is lowering of the SSS and reduction cranioplasty.

Operative Technique for Lowering the Superior Sagittal Sinus: Reduction Cranioplasty

(Figs. 13.12–13.17)

The purpose of the procedure, as described above, is to improve the venous drainage from the superior anastomotic veins into the SSS. It consists of lowering, and advancing, the SSS with its overlying sagittal suture, and performing a duraplasty. The duraplasty is performed so as both to advance the dura and reduce its redundancy, to fit it to the brain as snugly as possible, obliterating the subdural space and giving a normal course to the cortical bridging veins. The reduction cranioplasty which brings the reconstructed frontal, parietal, and occipital bones to rest upon the reduced dural envelope, is necessary to avoid the creation of an open epidural

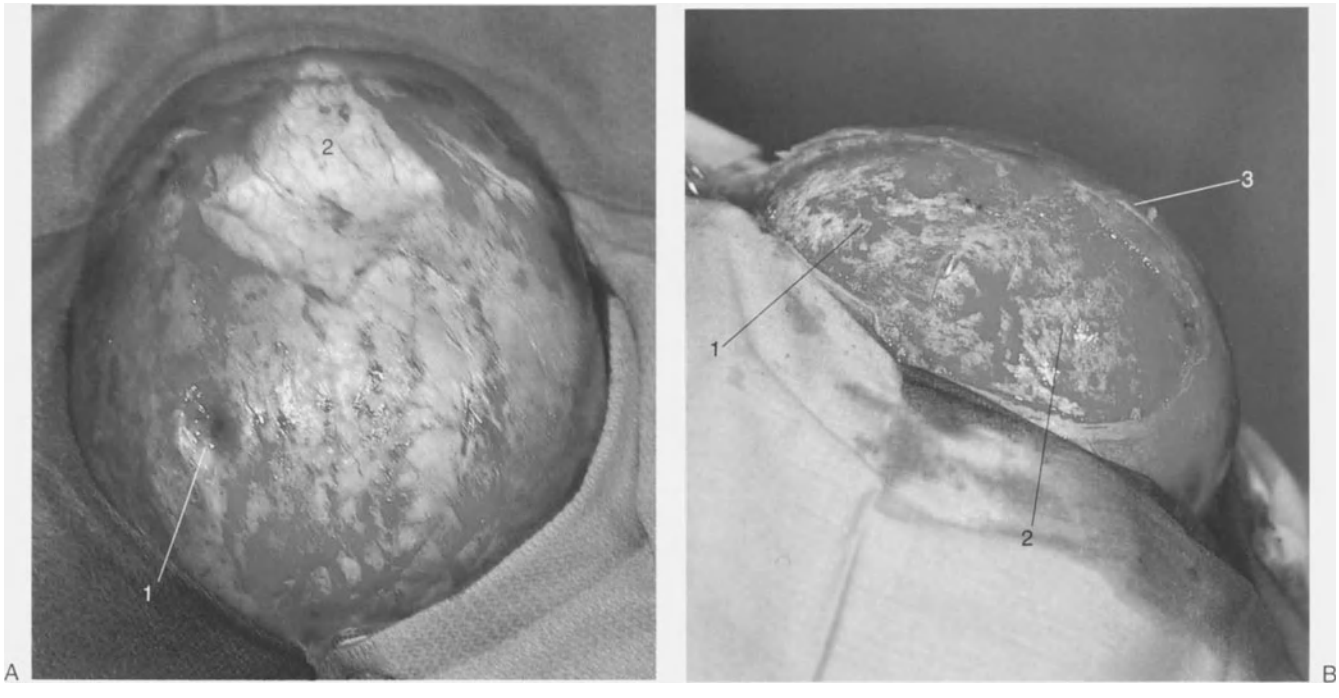


Figure 13.12. Technique for lowering the superior sagittal sinus and performing reduction cranioplasty. (A) After the scalp flap has been reflected, the bur holes (*1*) are made preparatory to lifting the bone flaps. Note the very large anterior fontanelle (*2*), which is, paradoxically, sunken. In fact, it is not unusual for children with chronic subdural collections of fluid to suffer progressive increase in head size and to have a sunken fontanelle. (B) The skull after the frontal (*1*) and parietal (*2*) dura has been exposed by reflection of frontal and parietal bone flaps. The strip of bone with the superior sagittal and metopic sutures (*3*) has been left in place and intact.

compartment. The net result is returning dura and bone to the convexity of the hemispheres, from whence they will once more grow with the brain.

The child is placed in the supine position with the head flexed at 20°. A bicoronal skin incision is extended from the antitragus of one side to the other, and then an S-shaped incision is extended posterior to this. This permits the reflection of three skin flaps: one forward and one laterally on each side. In infants the coronal suture is attached to the dura; consequently, sharp dissection must be used to separate the frontal and parietal bones from the dura. Also, the frontal and parietal bones may be separated from the anterior suture subperiosteally and extradurally. After the flaps are reflected, there remains a strip of bone 1.5 cm wide, overlying the SSS. This sagittal strip of bone, which includes the sagittal suture, is cut and freed posteriorly and anteriorly. It is not separated from the SSS. The dura is opened in Z fashion and the subdural space is irrigated with normal saline. No attempt is made to remove all of the subdural membranes, only those along the convexity.

Four drill holes are placed between the nasion and the glabella, depending on how much the sinus is going to be lowered and advanced. Similar drill holes are placed in the most anterior portion of the strip of bone overlying the SSS. Subsequently, the sagittal strip of bone with sinus is lowered, advanced, and anchored to the frontal bone using 3–0 suture material. Careful inspection of the cortical "hanging veins" through the dural opening permits the surgeon to bring these cortical veins and the lowered SSS into normal anatomical position vis-à-vis one another and the superomedial surfaces of the hemispheres. The surgeon who has never seen this procedure performed will remark immediately how low, *how very low,* the SSS and hemispheres have come to rest in the expanded cranial vault. Subsequently, the dura is tailored and closed in such a way that it is advanced and brought to fit the brain snugly, obliterating the subdural space.

The free frontal and parietal bone flaps are attached to the skull, using 3–0 sutures through multiple drill holes made along the bone edges. These flaps should be brought to rest over the dural surface, disregarding the apparent lack of cranial contour. There is often excess bone. One may find it advantageous to mold the lateral rim of the skull by cracking it first, then bending it into a desirable position before anchoring the flaps.

Follow-up imaging studies most often show normal arterial phases, the medullary system only minimally filled, and all normal venous drainage through the superficial cortical veins with no evidence of "hanging veins." It is of value to remark that the "hanging vein" appearance of the cortical bridging veins disappears and a normal course appears [10]. The ventricles diminish in size.

Bur Holes

In the very early stages of subdural fluid formation, simple bur holes (bilateral frontal) may suffice as a curative procedure. However, it is not uncommon for the fluid to reform, for the skin over the bur holes to bulge. Repeat drainage is attempted through the same bur holes, with or without insertion of drains into the subdural space. If the fluid reforms, one should proceed to imaging studies to learn whether "hanging veins" are present and, if so, plan to lower the superior sagittal sinus as soon as possible.

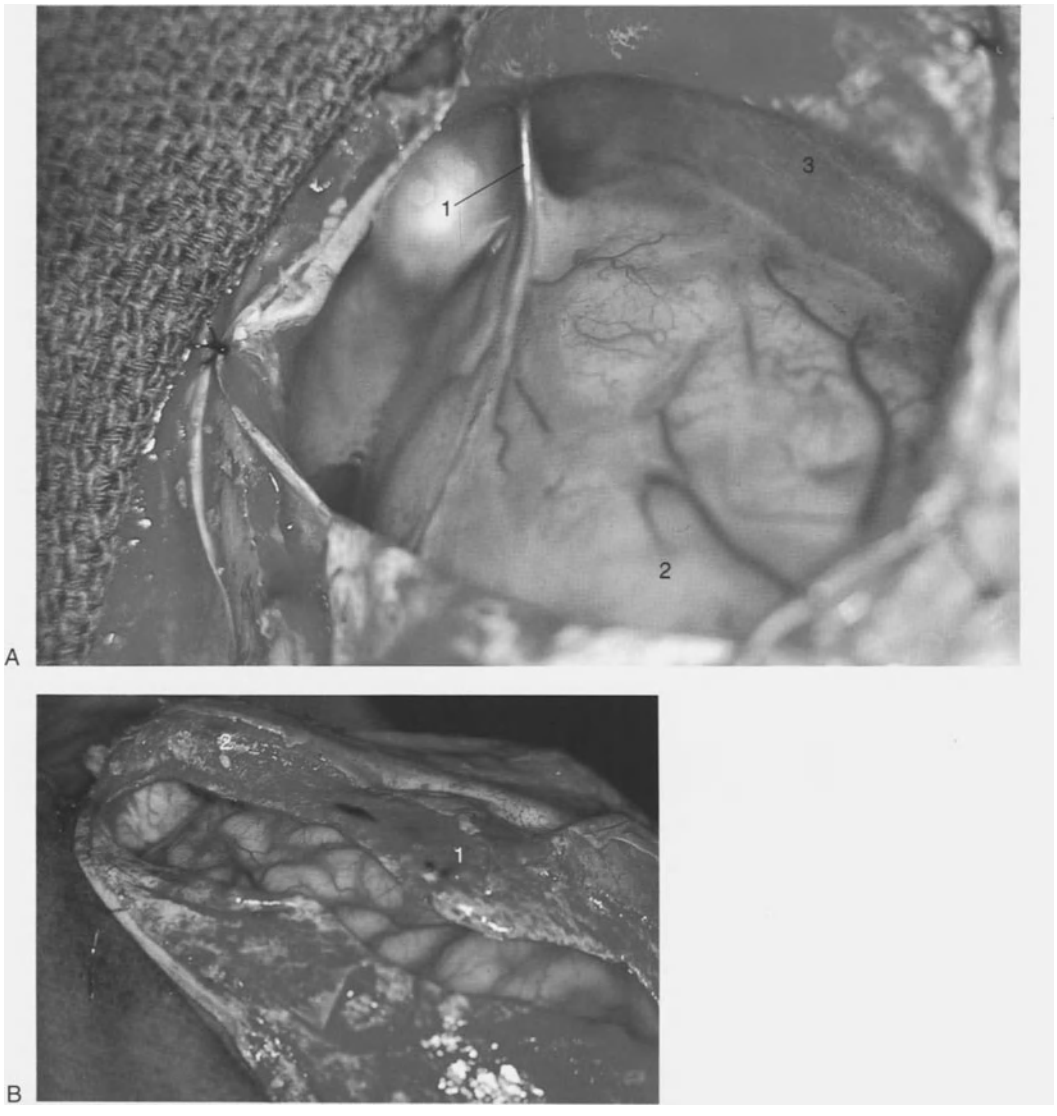
Subdural Peritoneal Shunt

The subdural peritoneal shunt has been used as a form of treatment for chronic subdural hematoma, though its failure could readily be predicted since the fluid within the subdural space, unless it is CSF, has a protein content greater than 500 mg%. Also, this fluid is not under a great deal of pressure. Therefore, its flow from the subdural space to the peritoneal cavity does not occur through a valve-regulated system. Open-ended tubes, however, allow such fluid to flow when the gravitational forces are favorable. In either event, this has not been a satisfactory treatment for the chronic subdural collection of fluid.

Cerebral Atrophy

In those instances where the cerebral atrophy has been progressive with increases in volume of subdural fluid and ventricular size, some surgeons have recommended the performance of *combined ventriculoperitoneal and subdural peritoneal shunting.* This, too, has not been attended by success.

Figure 13.13. Technique for lowering the superior sagittal sinus and performing *reduction cranioplasty.* (A) This photograph puts into relief the "hanging veins" (*1*), the opalescent outer subdural membrane (*2*), and the falx cerebri (*3*). (B) The dura has been opened bilaterally, and the sagittal strut of bone over the metopic suture (*1*) freed from the glabella (*2*), allowing the superior sagittal sinus (SSS) to fall upon the underlying, shrunken, cerebral hemispheres. (C) Forceps are being used to elevate the dura, allowing one to see the stretched "hanging vein" (*1*) anteriorly, and the most normal course of a bridging vein (*2*) posteriorly, where the dura and the SSS have been allowed to come to rest upon the convexity of the cerebral hemispheres. (D) In sum, the procedure consists of lowering the sagittal suture, so that the SSS comes to rest upon the hypoplastic cerebral hemispheres, and performing a duraplasty so that the lowered sinus is brought forward, allowing the cortical veins to resume normal caliber and configuration. The calvarium is remolded to come to rest upon the surface of the dura.



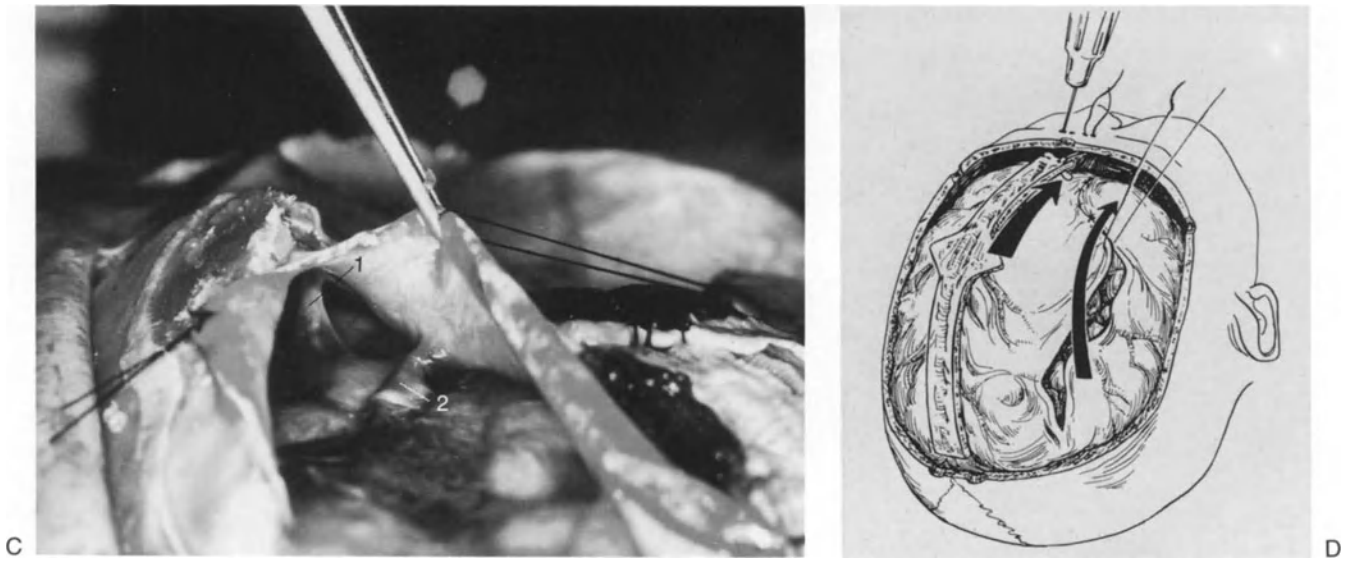


Figure 13.13. Legend see p. 434.

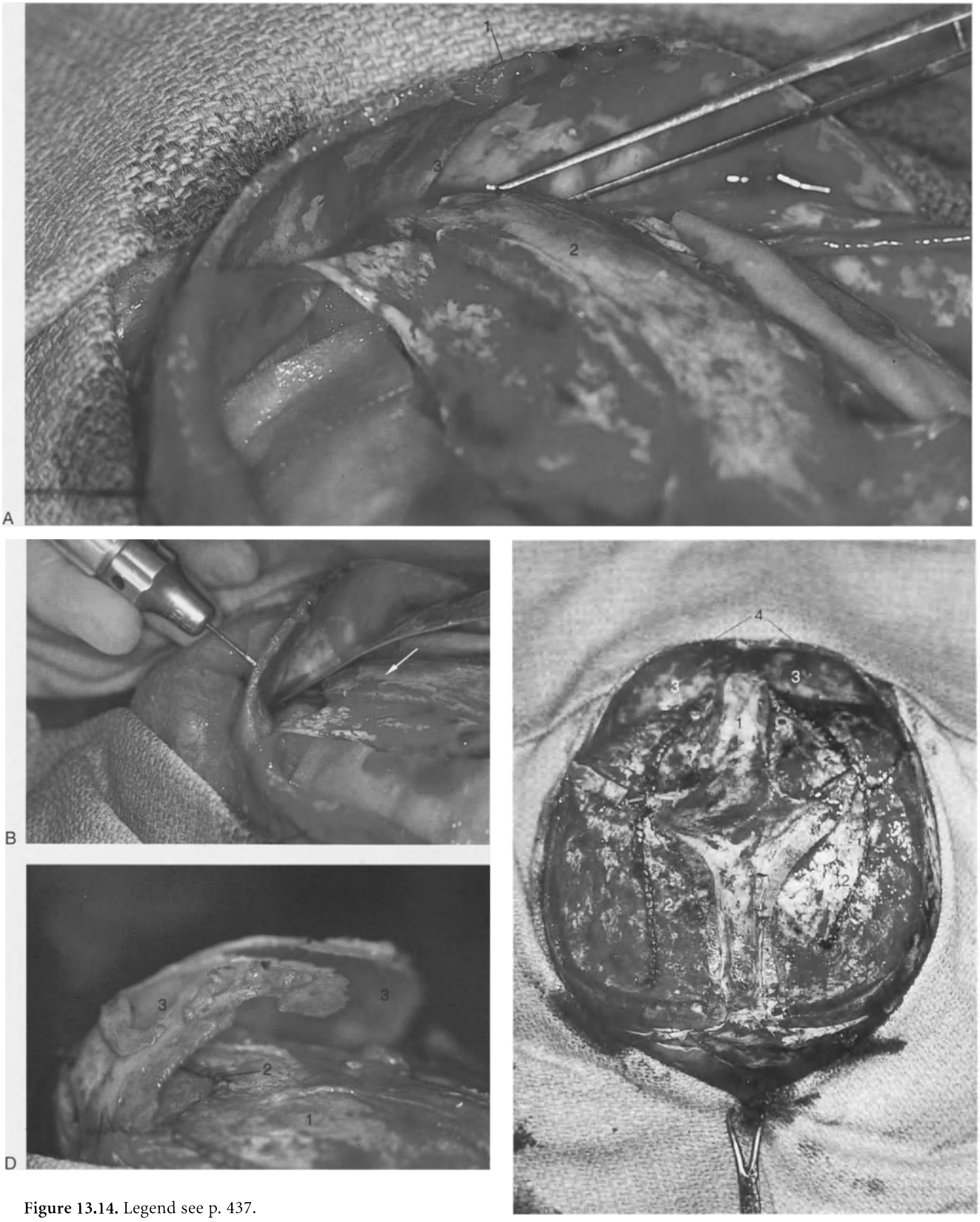
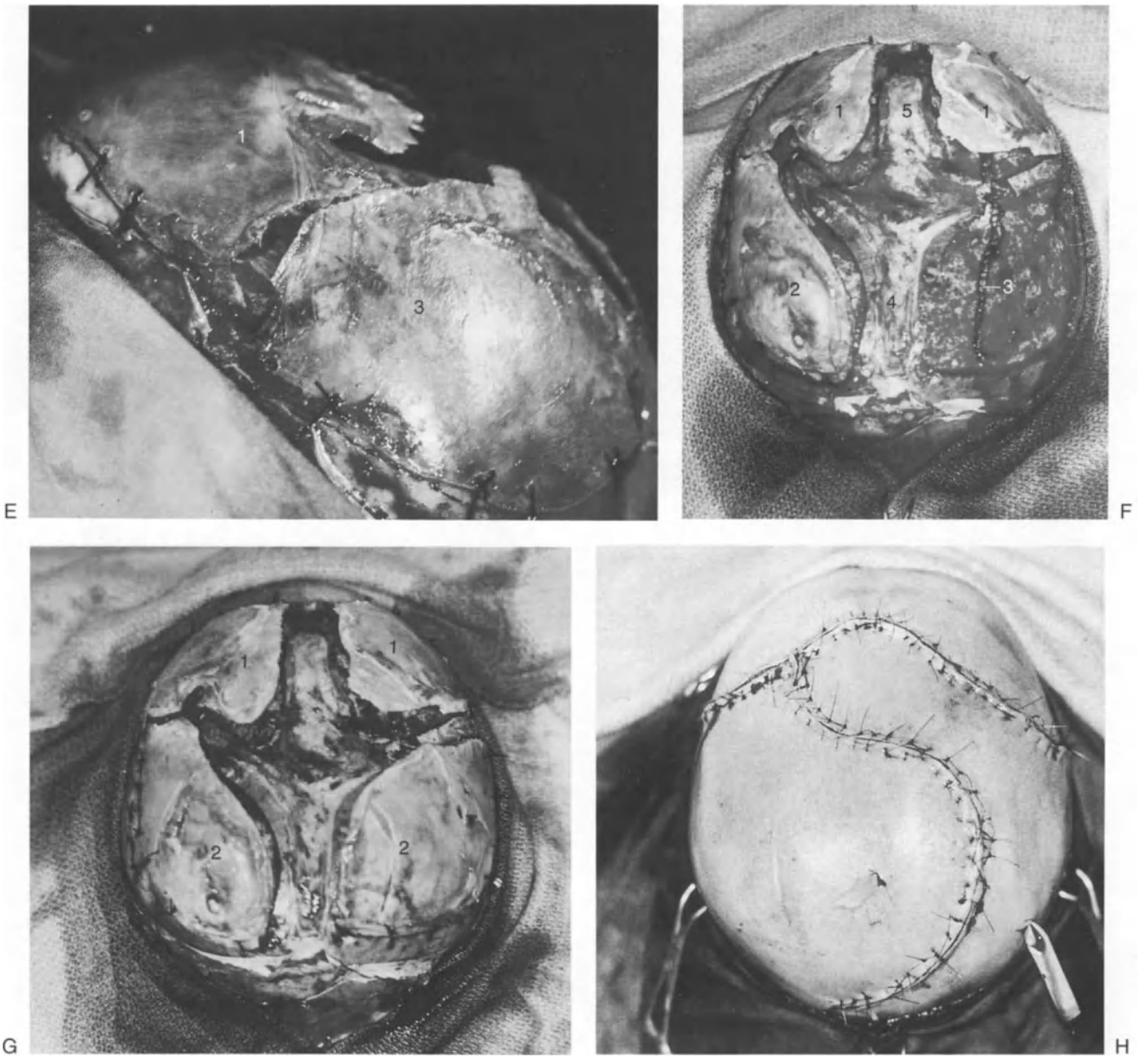


Figure 13.14. Legend see p. 437.



◀ **Figure 13.14.** Technique for lowering the superior sagittal sinus and performing reduction cranioplasty. Different stages in lowering the superior sagittal sinus (SSS) and reconstructing the skull. (A) The frontal bone has been osteotomized just above the glabella (*1*) and the metopic suture (*2*) is being brought to the crista galli (*3*). (B) The frontal bone is being perforated at the glabella with a high-speed drill so that the sutures may be passed through it and the strip over the metopic suture (*arrow*) to permit anchoring this latter in a lowered position. (C) The sagittal strip of bone (*1*) has been anchored anterior inferiorly to the inner table of the frontal bone at the crista galli, lowering the SSS into its normal anatomical position. One notes that after the dura has been closed (*2*), there remains an enormous dead (*3*) space behind the frontal bones (*4*). This should be reduced as much as possible by cutting grooves into it and then closing the "fan." (D) This lateral oblique view puts into relief the remarkable lowering of the dura (*1*) onto the surface of the brain, the dural suturing (*2*) to reduce dead space, and the contour of the frontal bones (*3*) prior to fracturing and molding them over the surface of the dura. (E) After the SSS has been lowered the frontal bones (*1*) are fractured, or osteotomized, along a line extending from the keyhole to the glabella (*2*). They may subsequently be molded over the suface of the dura and brain. The parietal flap (*3*) has now been brought back into position and anchored with periosteal sutures. (F) This vertex view shows the frontal (*1*) and parietal (*2*) bones molded over the lowered dura. The anterior and inferior portions of the metopic area (*5*) have been lowered and anchored to the glabella, the sagittal suture (*4*) with the underlying SSS lowered onto the brain surface, and the duraplasty (*3*) completed. (G) Before the skin closure, both the frontal (*1*) and parietal (*2*) bones are snuggled against the dura. (H) The skin incision has been closed and the Penrose drain left in the subgaleal space. Note that the contour of the head is quite acceptable.

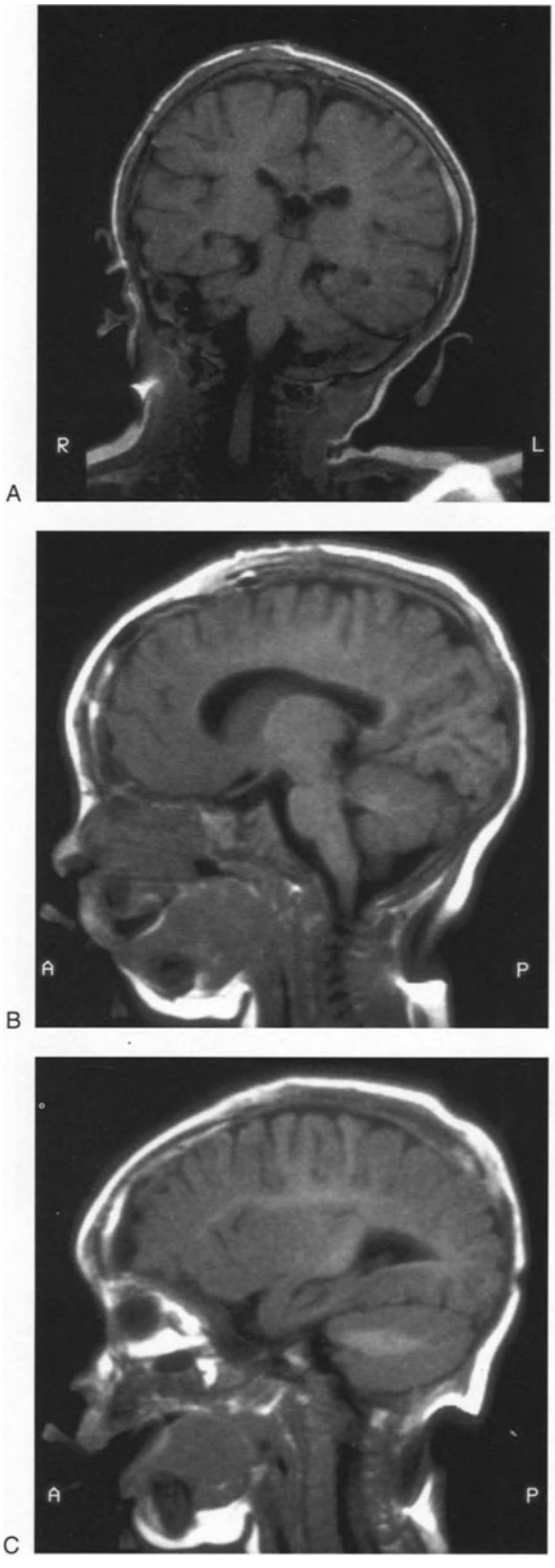


Figure 13.15. Legend see p. 439.

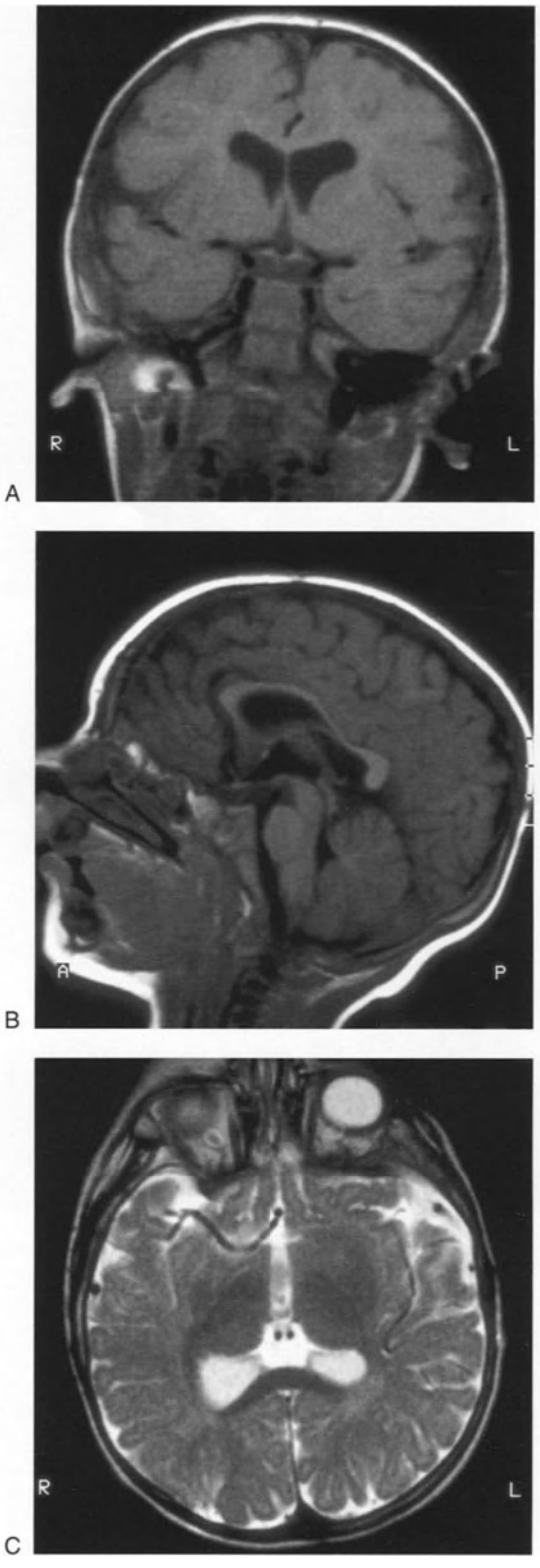
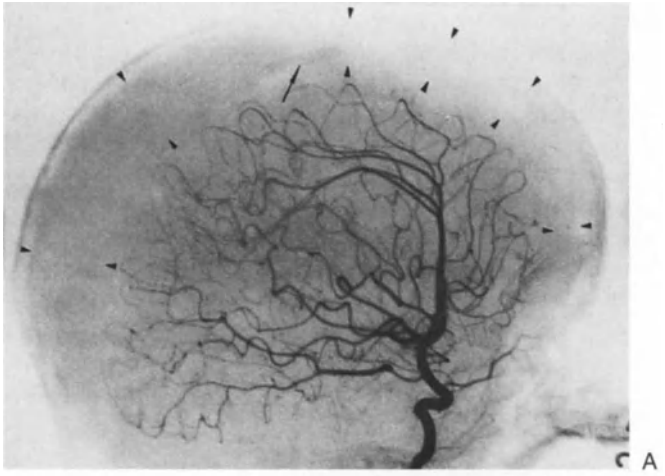
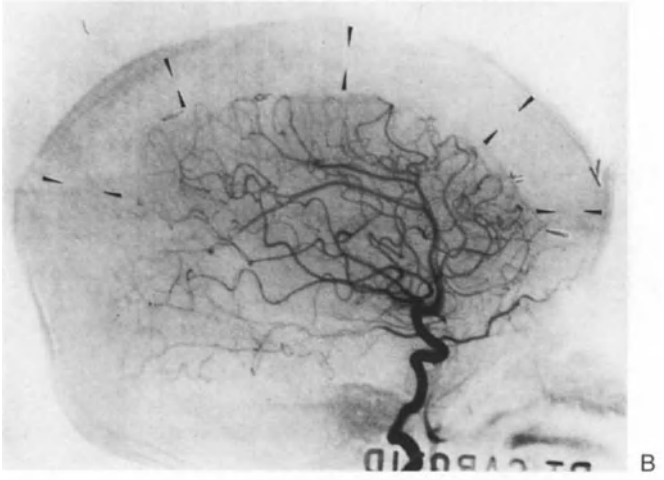


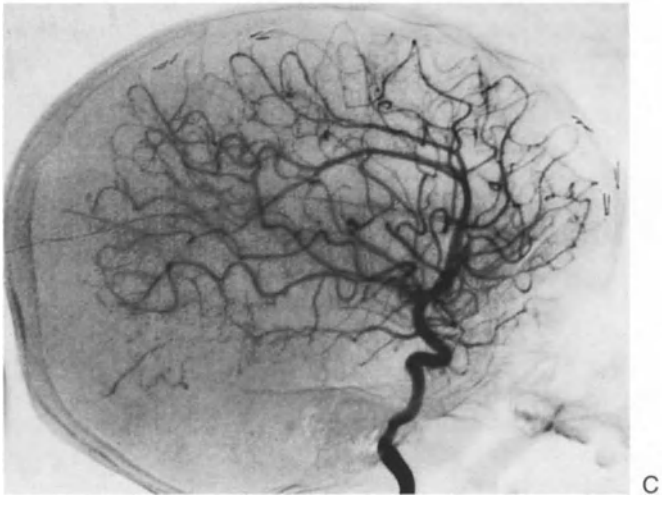
Figure 13.16. Legend see p. 439.

◀ **Figure 13.15.** This sequence of coronal (A), sagittal (B), and parasagittal (C) MR images was registered 3 weeks after reduction cranioplasty and lowering of the superior sagittal sinus. In (A) one sees return to normal of the ventricular system, reexpansion of the hemispheres, complete disappearance of the subdural collection of fluid except for a small film over the left parietal convexity, reapproximation of the dura and superior sagittal sinus to the convexity, and reformation of the scalp over the skull. In the sagittal, T1 weighted image section, one sees that the vertex of the brain is immediately adjacent to the vertex of the skull, there is no subdural collection whatsoever, and the scalp has almost completely remolded into a normal structure adjacent to the underlying skull. In the parasagittal section (C) there is only a questionable collection of fluid at the frontal pole, but the sulci and gyri are normal and there is no vertex collection of fluid in the subdural space.



◀ **Figure 13.16.** In this figure, the coronal (A), sagittal (B), and axial (C) MRI studies of the same child as illustrated in the previous figure were registered 3 months after surgery. In (A), the coronal T1 weighted image, one sees an almost perfectly normal cerebral parenchyma with minimal ventricular dilation and normal relationships between brain and dura, and dura and scalp. In (B), a sagittal section, one sees minimal dilation of the frontal sulci in an otherwise normal intracranial compartment. (C) This T2 weighted image puts into relief the normal cerebral hemispheres, subarachnoid spaces, and ventricles. It is concluded that reduction cranioplasy with lowering of the superior sagittal sinus performed when the bridging cortical veins have not all been destroyed permits the brain to reexpand normally and arrests the negative series of events resulting from the chronic subdural collection of fluid in infancy.





► **Figure 13.17.** These are three different arteriographic studies of the same child. (A) Before lowering the SSS the arteriographic characteristics of chronic collections of subdural fluid are those of a large space-filling defect over the convexity (*opposing arrowheads*) and bowing of the anterior cerebral system around dilated lateral ventricles; *the arrow* indicates the shunt tube in the subdural space. (B) Four weeks after surgery (lowering of the superior sagittal sinus and reduction cranioplasty). One may observe disappearance of bowing of the pericallosal system. The space over the convexity (*arrowheads*) is now epidural, not subdural. (C) Eleven months later one notes normal arteriographic characteristics indicating cerebral reexpansion and disappearance of the subdural fluid. (D–G) see p. 440.

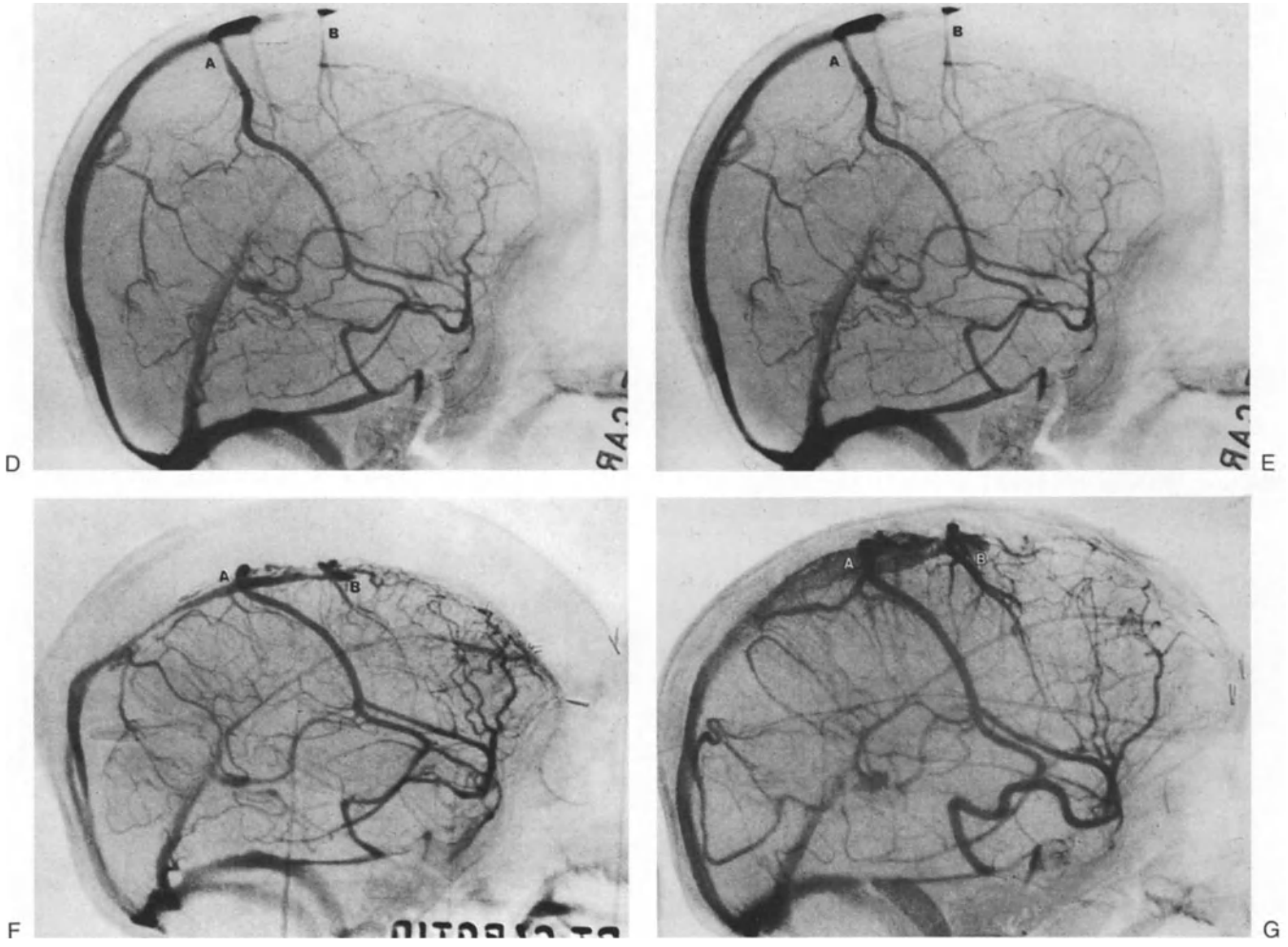


Figure 13.17. (D) One notes the subdural space-filling defect and the "hanging veins." It is of value for the reader to follow these hanging veins (labeled in *A* and *B*) through this figure and the subsequent three. (E) Three weeks after the study illustrated in (D), one notes progression in size of the subdural collection of fluid and further stretching of the hanging veins. (F) The immediate postoperative venogram illustrates how the superior sagittal sinus has been restored to its normal anatomical position and the hanging veins converted into normal bridging veins. (G) This study was performed 11 months postoperatively. One notes an almost entirely normal venographic pattern.

Membrane Resection

Resection of the membranes, which form over both the dura mater and the arachnoid, does not cure the child unless lowering of the SSS and reduction cranioplasty are performed. The subdural fluid reforms within hours of bilateral frontoparietal craniotomy and extensive resection of the membranes, and soon thereafter proteinacious membranes reform.

Cerebrospinal Fluid Leaks

Leakage of cerebrospinal fluid (CSF) into the periorbital tissue, directly through fractures of the mastoid, into the subgaleal space, is all self-arresting in that fistulization does not occur. However, CSF leakage through basilar skull fractures from the nose, into the nasopharynx, or from the middle ear, may all fistulize, thereby becoming permanent.

The site of drainage is of no value in indicating the site of leakage from the subarachnoid space and through the fracture in the skull: CSF rhinorrhea may result from a fracture of the posterior surface of the pe-

trous bone, the lateral surface of the body of the sphenoid, and the cribriform plate. Therefore, the diagnosis of CSF fistulae makes it incumbent upon the clinician to ascertain with precision the point of passage of CSF from the subarachnoid spaces, or cisterns, through the rent in the base of the skull. Otherwise, surgical closure of the dural opening is impossible. Very often, unfortunately, one is not able to identify the precise point of leakage.

We may speak in terms of two distinctly different operative procedures for correction of CSF leaks: (1) direct, when the surgeon knows where the precise point of leakage is, and (2) indirect, when the surgeon has not been able to identify the point of leakage, so that he proceeds with a "plugging" procedure.

Direct Approach

Direct repair may be performed through either the intradural or extradural approaches, depending upon the location of the leakage.

Intradural Approach

Breaches in the cribriform plate may be closed through either, though the intradural approach affords greater assurance of success and lesser risks of, paradoxically, a CSF fistula complicating surgery for a CSF fistula. This latter occurs when the dural covering of the rootlets of the olfactory nerve is pulled from the openings in the cribriform plate, resulting in fistulization between the periolfactory cistern and the nasal cavity.

Leaks through breaches in the region of the planum sphenoidale, the superomedial surface of the orbit, and the posterior surface of the petrous bone may be approached indifferently along the extra- or intradural routes.

Rents in the region of the medial aspect of the greater wing of the sphenoid, along the arc extending from the foramen spinosum through the superior orbital fissure, and over the tip of the petrous bone, are best approached by the intradural route.

Extradural Approach

The advantage of an extradural approach, when it is possible, is that it allows the surgeon to plug the fracture line with fragments of bone, to seal these in place with bone dust (or chips) and plastic, and then to cover this bony repair with periosteum before closing the dura. In order to accomplish this by the intradural route, one is obliged to strip dura from bone and then to lay a periosteal graft over the parenchymal surface of the dura.

Indirect Approach

The indirect approach consists of plugging the air sinus through which the CSF passes to reach the nasopharynx. This procedure should be resorted to only when all efforts to diagnose precisely the point of leakage have failed. The sphenoid sinus is plugged by the transsphenoidal route, packing it with fatty tissue, a muscle graft, and a plug of bone. The patient should be kept on continuous lumbar spinal drainage, in the horizontal position, for 10 days, during which time intravenous prophylactic antibiotics are given.

Meningocele Spuria

Sometimes called "growing skull fracture," the meningocele spuria results from a depressed skull fracture, which tears through the dura mater and penetrates the subarachnoid space. Consequently, the cortical surface is invariably damaged. The sine qua non for the pathogenesis of a meningocele spuria is incarceration of torn dural fragments between impacted bony spicules, thereby preventing dural healing. Cerebral pulsations erode the fracture edges and drive the cerebrum through the dural and bony rents. With time, the underlying ventricle may expand into a porencephalic cyst or dilate (entirely or just one horn).

The cure of the meningocele spuria entails reflecting a bone flap around the area of pathology, removing the bony spicules, opening the dura and freeing it from its adhesions to the underlying cortex, implanting a periosteal graft over the damaged dura so as to attain a tight closure. Either a small rib graft or methyl methacrylate is used to close the bony defect, and then the scalp is closed.

In those cases where the ventricle has dilated, with a porencephalic cyst extending from ventricular wall through the skull defect and into the subgaleal space, one is obliged to resect the gliotic surface of the cortex which borders upon the porencephalic cyst. This facilitates the formation of a cerebral cicatrix and a seal of the neural parenchyma. If this does not occur, CSF may accumulate within the subdural space, gaining access to it through the cerebral fistula. It is then trapped within the subdural space because of the collapsing cerebral mantle. In the postoperative period, lumbar spinal drainage may be indicated for approximately 2 weeks, with the patient receiving prophylactic intravenous antibiotics. CT or MRI scans will reveal whether parenchymal healing has occurred. If not, one may be obliged to insert a ventriculoperitoneal shunt.

Child Abuse

The material herein presented is taken directly from the review and analysis of the cases personally diagnosed and managed by us [11] (Tables 13.1, 13.2).

Child abuse is an infinitely complex phenomenon. Despite the continuous efforts to seek and prevent child abuse, it has been a protracted, painful burden on society. The physical and emotional insult occurring in child abuse ranges from deprivation of affection and love to fatal assault. Morse et al. [12] reported that 10% of trauma in the emergency department was related to physical abuse. Gregg and Elmer [13] determined that a higher incidence (20%) of children seen for accidents were actually child abuse victims.

Child abuse was recognized in all its clinical entities by Tardieu [14] in 1860. In 1946 Caffey [15] reported the first six cases of multiple fractures in long bones associated with chronic subdural hematoma. A few years later, Silverman [16] reported a similar observation and clearly defined it as a traumatic lesion. Wooley and Evans [17] in 1955 reached the conclusion that the trauma seen on the X-rays had been inflicted in a willful manner. The term "battered child syndrome" was first coined by Kempe et al. [18] in 1962, who were alarmed by the large number of children admitted to their pediatric service suffering nonaccidental, unrecognized trauma. The syndrome was defined as a clinical condition in young children who have received serious physical abuse which resulted in a permanent injury or death. Although comprehensive reports concerning the detection and prevention of child abuse have been published, few papers have dealt with traumatic mechanisms of the head injury. Guthkelch [19] and Caffey [20–22] reported that whiplash as a result of manual shaking of infants was a common primary type of trauma in the battered child. An effort is made here to identify traumatic mechanisms of head injury in the abused child.

Table 13.1. Study group at the Children's Memorial Hospital from 1970 to 1979

Year	Number of battered children	
	Total	With head injury
1970	82	6
1971	78	9
1972	69	13
1973	98	11
1974	187	17
1975	18	4
1976	13	8
1977	25	4
1978	26	3
1979	25	2
Total	621 (100%)	77 (12.4%)

The initial diagnosis was made by the admitting physicians on the basis of history, physical findings, and radiographic evidence such as periosteal hemorrhage and computed tomographic manifestations [23–25]. These data were eventually reclassified. Twenty cases out of the "suspected child abuse" and "accident" group had to be reclassified as "child abuse" or "suspected child abuse," respectively (Table 13.3).

The 77 assailants were classified according to their surname. Of this group, 64 (83%) had English surnames and 10 (13%) were Spanish. The racial distribution was 47 (61%) white to 30 (39%) nonwhite. The higher incidence of Spanish surnames in this study was due to the large number of Spanish-surnamed patients seen in our clinics. Consequently, it is not statistically significant.

More than half (53%) of the assailants were parents and the rest included relatives, baby-sitters, and mother's boyfriends (Table 13.4). The parents of 40 children (52%) were married, 24 had never married, and 13 were divorced. Sixty of the children (78%) were the result of 'unplanned' pregnancies. Eighty percent of the parents came from a low income group, some experiencing exceptional hardships, and most parents (87%) had completed high school.

The mental state of 45 children (58%) on admission ranged from lethargic to comatose, 25 patients (32%) showing some sort of motor disturbances and 24 (31%)

Table 13.2. Age and sex of the children studied

Age	Male	Female	Total	
			n	%
Less than 6 months	14	7	21	27
7 months to 1 year	9	9	18	23
1–2 years	17	10	27	35
3–4 years	4	1	5	7
Over 5 years	4	2	6	8
Total	48 (62%)	29 (38%)	77	100

having had episode(s) of seizures. Retinal hemorrhage was noted in 26 children (34%). The anterior fontanelle was overtly tense in 6 children (8%), 12 had anemia (16%), and 2 had a large head.

The 77 children showed 72 various skull fractures, the average being 0.9 fractures (Table 13.5). The fracture was most commonly observed at the cranium (60%) and out of 35 linear and depressed skull fractures 30 were found in the parietal bone. Eighteen patients showed fractures in long bones such as tibia and humerus; the "classical" picture of metaphyseal avulsion and/or subperiosteal reaction was observed on only four occasions.

The 77 children suffered from 126 craniocerebral injuries, averaging 1.6 per patient (Table 13.6). Skull fractures of different magnitude and intracranial hematoma were the most common finding, comprising 55% of the injuries. Subgaleal or subperiosteal hematomas also were encountered in 20.6%. Out of 33 intracranial hematomas, 30 were subdural.

Forty-seven children were discharged after medical treatment. The remaining 30 children underwent surgical intervention. Craniotomy was performed on 19 patients and subdural or epidural hematomas were removed. Further surgical treatment consisted of lobectomy (1), bur holes plus drainage (5), elevation of depression (4), and subdural taps (4).

Table 13.3. Comparison of original impression and status at the time of follow-up

	Number of patients at	
	Original impression	Follow-up
Child abuse	20	40
Suspected child abuse	46	37
Accident	11	0
Total	77	77

Table 13.4. Type of assailant

Assailant	Cases	
	n	%
Parent	41	53.2
Dad	17	
Mom	10	
Both	14	
Relative	12	15.6
Baby-sitter	7	9.1
Others	9	11.7
Unknown	8	10.4
Total	77	100

Table 13.6. Type of craniocerebral injuries

	Patients	
	n	%
Fracture of skull	37	29.4
Intracranial hematoma	33	26.2
Subdural	30	
Bilateral	22	
Unilateral	8	
Epidural	3	
Cephalohematoma	26	20.6
Contusion, cerebral	23	18.2
Concussion, cerebral	7	5.6
Total	126	100

Table 13.5. Radiographic findings

	Cases		Cases
Skull	43 (60%)	Long bone series	13 (25%)
Linear fracture	30	Tibia fracture	5
Parietal	25	Humerus fracture	5
Temporal	2	Metaphyseal avulsion	3
Occipital	1	Femur fracture	2
Frontal	1	Radius fracture	1
Multi-area	1	Ulna fracture	1
Depressed fracture	5	Subperiosteal reaction	1
Parietal	5	Others	11 (15%)
Diastatic fracture	2	Rib fracture	10
Splitting suture	6	Clavicle	1
Total			72 (100%)

Table 13.7. Recollection of original injury

Trauma	Injuries			
	According to original description		After recollection	
	n	%	*n*	%
Direct hit				
To head and/or face	15		37	
Other than head	17		25	
Total	32	36.0	62	53.9
Child "dropped"	4		5	
Child "fell off from height"	35		32	
Fell on head	8		18	
Other than head	26		13	
Unknown	10		1	
Child "thrown"	5		4	
Total	44	49.4	41	35.7
Shaken	2	2.2	9	7.8
Unknown	11	12.4	3	2.6
Total	89		115	

Table 13.8. Site of injury of 113 external wounds on 53 patients (2.1 external wounds per patient)

Site of injury	Wounds	
	n	%
Head and face	52	46.0
Neck	3	2.6
Chest	8	7.1
Abdomen	6	5.3
Dorsum	9	8.0
Buttock	9	8.0
Extremities		
Upper	11	9.7
Lower	15	13.3

Thirty-seven children were placed under the custody of the Illinois Department of Children and Family Service (IDCFS), and subsequently cared for in foster homes, nursing homes or the Rehabilitation Center. Thirty-two children returned home with or without an aid from the Visiting Nurse Association (VNA). Eight patients (10%) eventually died. Of these, four had acute subdural hematoma, three had severe cerebral contusion with edema, and one had skull fracture with severe cerebral edema. Six out of the eight deaths were under 1 year of age. Seven patients remained comatose until death; six showed retinal hemorrhage. At the time of the follow-up interview, 42 (54%) children were healthy with normal neurological findings, while 27 (35%) showed either mental retardation, neurological deficits, or delayed milestones.

Table 13.9. Distribution of external wounds and associated long bone injuries

	Intracranial hematomas (*n*=33)		Skull fractures (*n*=37)		Cerebral concussion or contusion (*n*=30)	
	n	%	*n*	%	*n*	%
Head	13	35.6	20	49.1	11	51.0
Face	3		9		8	
Neck	2	4.4	2	3.4	3	6.4
Chest	3		4		2	
Abdomen	3	35.6	4	23.8	3	27.7
Back	5		2		3	
Buttock	5		4		2	
Upper extremities	4 (1)	24.4	4	23.7	3 (1)	14.9
Lower extremities	7 (81)		10	3	4 (1)	
Long bone injuries = 0						

During the interview the parents were asked to recall the events; this in turn was compared to the original description of the injury. In order to assess the traumatic mechanism, the assailants were asked for their recollections of the incident. The injuries were then classified "direct hit," "dropped," "fell off from height," "thrown," "shaken," and "unknown." Table 13.7 compares the original descriptions at the time of hospital admission with the later recollections.

One hundred and thirteen external wounds were found among 53 patients, each child averaging 2.1 (Table 13.8). The head was by far the most common single site of injury (46%), then the trunk, limbs and neck.

Blunt injuries were evidently more common than nonblunt injuries. Ninety-six patients (85%) exhibited bruising, soft tissue contusion, regional subcutaneous hematoma, or periorbital ecchymosis (9). The other 17 patients showed nonblunt injuries such as excoriation (2), burns (11), and bite (4). External wounds were recounted among different intracranial pathologies to see if a cohort had more evidence of injuries over the head. Thirty-three children with intracranial hematoma showed 45 external wounds; 37 children with skull fractures showed 59 external wounds, and 30 children with cerebral contusion or concussion showed 47 external wounds (Table 13.9).

Children with skull fractures and cerebral contusions who were thought to have blunt injuries to those areas showed a relatively higher proportion of external wounds on the head and face, compared to the children with intracranial hematomas. For the remaining parts of the body such as neck, trunk and extremities the distribution of external wounds was statistically insignificant.

External wounds are more frequent on the head and face, particularly for patients with skull fractures, cerebral contusions, and/or concussions; patients with skull fractures showed more external wounds and associated bone fractures on the trunk and extremities, and patients with intracranial hematorna did not necessarily show any more tendency of long bone injuries or periosteal reactions.

Twenty-nine fractures were encountered in the long bones and chest cage. Fracture was most common in the ribs, the tibia, humerus, femur and clavicle. Among the different craniocerebral injuries, patients with skull fractures showed more associated bony fractures than patients with intracranial hematoma or cerebral concussion or contusion (Table 13.10).

Child abuse is not confined to any homogeneous group of society [12, 26, 27]. The parents belong to disparate age groups, ranges of intelligence, and social classes. Parents who are intellectually unfit, immature, self-centered, or dependent are unable to meet the needs of their children or provide reasonable child-rearing. Parents often strike out at their children at times of personal stress and emotional disturbance. These predisposing stresses are more prevalent among the poor and emotionally deprived groups (34%).

Children who are abused are often no different than other children. Nevertheless. they might be perceived by their parents as being restless, easily excitable, problematic, and excessive criers [12, 26]. Some mothers found that they could not control or discipline their children, and others were confused by the child's behavior. Friedman and Morse [26] expanded the discussion on the mother-child relationship in child abuse studies, and reported that 67% of children in the suspected abuse category came from families with notable behavioral and/or emotional problems.

Infants and younger children are most susceptible to child abuse. In other studies [27, 28], between 20% and 60% of children were under 2 years of age. The younger child is more vulnerable to injury, as it is incapable of evading its assailant. Indeed, in this series 27% of patients were under the age of 6 months and 85% were below the age of 2 years.

The alarming papers by Guthkelch [19] and Caffey [20, 21] described the essential clinical manifestations of child abuse as 'whiplash shaken infant syndrome' in which the battered babies are actually shaken babies. 'Habitual manual' whiplash shaking by grabbing the infants by the extremities may cause subdural hematoma [29], intraocular hemorrhage [21, 30–32], and multiple

Table 13.10. Associated bony fractures in craniocerebral injuries

Site	Intracranial hematoma	Skull fractures	Cerebral contusion, concussion	Total fractures	
				n	%
Claride	1	0	1	2	6.9
Rib	3	8	3	14	48.3
Humerus	1	3	0	4	13.8
Femur	0	3	0	3	10.3
Tibia	1	4	1	6	20.7
Total	6	18	5	29	100

traction changes in long bones. The pathogenesis of long bone changes [22, 33–35], metaphyseal avulsion [22], and subperiosteal hemorrhages [15, 22, 35] was thought to be caused by traction-stretching stresses on the periosteum, induced by grabbing the infants by the extremities or by the thorax, and shaking them, which in turn resulted in the whiplashing of the head onto the thorax. Caffey [20, 21] also stressed that the concurrent absence of external signs of trauma to the head and neck and to the extremities in more than half of their early cases supported a whiplash shaken mechanism.

One cannot provide any conclusive proof of the pathogenetic mechanism with which a head injury occurred. This study is basically an attempt to see if there is an overt clue of pattern of child abuse with which head injury occurred. Data from the confessed story of the original trauma in which head injury occurred are analyzed and compared to follow-up information which was available after interviewing the parents (assailants). The single, most common cause of injury was a "direct hit on the head and/or face" (55 occasions). A direct blow was thought to cause the craniocerebral injuries in at least half of this series.

The infantile brain and its blood vessels are relatively more vulnerable to repeated acceleration and deceleration injury [36, 37]. The pliable sutures and large fontanelles, subarachnoid space, and cisterns allow for an excessive tearing force at the attachment of the vessels. Thus a patient may have subdural hematoma without an obvious direct, violent injury to the head [29]. However, head and face are most commonly involved in the injuries of child abuse, and injuries of the limbs and trunk are relatively less common. This series showed that more than half of the patients had visible external wounds, and that half of the external wounds were found over the head and face. This fact probably implies that a traumatic force of an injury is directed at the head or face.

In the children thrown or hurled against the wall or to the floor, there are not only primary contusions adjacent to the scalp lesions, but also of contrecoup injury, reflected by contralateral subarachnoid hemorrhage and cerebral contusion. Examination of the galea frequently reveals numerous sharply outlined hemorrhages which may often reflect the outline of the weapon employed. This is also true even in the absence of conspicuous external bruising, abrasion, or laceration [38–41]. This observation of galeal hemorrhages is a clue that the child might have suffered a direct or indirect blow to the head or face. Particularly fractures of rib, clavicle, and long bones may reflect the impact of trauma much more accurately in those areas. Pathological studies done by forensic pathologists disclose the fact that internal visceral or organ injury and hemorrhages incurred by a direct blow may exist even without any overt external injuries [42–45]. It is difficult to determine the cause of a child's brain injury, and more so if the child has multiple injuries involving various parts of the body. In the latter case any milder force could also be a deadly blow to the child.

Post-traumatic Cerebrovascular Injuries

The range of vessels damaged from physical insult to the craniocerebrum is complete, though capillary damage (expressing itself clinically as cerebral contusion) in the newborn and venous damage (expressing itself clinically as subacute subdural hematoma) in the infant are most commonly encountered. Because of the redundancy of the arteries within the basal cisterns and fissures, the long course of the cortical bridging veins within the subarachnoid spaces, and location of the superior sagittal sinus beneath the metopic and sagittal sutures, these three vascular structures are particularly susceptible to the shearing forces which represent the characteristic pathogenesis of craniocerebral vascular damage in the fetus, newborn, and infant. This puts into relief the significance of traumatic vascular pathology in these age categories.

In this chapter, traumatic pathology of the vascular structures within the scalp, skull, pachymeninges, cerebral parenchyma, ventricles, and subarachnoid spaces will be discussed. Since there are no vascular structures within the leptomeninges, damage to this structure is not considered. Because of the particular anatomical characteristics of the vault and base of the cranium, vascular injuries resulting from calvarial and basal structures are discussed separately. The significance of shearing injuries of the walls of the lateral and midline ventricular systems, resulting in ependymal rupture, as the pathogenetic factor for intraventricular bleeding, as well as "central cavitation," are described mechanistically as forces responsible for tearing of ependymal and subependymal arteries.

Little attention has been given to *identification* of the bleeding sources: the primary orientation of the surgeon has always been to identify the *location* of the collection of blood so as to remove it. Closed head injuries presenting as transient losses of consciousness were not previously studied neuroradiologically with angiography, but now are studied with CT and MRI. Those remaining in coma and progressing to death or a vegetative state generally have such extensive parenchymal damage that a discreet vascular injury cannot be identified. Since serious head injuries almost invariably present as life-threatening emergencies, little time is spent analyzing the details of vascular damage. This is unfortunate, since capillary, diploic, arterial, venous, and sinus bleeding differ greatly from one another and demand equally different surgical approaches. Generally,

microvascular trauma is clinically not obvious, but if the volume of damage is considerable the typical clinical picture is that of a rapidly expanding intracerebral hematoma.

The mechanics of vascular injury are the same as those for cerebral laceration – shearing forces, explosive forces, and cavitation. These physical insults and stresses are indirect, resulting from different densities of individual anatomical structures and different responses to acceleration and deceleration rates. The vertical course of the brainstem within the basal cisterns predisposes the midbrain and pons to shearing forces. The "floating" corpus callosum (between large pericallosal cistern and III ventricle) is similarly subject to shearing and cavitation forces. The open fontanelles and sutures permit the cerebrum to flatten and expand beneath the skull as the membranous bones separate from one another. This permits much greater stretching of vessels, resulting in tears and stretch occlusion.

Age Categories for Post-traumatic Cerebrovascular Injuries

The nature of injury varies almost directly with the age of the child: perinatal injuries are incurred by the fetus in its passage through the birth canal; the newborn is susceptible to falls in the delivery room and nursery; the infant is subject to falls from the bassinet or high chair and automobile injuries; the toddler injures its craniocerebrum by falling while walking or running, being struck by an automobile, or bouncing around in the automobile as a passenger. All age categories within the first 2 years of life are subject to being battered or shaken. This does not appear to be more common in one cultural setting or another, though there are very real social influences.

Passage through the birth canal subjects the fetus to compressive craniocerebral injuries when uterine cervical dilation does not progress or there is a cephalopelvic disproportion. A precipitous delivery, on the other hand, may result in the child falling to the ground. Another very common cause for traumatic cerebrovascular injuries is forceps application, with high application being associated with a greater incidence of damage and low application a minimal incidence. The high and mid forceps applications damage much more commonly the superficial temporal, middle meningeal, and vertebral arteries. The latter are almost invariably damaged at the craniovertebral junction, in their passage from the foramen transversarium, through the pachy- and leptomeninges, and into the subarachnoid spaces. Therefore, vertebral and basilar artery injuries are incurred, as are vertebral and posterior inferior cerebellar artery injuries; carotid, anterior cerebral, and middle cerebral artery injuries are most uncommon, as are injuries to the deep venous structures. However, traumatic occlusion of the superior sagittal and transverse sinuses does occur.

Except for visible cutaneous evidence of injury (caput succedaneum, scalp or facial contusion), cerebrovascular damage suffered during the perinatal period is generally not identified clinically until much later. In fact, weeks, months, and years may pass before the true nature of neurological dysfunction is identified. The so-called "congenital" giant aneurysm (truly a post-traumatic false aneurysm) of the vertebrobasilar system is an example. The same applies for injuries which the newborn may suffer either in the delivery room or the nursery. Cerebral palsy, minimal brain damage, etc., all may result from this type of vascular injury.

Concerning the infant, falls from high chairs and bassinets are quite common, though a rarity when compared to automobile accidents. In the past several years, successful campaigns have been waged against use of the high chair and bassinet, so that this cause for injury has decreased remarkably. Unfortunately, despite the act that automobile accidents represent the most lethal noxae in the second and third trimesters of life, Western society still has not been able to respond positively. By the time the child becomes a toddler, it begins to fall regularly. Fortunately, the child instinctively falls by dropping to the buttock, so that head injuries from walking-falls are rare. However, the exploratory instincts of a toddler and lack of co-ordination combine to render the child susceptible to falls from chairs, beds, tables, and windows. Also, because of the child's low stature and total unawareness of danger, it suffers a high incidence of being struck by an automobile either in the streets in front of the child's home or, most tragic of all, while playing in the driveway of its own home. The child's high degree of mobility and rapid psychomotor development are responsible for it being allowed to play leisurely in the back seat of a car or in the deck area of a station wagon while the vehicle is being driven by one of the parents.

The first 2 years of life are those in which the child is vulnerable to senseless beatings [11] by the parents, siblings, and sitters. The "battered-child" syndrome is well known. What, apparently, is not well known is that beatings inflict low-velocity acceleration injuries upon the child's head and craniovertebral junction, just the type that most commonly causes linear and eggshell skull fractures, explosive forces upon the membranous bones of the skull, and hyperextension-flexion injuries at the craniovertebral junction. These are common causes for diploic epidural hematoma, petrosphenoid fractures resulting in traumatic occlusion or rupture of the internal carotid arteries, and stretching tears of the vertebral artery in its course from the foramen transversarium of the atlas to the basilar artery. The explosive forces resulting from low-velocity acceleration head injuries split the metopic, coronal, lambdoidal, and sag-

ittal sutures, creating shearing forces, which may either tear cortical bridging veins from the superior sagittal sinus or split the sinus itself for distances of 5–10 mm.

Anatomy of Post-traumatic Cerebrovascular Injuries

So as to put into relief the wide spectrum of vascular damages resulting from craniocerebral injury and to present an anatomicoclinical correlation, two classifications will be presented: (1) anatomical compartments and (2) bleeding sources.

The *anatomical* classification is predicated upon compartments, albeit potential rather than virtual, located between discrete anatomical structures. Thus, the highly mobile scalp overlying the mobile membranous bones provides a potential compartment into which a newborn or infant may bleed enough to present clinically with a picture of hypovolemic shock. Similarly, because of the loose adhesion of the outer layer of the dura to the membranous bones of the skull (with exception, of course, along the suture lines), diastatic fractures of the parietal or frontal bones often result in massive epidural-subgaleal hematomas, which both elevate the scalp and compress the underlying brain. The very dense adhesion of the periosteum to the membranous bones limits subperiosteal hematomas to the area surrounding linear or eggshell fractures. The amount of bleeding into the subperiosteal space, consequently, is of minimal clinical significance. Different from the subgaleal hematoma, it is exquisitely tender and very slow to resolve. The subgaleal hematoma results from tears of vessels located between the periosteum and the galea, the subperiosteal hematoma results from bleeding from the outer table of the membranous bone, and the epidural hematoma results most commonly from diploic bleeding, and only rarely from meningeal artery tears.

Collections of blood within the epidural space, with the exception of crushing and explosive injuries, do not cross the suture lines, since the outer layer of the dura, the suture, and periosteum are an anatomical continuum between membranous bones. Tears in the middle meningeal artery (anterior, middle, posterior branches) do occur during the first 2 years of life. However, this form of vascular injury is not the most common, since tears in the superior sagittal and transverse sinuses represent the large majority of vascular injuries resulting in epidural hematomas. For this reason, newborns and infants generally suffer epidural hematomas which are located along the midsagittal plane, at the vertex, expanding over the parasagittal surfaces of both hemispheres. Also, tears in the transverse sinus result in epidural hematomas which extend from the supra- to the infratentorial compartments. Both of these venous epidural hematomas are treacherous, in that the sinus is torn (establishing an anatomical basis for air emboli). Its repair often necessitates reflecting flaps to either side of the midsagittal plane or across the anatomical line between the supra- and infratentorial compartments. This technique may also provide for repair of the sinus laceration.

Burst fractures, secondary to low velocity acceleration injuries, may strip the outer layer of the dura from the membranous skull, causing bleeding from the inner table of the skull, thereby resulting in well-circumscribed epidural hematomas of very limited volume. This type of epidural hematoma is identical in pathogenesis and extends to the subperiosteal hematoma. The subdural compartment, especially over the convexity of the hemispheres, is most commonly recognized to contain very large (acute or chronic) collections of blood. Although the bridging cortical veins are most susceptible to damage from shearing forces (generally tearing them from the superior sagittal sinus), cerebral laceration and torn dural sinuses not infrequently are the pathogenetic factors. Only rarely do cortical arteries tear, other than in association with compound craniocerebral injuries in which scalp, skull, meninges, and brain are lacerated.

Because of the absence of pacchionian granules in the newborn and infant, the convexity, lateral, and basal bridging veins have lengthy courses within the subdural space. Because of the functional anatomy of the convexity bridging veins, one which brings the vein anteriorly as it exits from the subarachnoid space and then hooks upon itself posteriorly (hairpin style), prior to turning anteriorly once more immediately upon entering the superior sagittal sinus, acceleration injuries most commonly result in tearing the bridging vein from its insertion into the superior sagittal sinus. These are the most common injuries in the newborn and infant. Deceleration injuries, most common in late infancy and the toddler, result in tearing the bridging cortical vein from the cerebral convexity. Therefore, acute subdural hematomas, when removed, may present bleeding sources at either the dural sinus or the cerebral convexity. The same general mechanistic and pathogenic concepts apply to bridging veins entering the cavernous sinus (sphenoparietal system) and the transverse sinus at the sigmoid sinus (vein of Labbé) (Fig. 13.18).

Subarachnoid bleeding is an expression of either regional cerebral contusion or, very rarely, torn arteries. The regional cerebral contusion results either from deceleration injuries, or contrecoup acceleration injuries that damage cortical vessels. Shearing and stretching forces may rent or tear the vertebral, posterior inferior cerebellar, or middle cerebral arteries, permitting an outpouring of arterial blood into the basal cisterns. When the opening in these major vessels of the base of the brain is small, the subarachnoid hemorrhage may not be significant enough to announce itself clinically. Instead, there is a leakage of blood and then a false aneurysm forms around the rent in the artery. These false

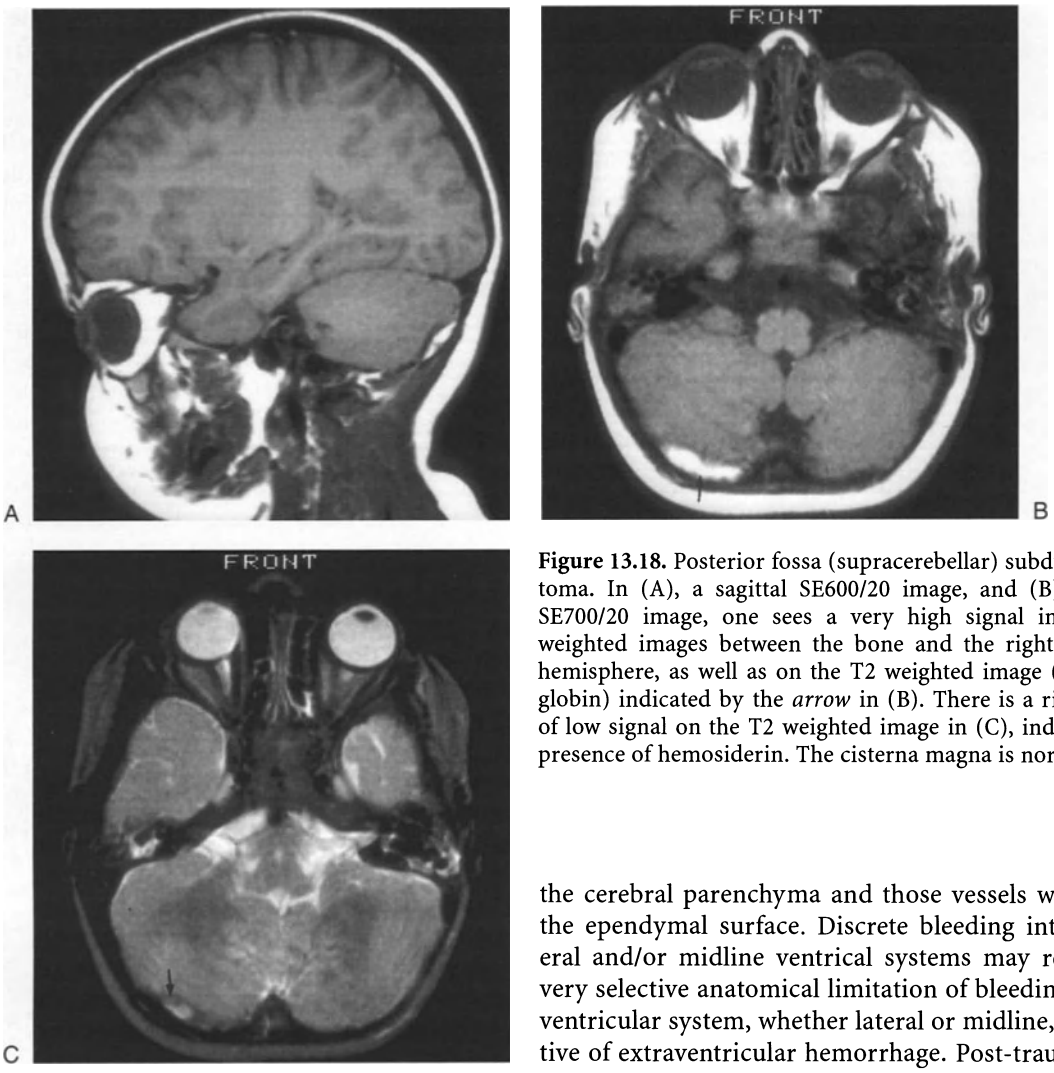


Figure 13.18. Posterior fossa (supracerebellar) subdural hematoma. In (A), a sagittal SE600/20 image, and (B), an axial SE700/20 image, one sees a very high signal in these T1 weighted images between the bone and the right cerebellar hemisphere, as well as on the T2 weighted image (methemoglobin) indicated by the *arrow* in (B). There is a rim (*arrow*) of low signal on the T2 weighted image in (C), indicating the presence of hemosiderin. The cisterna magna is normal.

aneurysms, over time, increase dramatically in size, developing into giant aneurysms or aneurysmal tumors. We doubt that the reported "congenital" aneurysms of the vertebral, posterior inferior cerebellar, and middle cerebral systems are anything other than post-traumatic (false) aneurysms. The fact that these aneurysms are most common on the vertebral basilar system is in support of their post-traumatic nature, because of the high incidence of extension/flexion movements (and injuries!) at the craniovertebral junction during passage through the birth canal.

Intraventricular hemorrhages, other than those occurring in the premature, extremely low birth weight newborn (intraventricular hemorrhage in association with respiratory distress syndrome), may be venous, arterial, or choroidal in genesis. They result from shearing forces across the lamina terminalis, foramina of Monro, and brainstem-cerebral hemisphere junctions. These forces, either direct or indirect (cavitation), tear the cerebral parenchyma and those vessels within it at the ependymal surface. Discrete bleeding into the lateral and/or midline ventrical systems may result. The very selective anatomical limitation of bleeding into the ventricular system, whether lateral or midline, is indicative of extraventricular hemorrhage. Post-traumatic occlusive vascular disease, less dramatic and, consequently, much less often diagnosed, is clinically obvious only when the carotid or middle cerebral systems are involved. Occlusion of a single vertebral or anterior cerebral artery in the newborn and infant is clinically silent. Post-traumatic occlusion of the anteromedial and anterolateral perforating systems, independent of occlusion of the internal carotid, may very well occur. However, it has not been reported and we have never documented a case. Similarly, post-traumatic occlusion of the posterolateral and posteromedial perforating systems, as discrete, isolated entities, does not occur. Basilar fractures, sincipital in this age category, do cause occlusion of the internal carotid arteries within the cavernous sinuses and the basilar artery distal to the anterior inferior cerebellar artery. When fractures extend across the body of the sphenoid, both internal carotid arteries may become stenosed and occluded superior to the cavernous sinuses, and uni- or bilateral caroticocavernous fistulae may occur.

The pathogenesis of traumatic vascular damage (Figs. 13.19, 13.20), whether hemorrhagic or occlusive,

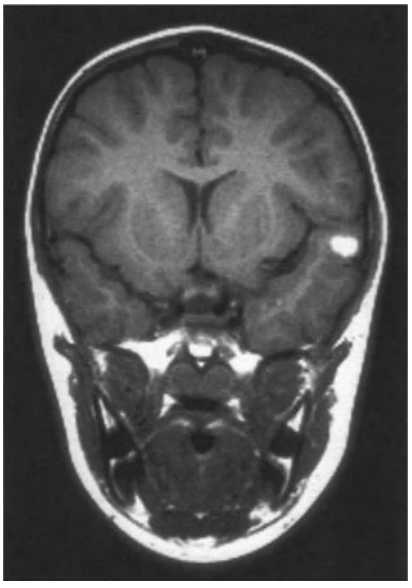


Figure 13.19. Intracerebral hematomas occur commonly following head injury, especially in young children in whom the skull is still somewhat "elastic," resulting from a combination of acute implosion and the consequences of rebound explosion: the parenchyma is damaged by the implosion, and then its reexpansion is so rapid that the microvasculature is torn during the explosion phase, resulting in an intracerebral hematoma which may be self limiting, but is generally progressive. In this illustration, a coronal SE560/15 image, one sees hyperintensity in the left superior temporal lobe. What is visible is meth-hemoglobin, expressive of subacute, post-traumatic intraparenchymal bleeding. The identification of such bleeding on an MRI or a CT study should not permit the surgeon to conclude that the volume of the lesion is defined: it may either expand or the cerebral edema around it become progressive.

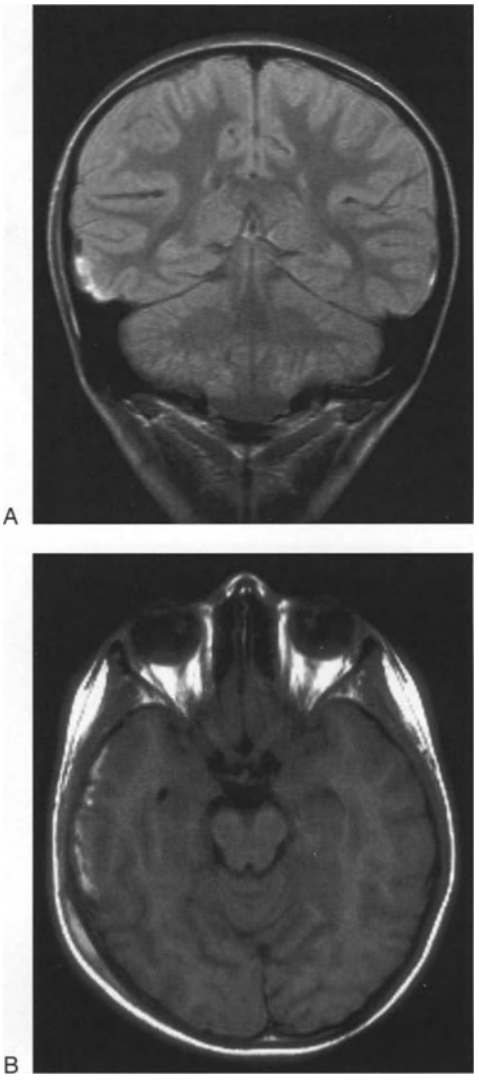


Figure 13.20. Cerebral contusion and subdural collection. This child was struck to the ground by a bicycle, suffering an immediate loss of consciousness which lasted approximately 5 min, then asymmetrical pupils with left-sided hyperreflexia. The clinical signs resolved in 36 h. This MRI image shows clear signs of cortical contusion of the convexity and pole of the right temporal lobe (t-3) with a collection of subdural fluid along the surfaces of T-3, T-2, and T-1 (A) seen best in the coronal projection, and extending from the pole posteriorly to the vein of Labbé (B) seen best in the axial projection. One also sees signs of contrecoup temporal contusion in the left temporal lobe, at T-2 level. Follow-up MRI scans 2 months later revealed no changes in the imaging characteristics.

rests primarily within shearing forces. Indeed, as already discussed, direct injury may be inflicted upon individual vessels. This is rare. Because of the high degree of mobility of the scalp on the skull, of individual membranous bones of the skull upon one another and over the dura, tearing of subgaleal vessels and subgaleal hematoma may result. The hypermobility of the membranous bones of the calvarium, which permits molding of the skull for passage through the birth canal causes, in instances of sudden impact, shearing forces to occur between the skull and the dura, forces that may tear the pachymeninges from the inner table of the skull or lacerate the dural sinuses, both of which result in epidural hematomas. The same forces, if severe enough, may result in tearing bridging cortical veins, subarachnoid arteries, and basal dural sinuses.

The brain of newborn and infants, not yet mylinated, is itself subject to the shearing forces at the cortical subcortical, parenchymal-ventricular, interhemispherical corpus-callosum, and cerebral hemisphere-brainstem levels. The resulting cerebral lacerations and/or cavitation damage parenchymal and vascular structures, with petecchial hemorrhages, contusion, intraventricular bleeding, or intracerebral hematoma occurring. Consequently, when the craniocerebral injury is of

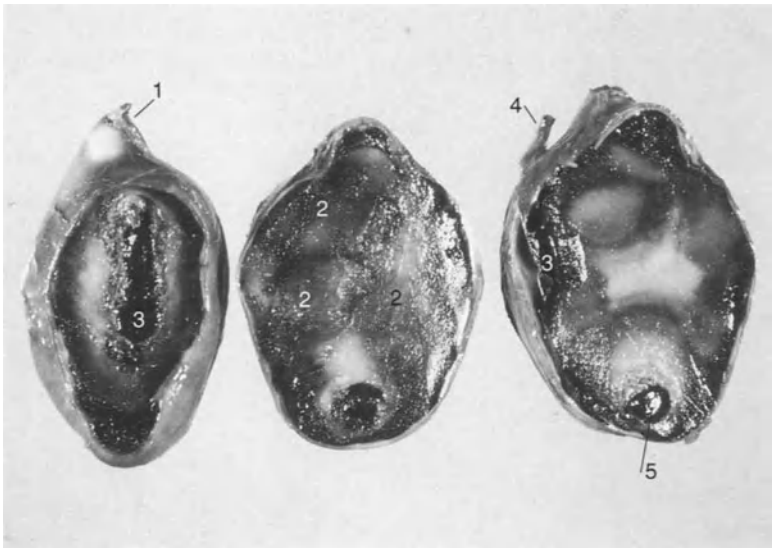


Figure 13.21. Anatomical specimen of a false aneurysm, the most common aneurysm of childhood. In essence, the false aneurysm is the firbrocollagenous capsule tamponading a rent in a vessel, but with a lumen at the center. One sees the parent vessel entering the false aneurysm (*1*), the lumen of the false aneurysm (*2*), the afferent lumen (*3*), stump (*4*), and efferent lumen (*5*) of the parent vessel, which had been damaged. This continuous passage of small amounts of blood into the lumen causes the false aneurysm to increase steadily in size without rupturing: aneurysmal tumor.

such nature and severity as to produce shearing forces, the presence or absence of intracranial mass lesions such as epidural, subdural, or intracerebral hematomas are epiphenomena of no clinical significance, in that the underlying brain damage is, in and of itself, so severe that removal of the intracranial clots will not alter the clinical course.

In sum, post-traumatic intracranial vascular injury may result in discrete, localized, hemorrhages or occlusion. If the hemorrhages occur into potential anatomical spaces (subgaleal, subdural, etc.) and are pathogenic, the removal may be curative. If the vascular damage results in the formation of a false aneurysm or an aneurysmal tumor, they must be treated surgically as soon as identified, since they increase in size and may cause brainstem compression or hemispherical destruction. If the injury causes vascular occlusion, no treatment exists. Cerebral laceration is a defined traumatic condition, which may exist alone or in association with other forms of intracranial vascular damage. Though one may describe its anatomical variants, there is no clinical pathological correlation that permits either prognosis or treatment.

False Aneurysms (Figs. 13.21–13.25)

"There are three types of intracranial aneurysms: (1) mycotic, (2) arteriosclerotic, and (3) congenital. Perhaps a fourth or syphilitic group should be included..." [46].

During the past two score years, intracranial aneurysms have been the subject of rather intense study from therapeutic, statistical, and technical points of view [47–53]. The pathological characteristics of these aneurysms were investigated morphologically [47, 54–59], biologically [60, 61], and hydrodynamically [62, 63]. All of these efforts are being directed toward a better understanding of the natural history and biology of aneurysms.

More recently, a spate of cases concerning the occurrence of aneurysms on the middle meningeal artery [64], the extradural/intracranial portion of the internal carotid artery [65, 66], and intradural branches of the internal carotid artery, secondary to surgical manipulation [67, 68], or in association with recurrent meningiomas [69], have been described. McKissock reported on the recurrence of an aneurysm after he had resected it [70]. Already recognized aneurysms have been observed to increase in size over varying periods of time, with and without repeated subarachnoid bleeds [71, 72].

With the steady increase in direct surgical approach to intracranial aneurysms of the berry variety, and with the advent of interventional neuroradiology and microvascular surgery, the incidence of manipulation of the cerebral vessels has augmented. If, therefore, it could be established that these vessels were prone to form aneurysms, or false aneurysms, following leaks, manipulation, or damage, it would become necessary to add a fifth type of aneurysm to the four described by Dandy, and the already cautious hand of the neurosurgeon would perforce become even more delicate in dealing with cerebral vessels.

The association between trauma to a blood vessel and the formation of a false aneurysm has been well established in all parts of the human body [67, 73–77]. The development of false aneurysms on the extracranial portion of the internal carotid artery has been the subject of case reports, although the exact description of the etiology and formation of a false aneurysm, plus the definition of this traumatic rupture of the internal ca-

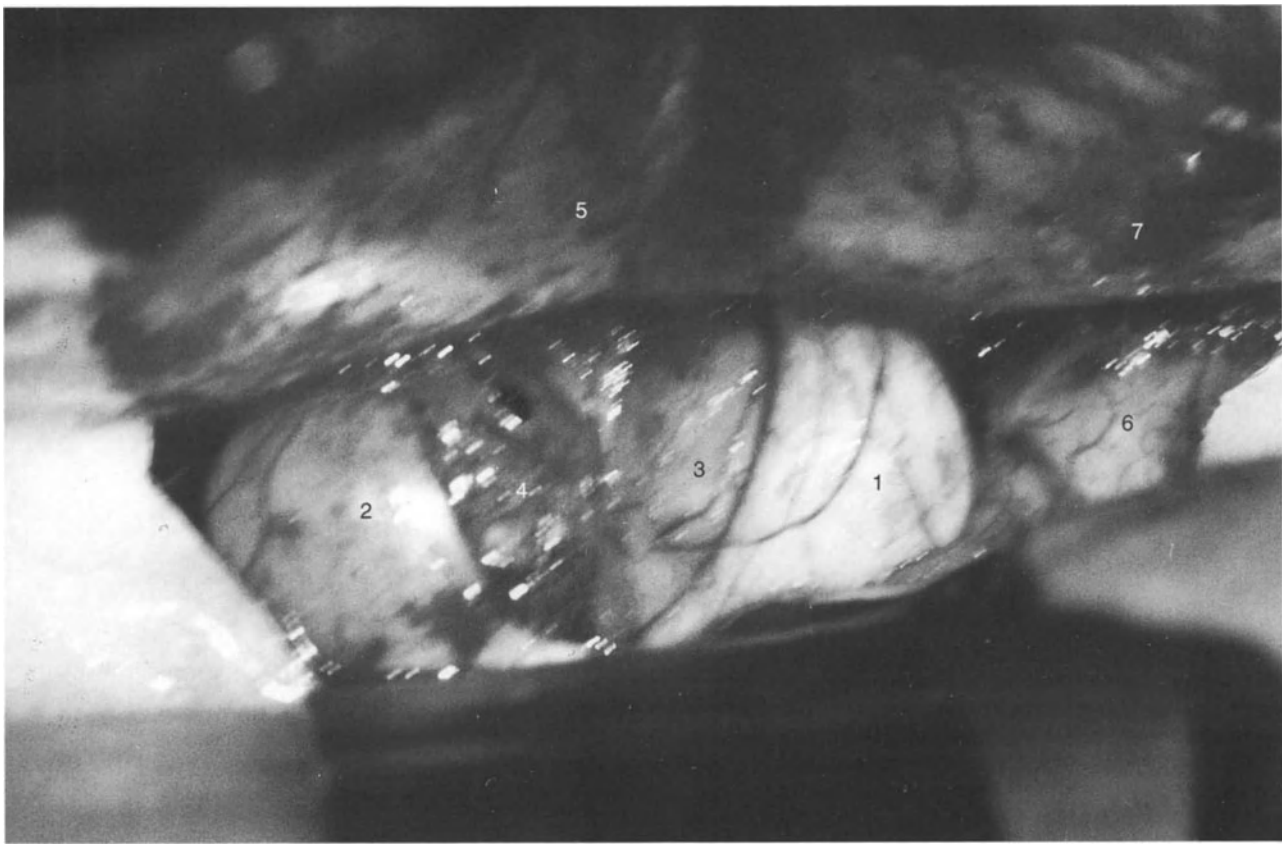


Figure 13.22. Operative photograph of bilateral internal carotid artery false aneurysms elevating and flattening the optic nerves. The right optic nerve (*1*) is more flattened than the left (*2*), and the right internal carotid false aneurysm (*3*) appears to be displaced laterally by a slightly larger left false aneurysm (*4*) which has a clot over its surface. One sees the planum sphenoidale (*5*), right temporal lobe (*6*), and sphenoid wing (*7*). The child's vision returned to normal after the false aneurysms were clipped and the clots evacuated.

rotid artery as such, has not been made. Rather, these publications have dealt primarily with traumatic rupture of the internal carotid artery into the sphenoid sinus as a cause of epistaxis [65] or carotid occlusion (illustrated here in Fig. 13.22). A variety of traumatic aneurysm has been described [78] in which an injury, generally a fracture of the base of the skull, has caused late hemorrhage and death. Birley and Trotter [66] reported a case of "traumatic" aneurysm of the internal carotid (intracranial/extradural) artery, and Goald and Ronderos [79] reported the formation of a traumatic aneurysm on the intracavernous portion of the internal carotid secondary to perforation by a steel spring.

Finkemeyer [67] reported on the formation of a "false aneurysm" on a branch of the middle cerebral artery following manipulation of that vessel during the removal of a meningioma, and Lunn [80] described the formation of a "post-traumatic false aneurysm" on the vertebral artery.

Angiographic diagnosis of extravasation from a torn middle meningeal artery, tamponaded by an expanding epidural hematoma, is both well established and well known [81–84].

Cressman and Hayes [78], reporting a case of traumatic aneurysm of the anterior choroidal artery, concluded that "it is inconceivable that the cerebral substance could provide a buttress strong enough to allow formation of a false aneurysm." However, Sadik and coworkers [72] concluded that a giant aneurysm resulted from "a small aneurysm which had bled and had been surrounded by a large hematoma filling the entire sylvian area and causing the shift." Furthermore, Sugar [77], reporting on the anatomy and angiography of vascular malformations, very clearly demonstrated, and documented, the existence of a false aneurysm that had developed from an intracerebral bleed from an arteriovenous malformation. He stated that a large "sac was associated with the anomaly" and that "this proved, on histologic examination of the surgical specimen, to be a false aneurysm, however, with no true vascular wall. There was a clot of many layers of various ages, surrounded by a thin fibrous tissue sheet, which was con-

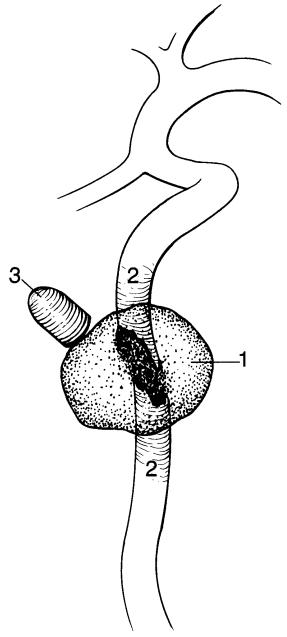


Figure 13.23. This is a composite drawing of the operative findings in a patient with a post-traumatic false aneurysm (gunshot wound), demonstrating the fresh clot (*1*) encircling both severed ends of the internal carotid artery (*2*) and thereby permitting the passage of blood across the intervening space. One bullet (*3*) caused the tear.

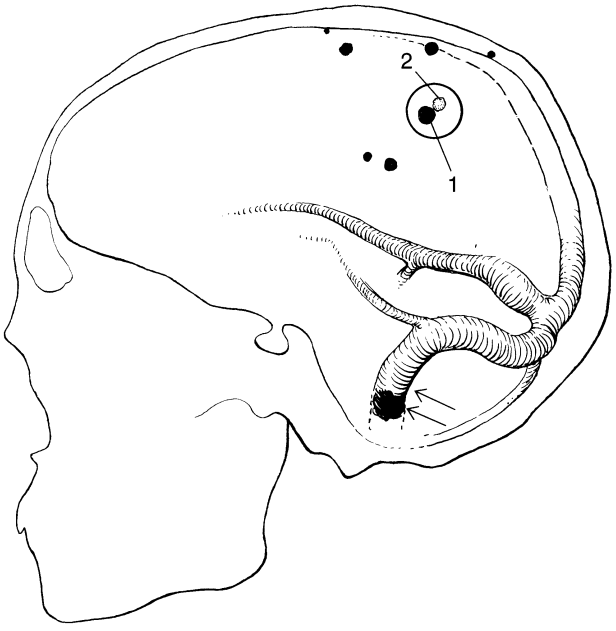


Figure 13.24. Composite drawing of a venous phase angiogram in a previously reported child who had suffered a buckshot wound to the head. *The black dots* indicate the pellets, (*1*) indicates a pellet which damaged a cortical artery, (*2*) indicates persistent filling of a cortical false aneurysm, and *the arrows* indicate the post-traumatic occlusion of the jugular bulb.

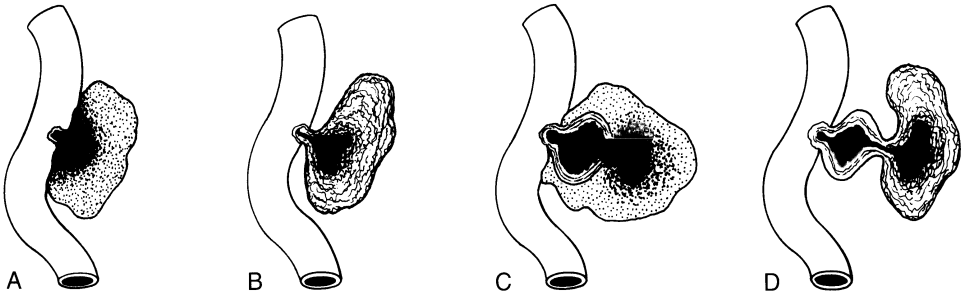


Figure 13.25. (A) The schematic drawing depicts a rent in an artery tamponaded by freshly formed clot. This is an *acute post-traumatic false aneurysm.* (B) The rent in the parent artery is tamponaded by fibrocollagenous tissue, which resulted from consolidation of the hematoma. This is a *chronic post-traumatic false aneurysm.* (C) In this figure a ruptured berry aneurysm is tamponaded by freshly formed clot. The aneurysm is schematically represented in cross section. This is an *acute spontaneous false aneurysm.* (D) In this schematic representation of a *chronic spontaneous false aneurysm*, the breach in the berry aneurysm is tamponaded by fibrocollagenous tissue which has resulted from consolidation of the surrounding hematoma. The berry aneurysm is depicted in cross section so as to show endothelial, elastic, and adventitial components, which stop abruptly at the point of transition into the false aneurysm. Also, the lumen of the berry aneurysm is continuous with the reservoir within the false aneurysm.

tinuous with the adventitia of a large vascular channel. Doubtless, this had bled many times, and formed a protective covering which contained the clot which compressed the brain."

Bjorkesten and Troupp [71], reporting on 700 intracranial aneurysms, cited 18 cases which had repeat angiography after an interval ranging from 2 weeks to 10 years. Fully 10 of these 18 cases had an increase in the size of the aneurysm. They cite Lofstedt as having stated that "exploration may be a trauma to the aneurysm, and may further its growth." Taveras and Wood [85], stressing the rarity of traumatic aneurysm, stated that "the angiographic enlargement of an aneurysm during a brief interval indicates a deterioration in the

wall of the sac which may be ominous." Figure 525 (p. 1733) in their book is an example of an acute spontaneous false aneurysm.

By false aneurysm, or pseudoaneurysm, it is understood that one means to describe a pooling of blood in a cavity whose walls are composed, in the fresh state, of clot which has tamponaded the egress of blood from the arterial rent, and, in the chronic state, by stratifications of the fibroconnective tissue which organizes from the clot. Therefore, it is possible to separate a false aneurysm from any of the other varieties of aneurysm by microscopic examination [86, 87]. In the former, one will find either fresh clot or fibroconnective tissue, but neither intima, nor adventitia, nor elastica. These latter are invariably present, though defective, in all of the other types of intracranial aneurysms, irrespective of the classification adopted [46, 51, 80, 86, 88, 89].

False aneurysm of the cerebral vessels may occur independent of trauma as, for example, in the case of a ruptured berry aneurysm, which serves as the focal point for its formation. The berry aneurysm, by rupturing, may bleed out into a lobe, and the resultant intracerebral hematoma serve as a tamponade to arrest the hemorrhage, without sealing the breach at the point of rupture. This, we believe, is the mechanism by which some of the giant aneurysms which have been described in the literature are formed [71, 90]. We have had cases of false aneurysm formation developing secondary to bleeding from a berry aneurysm and culminating in the formation of a true giant aneurysm, aneurysmal tumor [91].

One may, therefore, consider three basic forms of false aneurysms:

1. Traumatic
   a) Acute: consists of a rent in an artery and has a wall composed of fresh clot (Fig. 13.25A).
   b) Chronic: consists of a rent in an artery and has a wall composed of fibroconnective tissue, or laminations of collagen, which results from the organization of the clot (Fig. 13.25B).
2. Spontaneous
   a) Acute: consists of a ruptured berry, or arteriosclerotic, aneurysm, and fresh hematoma tamponading the breach (Fig. 13.25 C).
   b) Chronic: consists of a berry, or arteriosclerotic, aneurysm that has ruptured and is sealed by fibroconnective tissue and laminations of collagen resulting from the organization of the clot (Fig. 13.25D).
3. Aneurysmal tumor: a progressively expansile mass that develops from a chronic false aneurysm of either the traumatic or the spontaneous varieties, and whose enlargement results from the sum of the pulsatile forces of blood entering the aneurysmal reservoir and the deposition of laminations of fibroconnective tissue and collagen along the inner surface of the organizing clot (Fig. 13.21).

The therapeutic implications, from a surgical point of view, become readily obvious when we consider each of these various forms of false aneurysm in turn. In dealing with the false aneurysm that consists simply of a rent in a vessel tamponaded by fresh clot, of the acute traumatic variety, the surgeon must be prepared to sacrifice the parent vessel, for he will not find a neck he may clip or a fundus he may coat with plastic. An alternative, preferable, is the insertion of a stent. The approach to this type of aneurysm is best made through the clot so as to permit more direct visualization of the rent. In dealing with the acute spontaneous false aneurysm, the neck of the berry or arteriosclerotic aneurysm may be clipped after the hematoma has been evacuated.

I do not think that chronic false aneurysms, either traumatic or spontaneous, should be approached surgically. This conclusion is based upon the pathological nature of the aneurysm. Since there is already a firm fibroconnective capsule, walling off the rent in the vessel or the breach in the true aneurysm, one has little to gain by attempting either to exclude the aneurysm from the circulatory tree or to reinforce it with plastic.

The third type of false aneurysm is the most challenging, since it presents the problem of an aneurysm as the focus of a progressively expansile intracranial mass. Such an *aneurysmal tumor* has a relentless course; consequently, surgery becomes mandatory.

The realization that it is possible to produce false aneurysms by *manipulation* of the cerebral blood vessels or by the *application* of clips to them, even if only for brief periods of time, is sobering indeed. There is no definite information as to how long Finkemeyer [67] had occluded a branch of the middle cerebral artery in his "manipulative" false aneurysm. I conclude that what appears to be slight damage to the artery may very well result in the formation of a false aneurysm. The observations of Boldrey (cited by [92]) support this point, though the latter were careful to state that they did not visualize the anterior cerebral artery during resection of the meningioma. Similarly, the experience of Overton and Calvin [82] in their "iatrogenic cerebral cortical aneurysm" is strong supportive evidence that trauma may produce false aneurysms. The work of White and colleagues [93] offers an experimental basis for this conclusion.

Cranioplasty (Fig. 13.26)

Either autogenous bone (rib) or plastic (methyl methacrylate) are currently used for cranioplasty. Methyl methacrylate is discouraged for many reasons:

1. It is a foreign substance.
2. As a foreign substance, it cannot heal to the surrounding bone.
3. Its polymerization is an exothermic reaction that may damage the underlying dura and brain.
4. It may become dislodged and push into the underlying brain.
5. It is subject to cracking.

On the other hand, ribs are readily available and regrow within months after their removal; they may be molded into the proper contour, they fuse readily to the surrounding skull, and they heal into normal bone that affords the same protection to the brain as the skull. The only disadvantage of rib graft is that it entails an additional operative procedure: a thoracic skin incision and subperiosteal resection of the desired amount of rib.

The technical aspects of implanting the rib graft consist of splitting the bone graft into symmetrical halves by cutting through the cancellous bone in the long axis of the rib, and then molding the segments to the desired contour after cutting them in appropriate lengths to fill the defect. Once this is done, the graft should be put aside and the area of skull defect prepared for their implantation.

The incision is made and the scalp is reflected from around the defect, exposing the periosteum. This is incised, and the potential space between it and the underlying dura is opened into a virtual space by using Adson-Brown forceps to hold the periosteum and blunt-tipped tenotomy scissors to separate periosteum from underlying dura. At times this is simple, but at others it is quite a tedious dissection. One must take care not to cut through the dura and open into the subdural space. Once the potential space between periosteum and dura has been fully converted into a virtual space, a curette is used to scrape away the compact bone from the edges of the defect to expose cancellous bone, taking care not to poke through the underlying dura or the overlying periosteum. This facilitates healing of the edges of the bone graft to the surrounding skull.

After the defect has been prepared as described above, the individual pieces of rib graft are laid into the interval between periosteum and dura mater, each in its place. They are then placed into the interval between perisoteum and dura, as into a pouch. It is not necessary to anchor the edges of bone graft to the surrounding skull, since the periosteal/dural pouch holds them snugly in place and assists holding them in the proper contour. The opening in the periosteum is then closed. The use of wire to bind one rib to the other is unnecessary, and has the disadvantage of rendering subsequent CT or MRI scanning difficult or impossible.

When it is not possible to convert the potential space between periosteum and underlying dura into a virtual space without fragmenting the periosteum, one takes a periosteal graft from the surrounding skull and brings it over the positioned rib grafts, covering the defects. The periosteal graft holds them in place and assists bony healing.

Since it is my opinion that methyl methacrylate should not be used for a cranioplasty because of the above-described reasons, the technique for performing this procedure is not described. Rib or iliac crest are always available. There is no justification for the insertion of this undesirable material.

Vertebral Fracture Dislocation

For a multitude of reasons, ranging from anatomy through epidemiology, fracture dislocations of the lumbar and thoracic spines are, practically speaking, rarities in the newborn, the infant, the toddler, and the juvenile. Cervical fracture dislocations occur in the newborn as a result of birth injury. In the infant, toddler, and juvenile they are clinical curiosities. Unfortunately, cervical injury incurred during passage through the birth canal is fatal, and generally goes undiagnosed, since the newborn is apneic at birth. When correctly diagnosed, the outlook is grim, even though the child may be temporarily resuscitated by intubation and artificial ventilation.

There is no place for fusion in the management of cervical fracture dislocations, since children realign properly, and fractures heal spontaneously, with simple *immobilization* for periods of time ranging from 4 to 12 weeks. In fact, fusion is to be discouraged in newborns, infants, toddlers, and juveniles, since it arrests growth!

The developing spinal column is an extraordinarily complicated anatomicophysiologic structure, one which is best managed by a pediatric orthopod, a person familiar with techniques for immobilization and healing processes of growing and ossifying bone.

Figure 13.26. Legend see p. 457.

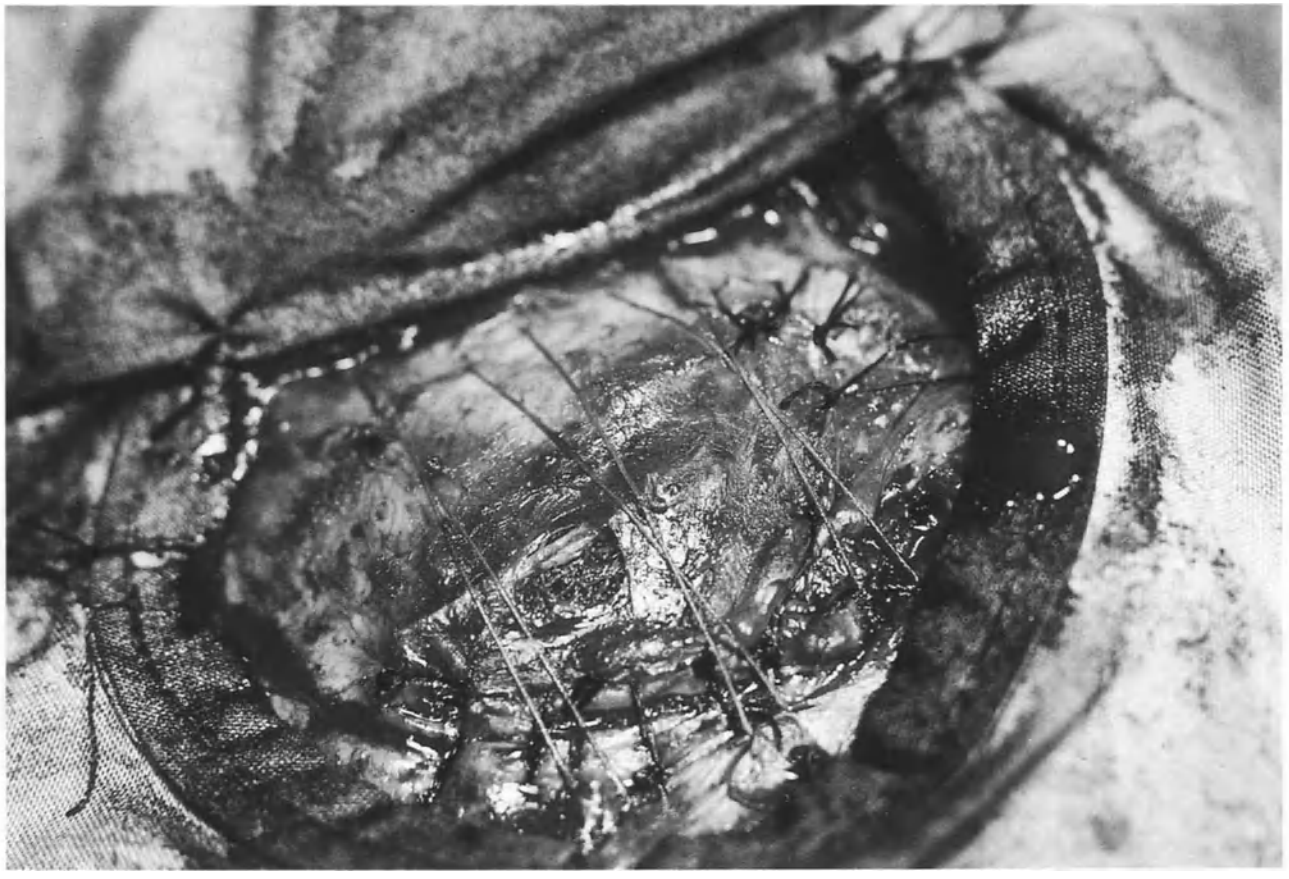


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Figure 13.26. The various stages in performance of a rib cranioplasty. (A) The periosteum (*1*) has been separated from the dura (*2*), opening up a large pocket between the two which will serve as a pouch into which the rib grafts may be inserted. The dura is then sewn tautly to the periosteum over normal skull (*3*) tightening it so as to diminish redundancy. (B) The rib grafts have been split (*1*). They are molded into the desired contour and cut the desired length prior to inserting them into the previously prepared pouch (*2*). (C) After all the rib grafts have been put into place, filling completely the defect, the periosteum over the rib graft pouch is sewn to the periosteum over normal skull. Absorbable sutures are then run across the rib grafts and its pouch, holding the former in place and maintaining the contour.

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14 Congenital Anomalies

"Here's your opposition! here's your two sets o'heirs to old Peter Wilks – and you pays your money and you takes your choice!"

MARK TWAIN, Adventures of Huckleberry Finn

Congenital Anomalies Involving the Craniocerebrum and Craniocervical Junction

Synostotic Cranial Anomalies

General

The hyperostosis of the sagittal and metopic synostotic sutures is generally more remarkable over the outer table, whereas coronal synostosis is characterized by exuberant hyperostosis along the inner table. Therefore, the synostosed sagittal suture may be lifted easily and safely from the superior sagittal sinus, since it is not adherent to this structure. The synostosed coronal suture, conversely, protrudes into the underlying dura and is generally adherent to it.

Whether single or multiple, the suturectomy consists of excision of the hyperotosis which occurs along the line of the prematurely closed suture. It must extend the entire length of the closed suture, and across the normal suture line perpendicular to it. The synostosis of the suture, in the pathological state, extends across all bones bordering upon it. Extending the suturectomy only into the perpendicular normal suture does not assure relatively permanent opening. For example, in performing a sagittal suturectomy, one must take care to bring the line of osteotomy *across the normal suture* and into the frontal bone anteriorly and the occipital bone posteriorly, for a distance of about 1 cm at either end.

The suturectomy for craniosynostosis may be performed along a single suture, the metopic in children with *scaphocephaly*; along two sutures, coronal and pterional, for children with *plagiocephaly*; or along multiple sutures: sagittal, coronal, pterional, and lambdoidal, for *pansynostosis*.

Some authors [1, 2] have continued to follow the recommendations of Ingraham and coworkers [3], who suggested applying foreign material to the linear craniectomy borders to prevent regrowth, though others [4] have found this technique to be of no value and to suffer the complicating factor of extrusion of the plastic from between the bone edges and through the wound. Anderson and Johnson [5] advocated the use of Zenker's solution and acetic acid, applied to the dura, to delay or arrest rapid growth of bone at the craniectomy sites. This has proven to be ineffective and complicated by temporary or permanent seizures in addition to persistent large cranial defects (often necessitating cranioplasty).

Suturectomy should be performed, ideally, between the 3rd and 7th months of life. This allows the child time to acquire its own immunological responses and to establish eating habits and diurnal pattern. Also, operating this early prevents deformity of the brain and skull, and diminishes risks of permanent skull deformity. The recurrence rate among children operated on before 3 months of age is much higher than those operated on between 3 and 7 months of age.

Metopic (Including Frontonasal) Synostosis: Trigonocephaly (Figs. 14.1–14.3)

Premature closure of the metopic suture results in a rather characteristic keel-like deformity of the frontal bone because of impaired expansion along the metopic suture and accentuated expansion along the coronal suture bilaterally. Brain growth occurs longitudinally, increasing the length of the skull anterior to the coronal suture. Since the sagittal suture is functional, there is both horizontal and longitudinal growth posterior to the coronal suture. No frontal eminences develop, and there is no protrusion posteriorly with ledge deformity of the occipital bone.

Surgical correction of metopic synostosis consisted classically of a suturectomy, extending from approximately 1 cm posterior to the coronal suture to 3 mm proximal to the frontonasal suture. For cosmetic and anatomical reasons, it had been considered impossible

to extend the suturectomy across the frontonasal suture. This technique suffices for immediate cosmetic appearance in the very mild cases, but does nothing to correct the hypotelorism.

Current technique, in light of the remarkable advances made in craniofacial surgery, consists of dissection of the periosteum from the coronal suture to about 2 cm on either side of the midsagittal plane. The coronal suture is then entered and the dura is separated from the inner table of the hyperostosed metopic suture with the use of a Penfield #1 or #2 dissector. The periosteum is removed from over the planned osteotomy line, 1 cm lateral to the midline on either side, from 1 cm behind the coronal suture to 2 mm across the frontonasal suture. Either a craniotome or a fine-tipped Leksell rongeur is then used to make the osteotomy, extending from the coronal suture into the frontonasal suture on each side, and then across the midline first posteriorly and then at the frontonasal region. The strip of synostosed metopic suture is lifted away from the anterior third of the superior sagittal sinus, which, at this age, holds minimal amounts of venous blood. The rongeur is used to nibble across the coronal suture posteriorly and frontonasal suture anteriorly. Finally, a small osteotomy is extended from one nasal bone to the other across the nasal suture.

The very severe cases, those accompanied by synostosis at the frontoethmoidal sutures, require *en bloc* removal of the entire frontal bone, with the supraorbital rims, freeing the frontal bone from the ethmoid. The orbital roof is opened posterior to the supraorbital rim. The removed frontal bone is then split down the synostosed metopic suture, cracked, molded into a cosmetically agreeable form, and repositioned over the frontal lobes. This procedure must be performed with an experienced plastic surgeon. The hypertelorism is managed by the plastic surgeon.

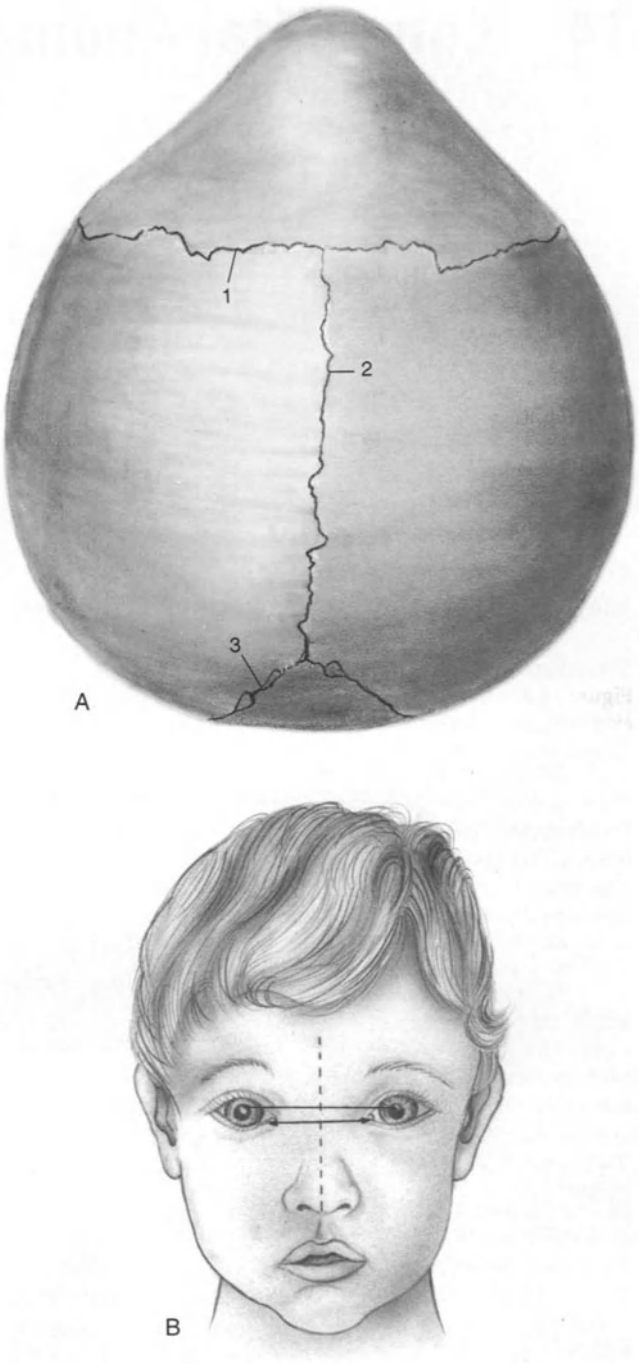


Figure 14.1. (A) Premature synostosis of the metopic suture (*viewed from above*). Though brain and skull growth in the longitudinal axis is much greater than normal, the resultant cramping of the frontal lobes displaces the brain posteriorly, across the line of the coronal suture *(1)*. Because of functional sagittal *(2)* and lambdoidal *(3)* sutures, that portion of the skull posterior to the coronal suture expands circumferentially. This is shown diagrammatically. (B) This is a drawing (one of three on ocular measurements redrawn from the book by Davis et al. [1]) illustrating measurements for intercanthal and interpupillary distances. In trigonocephaly, there is very often hypotelorism.

Sagittal Synostosis: Scaphocephaly

(Figs. 14.4–14.8)

Synostosis at the sagittal suture impedes growth and expansion at the junction of the parietal bones. The result is an elongated head with accentuated frontal bossing and protrusion of the inion: *scaphocephaly.* Consequently, in order to accommodate the expanding brain, there is compensatory growth between the parietal and occipital bones at the lambdoidal suture, the frontal and parietal bones at the coronal suture, and the frontal bone at the metopic suture.

Resection of the sagittal suture is performed after an S-shaped biparietal skin incision (permitting access to the anterior fontanelle and inion) has been made and the two portions of the scalp flap reflected to either side of the midline, exposing the hyperostosed area of the sagittal suture from anterior to the coronal to posterior

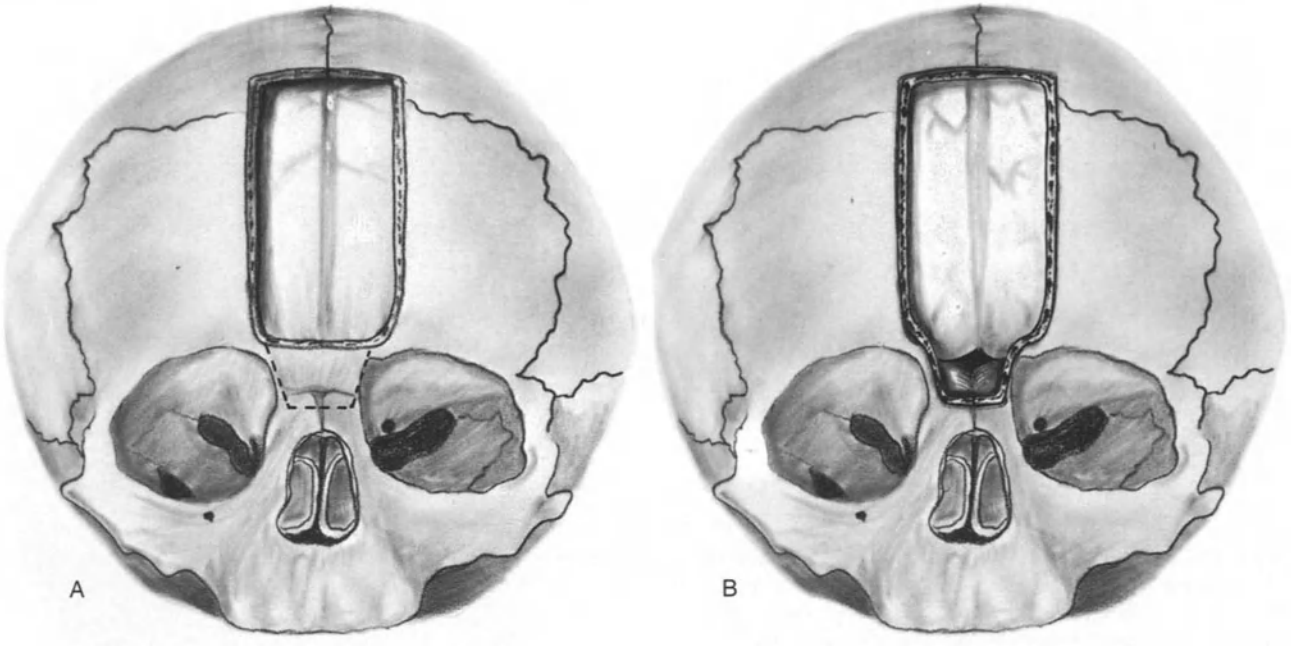


Figure 14.2. (A) Resection of the synostosed metopic suture exposes the underlying anterior third of the superior sagittal sinus. Note that it extends into the sagittal and across the coronal sutures posteriorly, but that it is not carried anteroinferiorly across the frontonasal or into the nasal sutures (*area within dotted lines*). Consequently, there will always remain an observable hyperostosis at the bridge of the nose. This limited procedure provides one with the illusion that the synostosis has been effectively dealt with because the keel-like deformity of the forehead is removed and the frontal bone may flatten as it reforms along the sagittal plane. However, the frontonasal synostosis persists, as does the hypotelorism. The cosmetic effect is less than desirable. (B) The only effective way to release completely the synostosed metopic suture is to extend the osteotomy across the frontonasal suture, and then to run it across the nasal suture. This releases the anteriormost portions of the medial orbital rims from one another, allowing the subsequent brain growth to correct partially the hypotelorism. However, since the hypotelorism is an expression of synostosis also of the frontoethmoidal, nasoethmoidal sutures, complete correction entails a much more extensive procedure.

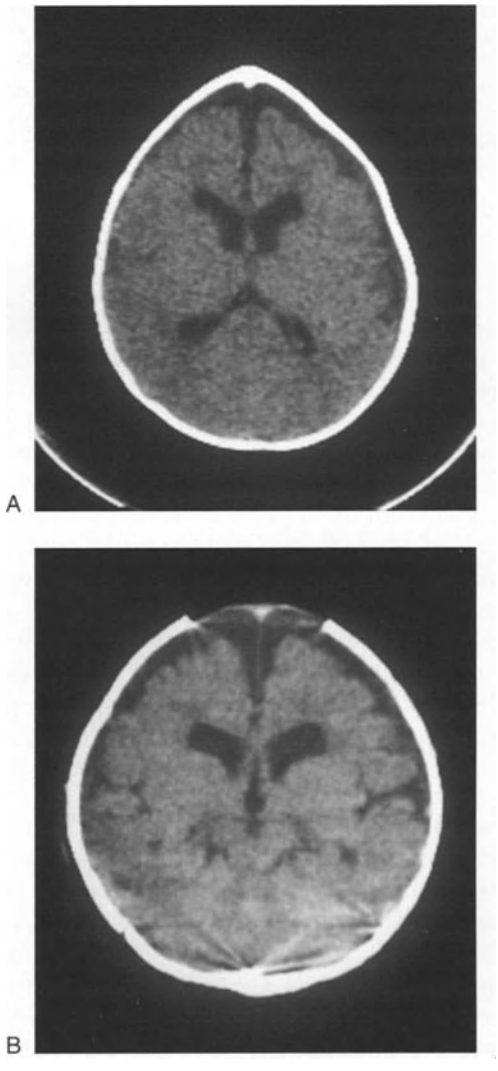
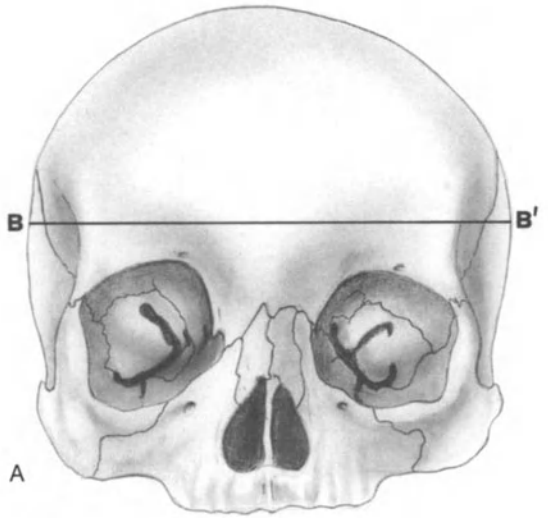
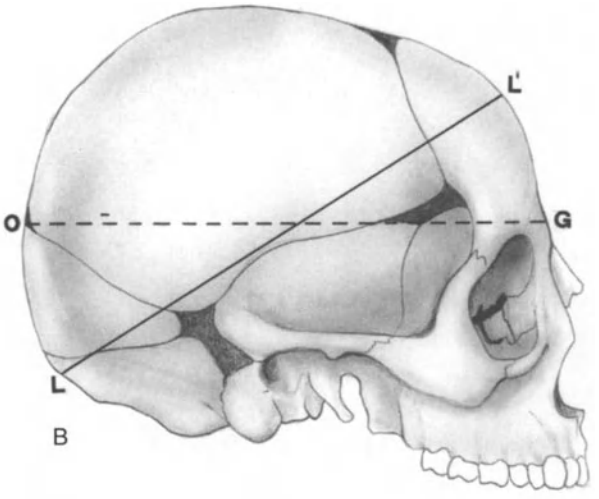


Figure 14.3. (A,B) These imaging studies reveal the pre- and ► postoperative appearance of the trigonocephalic skull operated neurosurgically as illustrated in Fig. 14.2B: the metopic suture was open across the frontonasal sutures. It is also important in these images to see the increased accumulation of cerebrospinal fluid in the subarachnoid spaces over the frontal lobes and the moderate ventriculomegaly. For more extensive treatment of the relationship between metopic synostosis and hydrocephalus, see Chap. 15, "Hydrocephalus."





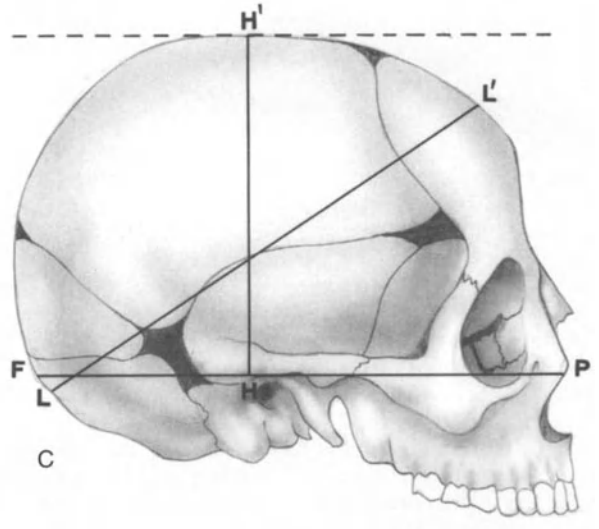


Figure 14.4. Cranial measurements. We have never done cephalometric calibrations, neither pre- nor postoperatively. Rather, it has been our approach to observe the form of the skull, its contour, orbital harmony and interrelationship, intracranial cerebral/cerebrospinal fluid relationships, and status of intracranial pressure. However, if one wishes to monitor skull sizes numerically, the criteria put forth in the book by David et. al. are quite adequate: the horizontal cephalic index (A), the anteroposterior diameter and occipital glabellar line (B), and the vertical cranial index measured as a perpendicular to the Frankfurt line from the superior border of the external auditory canal to the vertex (C).

to the lambdoidal sutures. The hyperostosis may be so significant at times as to result in an elevation of bone along the area of the sagittal suture.

One notes immediately that there is a ridge along the line where the sagittal suture had been, and that the coronal and lambdoidal sutures may be identified but not the sagittal suture. In fact, if the sagittal suture may be identified from coronal to lambdoidal sutures, the child does not have sagittal synostosis. Microcephalics have been confused with scaphocephalics.

After the skin flaps have been reflected to either side and before periosteal stripping, the flat edge of the unipolar thermocautery blade is used to cut the periosteum and expose the underlying skull. The periosteum is stripped from the parietal, frontal, and occipital bones, taking care to dissect it well across the coronal and lambdoidal sutures. It is not safe to use the thermocautery blade when crossing the coronal and lambdoidal sutures since it may coagulate through the suture and into the underlying brain, nor to use its cutting edge. Periosteum, dura, and suture are one fibromembranous continuum at the suture line.

Dissection across the coronal and lambdoidal sutures is performed with the periosteal elevator, after the peri-

Figure 14.5. The elongation of the head in scaphocephaly is an expression of longitudinal growth at the coronal and lambdoidal sutures, and vertical growth at the parietotemporal and sphenoparietal sutures (in the region of the pterion). (A) The scaphocephalic deformity. (B) Widening of the coronal and lambdoidal sutures. Because of arrested growth at the sagittal suture, the expanding brain volume is accommodated by growth at the coronal and lambdoidal sutures, with resultant lengthening of the parietal bone, posteroinferior displacement of the lambdoidal suture, and a ledgelike deformity at the inion (*arrow*). There is also anterior displacement of the coronal suture. (C) Note the bone ridge in the sagittal plane (*arrows*), which does not extend across the coronal or lambdoidal sutures. There are neither frontal nor parietal eminences, and the fontanelles are absent.

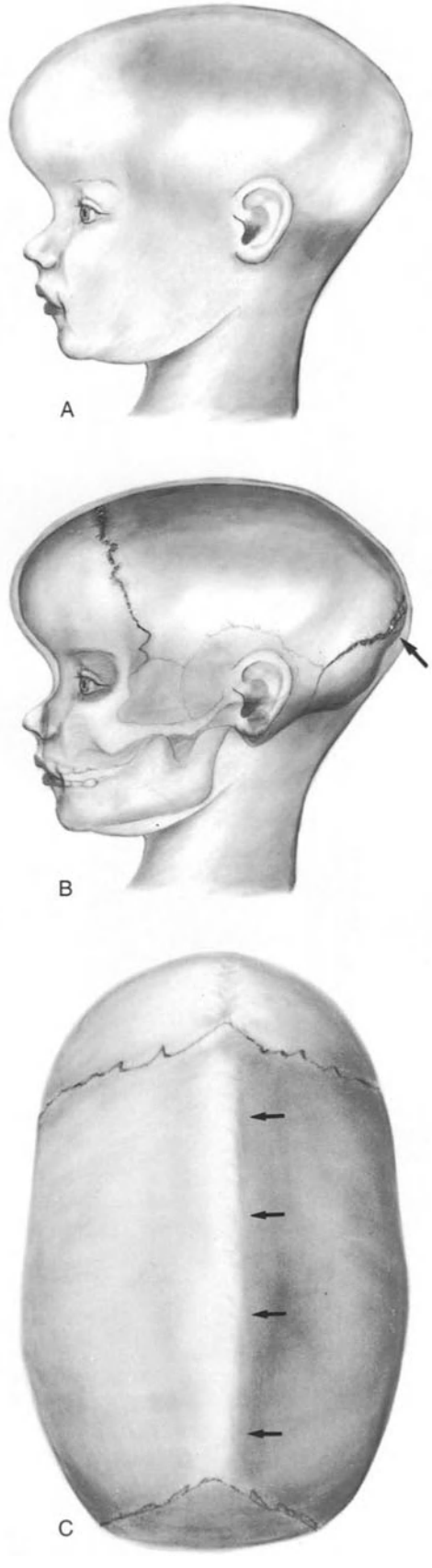


Figure 14.5 (A–C). Legend see p. 464.

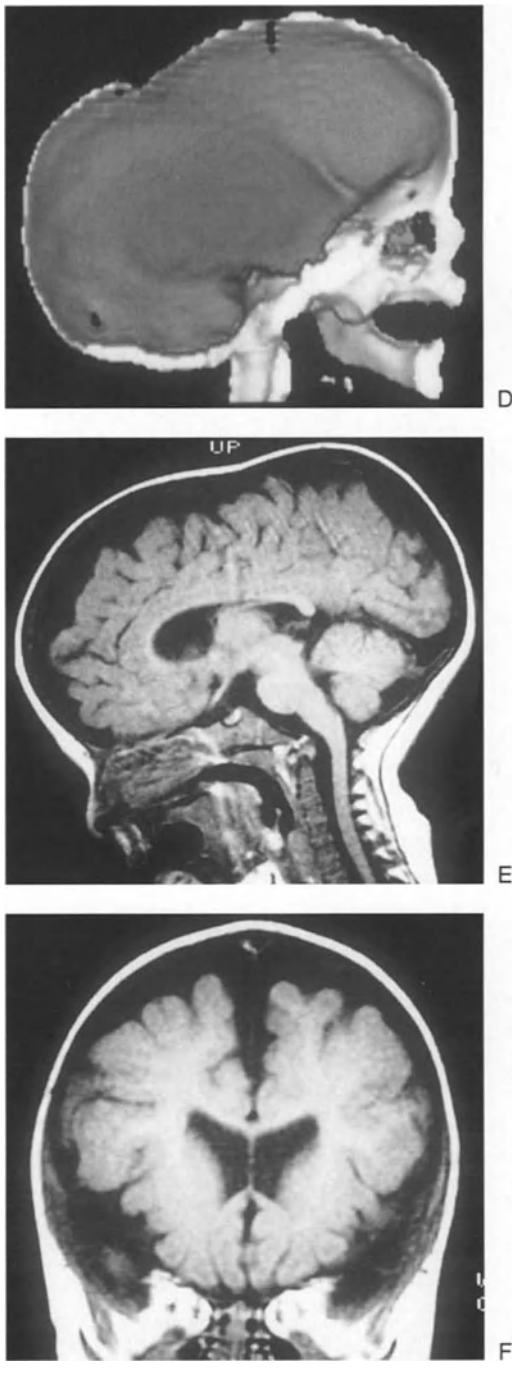


Figure 14.5. (D) This CT reconstruct illustrates the scaphocephalic deformity with a prominent frontal boss and an extremely prominent occipital boss plus overhanging of the inion. In (E), the T1 weighted MR image of the same child, one sees not only the scaphocephalic deformity but the elongation in the anteroposterior direction of the brain and, much more important, the remarkable accumulation of cerebrospinal fluid in the subarachnoid spaces over the convexity and along the medial surface of the hemisphere. The coronal T1 weighted image illustrated in (F) reveals ventriculomegaly and accentuates the visualization of the cerebrospinal fluid in the subarachnoid spaces and along the gyri.

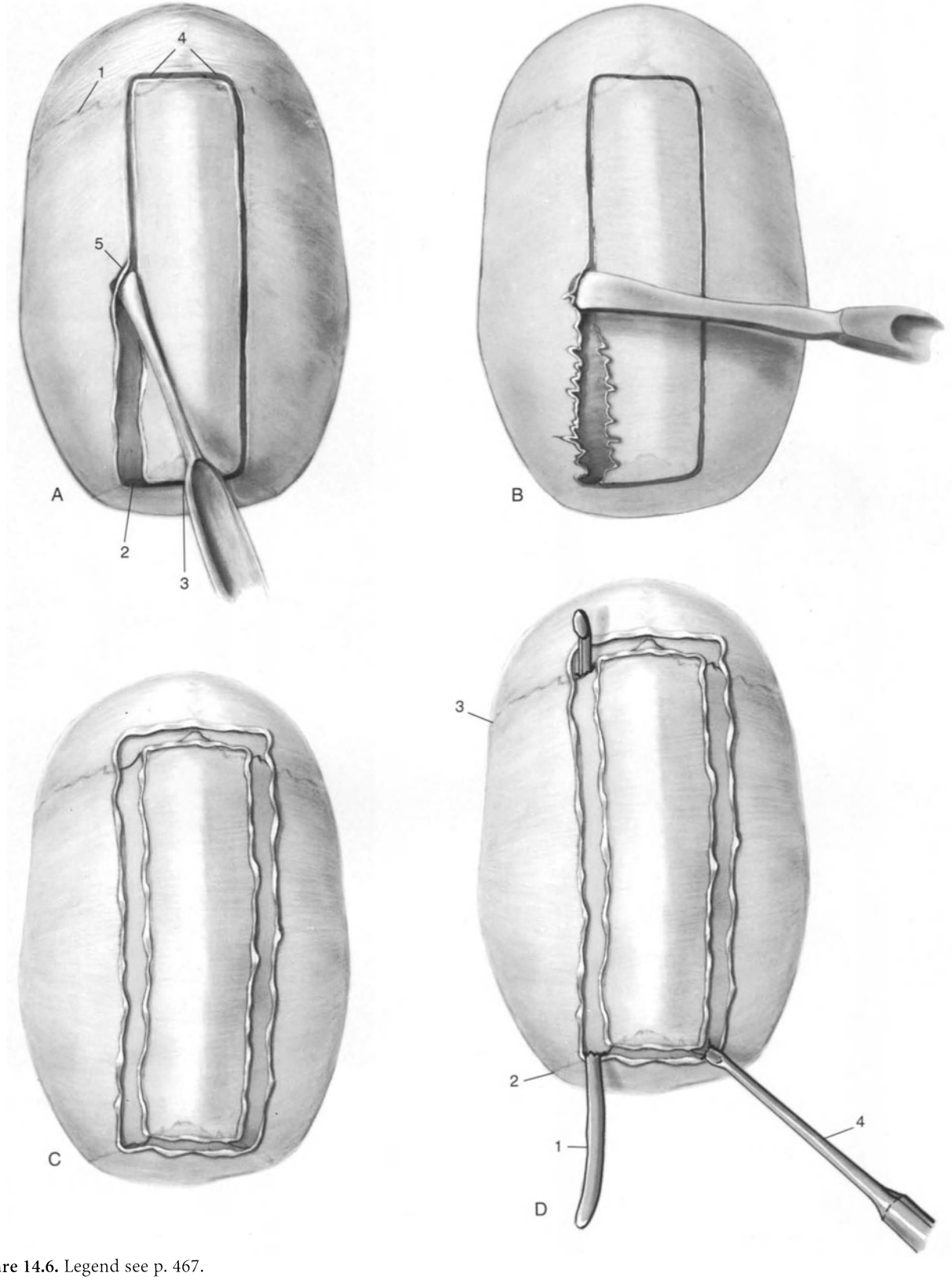


Figure 14.6. Legend see p. 467.

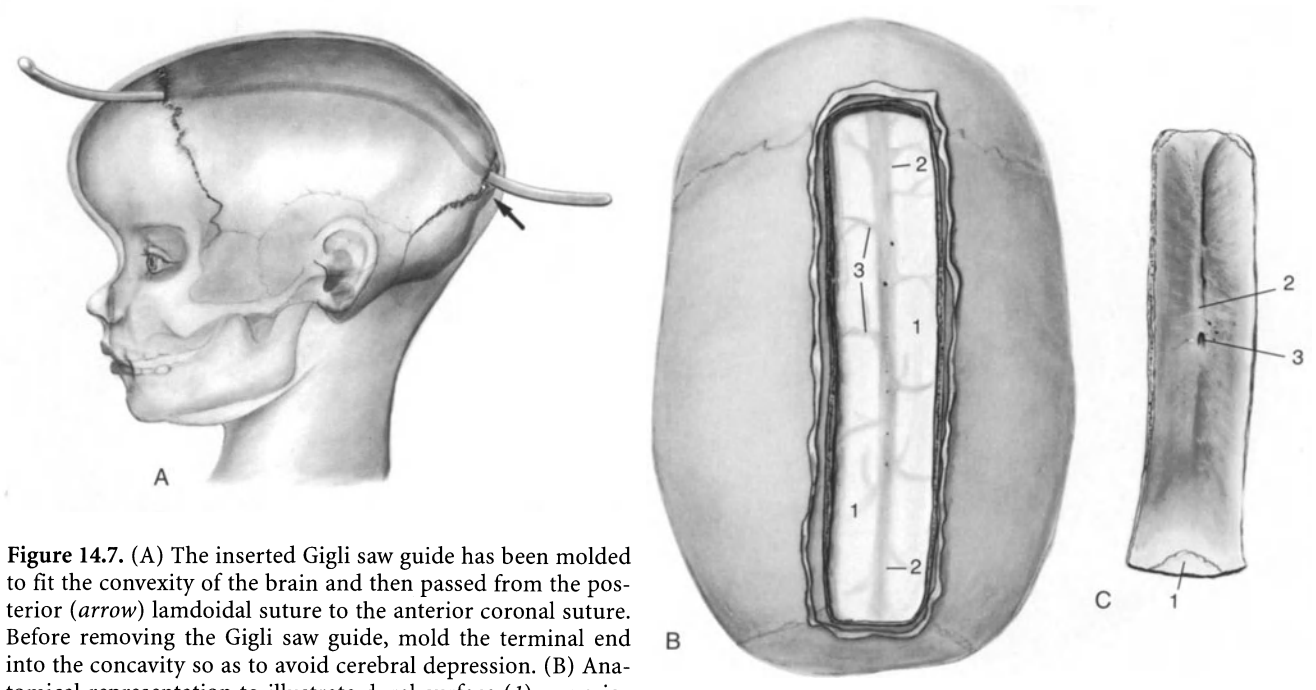


Figure 14.7. (A) The inserted Gigli saw guide has been molded to fit the convexity of the brain and then passed from the posterior (*arrow*) lamdoidal suture to the anterior coronal suture. Before removing the Gigli saw guide, mold the terminal end into the concavity so as to avoid cerebral depression. (B) Anatomical representation to illustrate dural surface (*1*), superior sagittal sinus (*2*), and intradural portions of the bridging cortical veins (*3*) after the sagittal strip of bone has been removed. (C) A wide strip of bone holding the hyperostosed superior sagittal suture has been removed from the underlying dura and the superior sagittal sinus (SSS). Note that the osteotomy has extended across the coronal suture anteriorly and the lambdoidal suture posteriorly (*1*). This strip of bone has an area of hyperostosis, immediately beneath the sagittal suture (*2*), which pushes into the SSS, often taking on the appearance of the keel of a boat. Small perforations in the dural surface of this flap (*3*) identify diploic draining veins passing from the diploë into the SSS.

◄ **Figure 14.6.** Stages in periosteal stripping. (A) The periosteum has been cut with the flat surface of the electrocautery knife, extending the incision from anterior to the coronal suture *(1)*, approximately 2 cm lateral to the sagittal plane, posteriorly across the lambdoidal suture *(2)* on either side. The periosteal incisions are connected to one another over the surface of the squamous occipital bone, posteriorly *(3)*, and to the edge of the anterior fontanelle *(4)*, anteriorly. When crossing the coronal and lambdoidal sutures, care must be taken to move rapidly, lest the coagulating current cut into the suture, causing bleeding.
The periosteal elevator is used to strip the periosteum from the skull *(5)*, with the elevator slightly angulated so that the stripping may be performed longitudinal to the cut edge of the periosteum, avoiding sawtoothed serrations. (B) This illustrates the incorrect technique for stripping the periosteum. (C) The periosteum has been stripped from the skull along the planned osteotomy lines, exposing only that surface area necessary to perform the osteotomy. The cuts across the frontal and squamous occipital bones are best made with a delicate rongeur. (D) The Gigli saw guide *(1)* has been passed from posterior to anterior, to separate the dura from the skull, before proceeding to use the craniotome to make the osteotomy. Note that the entrance into, and the exit from, the epidural space is at the lambdoidal *(2)* and coronal *(3)* sutures, respectively. No bur holes are made. Rather, the already described technique (Figs. 4.3, 4.4) is used to gain access to the epidural space, illustrated here by use of the curette *(4)* to open the suture.

osteum has been coagulated with the bipolar cautery forceps and incised. Proper use of the periosteal elevator is to angulate its cutting edge, so that a corner may be insinuated between periosteum and skull, and then to run the elevator in a longitudinal direction rather than making individual, sawtoothed strippings running perpendicular to the sagittal plane.

Bone wax, kept at body temperature, is applied to the bleeding surfaces of the exposed parietal, occipital, and frontal bones. Bipolar cautery is used to stop any oozing coming from the coronal or lambdoidal sutures. Take great care to stop all oozing from the bony surfaces, and to expose no more bone (strip no more periosteum) than needed to perform the osteotomy. This minimizes postoperative oozing and subgaleal hematoma.

The parietal bone is then separated from the lambdoidal and coronal sutures, using the suturectomy technique, immediately beneath the area of periosteal stripping on either side; the Gigli saw guide is passed from posterior to anterior, assuring protection of the underlying dura and brain during the osteotomy. Prior to inserting the guide for passage over the dura mater, it should be molded to conform with the curvature of the vertex of the skull. This avoids compressive damage of

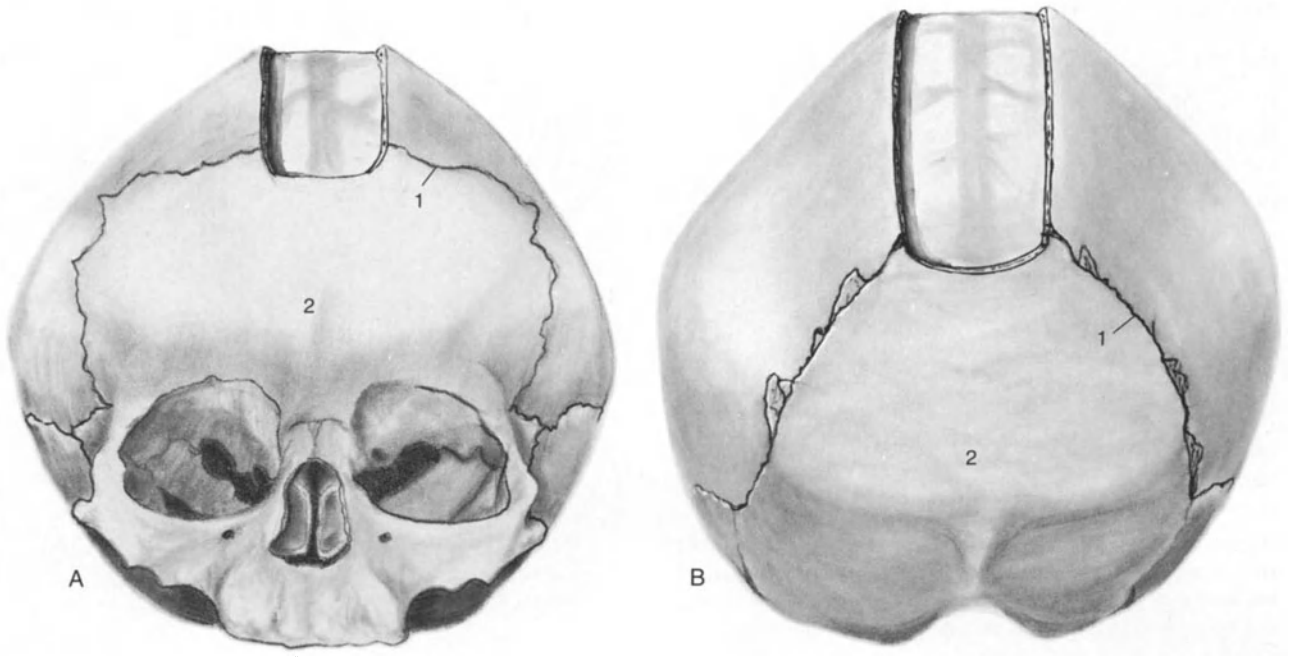


Figure 14.8. (A) Anterior extent of suturotomy across coronal suture (*1*) and into frontal bone (*2*). (B) posterior extent of suturotomy across lambdoidal suture (*1*) and into squamous occipital bone (*2*).

the underlying brain and, more importantly, stripping excessive amounts of dura from the overlying inner surface of the skull, which could result in severe blood loss secondary to oozing from the parietal bone. The molding should be such as to produce a concavity which complements the convexity of the cerebral hemisphere.

After osteotomies have been extended from lambdoidal to coronal sutures bilaterally, a rongeur is used to bite across the coronal and lambdoidal sutures into the frontal and occipital bones, freeing completely a single flap of bone. This is separated from the underlying dura and SSS with use of a Penfield #4 or #2 dissector. Carry the movements of the dissector from anterior to posterior. This exposes the underlying dura and the intact SSS. After the flap has been lifted away, tiny bleeding areas over the SSS may be identified and then covered immediately with soaked fluffy cottons. If one applies the sucker tip to the soaked fluffy cotton until it becomes glistening white, as it dries, and then leaves it in place for varying periods of time, most of this bleeding stops. If not, Avitene should be placed over the bleeding areas and a soaked fluffy cotton over it. One should try to avoid coagulating them.

The regrowth of bone takes place along the entire surface of the dura. It occurs first in small patches, then in larger islets. It *does not extend from the cut edges of the* parietal *bones medialward*! Consequently, no attempt should be made to apply caustic chemicals (such as Zenker's solution) to the bone edges, to sew the outer layer of the dura over the bone edge and onto the periosteum, or to place plastic (Silastic) cuffs over the exposed edges of the parietal bones. These procedures do not retard the regrowth of the skull, they certainly do not prevent it, and they put the child at a greater risk of complications. (In those cases where Silastic cuffing has been used, reoperation showed fully regrown bone completely surrounding the Silastic cuff.) The Zenker's solution is caustic; the Silastic cuffing is a foreign body. Reflection of the outer layer of the dura is a tedious procedure, one which increases risks of intraoperative bleeding.

A simple suturotomy, extending the full length of the sagittal suture from anterior to the coronal suture to posterior to the lambdoidal suture, extending approximately 2.5 cm to either side of the center of the SSS, is adequate to assure acceptable to excellent cosmetic results in these children. There is no indication for either extensive (radical) craniectomy or plastic reconstruction of the skull in newborn or infants.

The incidence, severity, and multiplicity of synostosis varies from one ethnic group to another so that one's experiences with Caucasian, black, and oriental children may be very different from what one would observe, for example, in Algeria.

Synostotic Craniofacial Anomalies

General

Gilles and Harrison, after treating a recessed compound malar fracture in an oxycephalic patient, reported in 1950 the first total facial osteotomy, which, unfortunately, resulted in only a partial correction [6]. They did, however, succeed in demonstrating that one must reposition and reconstruct the deformed and displaced bones of the facial framework in order to achieve acceptable cosmetic results. This experience, resulting only in correction of the exorbitism, led to Tessier's [7] studies of the facial skeleton, and his subsequent revolutionary concepts and techniques, which were predicated upon both LeFort's [8] experimental work and his own cadaver dissections [7]. His most fundamental concept was that one must move large, entire bones or segments of bones over extensive distances to reposition them around the globe, the nasooropharynx, and the frontal and temporal lobes, in order to achieve a functional structure and pleasing cosmetic result. He denuded these bony masses of their blood supply in relocating and reconstructing them, and molded them around the neural elements they protect.

In 1962, reporting on the management of congenital and traumatic hypertelorism, Converse and Smith [9] proposed reconstruction of the medial orbital wall. Again, developing upon this procedure, Tessier [10] made definitive contributions to the understanding and surgical correction of hypertelorism, proceeding to establish that corrective surgery should be a one-stage procedure. His one-stage facial advancements soon were expanded to include lateral orbital advancements as *en bloc* forward and/or medial repositioning of a single, or both, orbit(s) correcting completely the hypertelorism and the exorbitism. Tessier's work [11], as that of Converse and Smith [9], Munro [12], and others, illustrated that the preoperative evaluation of a congenital anomaly is complex and that it, as well as the surgery itself, must be undertaken by a *team of specialists* thoroughly versed in the diagnosis and management of craniofacial anomalies.

Surgical treatment of congenital anomalies of the craniocerebrum has classically been performed by specialists in pediatric neurosurgery for those malformations involving the calvarium and lesser wings of the sphenoid, and by plastic surgeons for malformations involving the bones of the face and base of the skull. Whereas all craniofacial work performed by pediatric neurosurgeons, with the sole exception of Jacques Rougerie and his colleagues [13], has been destructive in nature (craniectomies), the plastic surgeons addressed the problem from a reconstructive point of view. Their results have been remarkable. Therefore, operative procedures for the synostotic anomalies of the calvarium (scaphocephaly, trigonocephaly, plagiocephaly), all basically destructive procedures, are performed by neurosurgeons.

The procedures for the more complex craniofacial anomalies, basically reconstructive surgery, are outlined here in a purely descriptive sense. They are operations that require special training in reconstructive surgery, special knowledge of growth and development of the facial bones (including the dental arcade and the bony air sinuses), special instrumentation and techniques, and a well-coordinated craniofacial team. They are procedures for the craniofacial surgeon.

The following descriptions, with the exception of unilateral plagiocephaly, are meant only to be informative, so as to provide the student of pediatric neurosurgery with a clear and functional concept of what is entailed in the correction and reconstruction of the individual craniofacial anomalies. There is no intention to describe systematically the indications, technique, or complications of these procedures. The surgical treatment of unilateral plagiocephaly, however, will be described in detail, since it is within the scope of pediatric neurosurgery.

In 1974 Edgerton and coworkers [14] successfully operated on infants and toddlers with such complex craniofacial anomalies as Crouzon's disease and orbital hypertelorism, being able to perform cranioplasty, orbital repositioning, and midfacial advancements in a single procedure. It was their observation that significant advances in anesthesia and instrumentation, adaptation of osteotomies and knowledge of bone regenerative compacity to the younger child, and supportive postoperative care (available only in modern intensive care units) are the responsible factors for successful management of these cases in early childhood. The advantages of early surgery were considered to be:

1. A diminution in the stressful situation experienced by the parents
2. The ability to reconstruct soft tissue deformity once the bony abnormalities had been corrected
3. The attainment of normal milestones (eye movement, breathing, speech) after reconstructive craniofacial surgery

Neurosurgical aspects of craniofacial anomalies concern the presence and treatment of complicating hydrocephalus (as in the cloverleaf skull and some cases of Crouzon-Apert), excision of synostotic calvarial sutures, and the reflection of a bifrontal flap, preserving the orbital frontal band, measuring 2 cm in height as it extends from the supraorbital rims – Tessier's [15] linchpin for correcting hypertelorism and craniofacial dysostosis.

The skin incision invariably used is a bicoronal incision, which should be brought from antitragus to antitragus, running immediately behind the hairline.

Periosteal flaps, as already described, are reflected anteriorly (over the inferior frontal bone) but left adherent to the (frontoparietal) bones posteriorly; the anterior periosteal flap is a pedicle flap and is held in reserve to cover the frontal sinus if it is present and opened; the posterior flap will be used if it becomes necessary to reconstruct the dura along the olfactory rootlets.

Because of the severe anomalies of shape, thickness, and abnormal ridges of the inner table of the frontal bone in these children, one may not reflect the bifrontal bone flap described in Chap. 3. Instead, two parasagittal bur holes are placed, one to either side of the midline, just anterior to the coronal suture; two posterior frontal bur holes are placed on either side, 2 cm above the supraorbital rim and at the insertion of the temporalis muscle along the superior temporal line; and two anteromedial bur holes are placed, each 2 cm above the line of the supraorbital rim, and each in the same parasagittal plane as the posteromedial openings. It is extremely difficult to dissect the dura from the rootlets of the olfactory nerves, at the cribriform plate, with this orbital frontal band.

Separation of the dura mater from the inner table of the skull along the *orbital frontal band* is performed with fluffies and Telfa. At times, this separation is complicated by the presence of abnormal spurs of bone extending perpendicular to the orbital roof, ethmoid bone, and inner table of the frontal bone. There are generally two spurs, or keels, of bone extending into the dura, separating the underlying brain in two compartments from the inner table of the skull, located at the lateral borders of the frontal sinus, and sometimes a third at the midline in direct continuity with the crista galli. They indent the dura and underlying brain, rendering it difficult for the surgeon to remove them without nicking the dura mater. Dissection of the dura from the cribriform plate may be facilitated by resection of the crista galli. Its separation from the skull is best begun posterior to the lesser wing of the sphenoid, then extended lateral to the pterion and medially to the anterior clinoid: exposing completely the floor of the anterior fossa (orbital roofs, cribriform plate, planum sphenoidale). At times, these crests may be so large and so awkwardly placed as to oblige the surgeon to reflect the bifrontal flap in one, two, or even three segments. Individual openings in the dura should be repaired as they occur, using either simple closure, purse-string, or periosteal grafts.

Plagiocephaly (Improperly Called Coronal Synostosis)

(Figs. 14.9–14.14)

Plagiocephaly may be unilateral or bilateral, resulting either in the characteristic harlequin deformity of one orbit, or remarkable frontal bone changes with bilateral harlequin deformity and constriction, shortening, and verticalization of the anterior fossa. This latter causes compression of one or both frontal lobes, and displacement (proptosis or exorbitism) of the globe, so as to put stereopsis at risk. It is incorrect – incomplete – to consider this anomaly to be limited to coronal synostosis, a misnomer that resulted in incomplete surgical treatment (resection of only the closed coronal suture) between the time the first linear craniectomies were performed in 1890 by Lannelongue [16] and the time McLaurin and Matson [17] recommended, in addition to the linear coronal craniectomy, subtemporal decompression, extension of the craniectomy posteriorly along the fused squamosal suture line and, if fusion of the orbital bone had resulted in exophthalmus, resection of the roof and lateral orbital wall. Their work preceded the recommendation of Anderson and Johnson [18], who added resection of the sphenoid wing, an additional cut in the frontal bone extending from the lower end of the coronal craniectomy, immediately above the supraorbital ridge to the midline, and a vertical cut connecting this to the coronal channel. It was not until the 1970s that the purely destructive approach was abandoned and the entire superior and lateral aspects of the orbital rim were reconstructed [19]. When both coronal sutures are synostotic, the frontal lobes are compressed and dislocated posteriorly. Both globes are displaced from the normal line of vision. In either event, the exorbitism may be so severe as to impede lid closure, and thereby subject the cornea to ulceration and/or opacification.

This synostosis involves invariably the coronal suture, but may also involve the frontonasal and frontoethmoidal sutures as well as the zygomaticofrontal suture. Some authors have suggested that even the sphenopterygoid and pterygomaxillary sutures are involved. Consequently, the operative procedure must be designed to provide the surgeon access to the coronal suture from the anterior fontanelle to the anteroinferiorly displaced pterional region, the frontonasal and frontoethmoidal sutures, and the intracranial and intraorbital surfaces of the orbital roof.

The purpose of the operation to correct plagiocephaly is twofold: (1) to resect the synostosed coronal and temporosphenoidal sutures; and (2) to reconstruct the superior and lateral aspects of the orbital rim! If the frontonasal and frontoethmoidal sutures are synostosed, the procedure *must* be done in conjunction with a plastic surgeon experienced in craniofacial surgery.

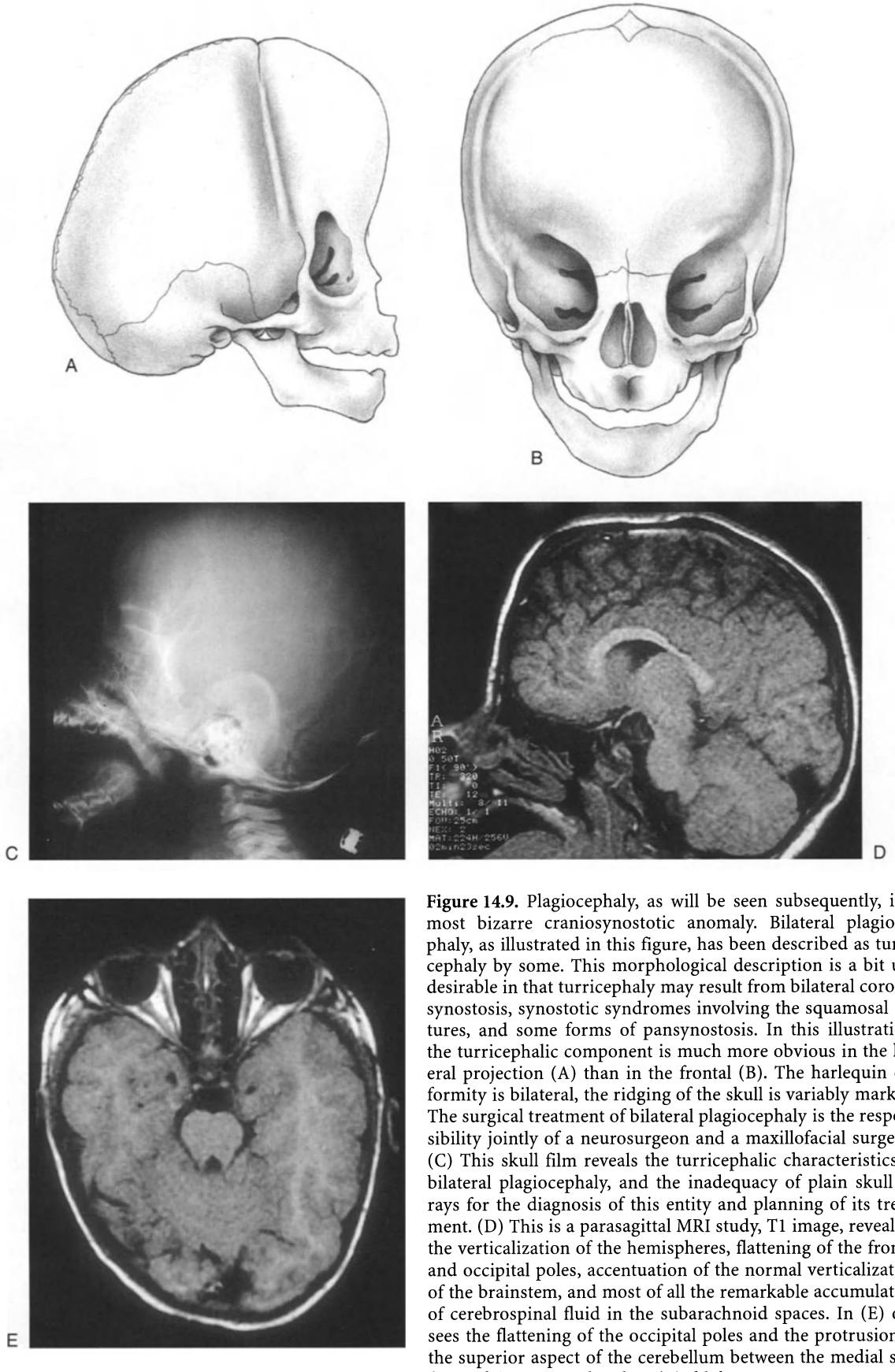


Figure 14.9. Plagiocephaly, as will be seen subsequently, is a most bizarre craniosynostotic anomaly. Bilateral plagiocephaly, as illustrated in this figure, has been described as turricephaly by some. This morphological description is a bit undesirable in that turricephaly may result from bilateral coronal synostosis, synostotic syndromes involving the squamosal sutures, and some forms of pansynostosis. In this illustration, the turricephalic component is much more obvious in the lateral projection (A) than in the frontal (B). The harlequin deformity is bilateral, the ridging of the skull is variably marked. The surgical treatment of bilateral plagiocephaly is the responsibility jointly of a neurosurgeon and a maxillofacial surgeon. (C) This skull film reveals the turricephalic characteristics of bilateral plagiocephaly, and the inadequacy of plain skull X-rays for the diagnosis of this entity and planning of its treatment. (D) This is a parasagittal MRI study, T1 image, revealing the verticalization of the hemispheres, flattening of the frontal and occipital poles, accentuation of the normal verticalization of the brainstem, and most of all the remarkable accumulation of cerebrospinal fluid in the subarachnoid spaces. In (E) one sees the flattening of the occipital poles and the protrusion of the superior aspect of the cerebellum between the medial surfaces of the temporal and occipital lobes.

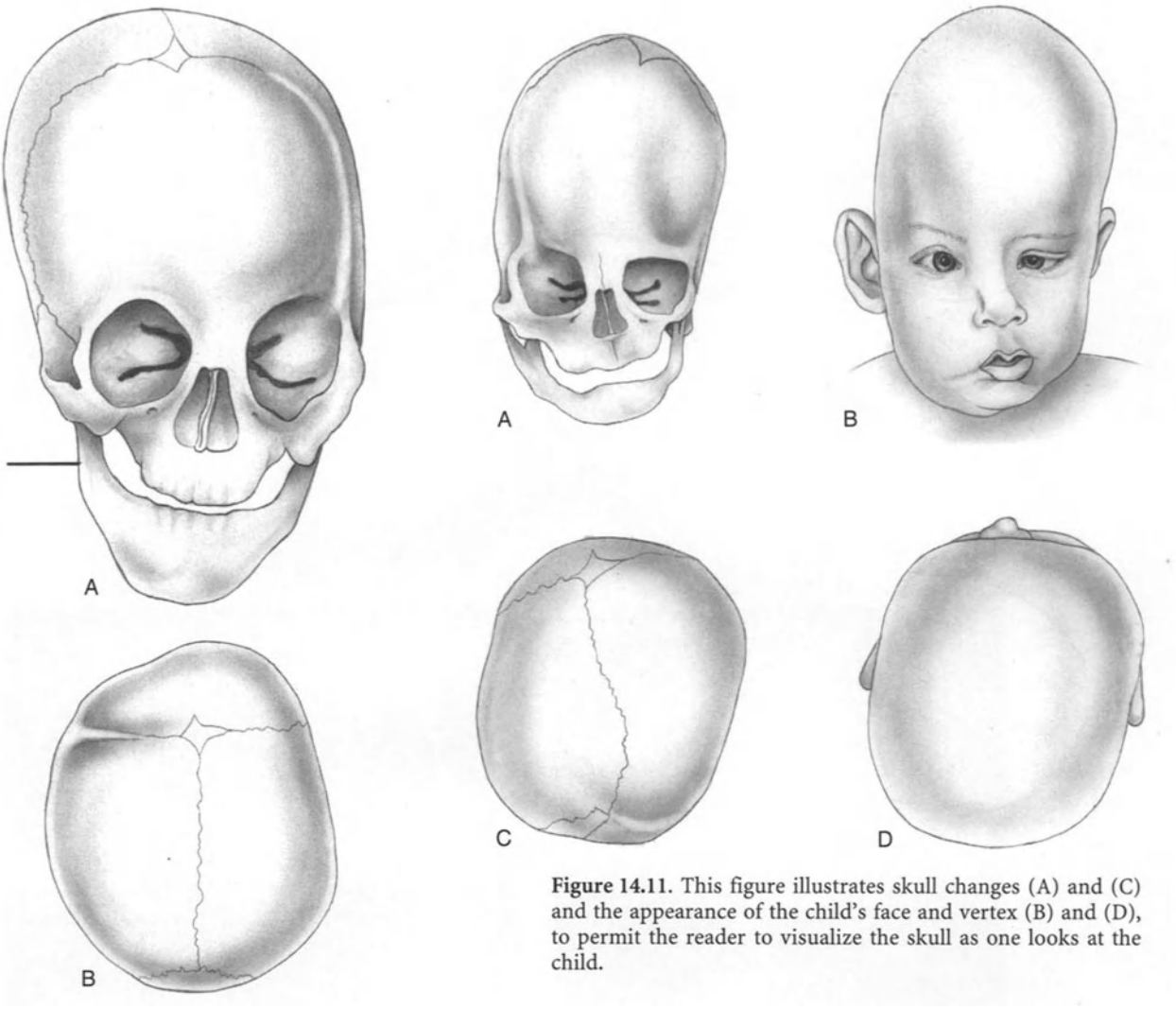


Figure 14.11. This figure illustrates skull changes (A) and (C) and the appearance of the child's face and vertex (B) and (D), to permit the reader to visualize the skull as one looks at the child.

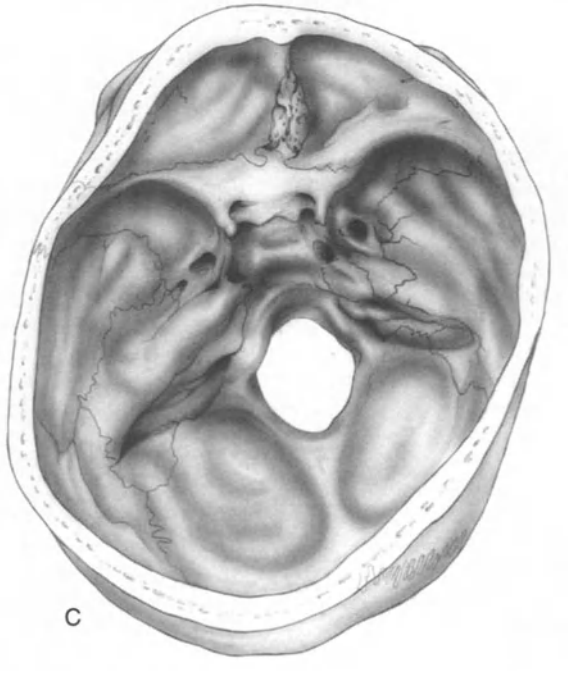


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Figure 14.10. Plagiocephaly. (A) In these drawings one sees the frontal deformity of plagiocephaly, with synostosis of the left coronal suture and the harlequin deformity on this side pulling the orbit superiorly, laterally, and posteriorly. There is an apparent compensatory deformity of the splanchnocranium: the nasal septum, line of the median incisiors, and mandible are angled to the right. In (B), a view of the vertex with the vault intact, one sees the flattening on the left which results from synostosis of the coronal suture extending inferiorly into the base of the skull. (C) A drawing of a three-dimensional MRI scan performed on a patient of ours illustrating flattening of the left frontal boss, compensatory increase in volume of the middle and posterior fossae on the left, anterior displacement of the right petrous pyramid and right lesser wing of the sphenoid, and angulation toward the left of the sagittal line of the foramen magnum.

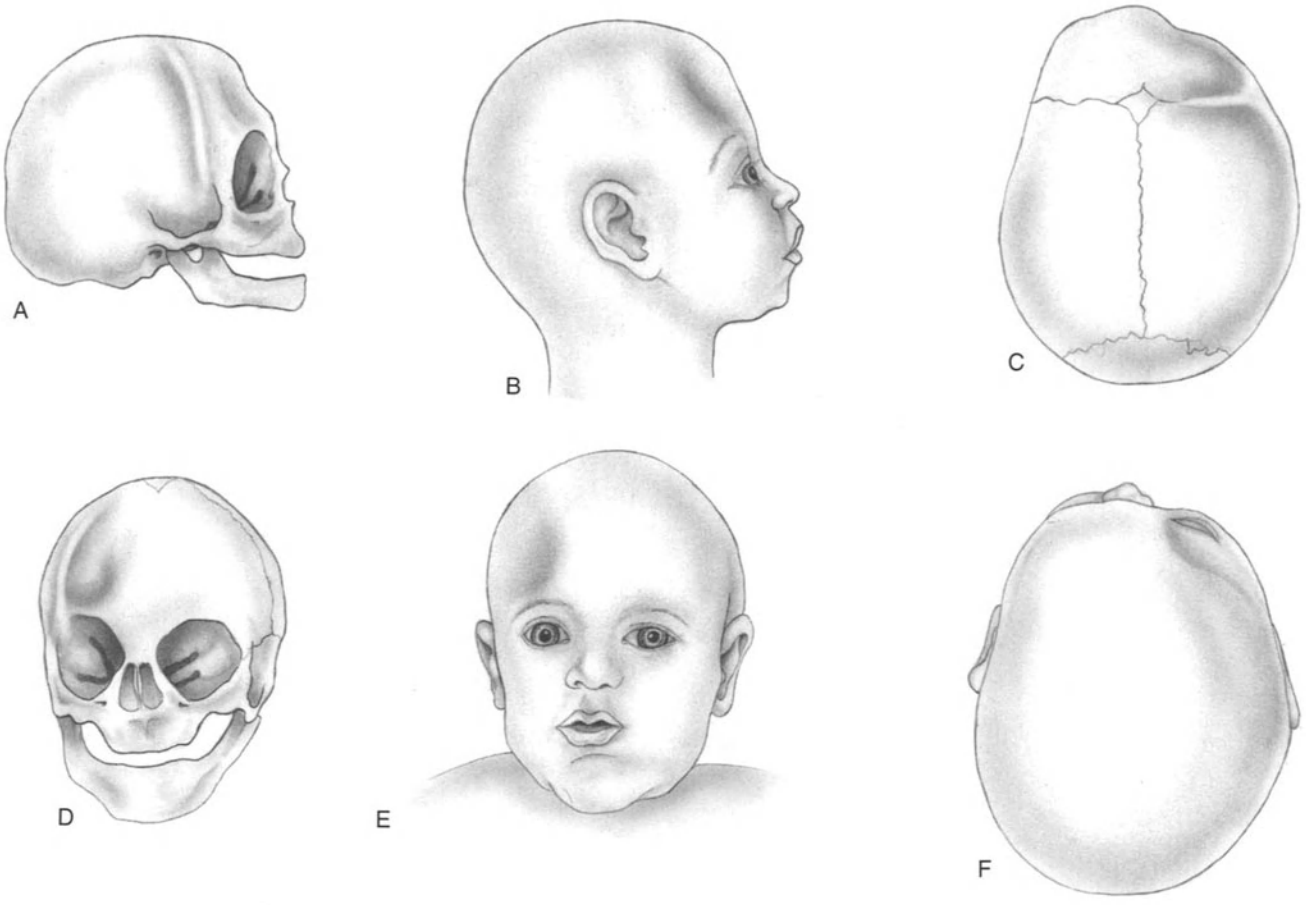
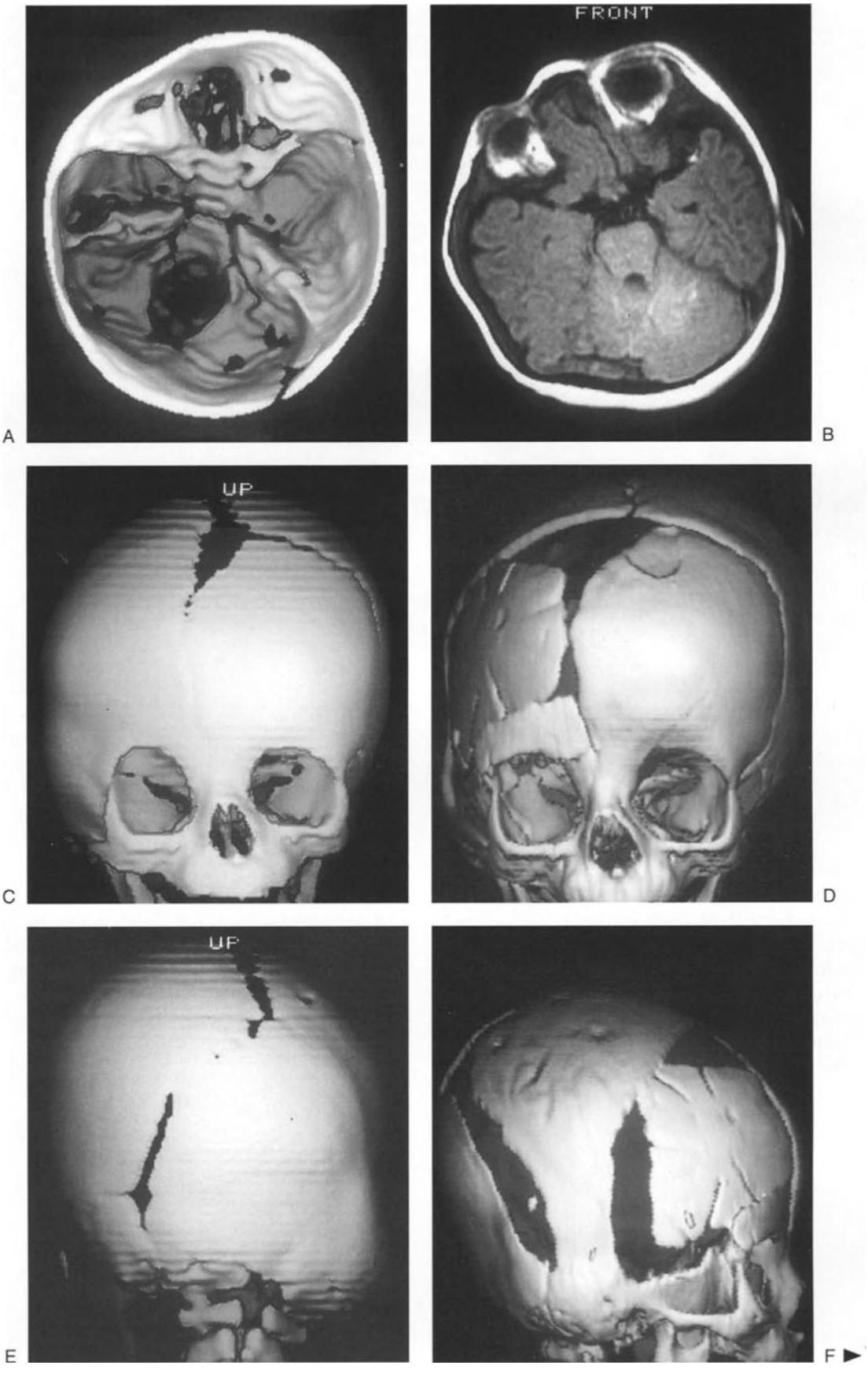


Figure 14.12. This figure compares relatively (A) the lateral projection of the skull, (B) and the child's face with the vertex view of the skull (C)... and the frontal view of the skull (D) with the frontal view of the face (E) to the vertex view (F) of the child's head. This permits the reader to compare horizontally and vertically skull with face and vertex.

For unilateral plagiocephaly, one should, therefore, reconstruct the superior and lateral aspects of the orbit, and on occasion the medial aspect. For bilateral plagiocephaly, the corrective procedure involves both orbits, the ethmoid and nasal bones.

The various involvements of individual sutures, and the degree of deformity resulting from their synostosis, are not to be conceptualized as isolated entites. Rather, they coexist to differing degrees and in different combinations. Clinical examination of the orbital and cranial deformities, as well as studying the plain X-ray films and computed tomography (CT) scans, assist in determining which sutures need resection and what procedure should be performed. Consequently, one is advised to read this section as a continuum. *It is not our intention to present these anomalies as independent and distinct anatomicopathogenic entities.*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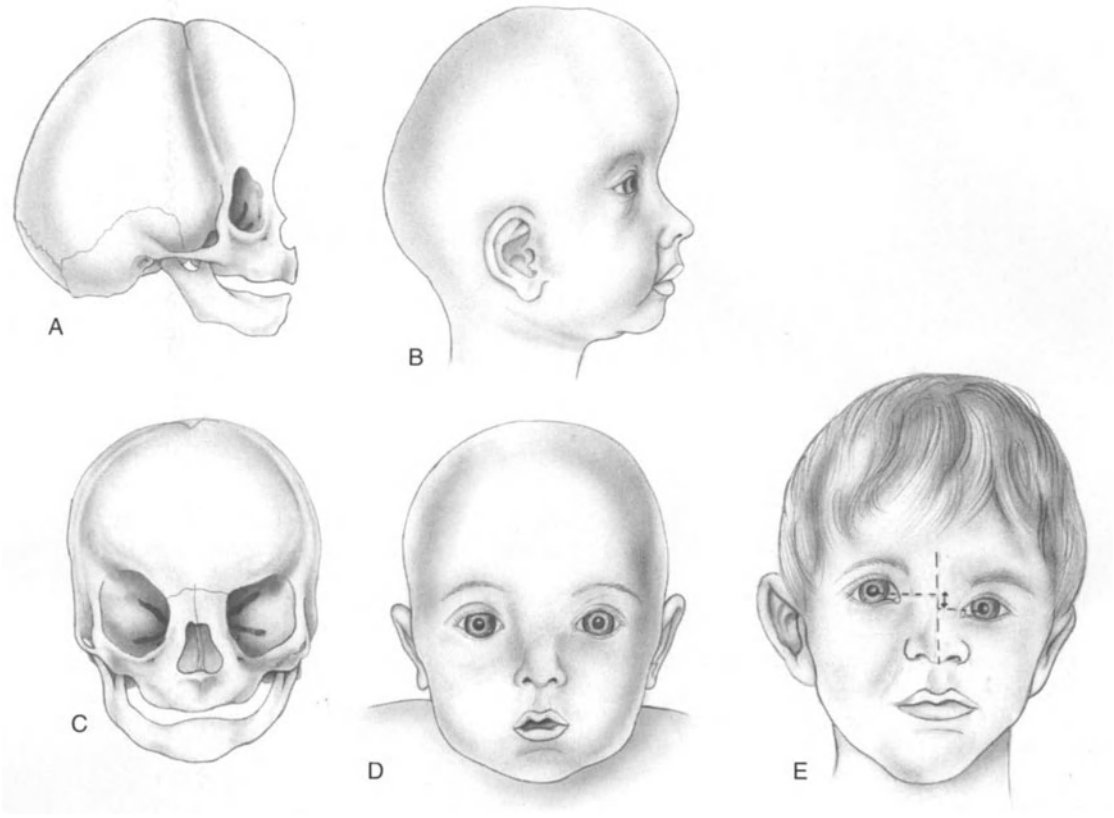

Figure 14.14. Plagiocephaly (bilateral). This is a series of schematic drawings used to illustrate the asymmetry so common in bilateral plagiocephaly (turricephaly) and the displacement of one orbit (globe) vis-à-vis the other, a truly disabling condition. (A) and (B) are lateral drawings illustrating the anterior tilt of the turricephalic deformity in the skull and the head, (C) and (D) illustrate the more remarkable obvious orbital deformity in the skull that one sees when looking at the child; and (E) is an exaggerated, but not very much, representation of the uneveness of the globes resulting from asymmetrical deformities such as these.

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Figure 14.13. Scaphocephaly. This is a series of imaging studies putting into relief the characteristics of skull base changes (A), secondary molding and deformities of the cerebral hemispheres and brain stem (B); the preoperative (C) and postoperative (D) CT reconstruction studies from the frontal projection; and the preoperative (E) and postoperative (F) CT reconstruction studies of the lambdoidal component of the plagiocephaly. One sees that the left coronal suture is closed and that the orbits are set at almost 45° to the coronal plane of the skull, with compensatory widening of the contralateral middle fossa and the homolateral posterior fossa. This study should be compared to the drawing illustrated in Fig. 14.14C to appreciate the variability of observations and pathological defomities one observes in the plagiocephalic skull, facts which strongly discourage one from drawing hard and fast conclusions concerning the dynamics of skull deformities resulting from such complex pathological entities as single or multiple suture synostosis. The brain displacements are equally dramatic with the only applicable adverb to these deformities being... "squash." The pre- and postoperative reconstruct studies provide the reader the opportunity to appreciate how very much may be realized and accomplished for the correction of these unsightly and at times disabling anomalies.

Unilateral Plagiocephaly

Frontosphenoidal Synostosis (Figs. 14.15–14.18)

Unilateral plagiocephaly, extending into the pterional sutures (parietosquamosal, sphenosquamosal, frontosphenoidal), is referred to as "frontosphenoidal." One need only resect the synostosed suture and reconstruct the superior and lateral surfaces of the orbit.

A unilateral frontal skin incision is used for this exposure. Once the flap is reflected, the anterior portion should be laid over a roll of gauze sponges and then retention sutures run along the line of junction of galea and periosteum, over the supraorbital ridge, as far laterally as the zygomatic process of the frontal bone. At this point sutures are placed along the anterior surface of the zygomatic process of the frontal bone as far inferiorly as the zygomatic arch. When these retention sutures are brought over the rim of the orbit and zygomatic process of the frontal bone and put on the stretch, they expose completely the glabella, supraorbital ridge

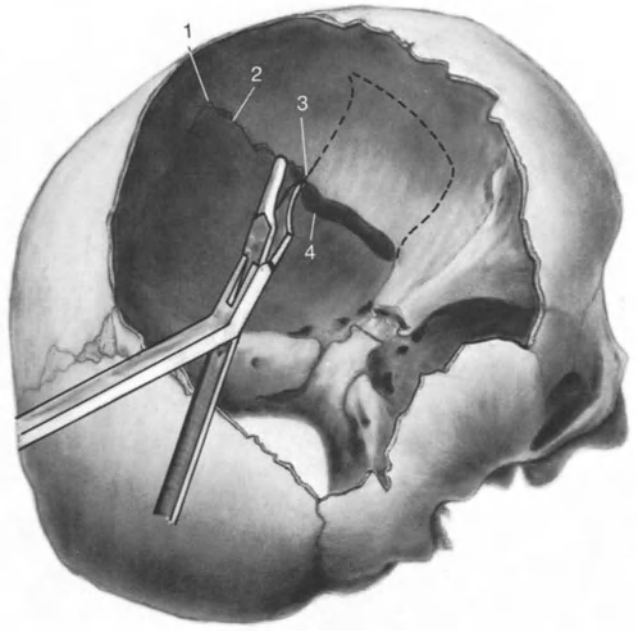


Figure 14.15. A three-dimensional artistic representation of the inner surfaces of the neurocranium, illustrating the extension of the craniectomy, along the synostosed coronal suture (*1*) from the pterional region (*2*), superomedially, to the lesser wing of the sphenoid. At this point it is extended into the unroofed orbit (*3*), and slightly inferomedially into the superior orbital fissure (*4*). The osteotomy for unroofing the orbit is *represented by the broken line.*

and sulcus, zygomaticofrontal suture, pterional region, and zygomatic arch. Posterior retraction of the scalp flap is brought far enough to expose the entirety of the coronal suture and the temporalis muscle. This latter will already have been cut parallel to and 0.5 cm beneath the superior temporal line, stripped from the underlying bones of the pterional region, to expose the squamous portion of the temporal bone and the lateral aspect of the greater wing of the sphenoid – as one would for reflecting a lateral frontal flap. Stripping of the temporalis muscle from these bones is followed by insertion of retention sutures into the temporalis muscle, sewing it out of the way.

At this point the frontal bone, greater wing of the sphenoid, and squamous temporal bones are exposed, as is the hyperostosed coronal suture from the region of the anterior fontanelle to the pterion. The periosteum is incised by using the flat edge of the electrocautery

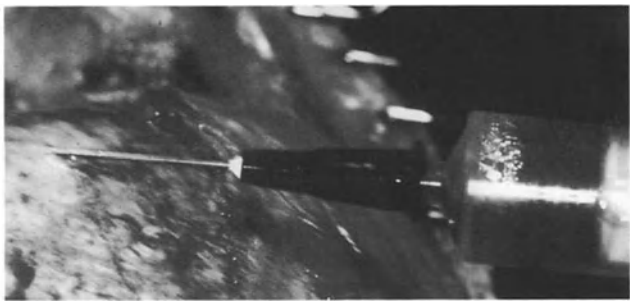
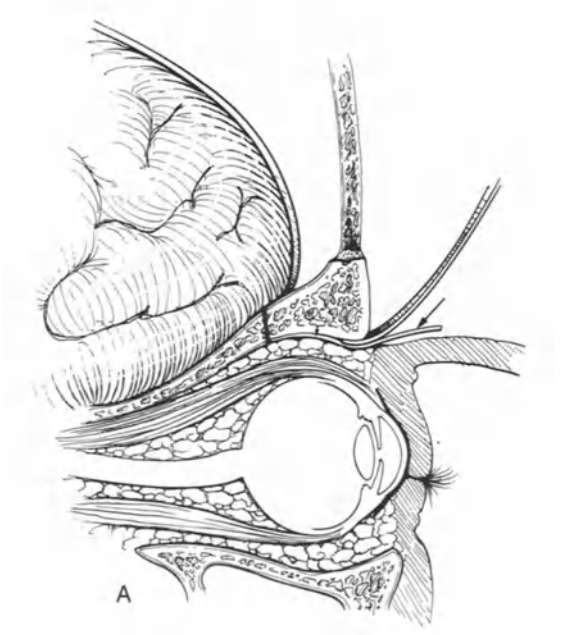


Figure 14.17. Fluid that accumulated in the subdural space, *after* the synostosed coronal suture and frontal bone were removed, is being aspirated.



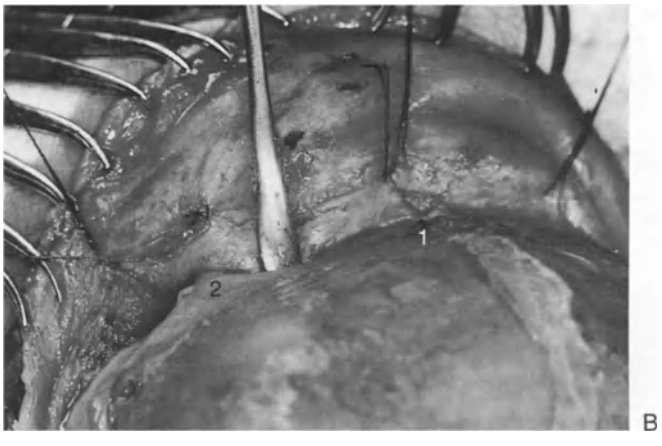


Figure 14.16. (A) The floor of the anterior fossa is exposed by retracting the frontal lobes. The periosteum (*arrow*) is dissected from the roof of the orbit with a Penfield #3 dissector, separating the periosteum and periorbita from the orbital rim and roof. (B) A Penfield #4 dissector is being used to separate the periorbita from the orbital surface of the roof of the left orbit. For orientation purposes, the glabella (*1*) and zygomatic process of the frontal bone (*2*) are indicated.

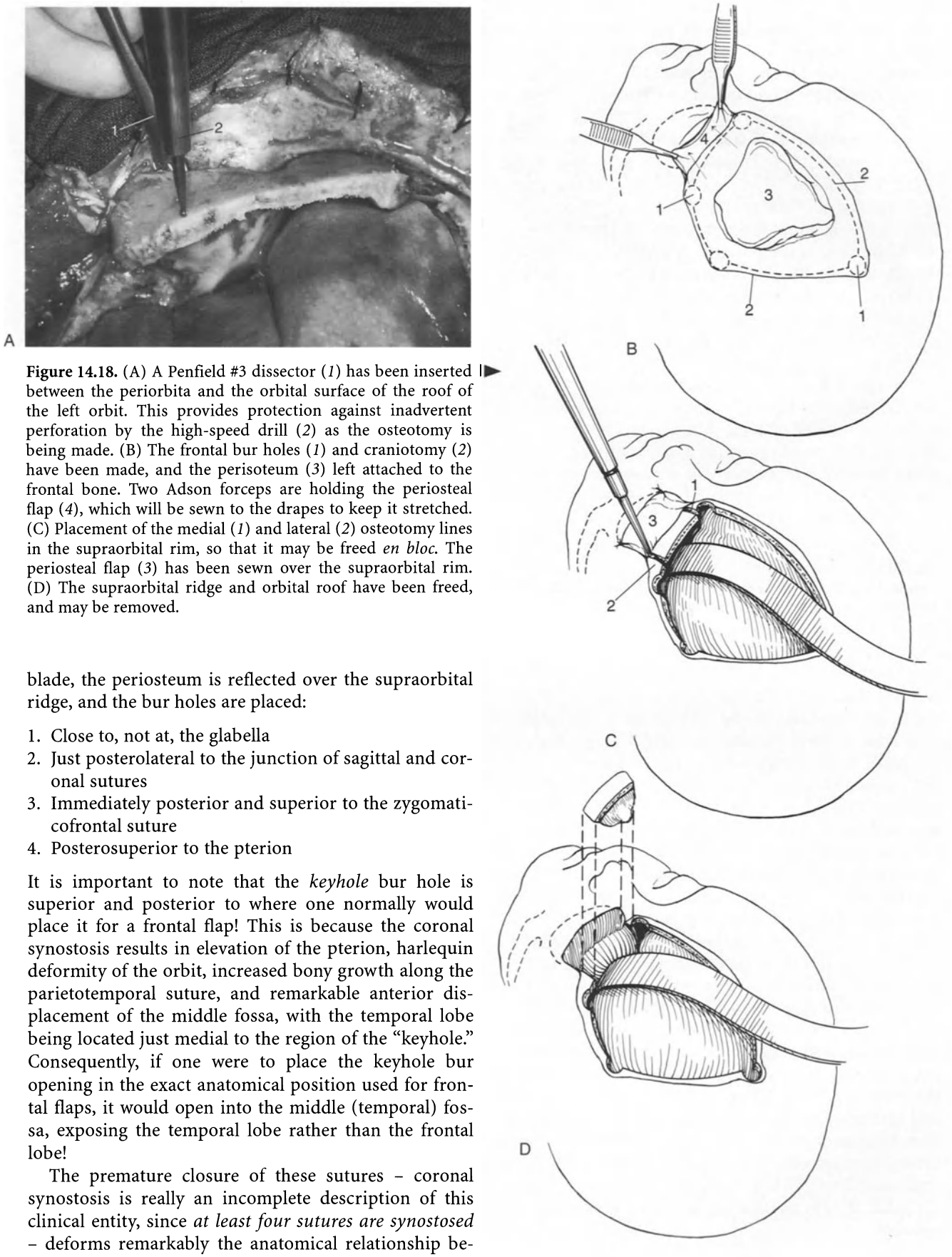


Figure 14.18. (A) A Penfield #3 dissector (*1*) has been inserted between the periorbita and the orbital surface of the roof of the left orbit. This provides protection against inadvertent perforation by the high-speed drill (*2*) as the osteotomy is being made. (B) The frontal bur holes (*1*) and craniotomy (*2*) have been made, and the perisoteum (*3*) left attached to the frontal bone. Two Adson forceps are holding the periosteal flap (*4*), which will be sewn to the drapes to keep it stretched. (C) Placement of the medial (*1*) and lateral (*2*) osteotomy lines in the supraorbital rim, so that it may be freed *en bloc*. The periosteal flap (*3*) has been sewn over the supraorbital rim. (D) The supraorbital ridge and orbital roof have been freed, and may be removed.

blade, the periosteum is reflected over the supraorbital ridge, and the bur holes are placed:

1. Close to, not at, the glabella
2. Just posterolateral to the junction of sagittal and coronal sutures
3. Immediately posterior and superior to the zygomaticofrontal suture
4. Posterosuperior to the pterion

It is important to note that the *keyhole* bur hole is superior and posterior to where one normally would place it for a frontal flap! This is because the coronal synostosis results in elevation of the pterion, harlequin deformity of the orbit, increased bony growth along the parietotemporal suture, and remarkable anterior displacement of the middle fossa, with the temporal lobe being located just medial to the region of the "keyhole." Consequently, if one were to place the keyhole bur opening in the exact anatomical position used for frontal flaps, it would open into the middle (temporal) fossa, exposing the temporal lobe rather than the frontal lobe!

The premature closure of these sutures – coronal synostosis is really an incomplete description of this clinical entity, since *at least four sutures are synostosed* – deforms remarkably the anatomical relationship be-

tween orbit, anterior fossa, middle fossa, and malar bone. Instead of being located entirely posterior to the orbit, the temporal lobe is stretched over and draped around its lateral surface. The pterion is located immediately behind the zygomaticofrontal suture, the greater wing of the sphenoid is converted into a vertical ribbon of bone, and the synostosed coronal suture extends anteroinferiorly to the lateral surface of the orbital rim.

On the intracranial surface the frontoparietal suture is hyperostosed, taking the form of a keel, deeply wedged into the dura. It exerts a constrictive force upon the underlying brain, one which is so compressive that there is a spontaneous accumulation of fluid in the subdural space within about 3–5 min of the time the frontal flap and hyperostotic suture are removed. This is remarkable. The fluid ranges in color from brownish red to yellow.

The lesser wing of the sphenoid runs in an almost vertical direction from the innominate line at the pterion to the anterior clinoid, compressing the frontal, parietal, and temporal operculae. The underlying dura is a dense band of tissue which also exerts a constrictive force, but does not need to be opened: it relaxes, becoming unfolded by the brain, after the synostosed sutures have been removed from the anterior fontanelle to the superior orbital fissure.

Knowledge of the location of internal hyperostosis along the line of synostosed sutures permits one to cut across the area of keel-like deformity, without damaging the underlying dura or brain. Connect the bur holes with a craniotome, not the Gigli saw, though it is safest to pass the Gigli saw guide from the posterosuperior bur opening to the one lateral to the glabella first, then to the posteroinferior hole. Lastly, passage of the guide from the glabella area to the *keyhole* allows the surgeon to separate dura from bone along three of the four osteotomy lines. Do not attempt to pass the saw guide along the line extending from the *keyhole* to the posteroinferior bur hole.This would oblige one to cross the keel-like hyperostosis. Rather, once the three osteotomies have been cut with the craniotome, to perform a suturectomy, rongeur away the synostosed sutures from the anterior fontanelle down to the pterion, and from here to the superior orbital fissure. The coronal suture is either incised or excised, depending upon the width of the hyperostosis. The bordering surfaces of the bones at the pterion are all removed with either a fine-jawed rongeur or a diamond bur, preferably the latter, extending the osteotomy along the lesser wing of the sphenoid from the pterion to the anterior clinoid and into the superior orbital fissure. This latter extension, because of the continuity of superior and inferior orbital fissures at the posteroinferior aspect of the orbit, automatically frees bony adherences from the floor of the orbit. It does not free synostosis involving the pterygoid plates.

This completes the first part of the procedure: removal of the prematurely closed coronal and temporosphenoidal sutures. It is accomplished after the frontal flap has been freed, and allows the surgeon to proceed to the second part of the procedure, reconstruction of the superior and lateral aspects of the orbital rim.

The reflected frontal flap is stored in sterile saline, and the dura is then separated from the roof of the orbit, exposing the intracranial surface of the orbital roof, from the cribriform plate medially to the zygomatic process of the frontal bone laterally, and from the superior orbital rim anteriorly to the lesser wing of the sphenoid posteriorly. At this time, one is advised to puncture the dura with a #21 needle and to aspirate the subdural accumulation of fluid. A simple nick in the dura may also suffice. The fluid which accumulates in the subdural space ranges from red to yellowish, may contain hematogenous elements, and expands rapidly in volume. After it has been removed, it does not reaccumulate.

The separation of the dura from the medial surface of the anterior fossa is accomplished with care, to avoid damaging the olfactory nerve and, more importantly, tearing dural sleeves from around the olfactory bulb outlets, risking creation of a cerebrospinal fluid leak. One need not proceed completely to the midline unless the frontoethmoidal and frontonasal sutures are synostosed, in which case the appropriate technique (described in the section on frontonasal, nasal, and frontoethmoidal stenosis in this chapter) is used. Once the dura has been freed from the bones bordering the anterior fossa, the next step, freeing of periosteum and periorbita, is taken, but retractors are not left over the frontal lobe.

Periosteum is now dissected from the supraorbital ridge, from medial to lateral, taking care not to damage the lacrimal gland, since this occupies the most superolateral portion of the orbit. A Penfield #4 dissector, or an Oldberg periosteal elevator, is desirable for completing the separation of periosteum from the rim of the supraorbital ridge. A Penfield #3 dissector, because of its curved blade, is helpful in separating periorbita from the orbital roof. Once the Penfield #3 dissector is inserted into the orbit, between periorbita and orbital roof, one may either see through the almost transparent orbital roof, or at least identify the location of the dissector tip by the indentation it causes when pressed against the orbital roof.

The Penfield #3 dissector is used to protect the intraorbital contents during osteotomy, medial and lateral, of the supraorbital rim and orbital roof. One now has fully exposed the orbital rim from just lateral to the glabella to the zygomatic process of the frontal bone, the periorbita from the region of junction of the periosteum to the superior orbital fissure.

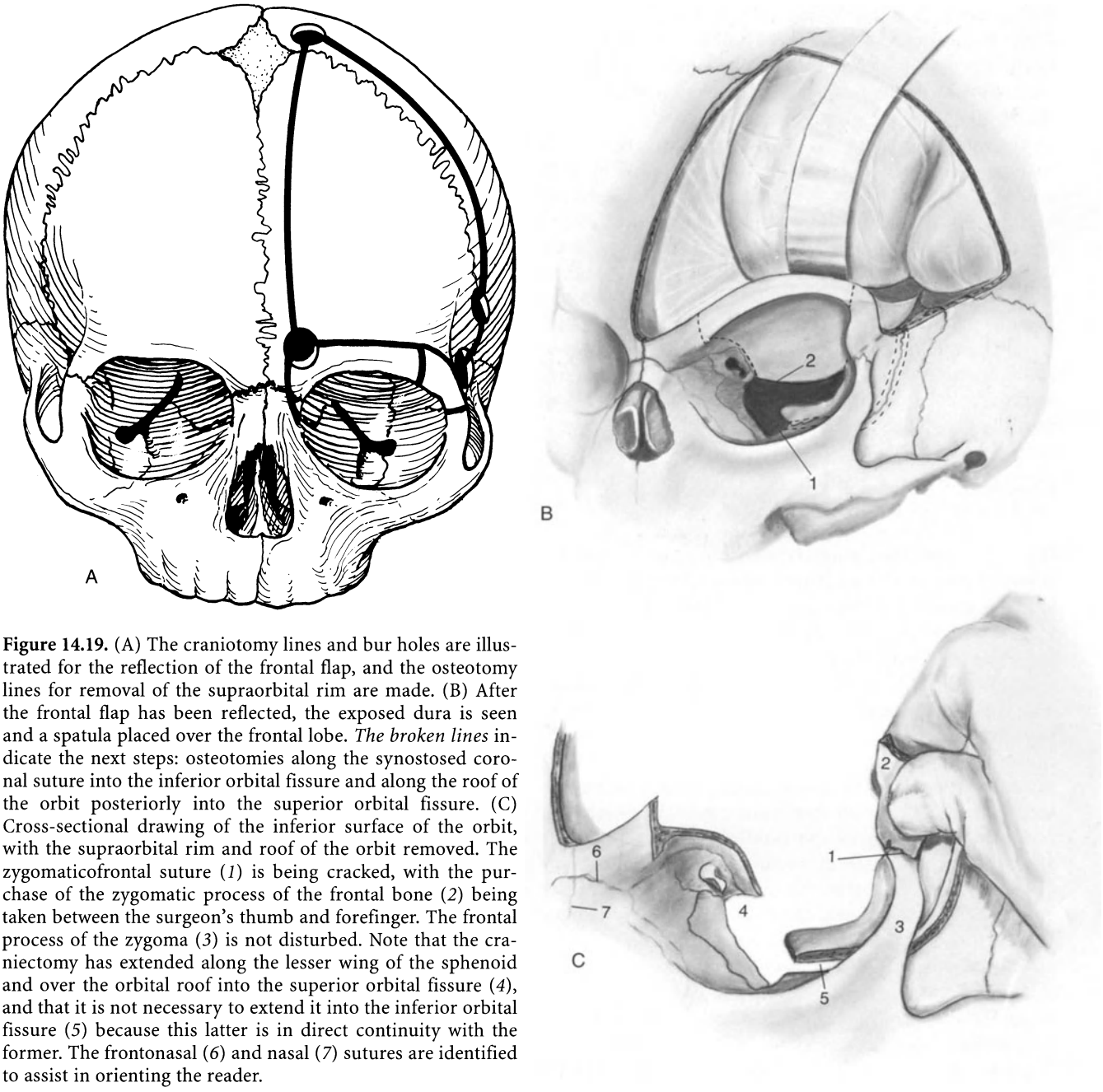


Figure 14.19. (A) The craniotomy lines and bur holes are illustrated for the reflection of the frontal flap, and the osteotomy lines for removal of the supraorbital rim are made. (B) After the frontal flap has been reflected, the exposed dura is seen and a spatula placed over the frontal lobe. *The broken lines* indicate the next steps: osteotomies along the synostosed coronal suture into the inferior orbital fissure and along the roof of the orbit posteriorly into the superior orbital fissure. (C) Cross-sectional drawing of the inferior surface of the orbit, with the supraorbital rim and roof of the orbit removed. The zygomaticofrontal suture (*1*) is being cracked, with the purchase of the zygomatic process of the frontal bone (*2*) being taken between the surgeon's thumb and forefinger. The frontal process of the zygoma (*3*) is not disturbed. Note that the craniectomy has extended along the lesser wing of the sphenoid and over the orbital roof into the superior orbital fissure (*4*), and that it is not necessary to extend it into the inferior orbital fissure (*5*) because this latter is in direct continuity with the former. The frontonasal (*6*) and nasal (*7*) sutures are identified to assist in orienting the reader.

Zygomaticofrontal Synostosis (Fig. 14.19)

Synostosis of this suture may occur as an isolated (extremely rare) anomaly or, most often, in conjunction with frontoethmoidal or frontonasal/nasal/frontoethmoidal synostoses. If it exists as an isolated entity a simple suturotomy suffices as treatment. If it exists in conjunction with frontosphenoidal synostosis, its treatment is incorporated into the overall freeing of the superior and lateral surfaces of the orbital rim.

The lateral rim of the orbit is freed by fracturing the zygomaticofrontal suture either with one's thumb and forefinger, or with the use of a rongeur. There is no need to discard the bone fragments; they are the lateral rim of the orbit. These steps free the superior and lateral aspects of the orbit, allowing the globe ample accommodation to assume a normal position vis-à-vis the optic nerve, freed optic canal, and optic chiasm. The freeing of the supraorbital ridge from its medial to lateral borders permits both the frontal lobe and orbital

contents to remold the bone into a circular, ample orbit. After the orbital roof is removed and its lateral surfaces freed, one may complete the removal of the middle and medial thirds of the lesser wing of the sphenoid, assuring opening into the superior orbital fissure.

The supraorbital rim is now taken from the saline soaking solution and reapproximated, loosely, into its normal position, either simply laying it in place, or using 4–0 sutures to moor it into place. The frontal bone is then brought back into position, laying it over the frontal lobe or, as for the supraorbital rim, mooring it into place. If it is severely deformed, as often occurs in older children, it is fractured, molded, and rotated 180° before being placed over the frontal lobe. This generally provides an agreeable contour and a pleasant cosmetic result. The scalp is closed and a firm, not tight, dressing is applied.

Frontonasal, Nasal, and Frontoethmoidal Stenosis
(Figs. 14.20, 14.21)

When the unilateral plagiocephaly involves the medial (frontonasal, nasal, and frontoethmoidal) *as well as the lateral sutures,* one frees completely the nasal and ethmoidal bones from the frontal bone. This is done along the line of the orbital roof *and* at the supraorbital rim. Consequently, the placement of the inferomedial bur hole is particular, as is the location and extent of the osteotomy. In sum, one reflects a frontal flap which includes the supraorbital rim and orbital roof, and which extends along the frontoethmoidal suture medially, across the zygomaticofrontal suture laterally, along the pterional/lesser wing of sphenoid plane to the superior orbital fissure and anterior clinoid process posteriorly.

The unilateral, frontal skin incision is used, and the periosteum is incised and elevated in the usual manner. However, the periosteal flap freed from over the supraorbital rim cannot be reflected in one piece because of its length and the need to extend it over the frontonasal and glabellar areas. It is necessary to incise it perpendicular to the medial rim of the orbit, so that it may then be sewn back over the reconstructed supraorbital rim at the end of the procedure.

The posteromedial (coronal) and posterolateral (squamosal) bur holes are placed as for reconstruction of the superior and lateral orbital rims, but the anteromedial (glabellar) and anterolateral (keyhole) openings are not. In fact, they are placed so as to permit access to the ethmoid bone medially and the inferior surface of the sphenoid wing, through the middle fossa, laterally. This placement permits *en bloc* removal of the frontal bone with the entirety of the superior half of the orbital rim if the surgeon so chooses, or a two-stage removal if this seems undesirable or inopportune at the time of surgery. One-stage reflection entails:

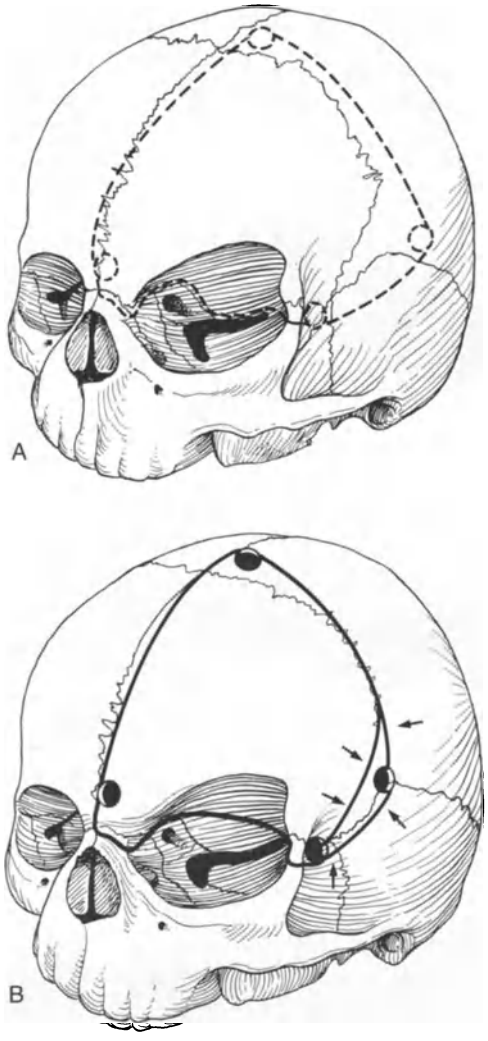


Figure 14.20. (A) Bur hole placement if one chooses to reflect the supraorbital rim, orbital roof, medial and lateral walls of the orbit in one piece (stage), after osteotomizing the frontoethmoidal suture and synostosed pterional sutures into the superior orbital fissure. *The broken line* indicates the osteotomy along the calvarium, into the orbital roof, and over the nasal bone. (B) Resection of the synostosed coronal/frontosphenosquamosal suture (*arrows*).

1. Freeing the dura from the orbital roof and frontoethmoidal suture
2. Osteotomizing the pterional synostoses and lesser wing of the sphenoid into the superior orbital fissure
3. Freeing the periorbita from medial, superior, and lateral orbital surfaces

In either event, performing a one- or two-stage reflection of the frontal bone and superior half of the orbital rim, one osteotomizes the frontonasal, nasal, and frontoethmoidal sutures. Synostosis along these suture lines is responsible for deformity of the medial orbital rim and the hypotelorism.

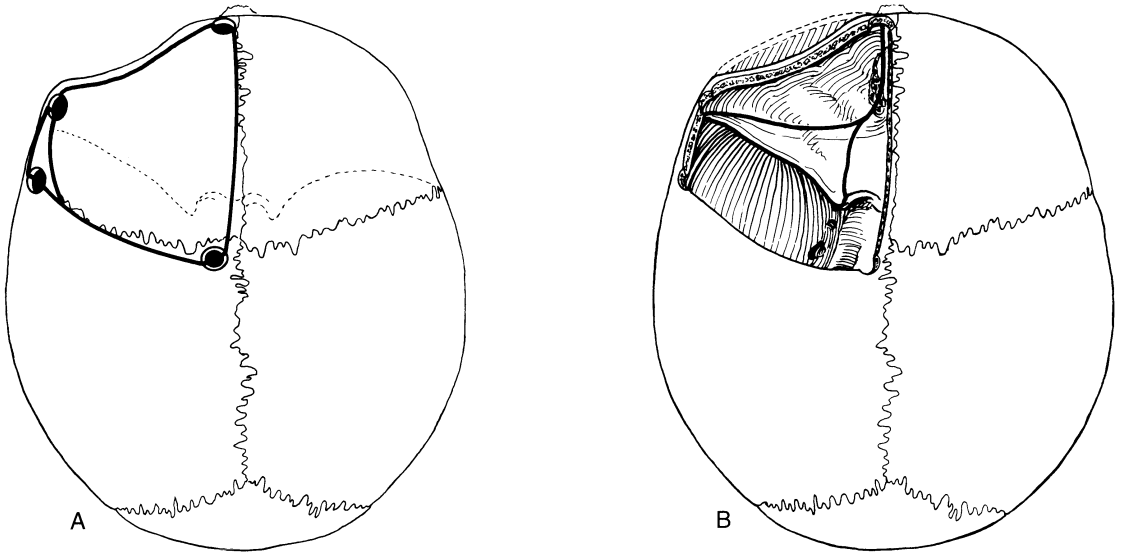


Figure 14.21. This view from the vertex illustrates the osteomies over the vault and along the floor of the anterior fossa. (A) *The solid line* indicates the osteotomy and illustrates the flattening of the frontal eminence. *The broken line* outlines the lesser wings and body of the sphenoid. (B) The frontal bone has been removed, permitting illustration of the osteotomized floor of the anterior fossa, over the sphenoid body and medial to the frontoethmoidal suture to the frontonasal suture. *The shaded area* illustrates restitution of the frontal eminence attained by rotating the frontal flap 180°.

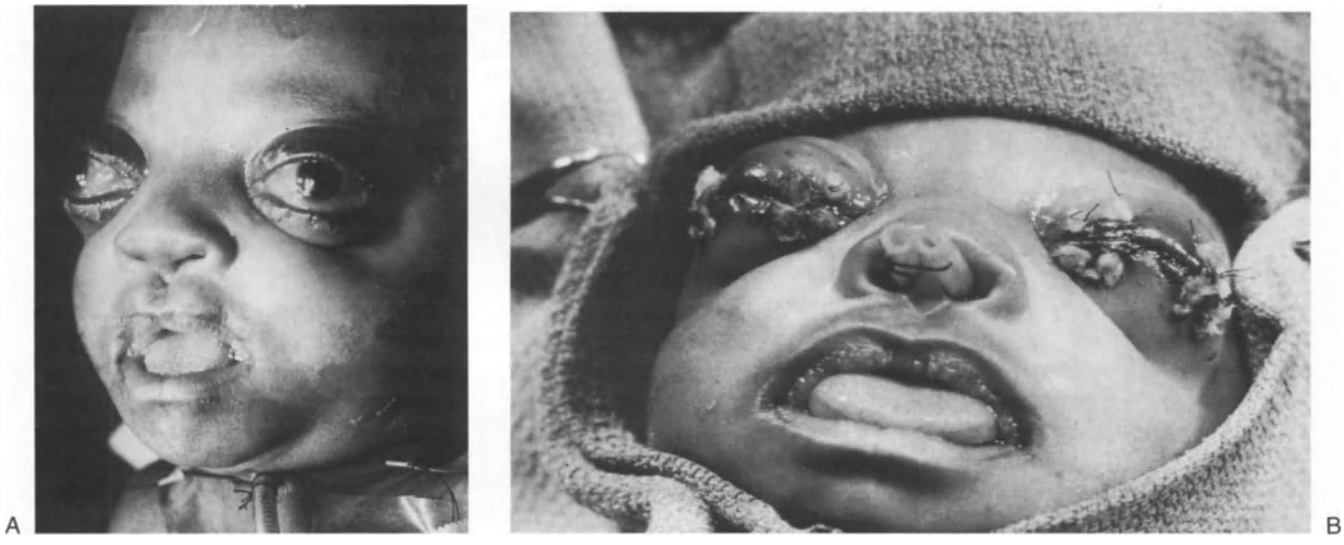


Figure 14.22. (A) The exorbitism which children with bilateral plagiocephaly suffer may be extraordinary. (B) In such instances, bilateral tarsorrhaphies are necessary.

If there is remarkable, unsightly, flattening of the squamous portion of the frontal bone, it may be separated from the orbital rim and rotated 180°, simply cracked and remolded, or buttressed with bone grafts before repositioning it. The orbital rim and squamous frontal bone are not tightly anchored at the time of closure; they are either loosely moored into position or just laid over the periorbita and the dura, allowing the growing brain to mold them.

Bilateral Coronal Synostosis: Plagiocephaly (Fig. 14.22)

Bilateral plagiocephaly may result in such severe exorbitism as to subject the sclerae and corneae to destructive damage. The operative procedure for the correction of bilateral plagiocephaly is quite different from the procedure for the unilateral deformity in that it involves opening the coronal, pterional, and medial (nasal, frontonasal, frontoethmoidal) sutures, the causative factors

of this severe craniofacial deformity. Its correction is undertaken with the participation of an experienced plastic surgeon.

The procedure consists of freeing, *en bloc,* the squamous portion and supraorbital rims of the frontal bone and then allowing it to be displaced anteriorly by the frontal lobes and superiorly by the globes. This corrects the exorbitism. The hypotelorism is corrected by the implantation of a bone graft between the osteotomized supraorbital rims. Standard plastic surgery texts should be consulted for details of the operative procedure. In the very young (less than 3 months of age) one may perform a radical craniectomy, removing and discarding the frontal and pterional bones: the skull grows over the dura.

Occipital (Synostosis) Plagiocephaly

This "skull deformity," with the very rarest of exceptions, is not the result of synostosis of the lambdoidal, or any other adjacent, suture. It occurs as a compensatory, or secondary, deformity in severe forms of coronal synostosis, but without lambdoidal synostosis. Few, very few, documented cases have been reported: *3.1% of the total number of children with other forms of neurocranial synostosis* and only 3.06% of those presenting with occipital plagiocephaly had synostosis of the lambdoidal suture [20]. The clinical picture referred to as "deformational (nonsynostotic) occipital plagiocephaly" (DOP) is controversial in management, and very surely the result, in the large (> 97%) majority of cases, of the prone position for sleep recommended for newborn by the American Academy of Pediatrics (AAP) in 1992. In fact, Kane et al. [21] reported a study involving three time frames, two preceding and one following this report, with an increase from 2.2 cases on average per year in the former to 52.3 in the latter ($P = 0.0001$)!

DOP should be treated with correct sleeping postures, keeping the child sitting, or a helmet. True lambdoidal synostosis should be treated with surgical excision of the synostosed suture from across the sagittal suture to across the occipitomastoid suture.

Hypertelorism (Fig. 14.23)

If one remembers that the term "ocular hypertelorism" is misleading, since it indicates the interpupillary distance, and that the term "orbital hypertelorism" is expressive of the true situation, then one has an understanding of what the plastic surgeon is attempting to accomplish. "Ocular hypertelorism" was coined in 1924 by Greig [22], "orbital hypertelorism" in 1972 by Tessier [23]. The earliest attempts to treat surgically orbital hypertelorism were really "patchwork," since the surgeons limited themselves to correcting extraorbital malformations of the epicanthus, eyebrows, root of the nose, and so on, by transposing skin or hair grafts and fat pads. In 1962 Converse and Smith [9] displaced the medial canthus and medial wall of the orbit along with the nasal rim, medialward. In 1963 Tessier did much the same but also displaced the inframedial angle with the lacrymal system, and then, in 1965, he first moved the floor of the orbit medialward. Subsequent to this, in 1973, Tessier [10] published his classic work on the surgical management of orbital hypertelorism, stating that "the objective is clear: to bring the eyes closer together without the risk of a meningeal infection, or affecting ocular, oculomotor, or respiratory functions. Since these deformities of the interorbital space lie on the cranial base, the intracranial route must be used in the majority of cases."

Reflection of a bifrontal flap for hypertelorism entails considerations that the crista galli may be remarkably hypertrophied, the dura may be deformed by an associated meningocele or meningoencephalocele, and that one may be able to preserve the olfactory nerves because the olfactory grooves are displaced laterally with the orbits. If a meningocele or encephalomeningocele is present, the olfactory grooves and nerves cannot be preserved. The various complications one may encounter in performing this procedure to correct hypertelorism are meningitis, blindness, oculomotor palsy, extraocular muscle weakness, and damage to the neurovascular bundle.

The steps in the performance of craniofacial reconstruction for hypertelorism, the A-3 Tessier procedure, through the intracranial route as discussed here are taken from the 1972 publication by Tessier, Guiot, and Derome [24].

Crouzon and Apert

The term craniofacial dysostosis may be used to describe both Crouzon's and Apert's deformities, since, for all intents and purposes, the clinical condition and surgical considerations are the same. It still is not clear, however, in either of the two genetic entities what the relationship is between the cranial synostosis and the very characteristic facial deformities, though some authors [25] have postulated that such facial hypoplasias as facial stenosis, independent of the presence of retrusion of the maxilla, are pathogenically identical to craniostenosis.

The aim of surgical treatment of craniofacial dysostosis is correction of the remarkable exorbitism, the malocclusion secondary to retrusion of the maxilla, the compressive obliteration of the nasal pharynx, and the recession of the frontal bone which is an expression of coronal synostosis.

In 1950 Sir Harold Gilles [6] was the first surgeon to perform a midface advancement for craniofacial dysos-

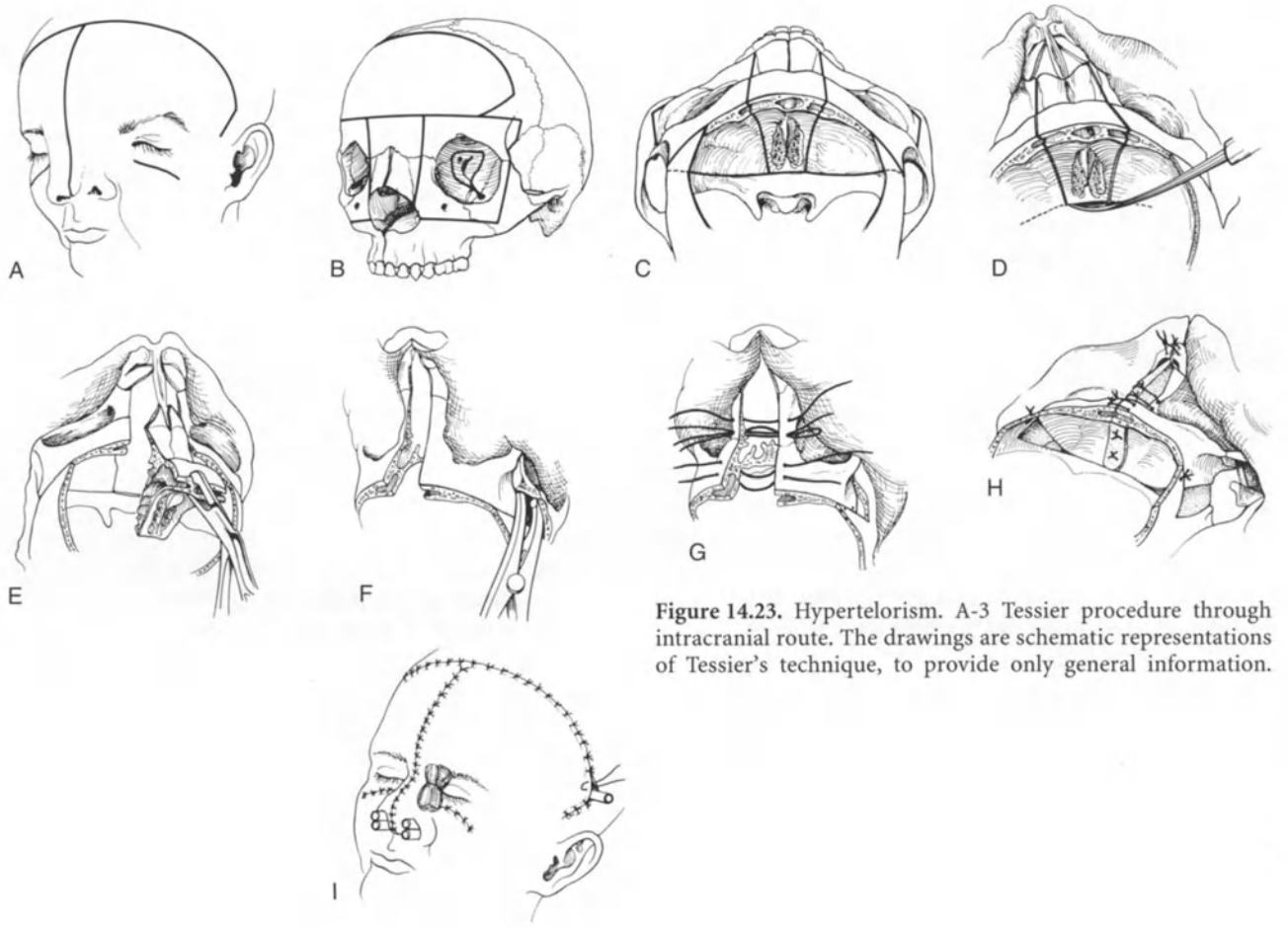


Figure 14.23. Hypertelorism. A-3 Tessier procedure through intracranial route. The drawings are schematic representations of Tessier's technique, to provide only general information.

tosis, leading the way for subsequent plastic surgeons to revolutionize completely the treatment of this and other congenital anomalies of the facial bones. In 1958 Tessier [26] performed his first midface advancement, which was followed by a period of 9 years of surgical experience. In 1967 Tessier [27] published these experiences and described his surgical approach to the treatment of craniofacial dysostosis. This is predicated upon a careful study of the stress lines of the facial bones as expressed clinically by facial bone fractures, and analytically by the LeFort [8] classification of these fractures: I, II, III. In fact, Tessier was successful in displacing the entirety of what he refers to as the "useful orbit" a minimum of 8 mm and a maximum of 10 mm off the orbital apex, in three directions, permitting him to treat hypertelorism, the facial retrusion of cranial synostosis and facial stenosis, orbital dislocation secondary to trauma, and orbital-facial clefts. The possible variations on manipulation of the orbit unlock the anatomical restraints for treating surgically a variety of facial and craniofacial abnormalities.

Though one may consider both Apert's and Crouzon's to be the same disease entity with regard to surgical planning and management, it may be of some value – if nothing else, academic – to note that Crouzon's disease is characterized by a less prominent supraorbital ridge, bregmatic bump, and recessed frontal bone. The exorbitism, consequently, is more remarkable in Apert's, especially in light of the fact that the recession of the supraorbital rim in Apert's is more severe than the facial retrusion. Also, patients suffering Apert's syndrome have a transverse frontal skin furrow, a crossbow deformity of the upper lip with anterior open bite, oculomotor paralysis, asymmetrical exorbitism and ptosis, and hyperseborrhea. These are relative distinctions. The only absolute distinction between Apert's and Crouzon's is the presence of syndactyly in Apert's disease.

The fundamental elements of Tessier's technique [28] for correction of craniofacial dysostoses are as follows:

1. Total correction of the orbital and maxillary anomalies
2. A monoblock osteotomy, extending to the pterygoid processes of the facial mass
3. Infrabasal and orbital rim osteotomies with sagittal splitting of the lateral walls of the orbits

4. Frontonasal and frontomalar triple osteosynostosis for fixation
5. Bone graft wedges used to fix the intercranial and facial separations used to correct inframaxillism, facial shortness, and vertical atresia of the orbit
6. Correction of hypertelorism, if present
7. Advancement of the inferior portion of the frontal bone via the cranial route if the frontal cranial deformity needs correction

Kleeblattschädel (Cloverleaf) Trilobed Skull Deformity

In 1960 Holtermueller and Wiedermann [29] reported 13 cases of cloverleaf skull deformity (12 of which had been previously published in the German literature but described as hydrocephalus, chondrodystrophic). The deformities these authors described consisted of a very characteristic cloverleaf appearance of the skull, accompanied by inferior displacement of the ears; hypertelorism, mandibular abnormalities, nasal flattening; micromyelia and skeletal abnormalities; *and hydrocephalus.* The trilobar skull (cloverleaf) was considered to be the result of synostosis of calvarial and basal sutures, compounded by hydrocephalus, which blows out the anterior fontanelle and the squamosal sutures. The authors considered the possibility of some relationship between kleeblattschädel and achondroplasia, Crouzon's craniofacial dysostosis, and Apert's acrocephalosyndactyly.

The surgical treatment consists of two distinctly different approaches: compensation of the hydrocephalus when present (it has recently been demonstrated – Turner and Reynolds [30] in 1980 – that hydrocephalus is *not* an invariable accompaniment of Kleeblattschädel) and cerebral decompression, either by performing a total craniectomy or resecting the synostosed coronal and lambdoidal sutures. Cases of synostosis of the sagittal suture, in addition to coronal and lambdoidal, have also been reported, in which instances of the remarkable bulging of the anterior fontanelle accounts for the vertical leaf of this cloverleaf deformity. Because of the severe deformities of the basicranium, it is best to manage the child with kleeblattschädel in collaboration with a plastic surgeon.

Arachnoidal Cysts

Midline Arachnoidal Cysts

The entrapment of cerebrospinal fluid within fibrosed arachnoid, around what was once a normal cistern, freely communicating with surrounding cisterns and subarachnoid spaces in the midline, occurs in the parasellar area, around the corpus callosum, within the region of the quadrigeminal cistern posterior to the colliicular plate, in the superior cerebellar cistern over the culmen monticuli, and in the cisterna magna. Cysts of the septum pellucidum or a cavum Vergae have ependymal walls, are in free communication with the ventricular system, and have not been reported to be pathogenic. No surgical procedure, consequently, should be performed on them.

At the present time, there are two schools of thought concerning surgical management of midline arachnoidal cysts.

Craniotomy

Craniotomy permits resection of the cyst membrane and reestablishment of communication between the cyst cavity and surrounding cisterns or subarachnoid spaces.

If one elects to resect the cyst membranes, then it is important to carry the dissection lateralward, past the line of coalescence between arachnoidal membranes, and into subarachnoid cisterns until the arachnoid may be identified as a single membrane completely separated from parenchymal structure. When this has been accomplished one may see cerebrospinal fluid well-up into the field intermittently, either with cardiac pulsations or respirations. The difficulty with this procedure is that the mesenchymal arachnoid tends to reform scar tissue postoperatively, once more sealing off a cystic cavity. The ideal would be to identify the ostium, through which cerebrospinal fluid gains access to the arachnoidal cyst, so as to seal it rather than resect the fibrotic cyst wall. "Theoretically, the CO_2 laser through a *ventriculoscope*, could be used more simply and effectively to vaporize the cyst wall and seal the ostium." "However, experience with the instrument and technique are wanting." This previous sentence was published as quoted in the first edition of this book, but since then both the instrumentation and technique have been perfected. In fact, this modality is now being used regularly... but we still do not know the results.

Cystoperitoneal Shunting

Cystoperitoneal shunts drain continuously the cerebrospinal fluid from ependymal or arachnoidal cysts, providing the possibility that the ostium may seal itself off spontaneously. The proximal end of the shunting system may be inserted into the cyst cavity either stereotaxically or through a ventriculoscope. Ultrasound is of great assistance for stereotaxic placement.

Chiari IV Malformation

Aplasia of the entire cerebellum with the exception of the flocculonodular lobes results in the *ex vacuo* dilation of the IV ventricle, cistern magna, and pontine cistern. There is no obstruction to the flow of cerebrospinal fluid. This primary dysplasia has no surgical treatment.

Lateral Arachnoidal Cysts

It is difficult to state precisely whether cerebrospinal fluid (CSF) containing cysts located over the cerebral or cerebellar hemispheres are arachnoidal cysts, ependymoarachnoidal cysts, or dysplastic areas of brain with leptomeninges bridging over them. In either event, these CSF-containing compartments are only partially covered by an arachnoidal membrane.

The surgical options of treatment of lateral arachnoidal cysts are the same as for midline cysts: resection of the arachnoidal membrane or insertion of a cystoperitoneal shunt. Arachnoid membrane resection for lateral cysts, however, has the added complication of possibly resulting in accumulation of increasing amounts of CSF in the subdural spaces: resection of the cyst wall permits the CSF to flow directly into the subdural space, thereby collapsing the cerebral hemisphere as increasing amounts of fluid pool within this compartment. Resection of the arachnoid membrane is followed by surgical opening into the lateral or medial cisterns, providing avenues for passage of CSF from over the cerebral hemisphere. One may not expect the CSF to percolate into the subarachnoid spaces.

Irrespective of whether the decision is made to resect the arachnoidal membrane or to shunt the cyst cavity, lateral arachnoidal cysts should be operated on per emergency if the child develops a clinical picture of an increase in intracranial pressure. Otherwise, the child may be operated on electively, *but then only if there is a clinical indication.* The success rate for permanent obliteration of the cyst and cerebral reexpansion is not such as to permit one to operate without definite clinical indications of impending intracranial hypertensive decompensation, progressive neurologic deficit, intractable seizures, or incapacitating episodes of headache.

Craniofacial Encephalomeningoceles

Ingraham and Swan [31], in their 1943 paper on spina bifida and cranium bifidum, defined *encephalocele* as "all meningeal protrusions, whether or not associated with nerve tissue, whose osseous defect was in the skull (cranium bifidum)," excluding dermoid cyst arising from the dura and extending through the bone. Etymologically speaking, this definition is undesirable, since it is limited to meningeal protrusions but, in fact, includes extrusions of cerebral parenchyma. It is preferable to speak of *craniomeningocele* when the protrusion is composed of dura mater, arachnoid, and subarachnoid CSF; to speak of *cranial encephalomeningocele* (adding, of course, the anatomical location) when the protrusion also includes cerebral parenchyma and/or ventricular CSF. This distinction is most important since it has prognostic value: parenchymal tissue, especially if a portion of the ventricle is included within the encephalocele, indicates an extremely poor prognosis, whereas, conversely, the absence of cerebral parenchyma and ventricular CSF indicates an excellent prognosis.

Since the time of Christian Fenger's [32] first operation on a basal encephalocele in 1895, the surgical technique has not changed, being limited to the development of the meningocele (or encephalocele) sac at its base, and then excising it (with neural elements if they are contained within the sac) and closing the dura tightly, before repairing the soft tissue over it and then closing the skin. The sole exception to this change in technique is the recent use by few pediatric neurosurgeons, but most plastic surgeons, of bone grafts to repair the bony defect. Plastic surgeons use the grafts invariably in their repair of the basal defects, neurosurgeons generally do not use bone to repair defects in the neurocranium. This is unfortunate, since bony defects over a repaired encephalocele may be responsible factors for progressive ventricular "blowout" which progresses to porencephaly.

These anomalies, fundamentally dysraphic in nature, may involve the base of the skull (chondrocranium) only, the vault and facial bones (with a concomitant anomaly of the ethmoid invariably present), or primarily the frontal bone. In its broadest terms, consequently, one may speak of basal and/or sincipital encephalomeningoceles, the former limited entirely to the chondrocranium and the latter involving the frontoethmoidal area.

From a purely surgical point of view, a modification of the 1974 Charoonsmith and Suwanwela [33] classification of craniofacial meningoencephaloceles is recommended. The terminology is predicated upon anatomical structures, permitting one to identify immediately the location of the encephalocele and to undertake appropriate clinical and diagnostic studies to determine feasibility and technique of surgical repair.

The treatment of basal encephaloceles has been controversial and, in general, disappointing. Blind removal of the encephalocele with a snare, under the *mistaken* diagnosis of a nasal polyp, has been performed in small lesions. Lewin and Schuster [34] successfully repaired a basal encephalocele by the transoral route. Upon opening the sac, the encephalocele contained no cerebral tissue. The sac was ligated, the stump replaced into the en-

docranial cavity, and the bony defect covered with an osteoperiosteal graft. It has been commonly agreed that the associated harelip or palatal defect should be repaired at a later date [31, 35, 36].

Blumenfeld and Skolnik [37] suggested not to treat asymptomatic small basal encephaloceles in neonates and infants. Robinson [38] recommended a tracheostomy in neonatal life when the encephalocele is large and interferes with sucking and breathing. The intracranial procedure can be performed later, under better conditions.

Surgery by the transnasal route should be discouraged in favor of craniotomy because of increased risk of cerebrospinal fluid infections. The intracranial approach to the encephalocele permits thorough inspection of the herniated cerebral structures, amputation if feasible, and repair of the bony defect with bone graft and fascial coverage for the dural defect [34, 39, 40]. Frequently, however, the size of the defect and the involvement of important cerebral tissue in the encephalocele limit the procedure to only an exploratory operation. The presence of vital structures such as hypothalamus and pituitary gland in the herniated sac precludes its excision and adequate repair of the bony defect, which would result in profound hypothalamic dysfunction and death. This proved to be the case in reported instances [41–43]. Therefore, the surgical treatment of basal encephaloceles has been frustrating and unsatisfactory, and is associated with a 50% mortality.

The repair of transsphenoidal, sphenoethmoidal, and spheno-orbital encephaloceles may be through a combined transfrontal and transethmoidal approach, with external excision of the encephalocele, duraplasty, rib graft to close the bony defect, and, where possible, mucosal closure. When associated with cleft palate, the approach should be primarily transoral. One must not attempt to reduce the encephalocele into the intracranial compartment through the nasal cavity. Preferably, use the transcranial route to dissect the ostium at the cranial end, the transoral/nasal route to resect the encephalocele, and then do the dural and bony repair through, respectively, the transcranial and transoral/nasal routes. This approach can be used for the sphenoethmoidal and spheno-orbital encephaloceles, but not for the transsphenoidal. This latter encephalocele must be repaired entirely by the transoral/nasal route.

The single most recurrent, and important, observation in a systematic review of the subject of basal (intranasal) encephaloceles is the high incidence of diagnosis by serendipity, usually with very negative effects upon the child's well-being: the nasal mass is diagnosed as a polyp and "polypectomy" is performed. Recurrent meningitis and persistent cerebrospinal fluid rhinorrhea, in fact, are so common following this procedure that Choudhury and Taylor [44] stated "the diagnosis of encephalocele should be considered in any patient with a nasal polyp, especially in children and in patients with recurrent bacterial meningitis, with or without rhinorrhea, in the absence of cranial trauma or surgery, or in the absence of external craniospinal anatomical defects." *One may safely add that the presence of CSF rhinorrhea following (nasal) polypectomy forces the physician to diagnose a basal encephalocele.*

Craniofacial Meningoencephaloceles: Basal Craniofacial Meningoencephaloceles

Basal craniofacial meningoencephaloceles result from defects in the clivus (basiocciput, spheno-occipital synchondrosis, basisphenoid), the sella turcia (body of the sphenoid), the planum sphenoidale, and the ethmoid.

It is not unusual to observe radiolucencies within the sphenoid bones of the newborn and infant, though they disappear by the 2nd or 3rd year. These are residua of vascular channels, considered by some to be the embryological and anatomical basis of congenital defects in the sphenoid bones, which result in craniopharyngeal meningoencephaloceles. Accordingly, Heineeke (1882) [45] identified the spheno-orbital and sphenoidal (which he called sphenopharyngeal) categories, also describing a third (sphenomaxillary) category, which we presently group with the sphenoethmoid since the mass occupies the posterior nasal cavity.

Since the first extensive review of the subject in 1947 by Gisselsson [46], the severity of the anomaly and the operative morbidity/mortality have been well known and progressively led surgeons to be cautious and conservative. Twenty-four of the patients reported by Gisselsson died either of associated anomalies or meningitis; there was a 50% mortality in the 13 patients who had been operated on. The first surgeon to treat successfully an intranasal meningoencephalocele was Fenger, in 1895, but it is not known whether his was an ethmoidal, sphenoidal, or a frontoethmoidal defect.

Sphenoidal Meningoencephaloceles

Encephaloceles enter the sphenoid sinus through either the sella turcica or basisphenoid. They may either remain within the sphenoid bone or, more commonly, bulge into the retropharynx.

Transsphenoidal Encephaloceles. The transsphenoidal encephalocele is characterized by a total absence of external signs indicative of the lesion, with the sole exception of those reported cases, by Lichtenberg [47] and Virchow [48], where the masses bulged through the nares and mouth.

Sphenoethmoidal Encephaloceles. Sphenoidal encephaloceles present in the posterior nasal cavity, extending

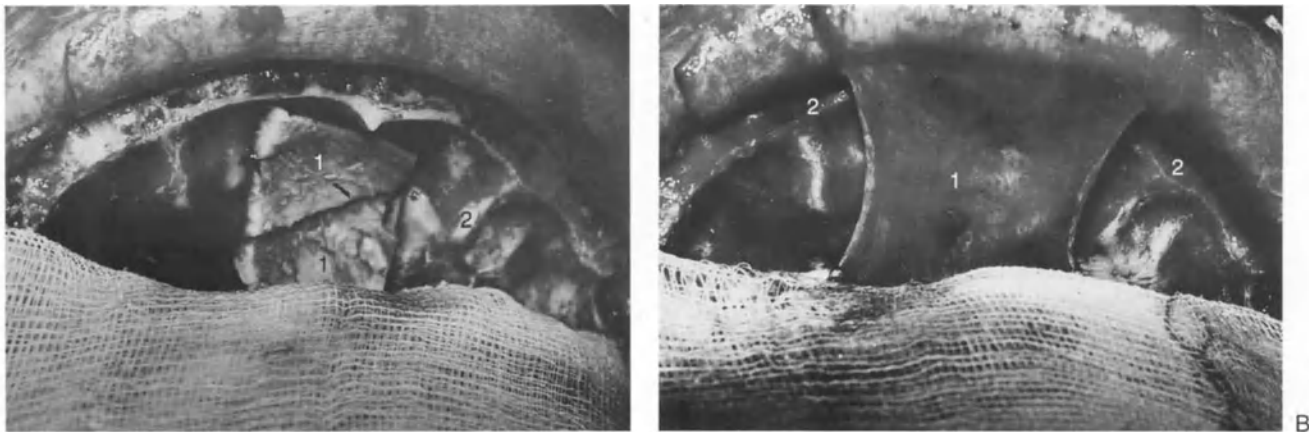


Figure 14.24. After the encephalocele has been resected, bone grafts should be lain into the defect and the dura reconstructed and/or grafted. (A) The defect in the ethmoidal bone is filled with bone grafts (*1*). The orbital roofs (*2*) are intact. (B) The prepared periosteal flap (*1*) is then brought over the frontal bone and bone grafts, before being sewn to the dura posterior and lateral to the grafts. As the frontal lobes come back into position to rest upon the orbital roofs (*2*) and cribriform plate, this periosteal graft comes to be interposed between the dura of the frontal lobes and bone graft.

through the interval between the sphenoid and ethmoid bones, at the planum sphenoidale.

Spheno-orbital Encephaloceles. Defects in the sphenoid bone, at the junction between the greater wing and the body, result in the encephalocele extending through the orbital fissures (superior and inferior) and into the orbit, causing remarkable exophthalmos. These defects in the sphenoid and orbital roof are common in, though not characteristic of, neurofibromatosis.

Ethmoidal Meningoencephaloceles (Fig. 14.24)

The encephalocele extends through the ethmoid bone and into the nasal cavity posteriorly.

Frontal Ethmoidal Meningoencephaloceles: Sincipital Encephaloceles

Frontal ethmoidal encephaloceles are characterized by a defect between the ethmoidal and frontal bones, at the foramen cecum, with the crista galli at the posterior rim of the defect. The facial bones (inferior frontal and orbital rim, nasal, lacrimal, and maxillary) are displaced in varying directions and destroyed to varying degrees, depending upon the specific route and length of the encephalocele.

Sincipital meningoencephalocele is the term used to describe those encephaloceles that protrude at the frontoethmoidal junctions, invariably bulging so much that they are readily visible and most often covered by intact skin. They are subclassified into nasal-orbital, nasal-ethmoidal, and nasal-frontal, so that they may be distinguished readily from the basal meningoencephaloceles previously described and the cranioschisis and cranioencephalomeningocele that are described later.

It was not possible to provide information concerning the geographic distribution of this particular congenital anomaly before the work of Suwanwela and coworkers [36], since this classification had not yet been suggested and authors reported cases as encephaloceles or meningoceles rather than subdividing them anatomically. Suwanwela and his colleagues state that "in Thailand, anterior encephalomeningoceles stand out as an exceptionally common and most interesting malformation of the nervous system." In an 8-year period they treated 100 cases of sincipital (frontoethmoidal) meningoencephalocele, but only 1 temporal, 3 vertex, 11 occipital, and 35 spinal meningoceles, stating that the incidence of sincipital meningoencephalocele is one in 5000 living people in Thailand. They do not give information concerning prevalence of the anomaly among live births or stillbirths.

At birth, the sincipital meningoencephalocele generally (though certainly not invariably) is quite small, but it increases steadily in size as the facial deformity worsens with increasing age, suggesting the advantages of early surgery. Another indication for early surgery is the presence of exposed parenchymal tissue or the absence of full-thickness skin covering.

Two different surgical approaches may be used, *extracranial* and *intracranial,* with the former being recommended when the meningoencephalocele has a short stalk and an ostium wide enough to permit access to the intracranial component (forehead and frontal nasal junction defects). The intracranial approach, conversely, is indicated when the stalk is quite long and the ostium small, so that dural repair must be performed

from the intracranial surface. One may reconstruct the entire craniofacial anomaly in one sitting if the extracranial approach is used, but a second operation is necessary to correct the facial anomaly if the intracranial approach is used.

If a complicating hydrocephalus is present, it should be treated with ventriculoperitoneal shunting before any attempt is made to resect the meningoencephalocele (and reconstruct the facial anomalies), since the diminution in ventricular volume and pressure minimizes the anomaly and arrests its progression. Of course, a shunt should not be inserted if parenchymal tissue is exposed, the surface of the meningoencephalocele is infected, or there is a cerebrospinal fluid leak. In these instances, one may find it advantageous to insert an external ventricular drain, which has the same effect as a shunting system but provides added protection against infection. Once the infection is cured or the skin defect repaired, one may proceed to internalize a sterile shunting system and then wait for an appropriate time to resect the meningoencephalocele and reconstruct the craniofacial anomaly.

Nasal Orbital Meningoencephalocele

The mass generally protrudes between the medial rim of the orbit and the nasal bone, obliterating the nasolabial fold and displacing the globe superolaterally. The frontal bone, glabella, and nasal bone are normal, but defects are present within the maxilla, at the nasal and frontal junctions, the lachrymal bone, and the cribriform plate of the ethmoid. Parenchymal contents of the fronto-orbital meningoencephalocele include the olfactory bulb, nerve, and gyrus.

The nasal bone forms the anterior rim of the ostium, along with the maxillary bone, whereas the lachrymal bone and lamina papyracea of the ethmoid form the posterior rim, with the stalk being quite long. The extrusion extends from the frontoethmoidal junction intracranially, across the maxillary rim of the orbit and along the midfacial plane.

The surgical repair consists of performing a bifrontal craniotomy and then using an extradural approach to separate the dura mater from both orbital roofs and the cribriform plate. Coagulate, one at a time, the dura and arachnoid covering the individual olfactory rootlets as they penetrate the perforations in the cribriform plate on the normal side, if the nasal orbital meningoencephalocele is not bilateral. The dura is separated well behind the posterior rim of the frontoethmoidal defect(s), and then the dural covering of the meningoencephalocele is separated from adherent tissue using fluffy cottons, as the dissection is extended anteriorly, inferiorly, and slightly laterally around the sac, into the medial surface of the orbit, and from the lateral surface of the nose. As soon as the horizontal plane of the ostium (orbital roof, supraorbital rim, ethmoid) has been identified and freed of dural adhesions, enough of the dural protrusion to permit a sturdy and watertight closure is reduced into the intracranial compartment. The stalk is double-ligated (with the ligatures placed at a 4-mm distance from one another) and transected between the two ligatures. The proximal, cranial, portion of the stalk is then invaginated and the redundant dura imbricated over it and sewn snugly to normal dura, obliterating the pouchlike extrusion and reinforcing the area of herniation.

Before placing the bone graft over the ostium, the distal portion of the meningoencephalocele is removed from its bed, if this may be easily performed, or freed as extensively as possible and then advanced into the subcutaneous space at the region of the nasolabial fold, if not. A periosteal graft (taken from the surface of the frontal bone at the time of craniotomy) is then lain along the intracranial surface of the ostium, covering the entirety of the ostium and the floor of the anterior fossa, and anchored at its periphery to the dura, leaving an opening large enough for insertion of the bone graft. This creates, in essence, a pouch of periosteum: the outer layer of the dura, which is the inner layer of the cranial periosteum, and the periosteal graft, which will be the outer layer of the cranial periosteum. The bone graft is then inserted into the pouch, over the ostium, covering completely the frontoethmoidal defect, and then the opening in the pouch is closed, sealing the bone graft and locking it into position over the defect. It is preferable to insert a graft that is much larger than the surface area of the defect, to cover it as a lid and not fit snugly into the defect. This minimizes risk of slough or dislodgement of the bone graft.

If the distal portion of the meningoencephalocele has not been removed through the intracranial approach, one proceeds to bring the frontal bone flap back into position, anchor it into place, and close the skin, before removing the distal portion of the encephalocele through a facial incision made along the nasolabial fold. Considerations of the feasibility and technique of reconstruction of the orbital rim are matters for the plastic surgeon.

Nasal Ethmoidal Meningoencephalocele (Fig. 14.25)

The meningoencephalocele extends through the frontal ethmoidal region, behind the nasal bones and the frontal process of the maxilla, and in front of the nasal septum and cartilage at their attachment to the ethmoid nasal mucosa. This forms the medial surface, the medial wall of the orbit forms the lateral surface. The nasal ethmoidal encephalocele has a long stalk that extends from the frontoethmoidal defect to the junction of nasal bones and cartilage, between the inner canthi of the eyes on either side. The nasal bone is displaced super-

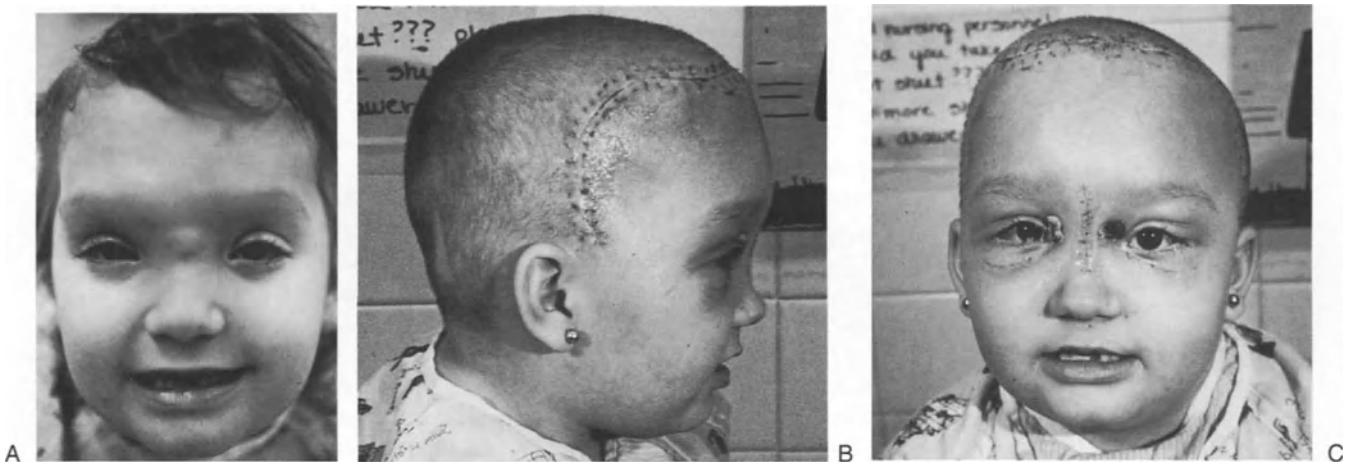


Figure 14.25. (A) Preoperative nasal ethmoidal encephalocele. (B, C) Postoperative results. Note that the procedure was performed by a combined intracranial and nasal-frontal approach.

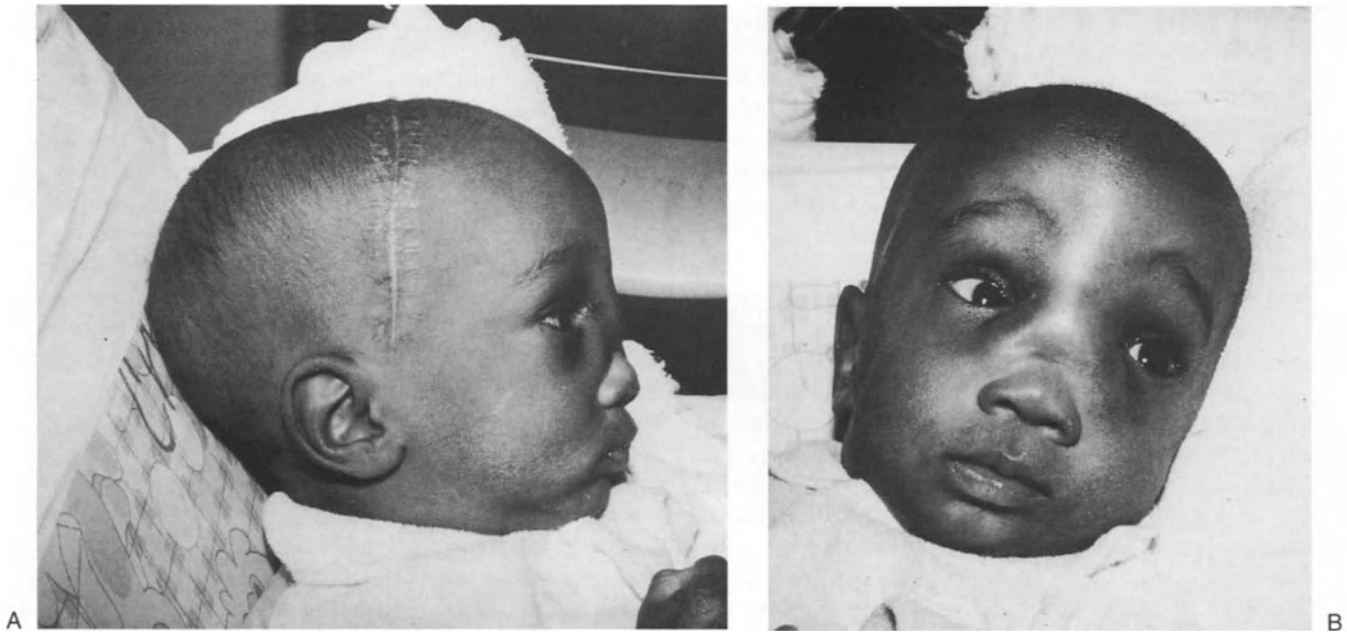


Figure 14.26. Postoperative result of a repair of the nasoethmoidal subgroup of sincipital (frontal ethmoidal) cranioencephalocele. (A) The repair was performed by the intracranial route, and all of the encephalocele was removed. (B) The dura is repaired and the bony defect is grafted with autogenous bone. Reconstructive nasal surgery still needs to be performed.

iorly, and the nasal cartilage inferiorly along with the most anterior portion of the ethmoid at the foramen cecum.

The operative procedure is identical to the one described for the nasal orbital subcategory of frontal ethmoidal meningoencephaloceles. Because of the superior displacement of the nasal bone and the midline location of the meningoencephalocele, there is considerable bony deformity around the bridge of the nose and medial aspect of the orbit, necessitating reconstructive surgery in most, but certainly not all, instances (Fig. 14.26).

Nasal Frontal Meningoencephalocele

The extrusion of the nasofrontal meningoencephalocele is through an interval between the frontal and ethmoidal bones intracranially, and the frontal and nasal bones

at the face, with a resultant short stalk. The medial orbital walls, at the lachrymal bones, are displaced laterally, and the ethmoidal bone is displaced posteroinferiorly. The crista galli is wedged into the stalk at its posterior inferior surface, both orbital roofs being displaced inferiorly and lateralward. The nasal septum, cartilage, and bones have normal relationships with one another and the frontal processess of the maxilla. Thus, the meningoencephalocele presents as a mass within the region of the glabella. It varies in size, form, and content much more than either the nasal orbital or nasal ethmoidal subgroups.

Because of this great variation in form, size, and content of the nasal frontal meningoencephaloceles one may expect olfactory nerves and gyri, frontal poles and lobes, optic nerves and chiasm, internal carotid and anterior cerebral arteries, and even the ventricular system and temporal lobes to be herniated into the sac to varying degrees, depending upon the volume of the nasal frontal meningoencephalocele. Consequently, the surgery must vary both in extent and technique, since the small defects are compatible with life and, in some cases, a normal life. The very large defects tend to be fatal in a brief period of time. Surgery on the larger defects should be undertaken only to facilitate nursing care and to render the child less grotesque, so that the family and medical personnel caring for him may do so with some degree of ease and with a minimum of psychological shock. The technique for the smaller defects is the same as already described for the nasal orbital and nasal ethmoidal subgroups. Orbital reconstruction is reserved for those children who will be viable.

Facial deformities in all three frontal ethmoidal meningoencephalocele subgroups tend to become less severe over the months and early years following resection of the meningoencephalocele and repair of both the dura and bone. One should not, therefore, undertake reconstructive surgery of the face at an early age. When the facial reconstructive surgery is performed, whether in a single stage with the resection of the meningoencephalocele, or in two stages, the orbital reconstruction is performed only after the meningoencephalocele has been resected, the dural defect repaired, and the bony defect closed.

Cranioschisis

Cranioschisis describes clefts in the neurocranium, basicranium, facial bones, and acrania or anencephaly. One may subdivide cranioschisis into four developmental anomalies:

1. Cranial: upper facial cleft
2. Basal: lower facial cleft
3. Acrania
4. Anencephaly

The upper and lower facial cleft anomalies are not seen by the pediatric neurosurgeon. Anencephaly has no surgical treatment. We have never seen a case of acrania.

Cranial Meningoencephalocele

Cranial meningoencephaloceles are limited to the neurocranium, occurring along the sagittal plane from the region of the metopic suture anteriorly to the craniovertebral junction posteriorly, so that one may encounter interfrontal, anterior fontanelle, interparietal, posterior fontanelle, and occipital (cervical) meningoencephaloceles. As the craniofacial meningoencephaloceles, they vary considerably in presence or absence of skin covering, form, volume, and content. Also, the mortality and morbidity are directly related to the size of the meningoencephalocele and the cranial ostium.

Orbital Encephaloceles

Orbital encephaloceles most commonly protrude from the medial aspect of the orbit, through the lachrymal and ethmoidal plates, displacing the globe laterally, swelling somewhat beneath the nasal bone. Occasionally, an encephalocele may protrude through a dysplastic orbital roof, displacing the globe inferiorly and expanding beneath the superior palpebral fissure. Bulges at the lateral aspect of the orbit are suspect when considered encephaloceles. Rather, thought should be given to the most likely probability of an orbital tumor of one kind or another ranging from dermoid (Fig. 14. 27) through lachrymal gland, or, more likely, von Recklinghausen's disease.

The medial or superiorly located orbital encephalocele should be repaired through a medial frontal craniotomy, extradural approach, with the child in the supine position. Removal of the superior and medial aspects of the orbital rim, as when approaching an intraorbital tumor, exposes the encephalocele. One need not open the dura. It is sufficient to imbricate the redundant dural prolapse, layer over layer, reinforcing the area of defect. The bony orbital rim is then replaced and securely anchored. If the bony defects are extensive, one may choose to take a small graft from the posterior frontal or anterior parietal bones, mold it into the desired position, and thereby reinforce the areas where one expects prolapse. The dura must be imbricated and bone grafts placed in the superior and medial aspects of the orbit. If this is not done, the intraventricular pulsatile force may result in the formation of a porencephalic cyst and displace compressed cerebral mantle through the bony defect.

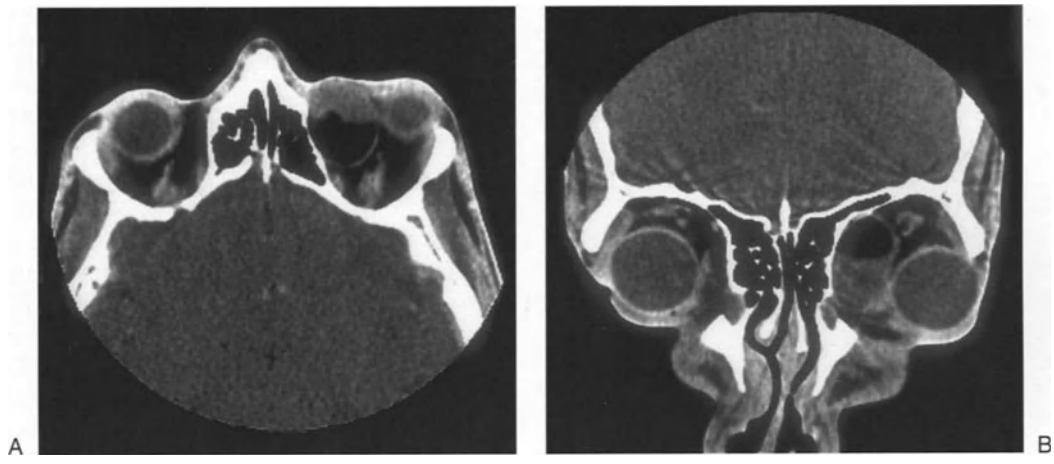


Figure 14.27. Orbital dermoid – this CT scan, axial (A) and coronal (B), is an imaging study of an extraconal mass in the superomedial portion of the left orbit. It has fatty and solid portions, with the fat located inferiorly in the axial image and superiorly in the coronal image. The lesion is displacing the globe lateralward.

Interfrontal Meningoencephalocele

Interfrontal meningoencephalocele occurs within the region of the metopic suture, so that the two frontal bones are displaced from the midline, forming the lateral borders of the defect. The glabellar region forms the inferomedial border, and the two parietal bones at the sagittal suture the posterior border. The frontal and a portion of the parietal lobes herniate into the dural sac.

The surgical procedure consists of resection of the encephalocele sac and its contents, watertight repair of the dural defect, and skin closure. If the defect is small and the underlying cerebrum is normal or only minimally dysplastic, one should place a bone graft over the defect. This minimizes the risk of recurrence or the development of an underlying porencephalic cavity.

Anterior Fontanelle Meningoencephalocele

The frontal and parietal bones, along with the metopic, coronal, and sagittal sutures, form the borders of this defect. The surgical implications are the same as those already described for interfrontal meningoencephalocele, with the exception of consideration of implantation of a bone graft.

Interparietal Meningoencephalocele
(Figs. 14.28–14.29)

The anterior and posterior fontanelles represent, respectively, the anterior and posterior extremities of the defect, which is bordered laterally by the parietal bones. These defects are associated with severe cerebral dyplasia and herniation of the hemispheres into the encephalocele. The arterial and venous structures are also displaced into the encephalocele sac.

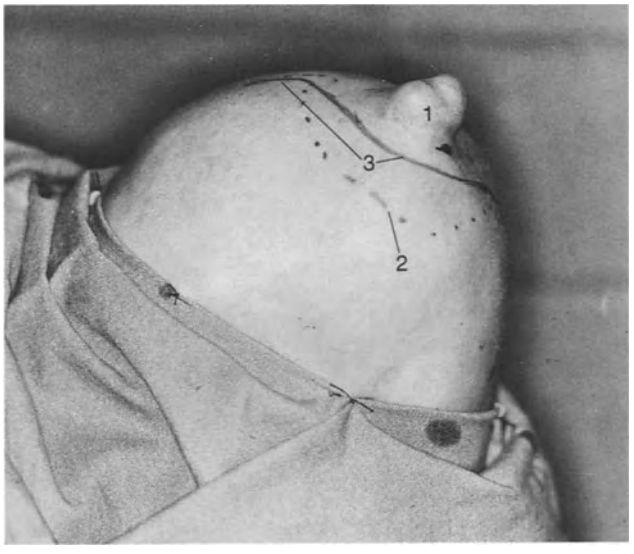


Figure 14.28. This is an interparietal meningoencephalocele that was operated on because the underlying brain was intact (as determined by cerebral angiography and pneumoencephalography). The meningoencephalocele (*1*) is bilobed and quite small, whereas the bony defect (*2*) is large. An S-shaped incision (*3*) was used to expose the sac and stalk of the meningoencephalocele.

Surgical considerations should be limited to serving humane, social, and psychological purposes.

Posterior Fontanelle Meningoencephalocele

Posterior fontanelle meningoencephaloceles are as severe as the interparietal, from which they are separated only by the presence of parietal bones along the anterior surface of the herniation.

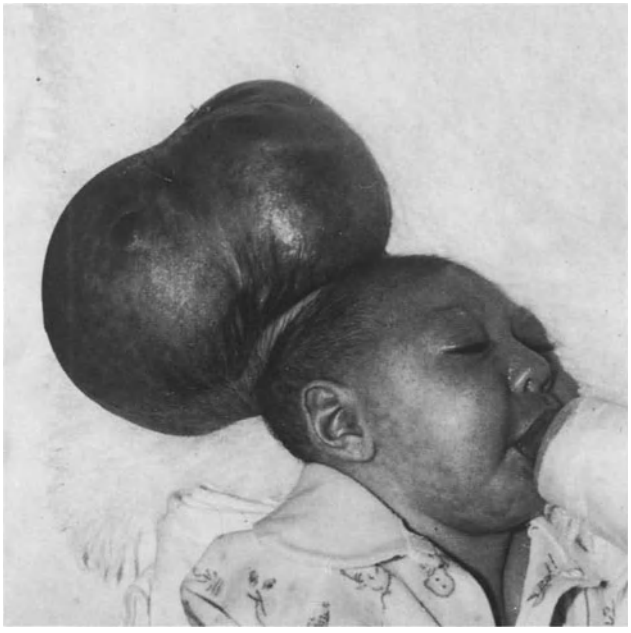


Figure 14.29. This child has an enormous interparietal meningoencephalocele, which contained most of both cerebral hemispheres, the anterior and middle cerebral arteries. This type of encephalocele is not a surgical entity...except for nursing and psychosocial reasons.

Chiari III Malformation: Occipital or Cervical Meningoencephalocele

Originally, the Chiari III malformation was considered to be a superior (occipitocervical, or upper cervical) meningoencephalocele or meningocele. Subsequently, authors included occipital encephaloceles under this terminology. Consequently, it is best to use the term Chiari III malformation for occipital, occipitocervical, and upper cervical meningoceles or meningoencephaloceles. In essence, the anatomical changes are not all that different, with the exception of the (very rare) pure meningocele, often covered by skin, of the upper cervical region.

The anatomopathological complexities of these dysraphic malformations are very variable, ranging from a simple occipital encephalocele containing fragments of cerebellar tissue, through coexistent Chiari III and Dandy-Walker malformations with cystic transformation of the IV ventricle, to displacement of portions of the cerebrum and hindbrain into the encephalocele.

The operative procedure consists simply of resecting the dermal and dural components of the encephalocele, along with the dysgenetic neural elements. Major vascular structures, identified preoperatively, should be separated from dysgenic brain and preserved. The dura mater and skin are closed as separate layers. No attempt is made to place the herniated dysgenetic brain in the cranial cavity.

Craniocerebral Disproportions: Chiari Malformations (Figs. 14.30–14.38)

Actually, it was Cleland [49], in 1882, who first gave a detailed description of what is now referred to as the Chiari II malformation. Arnold [50] did describe a case of spina bifida, in which there was downward displacement of the cerebellum into the cervical canal, but his description of the hindbrain changes was limited.

As we know them today, there are four Chiari malformations (the author prefers to define them as Chiari malformations and not Arnold-Chiari malformations), all of which have in common only one thing: *varying degrees of changes in content, relative location, and form of the posterior fossa or the structures within it and the upper cervical canal.* The classification of these anomalies, on the basis of changes in anatomy of the hindbrain associated with congenital hydrocephalus, is, of courses, incorrect: the Chiari I and IV malformations are only *rarely* associated with congenital hydrocephalus. The Chiari II malformation is associated with hydrocephalus in approximately 78% of children, the Chiari III malformation in approximately 50%. Thus, the Chiari malformations are discussed independent of the associated hydrocephalus which, in fact, is either an independent entity or a complication.

These four distinctly different anomalies are as follows:

1. Chiari I: characterized by variable displacement of the tonsils into the upper cervical canal.
2. Chiari II: variable displacement of the inferior portion of the cerebellar vermis and hemispheres, plus similar caudal displacement of the medulla oblongata, the elongated IV ventricle, and the lower portion of the pons Varolii into the upper cervical canal.
3. Chiari III: downward displacement of the medulla oblongata and herniation of portions of the cerebellum and, in some instances, the hindbrain into a high cervical or suboccipital meningocele.
4. Chiari IV: hypoplasia of the cerebellum, with the IV ventricle expanding into the entire posterior fossa and the pons Varolii being absent. Only the flocculonodular lobes are present.

Any discussion of surgery to treat either the Chiari I or Chiari II malformations must be preceded by an explanation of the anatomical pathology and the pathogenesis. This is especially true in light of the fact that the operative procedures are decompressive, designed to remove what is considered to be a constrictive force from the cerebellum, medulla oblongata, and medulla spinalis.

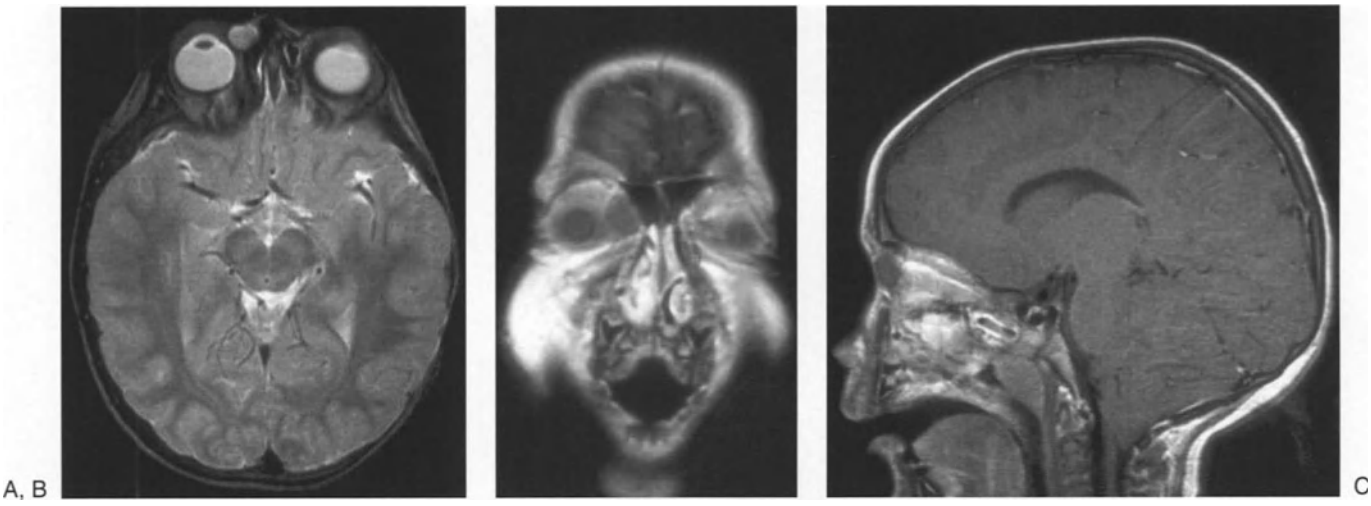


Figure 14.30. This is a very interesting example of combined congenital anomalies: an orbital dermoid tumor and a Chiari I malformation. (A) An axial SE2400/90 image; (B) a coronal SE640/15 post-gadolinium image; and (C) a sagittal SE560/15 post-gadolinium study reveals the orbital dermoid tumor and the Chiari I malformation. The tonsils are displaced well below the rim of the foramen magnum. There is a round cystic lesion in the superomedial margin of the right orbit, extraconal in location, which has a medium signal intensity on the T2 weighted image and which does not enhance.

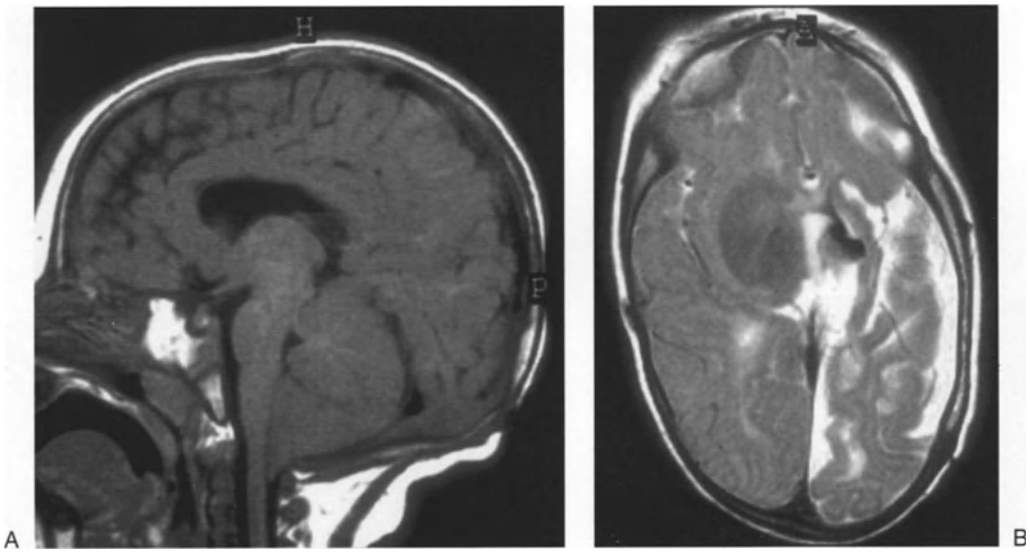


Figure 14.31. Left hemisphere hypoplasia. This is another example of combined congenital anomalies of the central nervous system: hypoplasia of the left cerebral hemisphere and the Chiari II malformation in a child with meningomyelocele. The sagittal section (A) is an SE500/15 image and the axial section (B) is a 3500/90 image.

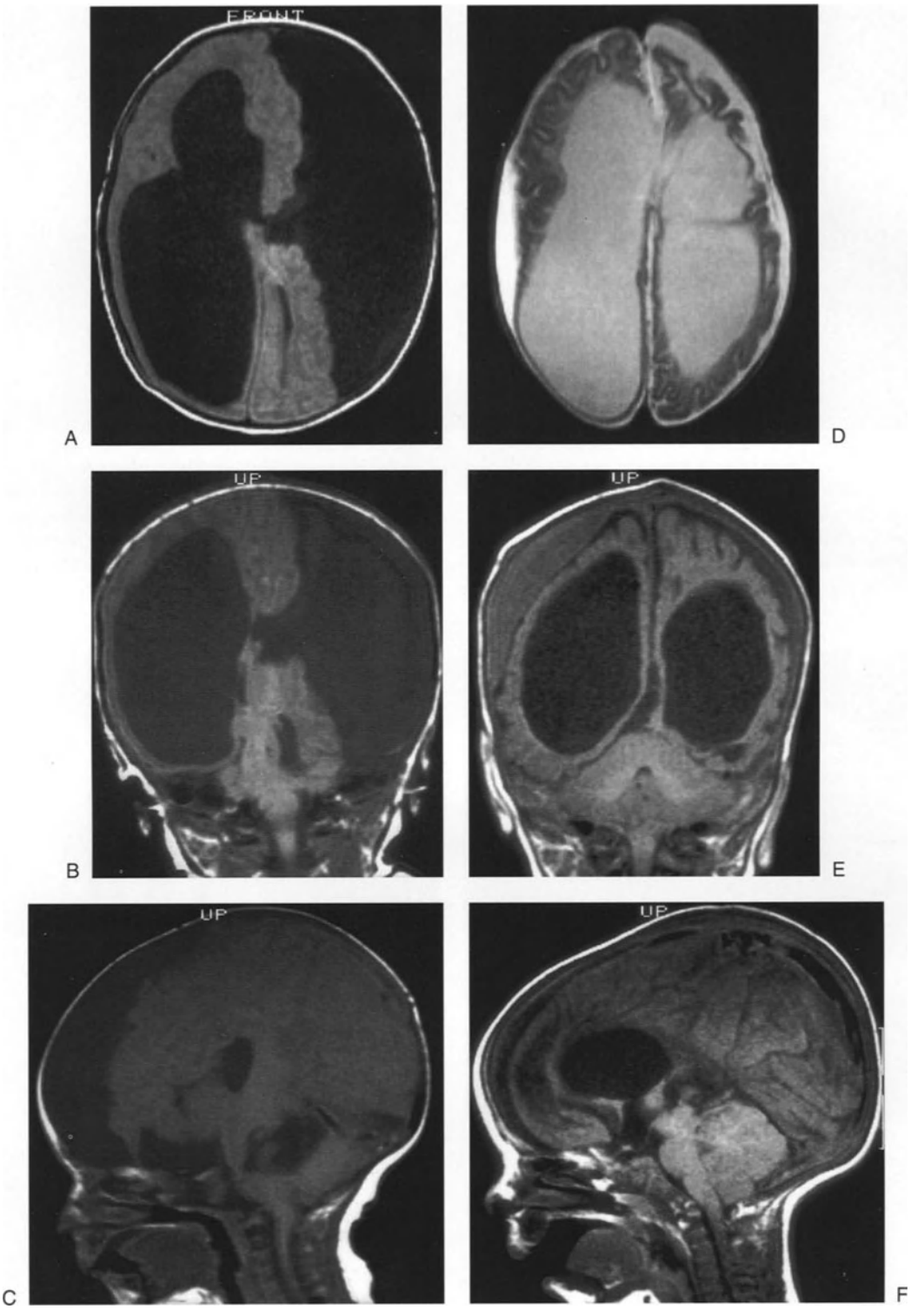


Figure 14.32. Congenital ventricular dysmorphism associated with aqueductal stenosis. Pre- and postshunt studies. (A) An axial SE700/20 image, (B) a coronal SE750/22 image, and (C) a sagittal SE650/20 image are the preshunt studies. These were all done in the patient at 5 days of age. They reveal a left extra-axial CSF collection compressing the underlying hemisphere and communicating with the right lateral ventricle which, in turn, is enlarged. The IV ventricle is not visualized. In the axial (D) SE2400/90, coronal (E) SE560/5, and sagittal (F) SE560/15 postshunt studies one sees that the left hemisphere is no longer compressed by the extra-axial mass, the ventricles remain large but have now assumed normal morphology, and that there is a subdural collection over the right parietal hemisphere which is probably secondary to the shunt. Though the IV ventricle is now visible, the aqueduct of Sylvius remains occluded.

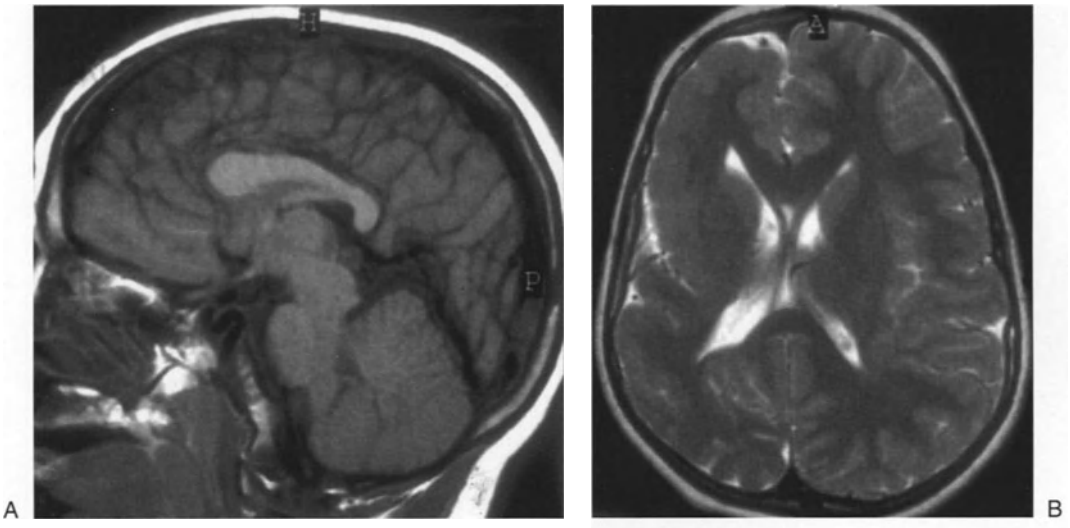
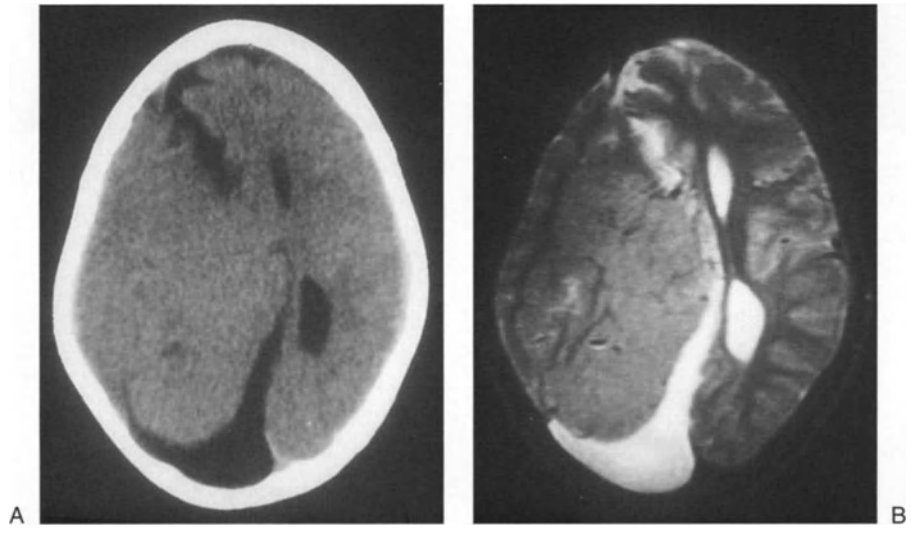


Figure 14.33. Migration anomaly with right cerebral hemisphere hypoplasia. The sagittal (A) SE600/20, axial (B), and (C) TSE3000/90 images reveal a hypoplasic right hemisphere which has thickened cortex and flattened sulci, a dilated right lateral ventricle, and hypoplastic right cerebral peduncle, and a dysgenetic corpus callosum with an absent rostrum but an enlarged genu.

Figure 14.34. Agenesis of the corpus callosum and migration defect. These [T1 (A) and T2 (B) weighted MRI] images reveal the very characteristic changes of migration defect in the right cerebral hemisphere with an apparently normal, though markedly compressed, left hemisphere.



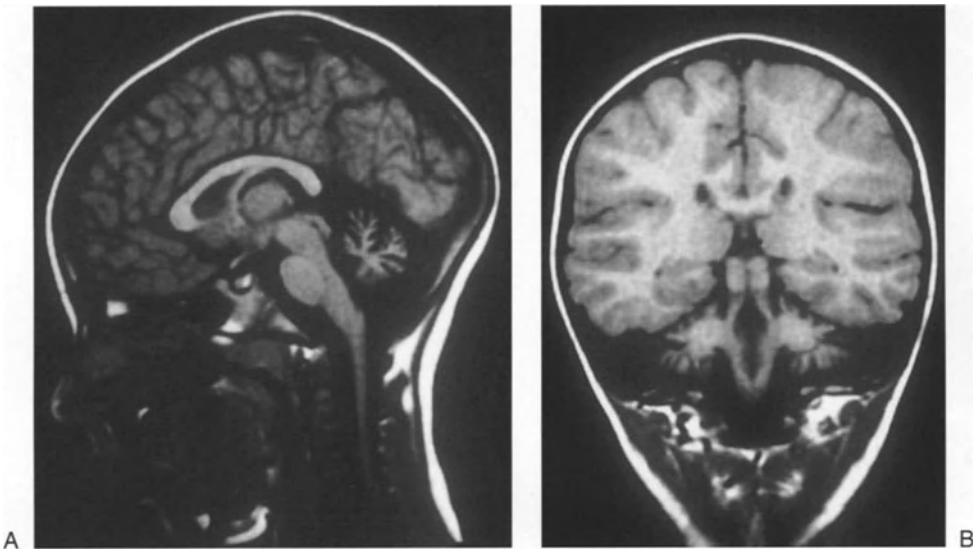


Figure 14.35. This T1 weighted MRI study in the sagittal (A) and coronal (B) sections reveals a rather remarkable degree of congenital cerebellar atrophy. It is interesting to note that by the time this child arrived at 7 years of age she had compensated very well for the truncal ataxia, learning to walk by looking at the horizon.

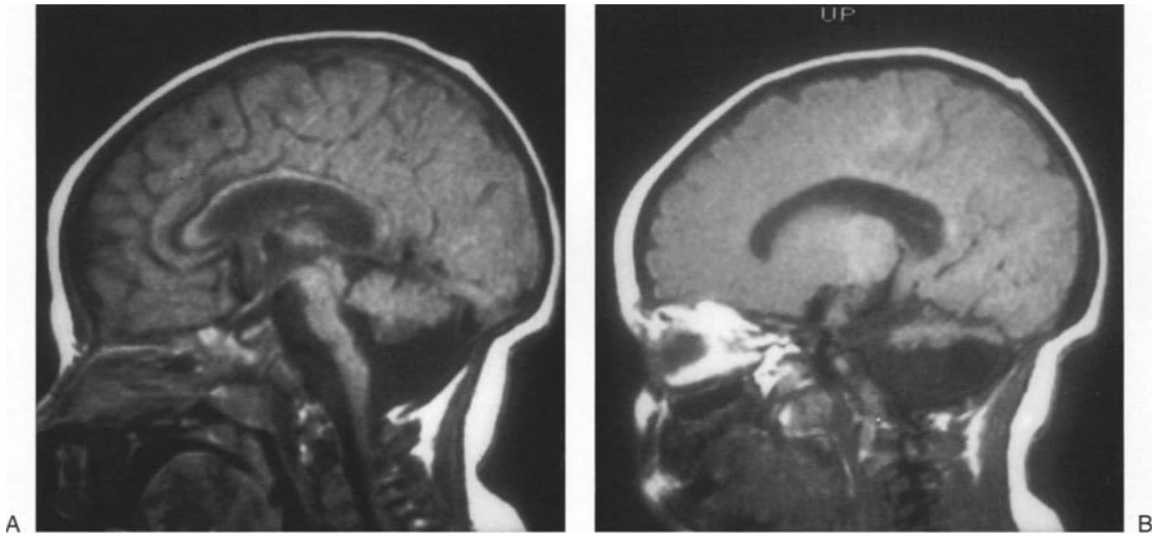


Figure 14.36. Hypoplasia of the cerebellar vermis and hemispheres. These sagittal (A) and parasagittal (B) MRI T1 weighted images reveal the cerebellar hypoplasia and apparently dilated cisterna magna.

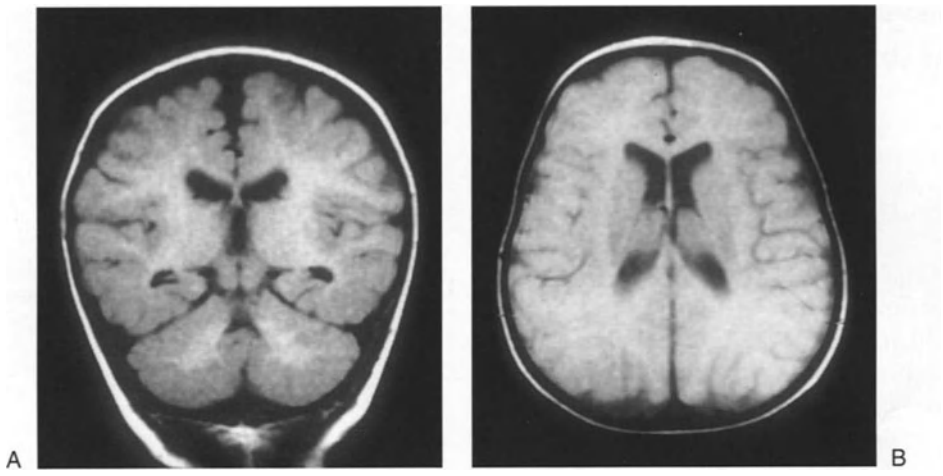


Figure 14.37. Megacephaly with minimal ventricular dilation. This T1 weighted MRI imaging study of the coronal (A) and axial (B) slices reveals the overall increase in cerebral and cerebellar bulk, gray and white matter, plus minimal ventriculomegaly.

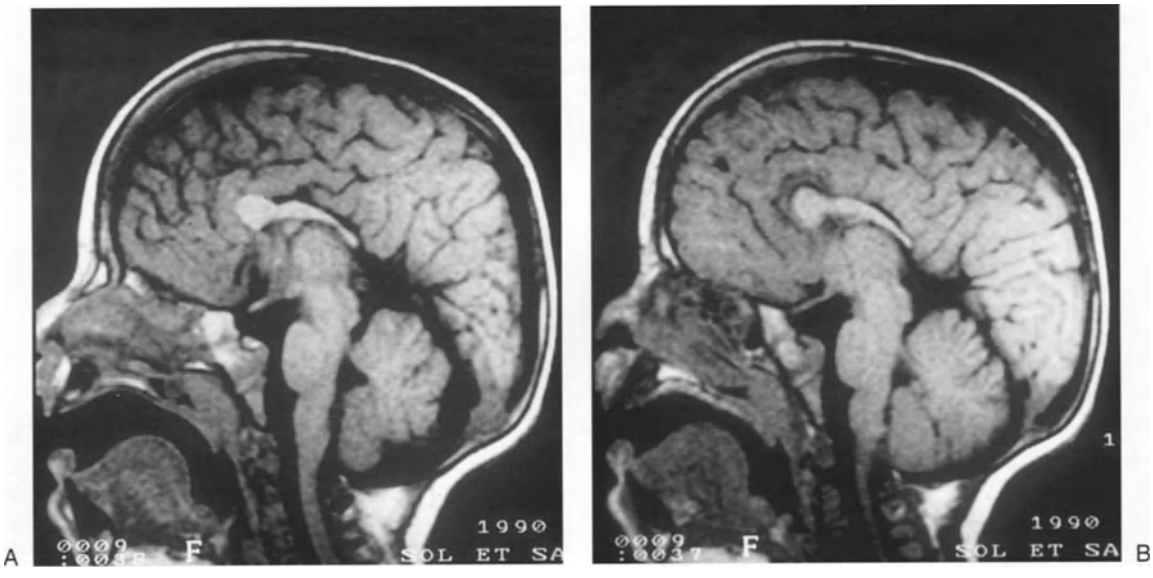


Figure 14.38. Dysgenesis of the corpus callosum. These (A) sagittal and (B) parasagittal T1 weighted images reveal aplasia of the splenium of the corpus callosum.

Chiari I Malformation (Figs. 14.39–14.46)

The Chiari I malformation is not invariably a malformation! It is at times an anatomical variant, present in a small percentage of human beings (children of all ages, and adults), who suffer neither cerebellar nor medullary compression, who have neither hydrocephalus nor syringomyelia. At other times, it is a real pathologic entity resulting from incarceration of the cerebellar tonsils within the cisterna magna, hind-brain compression, associated hydrocephalus and hydrosyringomyelia. Its diagnosis is by either MRI or CT. It may, consequently, be encountered in normal and asymptomatic children as well as an individual who suffers from hydrocephalus and syringomyelia.

By definition, a Chiari I anatomical variant is a neuroradiological diagnosis characterized by location of the caudal loop of the lateral medullary segment of the posterior inferior cerebellar artery (PICA), or the tonsils, inferior to the horizontal plane of the foramen magnum. However, when this neuroradiological picture is present in a child with symptoms and/or signs of cerebellar impaction, hydrocephalus, dissociated sensory deficit with muscle weakness, and atrophy, it is pathological and pathogenetic. A shunt, posterior fossa decompression with duraplasty around the cisterna magna, and/or at times resection of the tonsils may be indicated in these children.

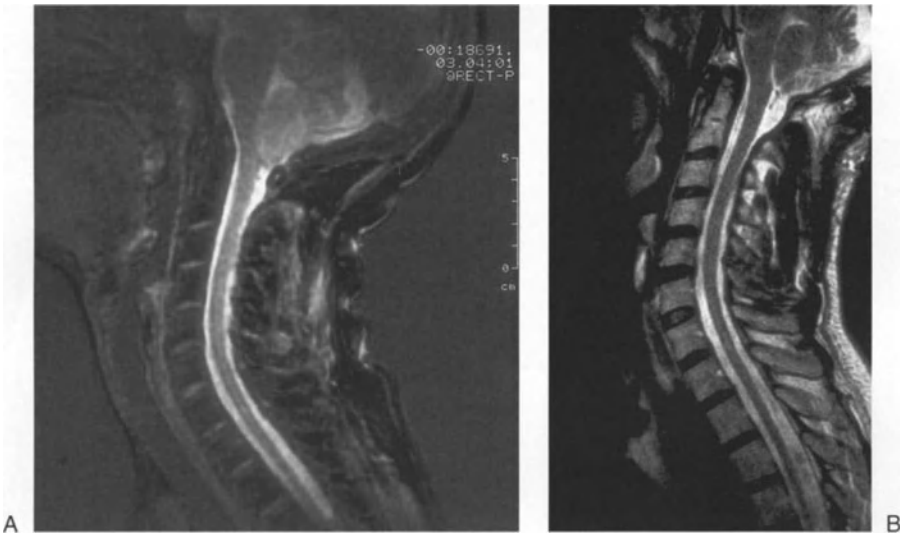


Figure 14.39. Chiari I malformation. This MRI study reveals (A) in the T2 weighted image that the cerebellar tonsil bulges beneath the foramen magnum, pushing into the medulla oblongata and upper medulla spinalis. There is no foramen magnum visible beneath the inferior surface of the cerebellar hemispheres, but a very small one posteriorly located. This patient presented with nuchal rigidity, episodes of loss of consciousness, persistent headache, hoarseness, and papilledema. In (B) a T2 weighted image performed 1 week after suboccipital craniectomy (without opening of the dura) and resection of the arch of C1, one sees an expansion of the subarachnoid space posterior to the upper cervical cord, return of the tonsils into the posterior fossa, and an increase in the volume of CSF along the posterior and inferior surfaces of the cerebellum. It should be noted that this patient did not have syringomyelia.

►

Figure 14.41. Chiari I malformation in the nonoperated state. In (A), a sagittal SE600/20 image, one sees the cerebellar tonsils in an extremely descended position: C2–3. In (B) a sagittal turbo SE3500/120 image of the cervicothoracic spine, one sees a bisegmental syrinx: C5–T2 and then T3–T5.

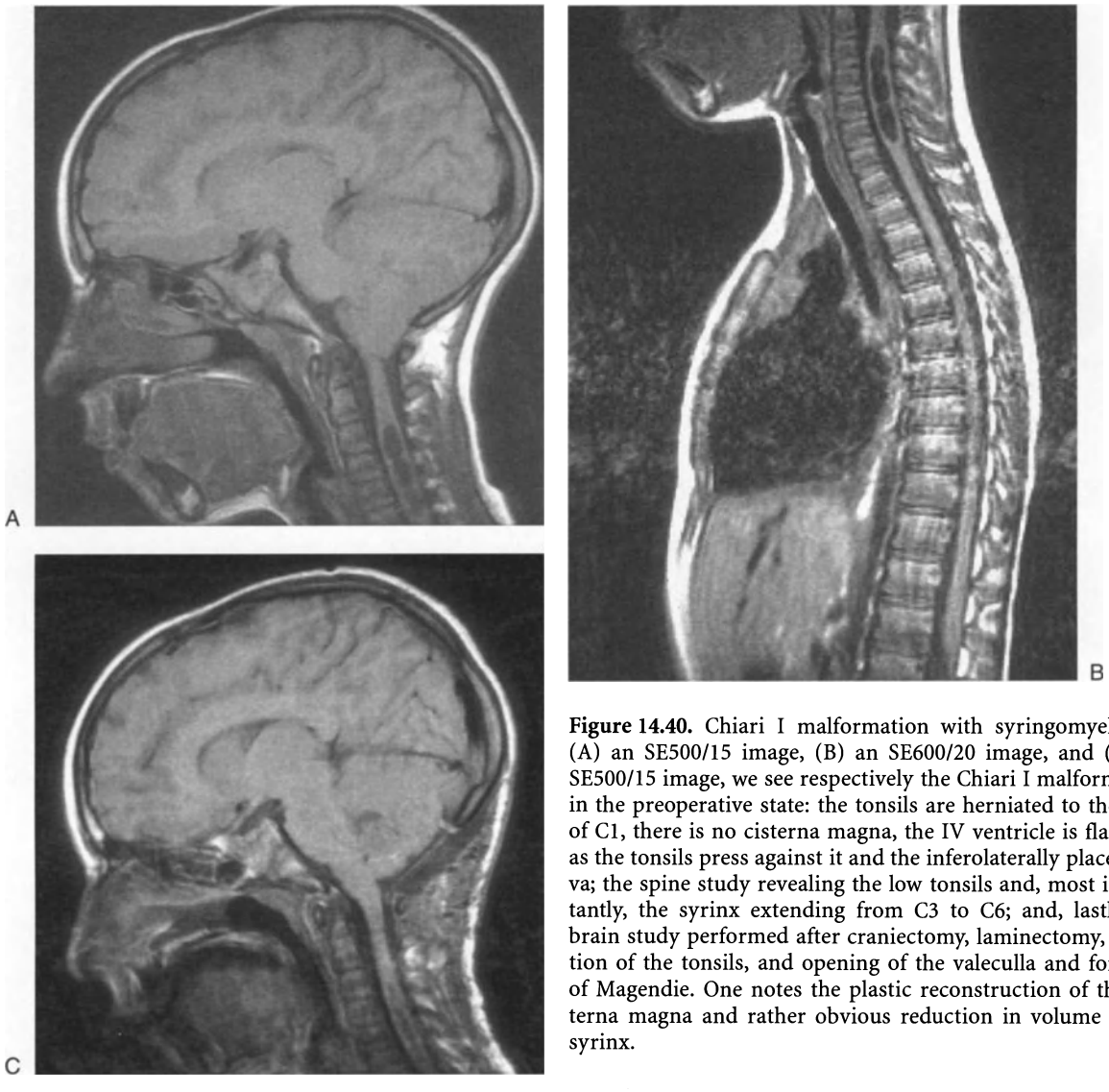


Figure 14.40. Chiari I malformation with syringomyelia. In (A) an SE500/15 image, (B) an SE600/20 image, and (C) an SE500/15 image, we see respectively the Chiari I malformation in the preoperative state: the tonsils are herniated to the level of C1, there is no cisterna magna, the IV ventricle is flattened as the tonsils press against it and the inferolaterally placed clava; the spine study revealing the low tonsils and, most importantly, the syrinx extending from C3 to C6; and, lastly, the brain study performed after craniectomy, laminectomy, resection of the tonsils, and opening of the valeculla and foramen of Magendie. One notes the plastic reconstruction of the cisterna magna and rather obvious reduction in volume of the syrinx.

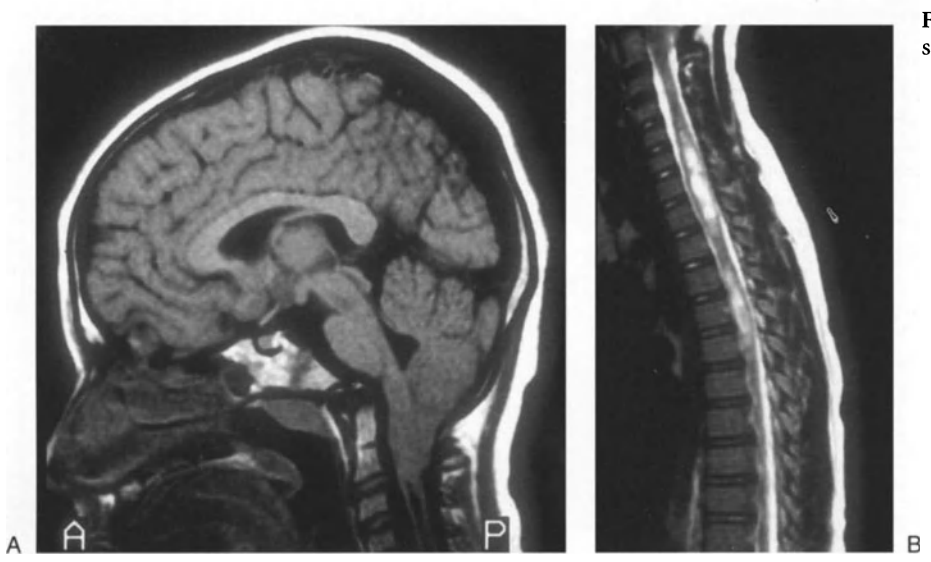


Figure 14.41. Legend see p. 498.

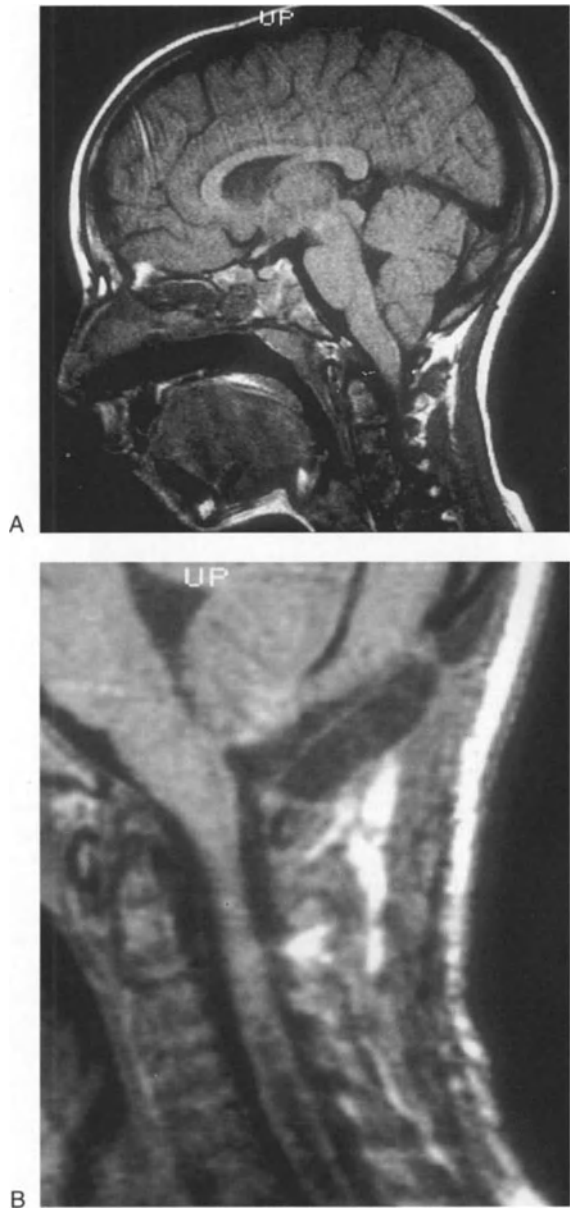


Figure 14.42. Chiari I malformation – normal and postoperative. In (A), a sagittal T1 weighted image, one sees the preoperative condition of the symptomatic Chiari I malformation with the tonsils located below the rim of the foramen magnum and compressing the medullospinal junction. Note that there is no cisterna magna beneath the tonsils. (B) The postoperative state, however (2 weeks after suboccipital craniectomy, resection of the arch of C1 opening widely of the dura, resection of the cerebellar tonsils and sewing them superolaterally, opening of the vallecula, and plastic reconstruction of the dura) reveals the pseudo cisterna magna created by surgery and free passage from the IV ventricle through the foramina of Magendie and into the pseudo-cisterna magna. Note that the syrinx is very small.

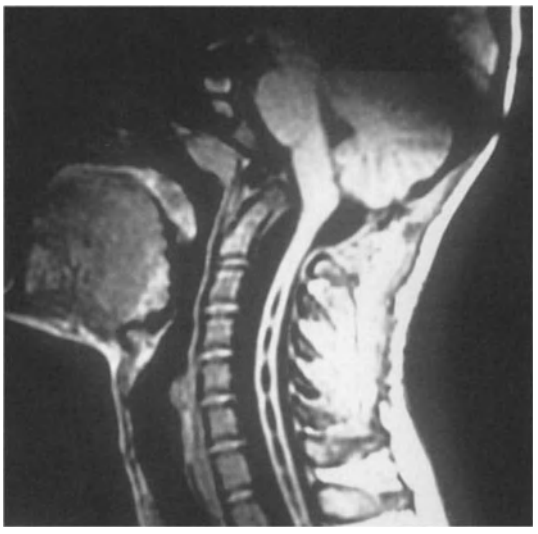
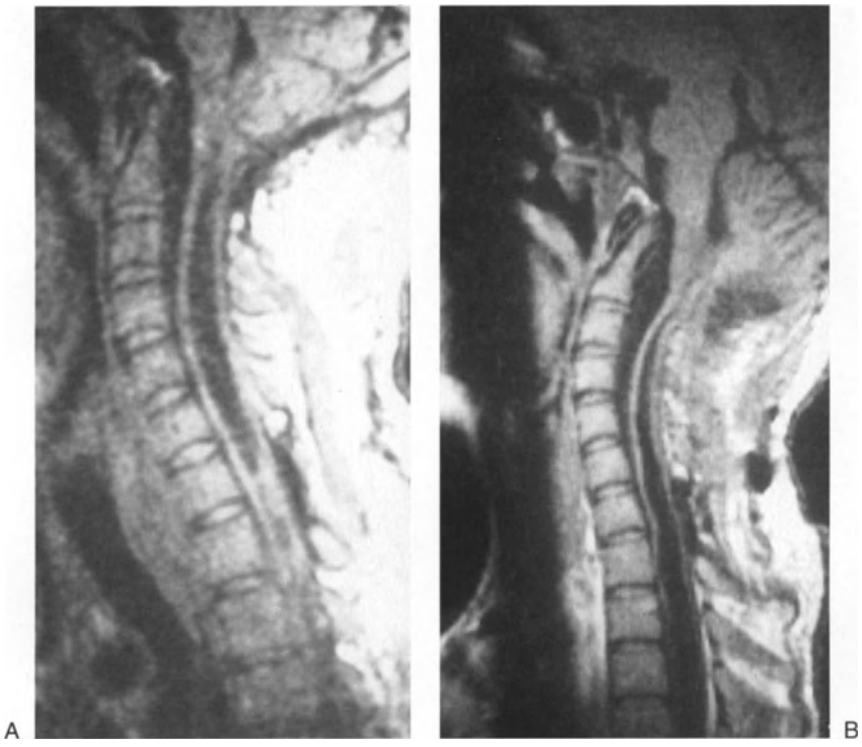


Figure 14.43. Chiari I malformation – 1 year after craniectomy/resection of the arch of C1/resection of the tonsils and opening of the vallecula/duroplastic reconstruction of a pseudo-cisterna magna. This child's clinical picture resolved completely, and she assumed fully normal activies. One notes persistence of the pseudo-cisterna magna, which very probably acts as a buffer for absorbing the cerebrospinal fluid pulsations as they jet into the region of the foramen magnum. One also notes persistence of hydromyelic compartments: a very common observation in postoperative patients.



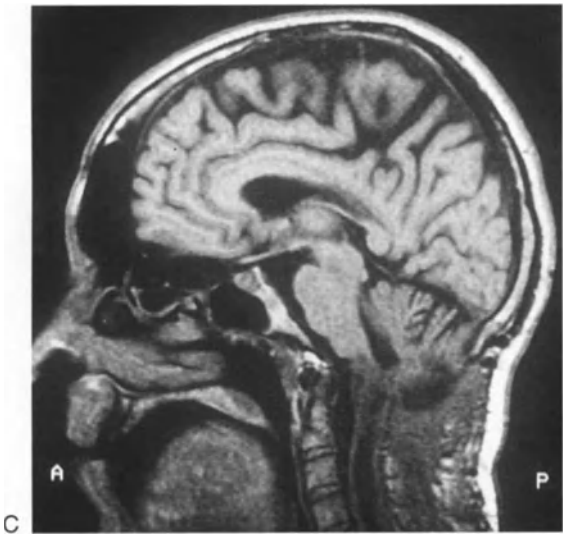


Figure 14.44. Syringomyelia – in a 30-year-old man. This patient, a restaurant owner and waiter, had suffered symptoms and signs characteristic of the Chiari I malformation and cervical syringomyelia since the age of 6 years. These were slowly progressive until 3 years before this study, when they became incapacitating. The patient at this age is included in this book just for the purposes of putting into relief the developmental characteristics of the Chiari I malformation and, more importantly here, syringomyelia. In (A) a sagittal SE360/20 study, one sees that the tonsil and bulbomedullary junction have become a single mass of tissue, and (B) that the syringomyelic cavity extends from the bulb to the level of C7. In (C) one is confronted with a paradox: the sagittal T1 weighted image reveals further dilation of the IV ventricle and an ample pseudocisterna magna. However, the syringomyelic cavity has dilated and extended to the medulla oblongata. The paradox is that the patient has continued to improve neurologically and functionally.

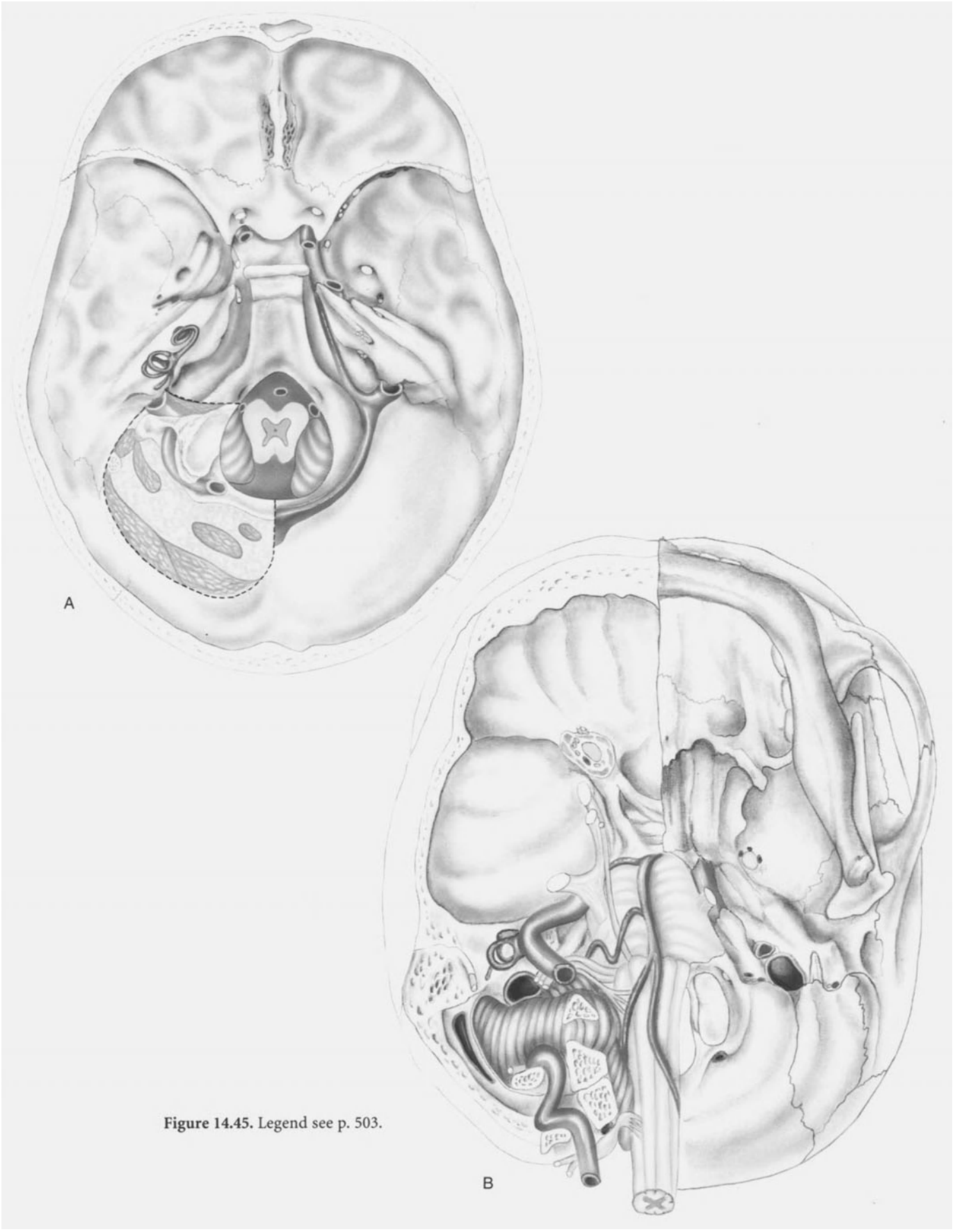
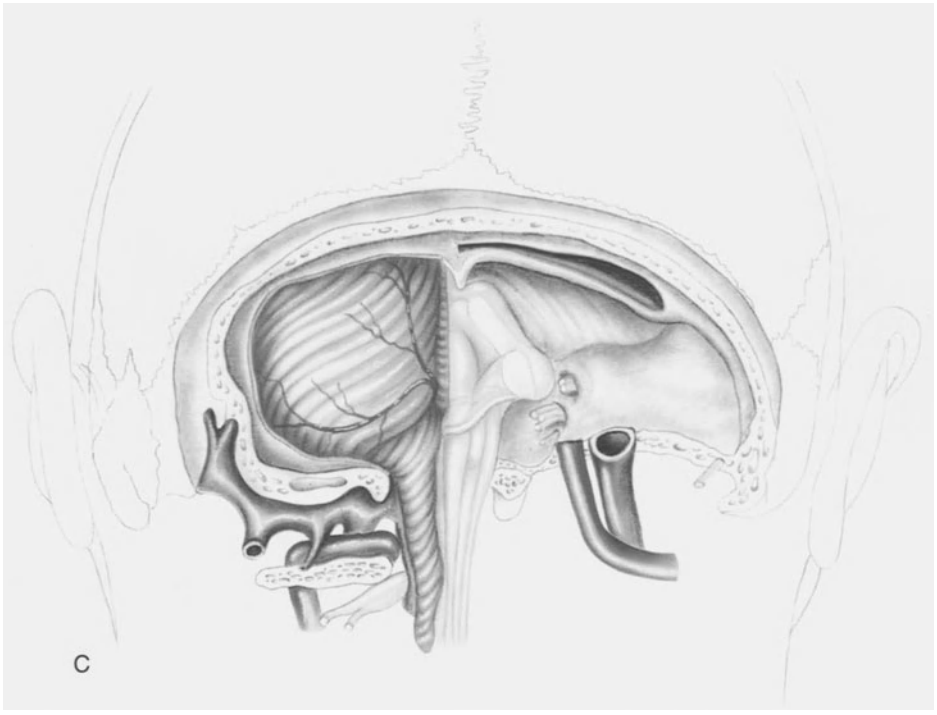


Figure 14.45. Legend see p. 503.



◀ **Figure 14.45.** Having provided examples of the Chiari I malformation with and without such other associated congenital anomalies of the nervous system as dermoid tumors and cerebral dysplasia, it is best now to present diagrammatic drawings illustrating the pathological anatomy of the Chiari I malformation. This is, very probably, one – and surely the most commonly important – of the very few developmental anomalies which are pathogenetic purely because of disturbances in normal anatomical structural interrelationships. Consequently, the precision of surgery (technically and with regard to correcting or eliminating anatomical disproportions is most important. (A) This drawing of the base of the skull is meant to illustrate that in the Chiari I malformation there is no identifiable bony anomaly, *on the left*; but that, *on the right*, an observation very common in the Chiari II malformation, the transverse sinus borders the rim of the foramen magnum. In both the Chiari I and Chiari II malformations, the tonsils descend beneath the rim of the foramen magnum and constrict the lower medulla oblongata and the upper medulla spinalis. *On the right*, the tentorium is not present, a very common event in the Chiari II malformation which does not occur in the Chiari I malformation (where the tentorium is normal). *On the left the broken line* indicates an opening in the squamous occipital bone, permitting visualization of the erector capiti, semispinalis and splenius muscles, as well as the vertebral artery and condyle of C1. Laterally, one notes the internal jugular vein in normal position, as occurs in the Chiari I malformation. *This comparative description of Chiari I and Chiari II anatomical disproportions will be followed throughout this entire series of drawings, represented here in four figures.* (B) This axial artistic representation of the disproportions in anatomical relationships between posterior fossa and upper cervical cord structures is, again, bony skull base *on the right* and skull base with involved parenchymal structures *on the left*. The middle and anterior fossae are not altered but in the posterior fossa of the Chiari II malformation one often finds the condyle in a parasagittal plane and the foramen magnun converted into either a triangular or, as in this case, an oblong structure. The horizontal portion of the squamous occipital bone in the Chiari II malformation is lengthened and the inion is more posterior than normal. Comparing the skull base *on the right* with that *on the left* permits the reader to understand these descriptions. In both Chiari I and Chiari II, much more prominent in Chiari II, the basilar artery is lengthened as are the subarachnoid portions of the vertebral arteries, the inferior portion of the cerebellar hemisphere is jammed against the posterior rim of C1, and the cerebellar tonsils extend for varying distances by insinuating themselves between the posterior arches of the cervical vertebrae and the spinal cord. In this ventral view, one appreciates that the tonsil may also compress the lateral surface of the medulla (olive) and the spinal cord, compressing and stretching the ventral cervical roots. (C) This illustration is to be read with the text of that just described in (B). *On the right* one sees a transverse sinus in the normal position, the three cerebellar peduncules and lateral recess of the IV ventricle normally located, and the obex and clava and gracilis in normal position: the situation in the Chiari I malformation when, as one sees *on the left*, the tonsils are herniated through the foramen magnum, in this case, to approximately the level of C2: not unusual in Chiari I patients. It is not the level of herniation of the tonsil which distinguishes Chiari I from Chiari II, but the structures which are herniated through the foramen magnum (bulb and IV ventricle along with tonsils) which make this distinction.

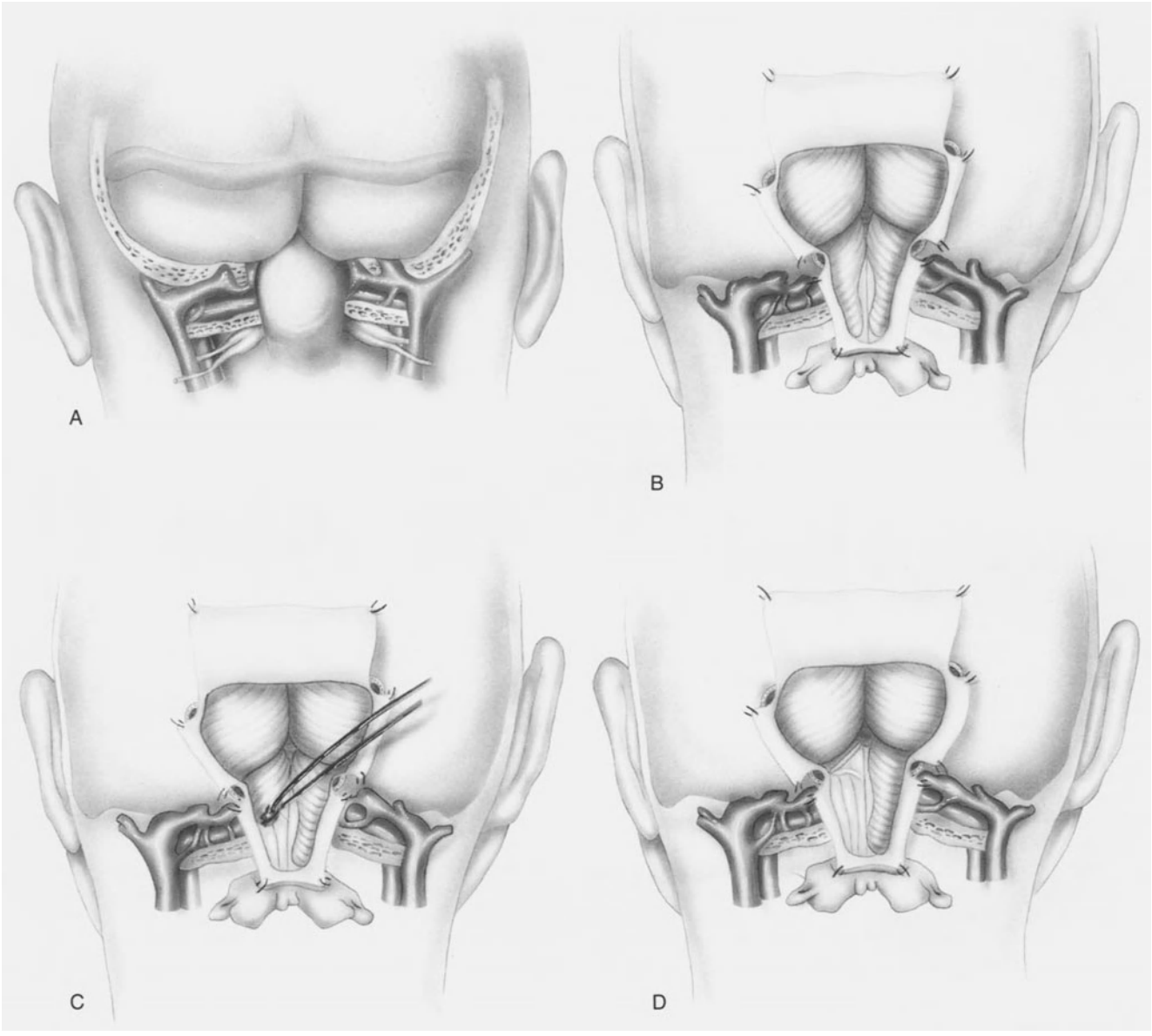


Figure 14.46. (A) This is an artistic representation of the surgical observations in a Chiari I child after the suboccipital craniectomy and resection of the arch of C1 have been performed. The craniectomy was not extended across the line of the transverse sinuses and torcula Herophili: they are illustrated here only for the purposes of placing into relief the fact that in Chiari I malformation patients the transverse sinuses are in normal position. The remarkable bulging at the foramen magnum and down to below C1 is expressed in yellow tints, just as they appear to the surgeon, and is expressive of extreme cramping of the bulbomedullary structures caused by the displacement of the tonsils across the line of the foramen magnum and into the cervical canal. (B) The second stage in surgery for Chiari I malformation: the dura has been opened in a "Y" manner and reflected superiorly, laterally, and inferiorly. We prefer to anchor the dura taughtly so as to prevent shriveling. The arch of C1 has been resected and the foramen magnum has been opened as far posterolaterally as the posteromedial extensions of the condyles. One sees the lingular-like extension of the tonsils and interprets their adhesion to the bulbomedullary structures, identifying that the uvula of the cerebellar vermis has occluded the vallecula and foramen of Magendie. *There is no cisterna magna!* (C) The tonsilar resection is carried out slowly, using microbipolar forceps to coagulate the lingular extensions of tonsil starting at the most inferior pole, in such a manner as to turn them posterosuperiorly before activating the coagulation. This causes shrinkage and retraction of the tonsilar tissue from the underlying spinal cord and /or bulb. (D) The resection of tonsilar herniation is completed when the entirety of the tonsil has been resected and brought posterolaterally to the cerebellar hemisphere from which it originated. This frees the inferior portion of the IV ventricle, opens the vallecula, and permits flow of cerebrospinal fluid out of the foramen of Magendie. In this illustration only the tonsil on the right has been resected, the resection on the left has yet to be begun.

Chiari II Malformation (Figs. 14.47–14.62)

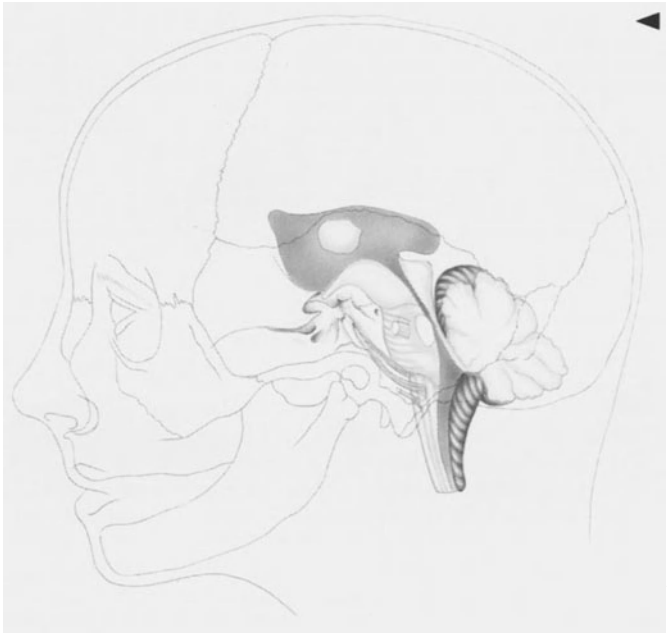


Figure 14.47. This is a drawing of a composite of our experiences with the pathological anatomy of Chiari II children extrapolated from cerebral angiography, CT reconstruct studies, MR images, angio MRI studies, surgical observations, and dissections in the morgue. From superior to inferior one notes the very bizarre III ventricle with a large massa intermedia and the beak of the midbrain resting immediately dorsal to the posterior clinoids: there is almost never a membranous floor of the III ventricle and the interpeduncular cistern is rarely visible. The midbrain is angled anteriorly and buckled moderately at the pontomesencephalic junction. There is beak-like deformity of the collicular plate and a remarkable dilation of the suprapineal recess. A pineal recess is seldom identifiable. The aqueduct is very much elongated, the IV ventricle is also elongated and displaced to the line of the foramen magnum, bordered posteriorly by the herniating tonsils. Ventrally, the pons is elongated and the bulb is either within or below the foramen magnum: VII, VIII, IX, X, and XI are seen coursing superiorly to their exit foramina at the base of the skull. A cisterna magna is seldom visible; the tentorium is almost invariably displastic and very often absent. Not shown in this illustration, the transverse sinuses, functionally at least, are located at the rim of the foramen magnum.

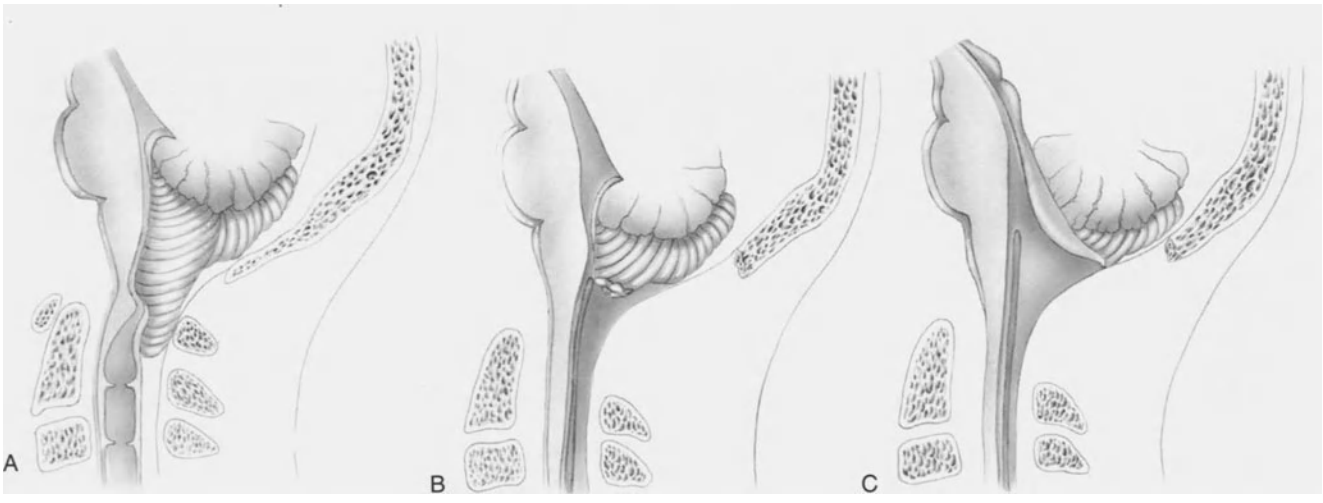


Figure 14.48. This illustration should be compared to Fig. 14.47 to appreciate the anatomical interrelationships which permit the distinction between the Chiari II malformation (Fig. 14.47) and the Chiari I malformation (Fig. 14.48). (A) The cerebellar tonsils are herniated across the rim of the foramen magnum, most commonly as in this illustration with their point of origin from the posteroinferomedial surface of the cerebellar hemisphere. The herniation is very variable in length. There is tonsilar compression of the bulb, the bulbomedullary junction, and the superior portion of the spinal cord with dilation of the central canal generally in the form of link-sausages, as illustrated here. Note that there is no cisterna magna at the foramen magnum. (B) In this illustration, the cerebellar tonsil has been coagulated throughout the entirety of its herniated area to the point of its origin from the posteroinferomedial surface of the cerebellar vermis. There is now a subarachnoid space dorsal to the spinal cord, the central canal is open, but a pseudo cisterna magna has not yet been formed. (C) This is a diagrammatic representation of the pseudocisterna magna which forms after the duroplastic closure has been completed. Since fascia lata, never dural substitutes of any kind, is used, the healing is complete and the closure invariably watertight: two essentials for success of this procedure.

The Chiari II malformation is an extraordinarily complex and variable anomaly, one which is still neither understood nor has an effective surgical treatment or palliation. Attractive though it may be to the neurosurgeon, compression of the neural structures within the posterior fossa and foramen magnum probably is neither the sole nor most significant pathogenic factor in the Chiari II brainstem and upper spinal cord anatomicopathology. Consequently, surgical decompression does not often result in a cure, an arrest, or amelioration of the neurological defects secondary to ischemic cellular changes of the brainstem (motor and sensory V, hind cranial nerve, superior olive, nuclei gracilis and cuneatus, tractus solitarius, reticular formation), or spinal cord nuclei. Similarly, it is not generally followed by disappearance of *the myelomalacia, which is diagnosed clinically in its end stage, as hydromyelia.* The parenchymal changes, gliosis of the optic tracts and geniculate bodies, and aplasia of the periaqueductal gray matter are most probably elements of the primary dysgenesis. The latter often results in nodular hypertrophy of the tectal area, occlusion of the aqueduct, cardiorespiratory irregularities, vocal cord paralysis, and disturbances in diaphragmatic tone and function.

Ischemia, anoxia, and growth of the child are much more likely causes for the progressive changes in function and structure than bony compression. The ischemia results from diminution in arterial lumen and diminished arterial flow secondary to stretching of the vertebrobasilar system and its perforating and circumferential branches on the arterial sides, and from diminished venous drainage secondary to dural sinus anomalies on the venous side. The tissue anoxia is a result of the ischemia. Growth puts greater demands on the neuromuscular structures and further stretches the vessels, closing the vicious circle.

A shunting procedure, when hydrocephalus is present, is not adequate treatment for the symptoms and signs of the Chiari II malformation (resulting from constrictive and distractive forces centering around the brainstem, from the midbrain to the medulla oblongata). This is attested to by the fact that it does not palliate the symptoms and signs of bulbocervical deficits. The shunting only compensates the hydrocephalus. In fact, in many instances the clinical picture (stridor, nuchal rigidity, paresis of the upper extremities, aspiration) does not appear until after a shunting procedure is performed.

Some surgeons have suggested resecting the posterior arches of the upper cervical vertebrae, some craniectomy of the squamous occipital bone immediately around the posterior rim of the foramen magnum, others a combination of the two, and still others a combination of the two with opening of the dura mater over the cervical cord. There is presently unanimity concerning the extreme difficulty and dangers of opening the dura mater subjacent to the squamous occipital bone. It has been postulated that opening the dura over the cervical cord relieves the compressive forces from the herniated cerebellar tonsils and medulla oblongata. However, very little may be accomplished, at great risk, with no reason to expect clinical improvement. There is great controversy over the efficacy of cervical laminectomy and dural opening. I see no justification for this latter procedure.

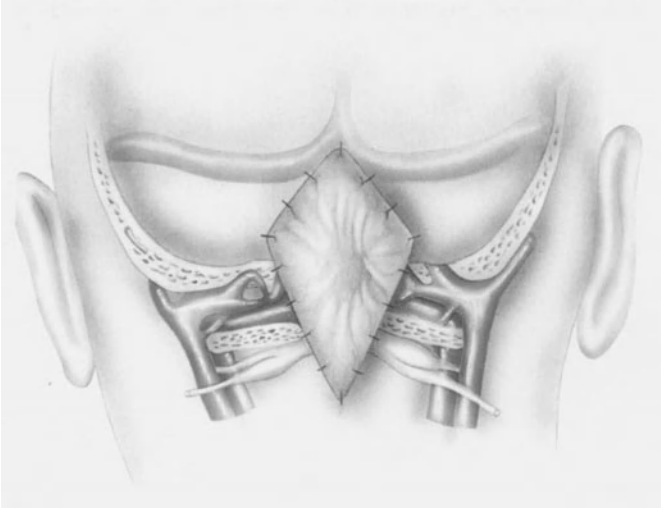


Figure 14.49. Apart from the "artistic enthusiasm" in illustrating sutures brought through the torcula Herophili, transverse sinuses, epidural venous plexus, vertebral artery, etc., one is able to appreciate here the form and surface area of fascia lata graft brought in to create the pseudocisterna magna with the dural closure. This affords an ample compartment in which cerebrospinal fluid may pool to serve as a buffer for the pressure oscillations occurring during systole and diastole.

►

Figure 14.50. The Chiari II malformation. The anatomical descriptions of individual structure deformities and interrelationships between parenchymal and skull structures have already been described. In this figure, MRI images of pre- and postoperative procedures are illustrated. The complexity of the Chiari III malformation is compounded by the almost invariable tethering of the terminal spinal cord and roots to the dura and other mesenchymal structures both before and after operative repair of the meningomyelocele and/or complicating hamartomatous formation; the constrictive process "developing" progressively around the craniovertebral junction, involving cerebral parenchymma, dural sinuses, CSF pathways, epidural veins, hind cranial nerves; and the biventricular (dilation of the III ventricle is rare and the IV ventricle is almost invariably converted into a tubular structure more reminiscent of a lengthened aqueduct than anything else) hydrocephalus with variable parenchymal anomalies such as polymicrogyria, enlarged massa intermedia, and beaking of the collicular plate.

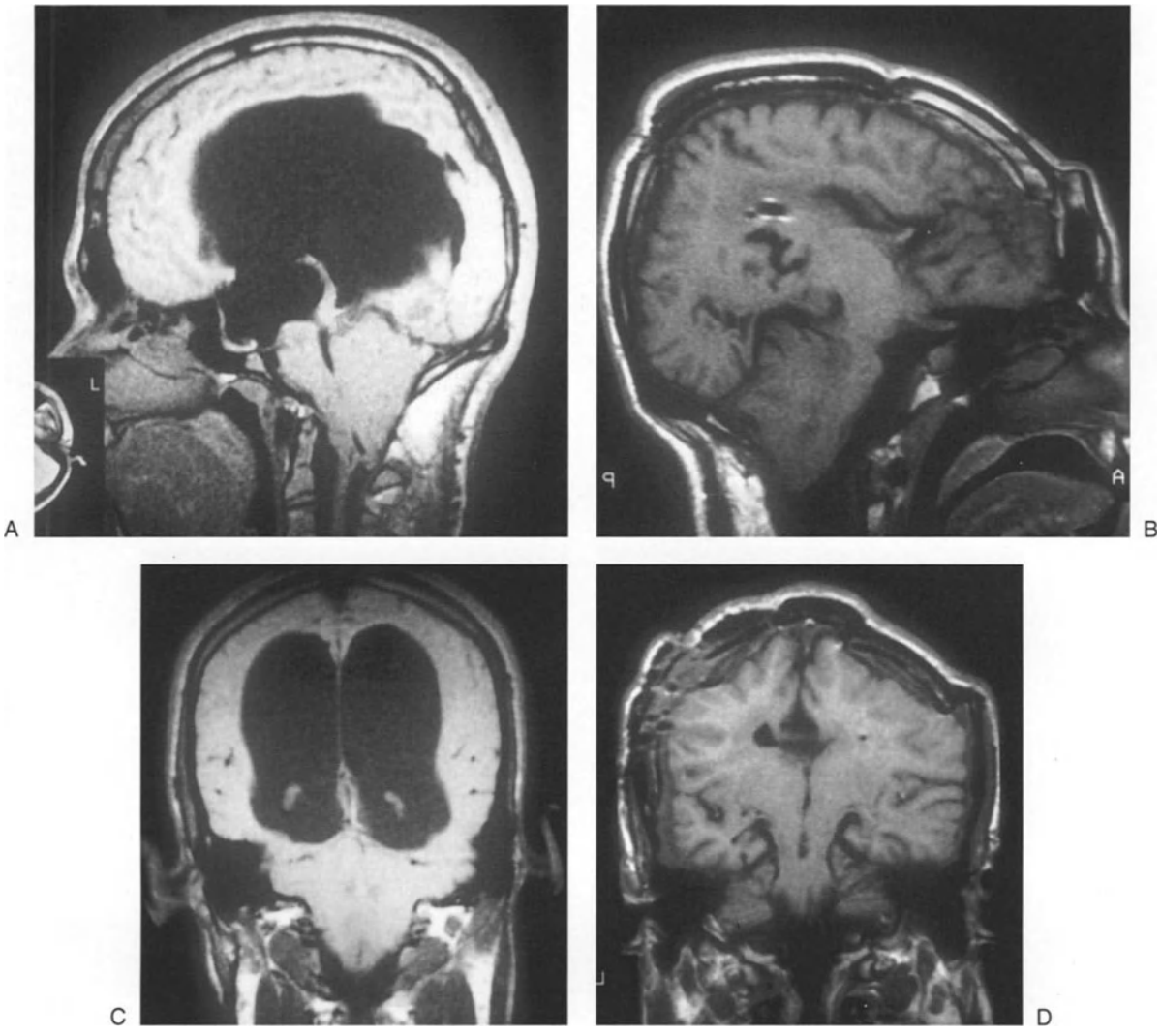


Figure 14.50 (*continued*). Consequently, we have not found that there is any single surgical procedure which may resolve all the problems encountered, nor that there is a single sequence of surgical procedures which may be used. Rather, each child is treated step by step, as problems arise; each adolescent presents different craniocerebral and vertebromedullary problems. (A) This is a 19-year-old boy whose meningomyelocele was repaired at birth and who then carried a ventriculojugular shunt until 16 years of age, suffering all of its complications: repeated failures, shunt nephritis, septic endocarditis, occlusion of both internal cerebral veins and subsequent porta-portal transthoracic collateral flow, etc. The biventricular hydrocephalus, III ventricle almost within the sella turcica, beaking of the midbrain, linear transformation of the IV ventricle, herniation of cerebellum, and medulla oblongata into the cervical cord are all visible. (B) After a suboccipital craniectomy, resection of the arch of C1, resection of the tonsils and opening of the vallecula and foramen magnum, and fascia lata duroplasty creating a pseudocisterna magna, the operation was performed. The boy improved for a few days and then suffered massive bilateral subdural hematomata from parenchymal collapse secondary to cerebrospinal fluid circulation through the normal pathways. In order to preserve the functioning status of the bridging cortical veins and avoid repeated hematomas, a reduction cranioplasty with lowering and advancement of the superior sagittal sinus was performed. Within a matter of 1 week, the boy was awake; within 1 month he was intellectually normal; within 3 months he was ambulating and totally independent in all of his activities. (C) This is a coronal T1 weighted image MRI study performed before the decompression of the posterior fossa and plastic reconstruction of the cisterna magna. One sees the biventricular hydrocephalus. Subsequent to the posterior fossa decompression, the massive bilateral subdural hematomas formed. (D) Here, one may appreciate the presence of a relatively normal ventricular system, opening of the subarachnoid spaces, and circulation of cerebrospinal fluid through the basal cisterns and over the hemispheres, the almost complete resolution of subdurals, and the skin redundancy still present in this very early stage after the reduction cranioplasty and lowering of the superior sagittal sinus. Approximately 1 year after these procedures the skull form was quite acceptable.

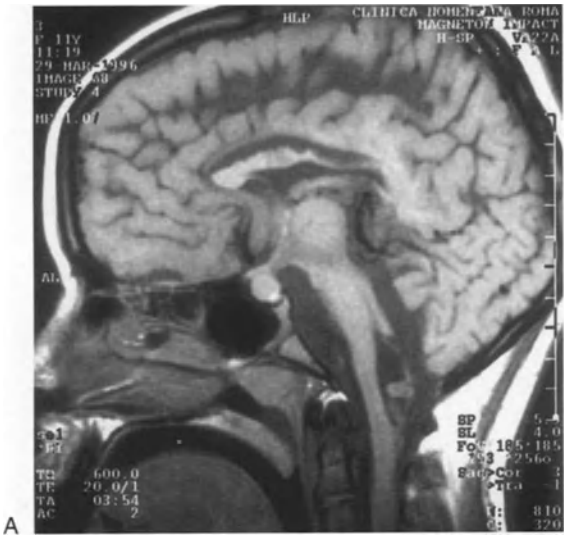


Figure 14.51. Chiari II malformation. This is a most unusual, and instructive case. In (A) a sagittal SE600/20 image, one sees the Chiari II malformation and very severe hypoplasia of the brainstem with the IV ventricle transformed into an ovoid structure and located within the foramen magnum. (B) An axial SE2400/90 image reveals the cerebellum to be severely hypoplastic and the vermis located within the cervical canal at the C3–4 level. (C) Axial TOF2D venous imaging reveals the transverse sinus located at the foramen magnum, and (D) a sagittal SE786/17 image along with (E), a sagittal turbo SE5000/90, and (F), an axial TSE4461/90 image, reveal compression and flattening of the medulla spinalis with a posteriorly located arachnoidal cyst. In (G) a venous angiogram, coronal views, the hypoplasia of the transverse sinus may be appreciated and in (H), a three-dimensional CT reconstruct, we may document the left rotatory deformity producing asymmetry during growth of C2.

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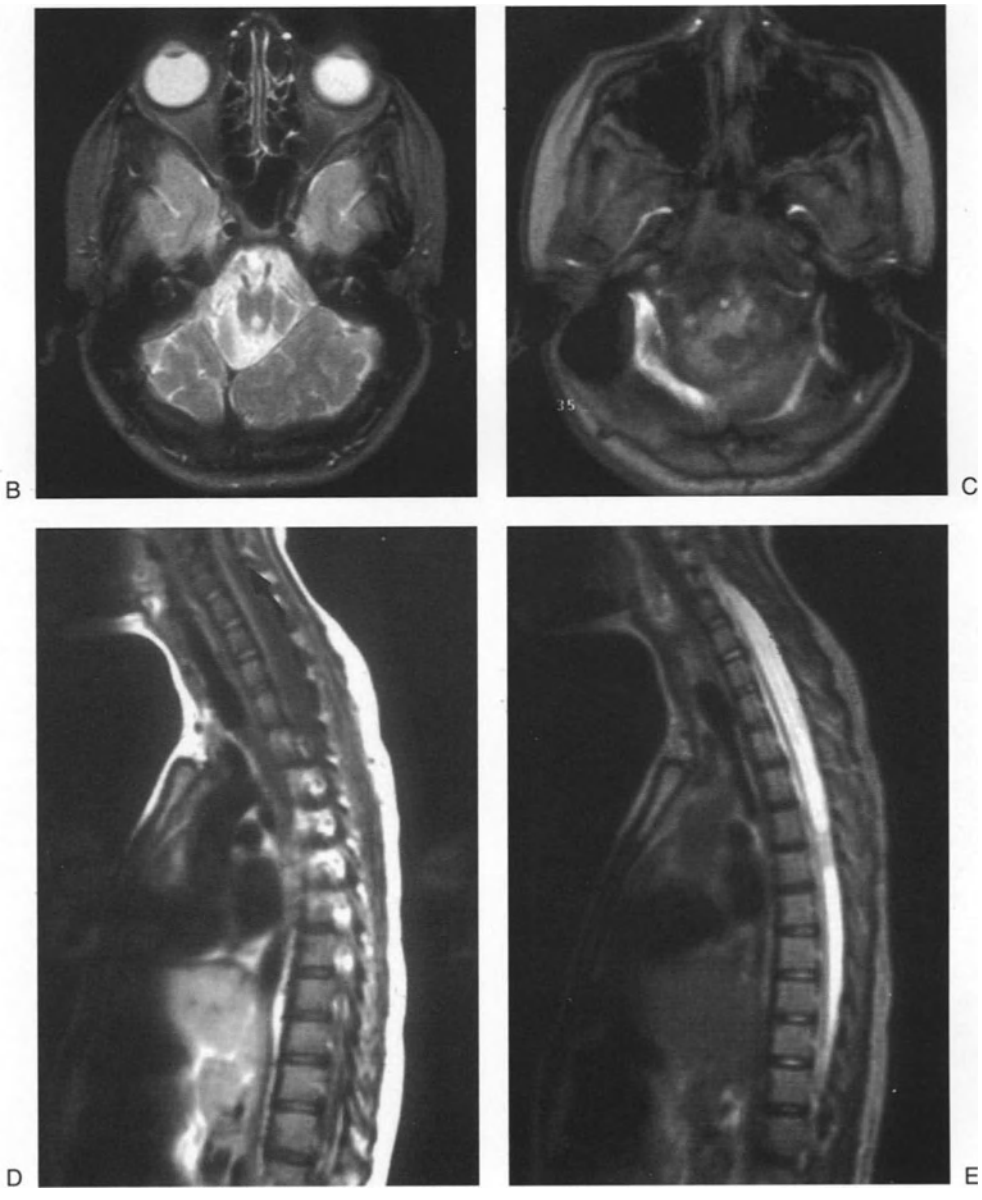
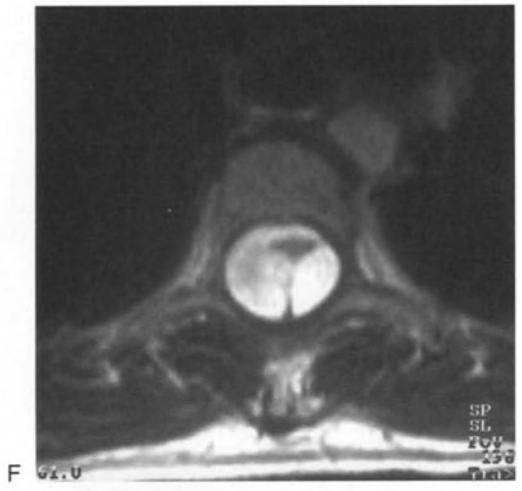
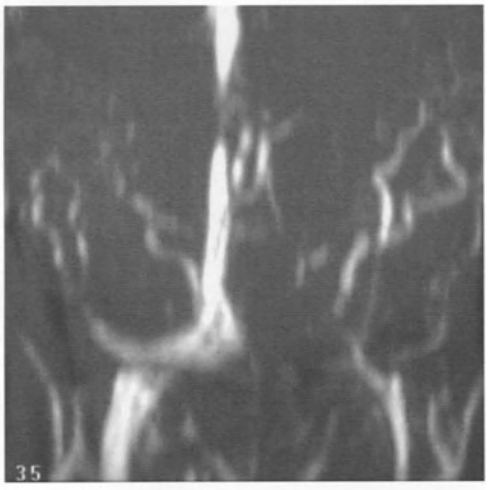






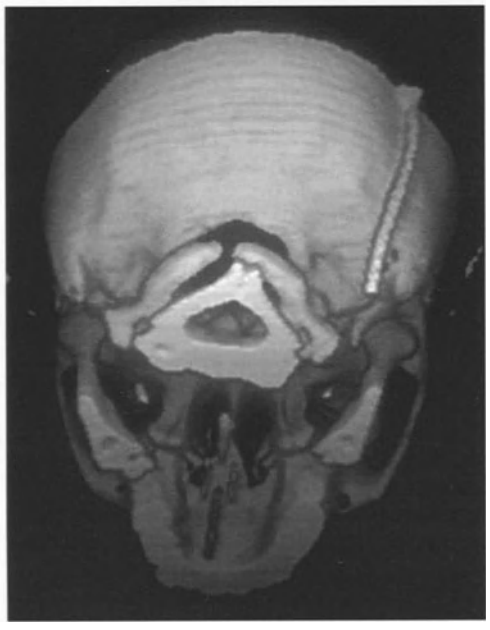
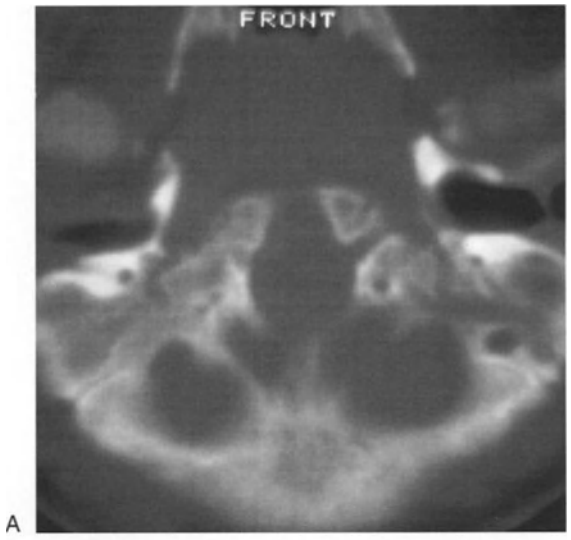


Figure 14.51. Legend see p. 508.



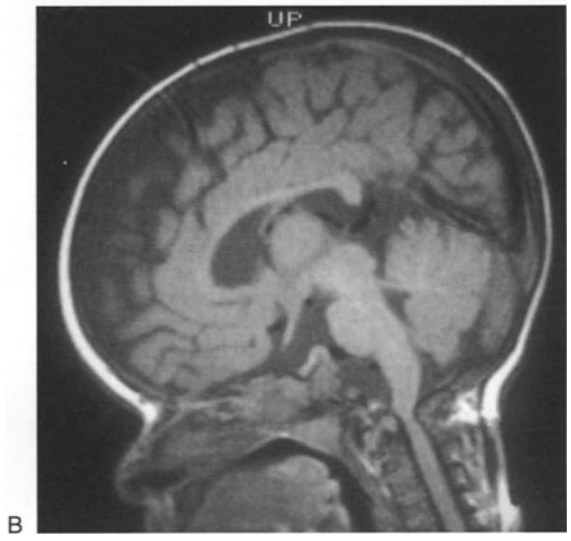


Figure 14.52. Stenosis of the foramen magnum. This clinical entity is related to fibrous dysplasia and causes very different anatomical changes from what one sees in the Chiari I and Chiari II malformations. The pathological problem is simply a stenosed foramen magnum, generally circumferential in form, but most easily approached surgically along the posterior midline. In (A) one has an axial projection of the foramen magnum which permits the appreciation of its remarkable state of stenosis. (B) is a sagittal T1 weighted image which permits the reader to appreciate the fact that the cerebellum is entirely within the posterior fossa, there is a cisterna magna, and that the "strangulation" is of the cervical cord at its uppermost limit and is secondary to the bony stenosis. There is minimal lateral ventricular enlargement, but remarkable accumulation of cerebrospinal fluid within the subarachnoid spaces and the basal cisterns.

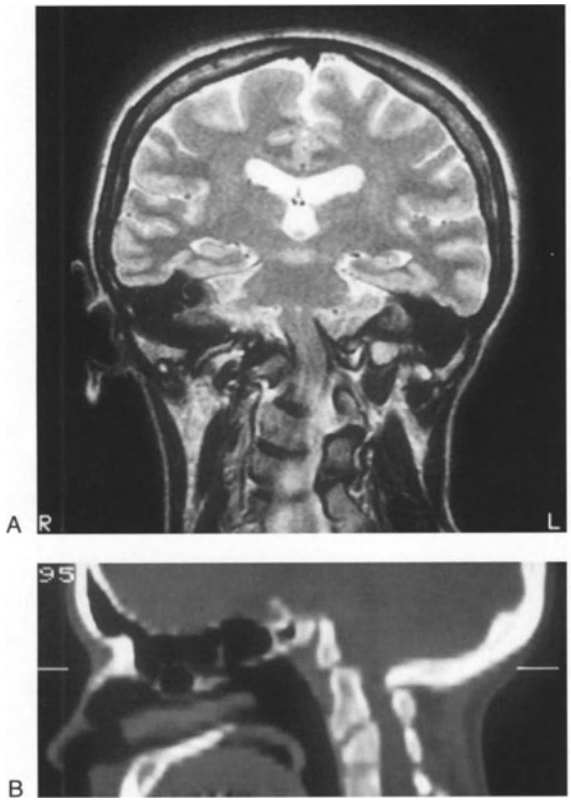


Figure 14.53. Basilar impression. In (A), the coronal T2 weighted image, one sees moderate ventriculomegaly, marked pooling of cerebrospinal fluid in the cortical sulci and curvilinear distortion of the pontomedullary portions of the brainstem. (B) This sagittal CT reconstruct reveals the typical components of basilar impression and the narrowing they impose upon the foramen magnum.

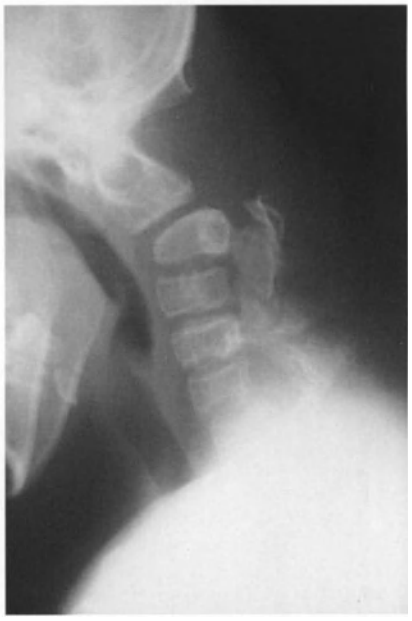


Figure 14.54. Vertebral anomaly. This is another example of congenital instability of the vertebral column, in this case at C2–3 level, which was very probably treated improperly: a fusion was attempted. In children, especially very young ones, stability most often may be attained by prolonged immobilization. Since in this child the posterior arch of C2 was congenitally absent, one does not understand the surgical decision to attempt to obtain fusion by posterior wiring.

If one undertakes to perform a decompressive procedure for treatment of the Chiari II malformation (a procedure we discourage), one should be alerted that (1) the squamous portion of the occipital bones is so deformed as to become an almost horizontally placed structure; (2) the dura over the posteroinferiorly displaced occipital lobes and cerebellar hemispheres represents the parieties of a labyrinth of venous sinuses; (3) the arc of the posterior rim of the foramen magnum that can be craniectomized measures no more than 1 cm; and (4) the dura mater over the inferiorly herniated cerebellar tonsils and medulla oblongata is adherent to the underlying leptomeninges.

The operative procedure for occipital, occipitocervical, or cervical bony (and dural) decompression is performed with the child prone. *Flexing the head on the neck may add to the distracting and constrictive pressures upon the brainstem, so it is safest to keep it neutral.* After the skin incision has been made and the muscles dissected from the squamous occipital bones and the desired posterior cervical arches, self-retaining retractors maintain adequate exposure of the bony structures. Because of the "horizontalization" of the squamous occipital bones, this structure becomes vertical to the horizontal plane when the child is prone. Therefore, the surgeon, working at the head of the patient, has a ledge obstructing his line of vision to the region of the foramen magnum. Lowering the child's head diminishes this somewhat, but never enough to have either an adequate view of the region of the foramen magnum or control of bleeding that may occur within the area. The view of the upper cervical spines, however, is complete.

After the periosteum has been stripped from the squamous occipital bones and the posterior arches of the cervical vertebrae, one may perform the craniectomy and laminectomy most precisely with the use of power driven diamond head burs, extending the craniectomy on each side to the most medial aspect of the condyles of the occipital bone.

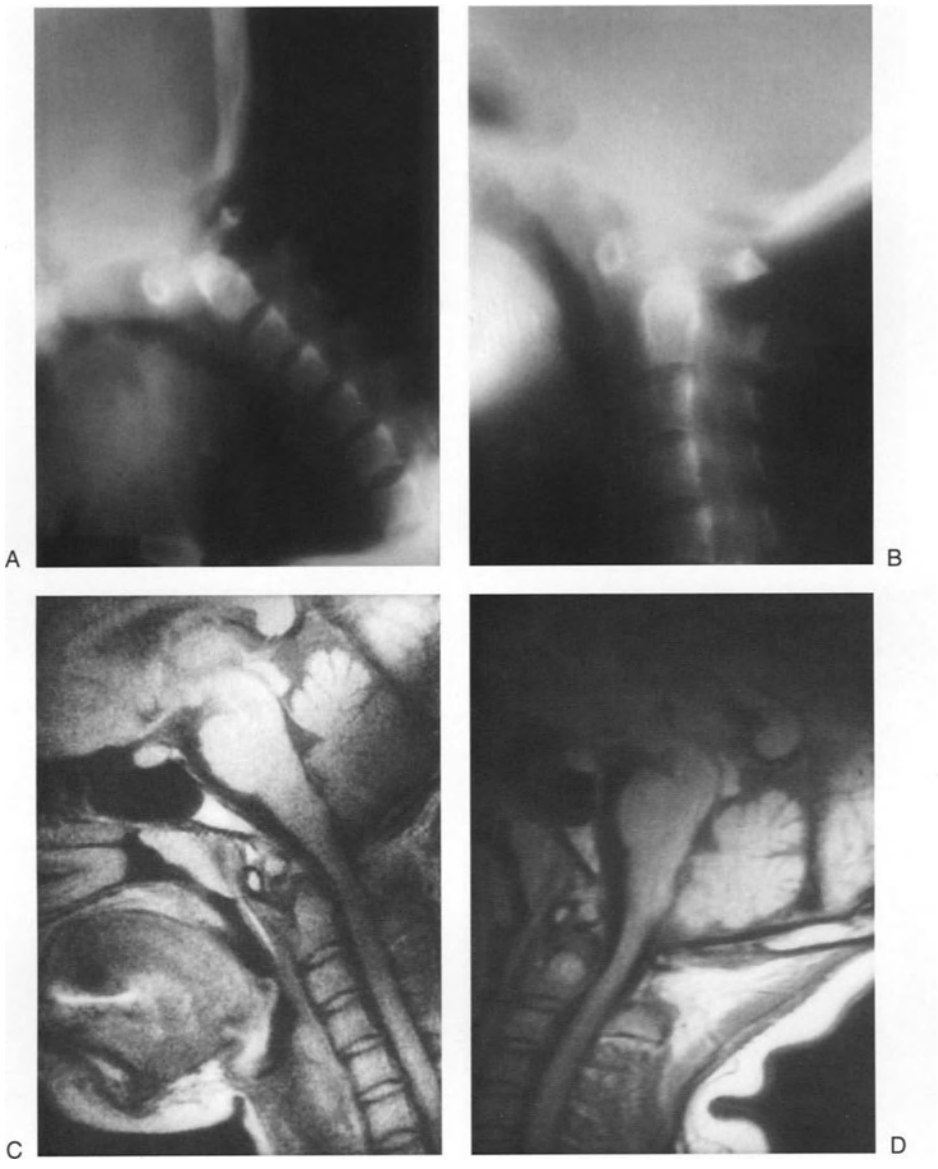


Figure 14.55. Congenital malformation, C1–C2 of ligamentous structures. (A) and (B) are, respectively, flexion and extension tomograms illustrating the abnormal movement of C1 and the skull forward on C2 and the remaining vertebrae during flexion without a full return to normal anatomical relationships (B) during extension. (C) and (D) are T1 weighted image MRI studies during flexion and extension, illustrating decompression deformity at the upper cervical cord during flexion and relaxation of this compression during extension.

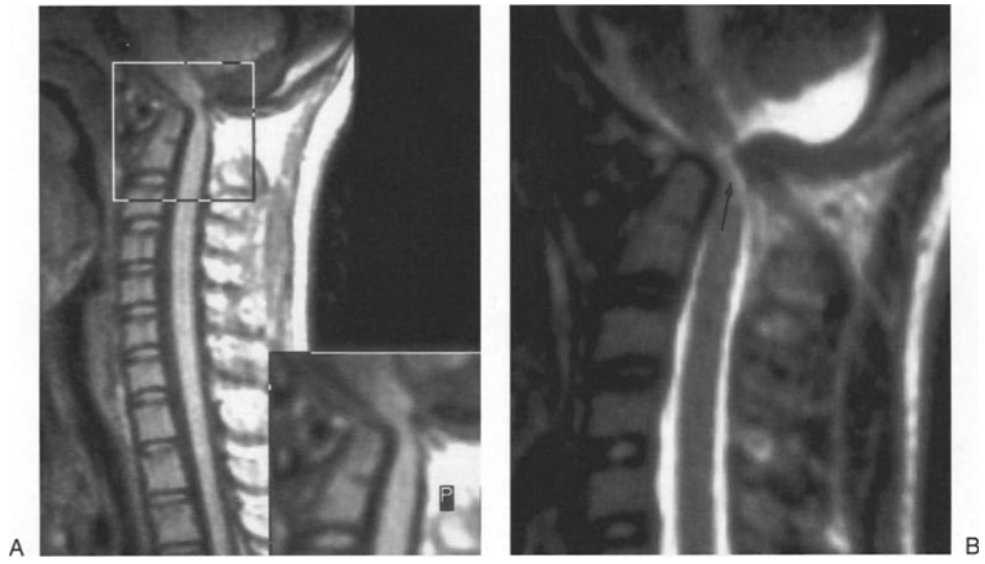


Figure 14.56. Craniovertebral junction anomaly with cord compression in Down's syndrome. In (A) one sees the severe constrictive effects of the mesenchymal dysplasia at the craniovertebral junction. *In the inset*, one is able to identify the major compressive changes at the C1–2 junction, with the dens pushing into the ventral aspect of the cervical cord and the foramen magnum compressing this structure dorsally. (B) Sagittal TSE2857/150 image shows posterior dislocation of the dens with cord compression of the medulla spinalis. In the cord, one sees high signal intensity alteration at the level of compression, secondary to edema (*arrows*).

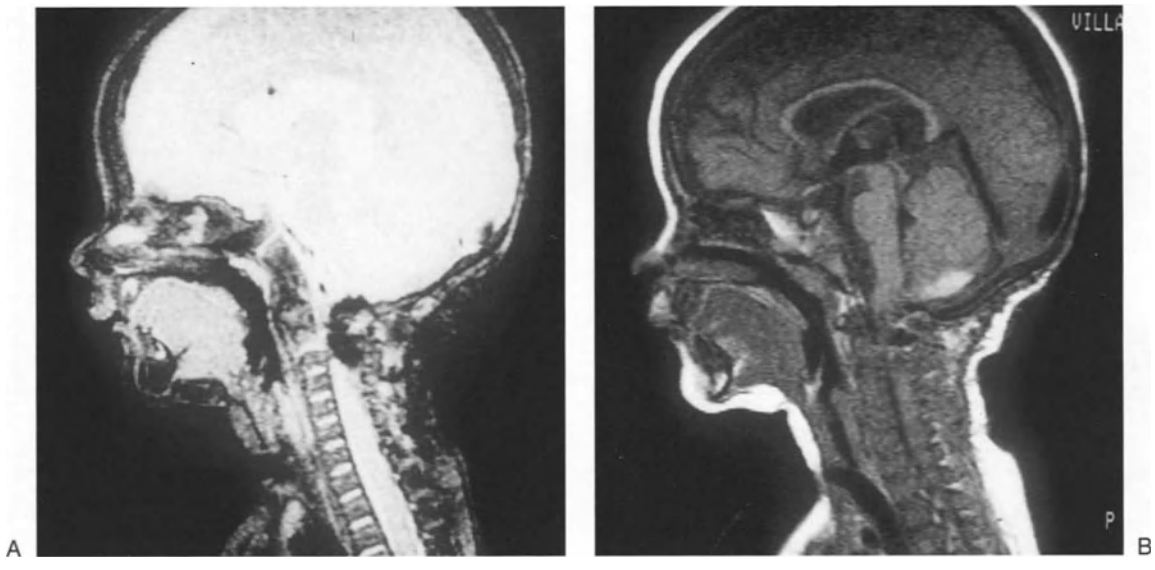


Figure 14. 57. Fracture dislocation C1–C2. This clinical situation, extraordinarily rare (the only example in our experience) is included here, rather than in trauma, since the clinical presentation is similar to that of the other entities heretofore described. (A) One sees the acute effect of the trauma: fracture dislocation of C1–C2, perifracture hematoma, and acute cord compression. (B) In this T1 weighted image one sees hematoma within the posterior fossa and *almost* complete transection of the medulla spinalis. As a result of the craniovertebral trauma this child developed a false aneurysm of the vertebral artery which was repaired surgically.

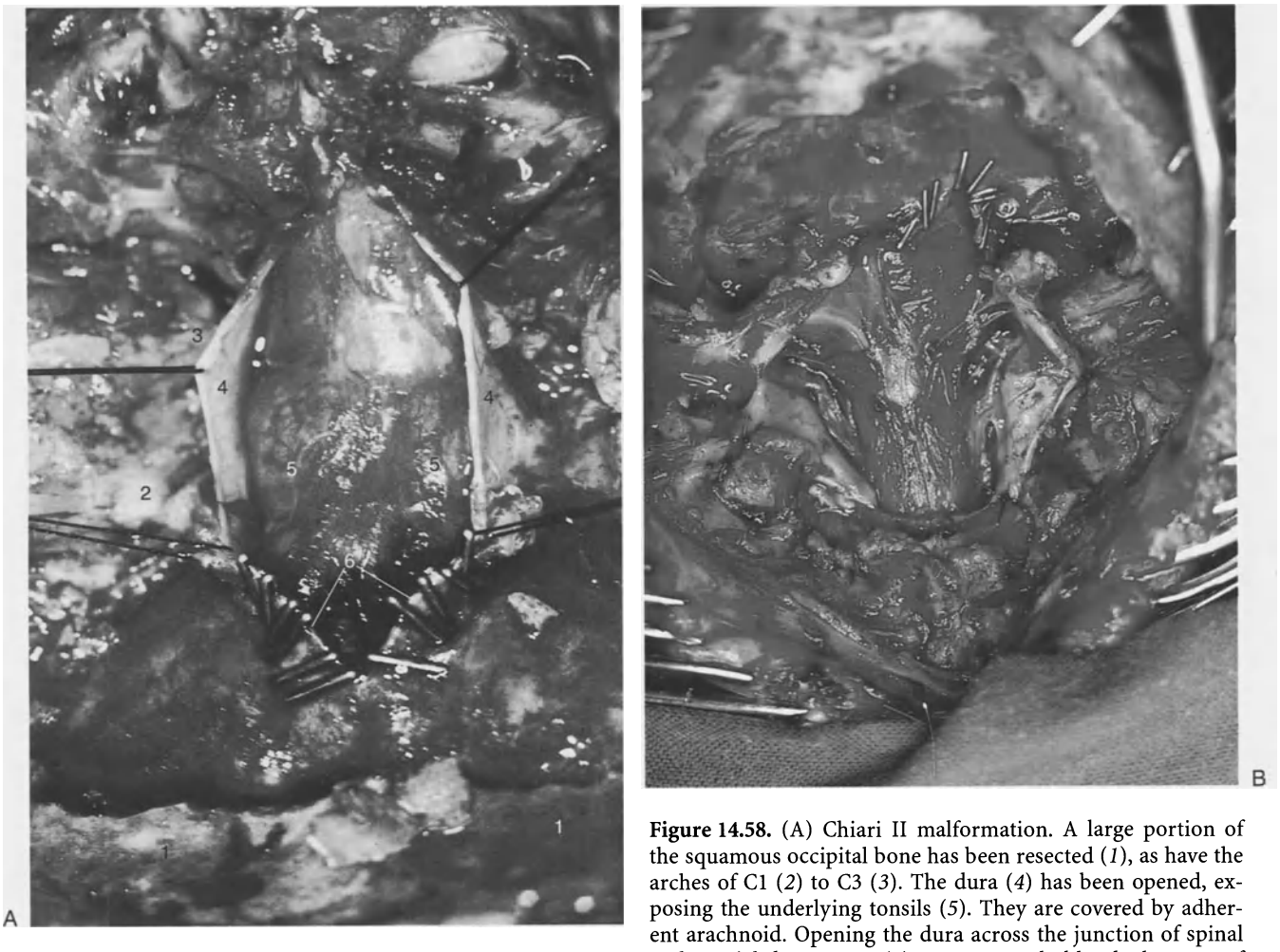


Figure 14.58. (A) Chiari II malformation. A large portion of the squamous occipital bone has been resected (*1*), as have the arches of C1 (*2*) to C3 (*3*). The dura (*4*) has been opened, exposing the underlying tonsils (*5*). They are covered by adherent arachnoid. Opening the dura across the junction of spinal and cranial dura mater (*6*) was extremely bloody, because of the labyrinthine dural sinuses, necessitating application of hemoclips along the entire opening. Cephalad is inferior: the surgeon's view. (B) This is the same child as in (A). Attempts to open the dura from over the cerebellar hemisphere were fraught with severe bleeding, necessitating application of hemoclips and preventing further dural opening. Cephalad is superior.

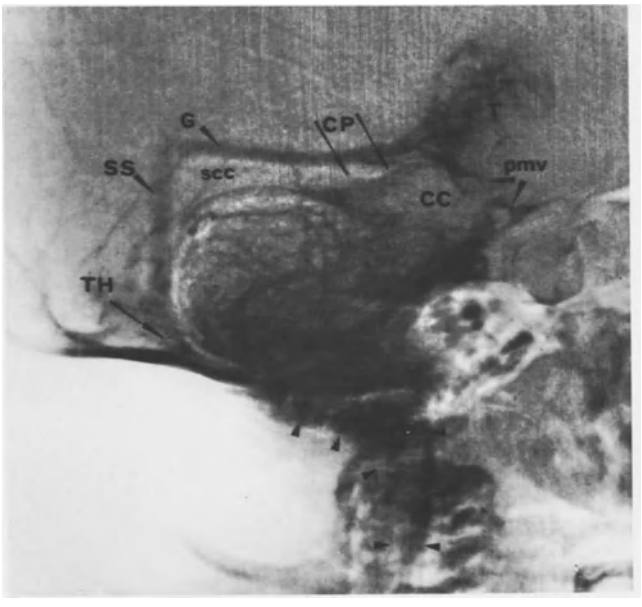


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Figure 14.59. Posterior fossa venographic study of the child illustrated in Fig. 14.43. Note the staining of the posterior two-thirds of the thalamus (*T*), in order to appreciate fully the anteroinferior displacement of the thalamus so typical of aqueductal stenosis. The perimesencephalic vein (*pmv*) is well shown as is the hypertrophy of the crux cerebri (*CC*). The colicular plate (*CP*) outlines the inferior surface of the quadrigeminal cistern. The great vein of Galen (*G*) is stretched, flattened, and horizontalized. It enters the straight sinus (*SS*) at right angles. This represents the sinus changes typical of aqueductal stenosis. The torcular Herophili (*TH*) rests in the same horizontal plane as the horizontalized squamous portion of the occipital bone. The very dramatic herniation of the inferior half of the cerebellar hemispheres and the hemispherical tonsils through the foramen magnum and down to the level of C4 is outlined by the *arrowheads*. The remarkable dilation of the superior cerebellar cistern (*scc*) may be identified.

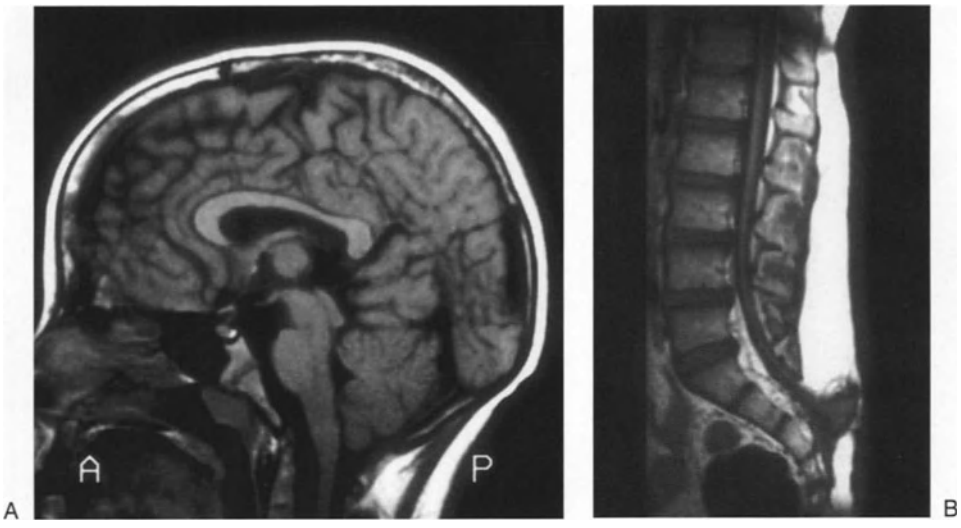


Figure 14.60. Chiari II: tethered cord, meningomyelocele, and agenesis of the tentorium. (A) Sagittal 28/4 3-mm slice thickness image revealing posterior dislocation of the temporal lobe (temporo-occipital junction) and upward displacement of the straight sinus secondary to aplasia of the tentorium. In (B) a sagittal SE600/20 image, one has an image of the most distal portion of the medulla spinalis, revealing posterior schisis of L5 and the visible totality of the posterior sacral arches with dilation of the sacral sac from S3 to S4.

The laminectomy is extended to the medial surfaces of the pedicles. The bone edges should be waxed often, with very soft beeswax, to diminish the risk of air emboli.

If one chooses to open dura mater, it is safest to start at the lowermost cervical level, performing the durotomy from inferior to superior, with a right-angle dural scissors. The greatest danger of the procedure is the extension of the durotomy across the point at which the outer layer of the cranial dura and the inner layer of spinal dura separate: the annular sinus at the rim of the foramen magnum. This is because of the extensive intradural venous sinuses and the inferior displacement of the transverse sinuses in children with the Chiari II malformation. Opening the dura in this area is discouraged!

Those who recommend only opening the dura over the herniated (into the cervical canal) cerebellar tonsils justify this with the observation that the medulla oblongata is herniated into the upper cervical canal, a conclusion drawn from the observation that the upper cervical cord may be both stretched and buckled, and the IV ventricle elongated. If this is done, the dura should not be left open (pseudomeningocele may result) and one should not attempt to separate the tonsils from the underlying medulla and spinal cord. Adhesions between the leptomeninges and the remarkably constricted cerebellomedullary structures are not easily freed. A duraplasty, using fascia lata or periosteum, is advisable.

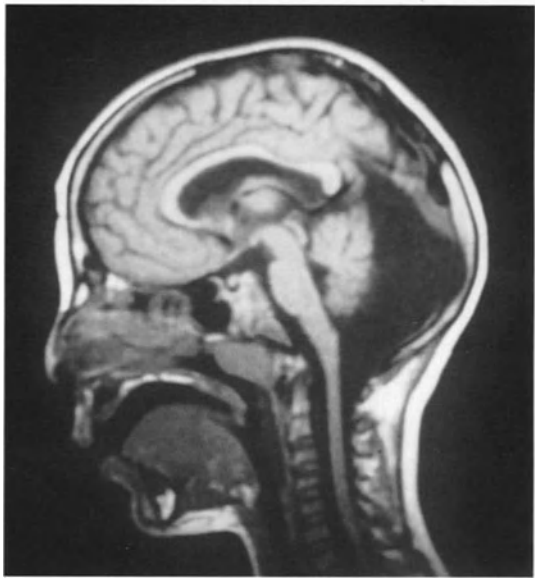


Figure 14.61. This is a sagittal cranial MRI T1 weighted image, in a child with congenital meningomyelocele, with no hamartoma present. A most unusual observation is what we see here: no Chiari II, cystic transformation of the cisterna magna, and hypotrophy of the cerebellum.

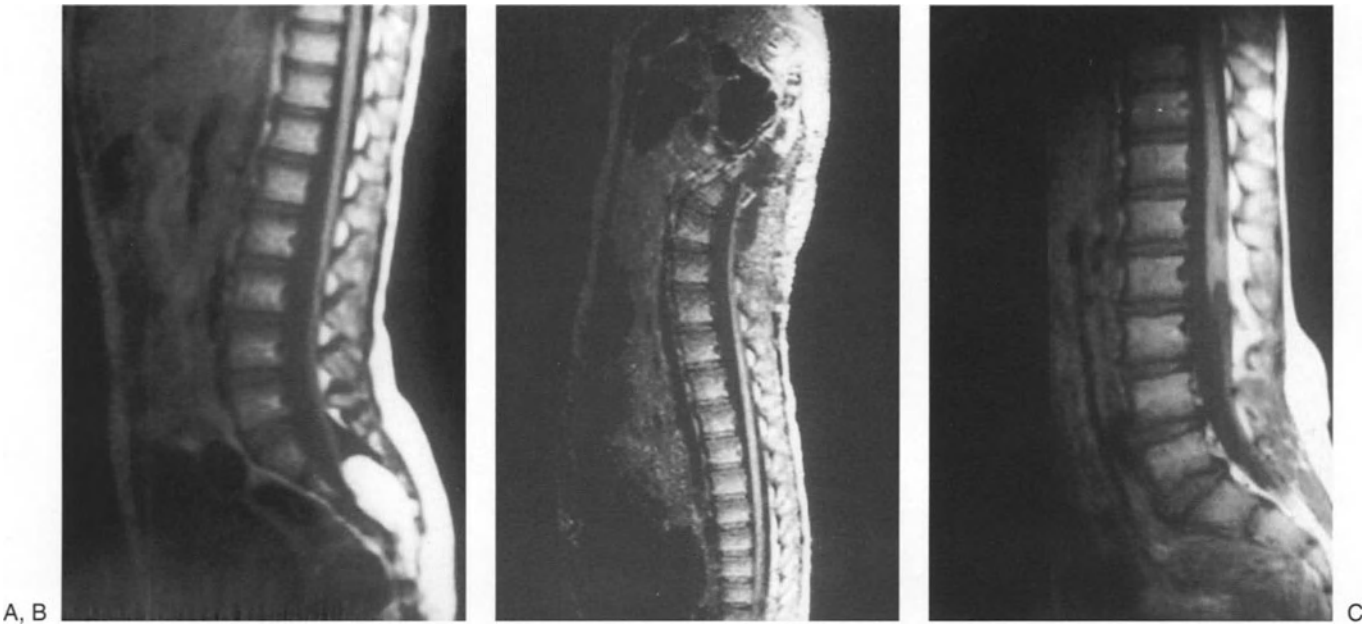


Figure 14.62. Lipomatous hamartoma. This child, with a clinical entity commonly in our opinion classified as a lipomeningocele, had a congenital meningomyelocele with a lipomatous hamartoma. This is the child illustrated in Fig. 14.61. (A) This is the preoperative study revealing the lipoma of the filum terminale, lodged within the remarkably dilated sacral spinal canal. There is anchoring of the most caudal element of the spinal cord to the lipoma. (B) shows the postoperative study. One notes that the lipoma has been resected and that the spinal cord has been freed. There is a terminal hydromyelia at the conus. (C) Three years later, one notes the terminal portion of the conus to have risen to the level of L3. In this interim, the child's clinical condition was one of progressive improvement.

Separation of Craniopagus Twins

(Figs. 14.63–14.69)

No single neurosurgeon has reported more than two personal cases of craniopagus separations; no two reported craniopagus cases reported have been anatomically identical; and we have never seen a craniopagus anomaly. Hence, for whatever value it may have, the following is a transcription of the paper "Surgical Separation in Craniopagus Twins" by H. Grossman, O. Sugar, and P. Greely [51].

On 16 September 1951, craniopagus twins were born in Rock Island, Illinois. The record of birth and course during the first 6 weeks have been adequately described by Durr [52]. The present preliminary communication will give a resume of the sequence of events that led to the eventual surgical separation of the twins and of the postoperative course.

The babies were admitted to the University of Illinois Hospitals on 2 October 1951. They were in a conjoined state of *craniopagus parietalis* (Fig. 14.63). Their general condition was good, with normal increments of growth and neuromuscular development. Their measurements on the Stuart-Vickers tables ranged form the 25th to the 50th percentile for their age. The babies had obviously independent activities; they slept at different

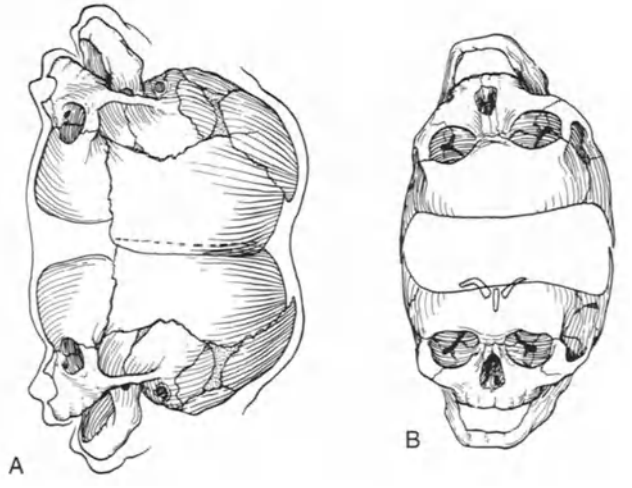


Figure 14.63. (A) Roentgenograms of the skull made at the age of 5 days. Note absence of bony sectum and overriding of parietal bones. (B) Pneumocephalograms made at the age of 6 weeks. Air was injected into the lumbar subarachnoid space of R.D.B. The subarachnoid air is confined to one brain. Note the height of the III ventricle between the lateral ventricles. (There was questionable agenesis of the corpus callosum).

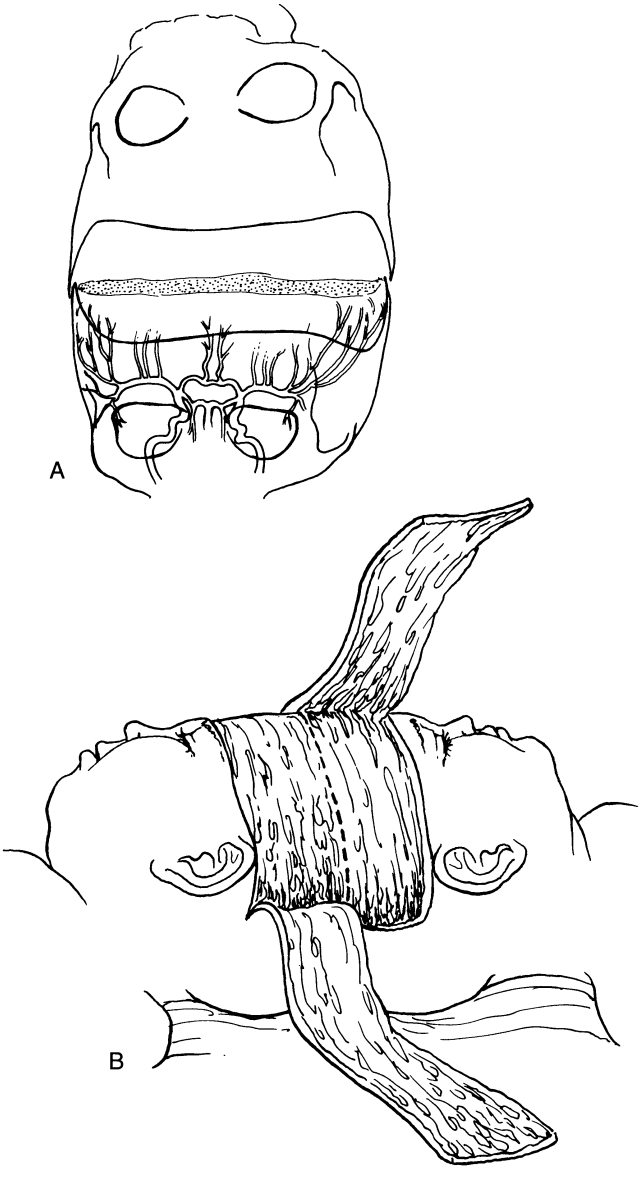


Figure 14.64. (A) Angiogram in R.D.B. at age 5 months. Iodopyracet (Diodrast) in the middle cerebral artery distribution appears to cross over to the opposite hemisphere, but the midline dye is actually from the anterior cerebral arteries. No dye appears in the cerebrum of R.L.B. (B) Pedicle flaps, which were replaced after the first attempt at separation on 26 November 1952.

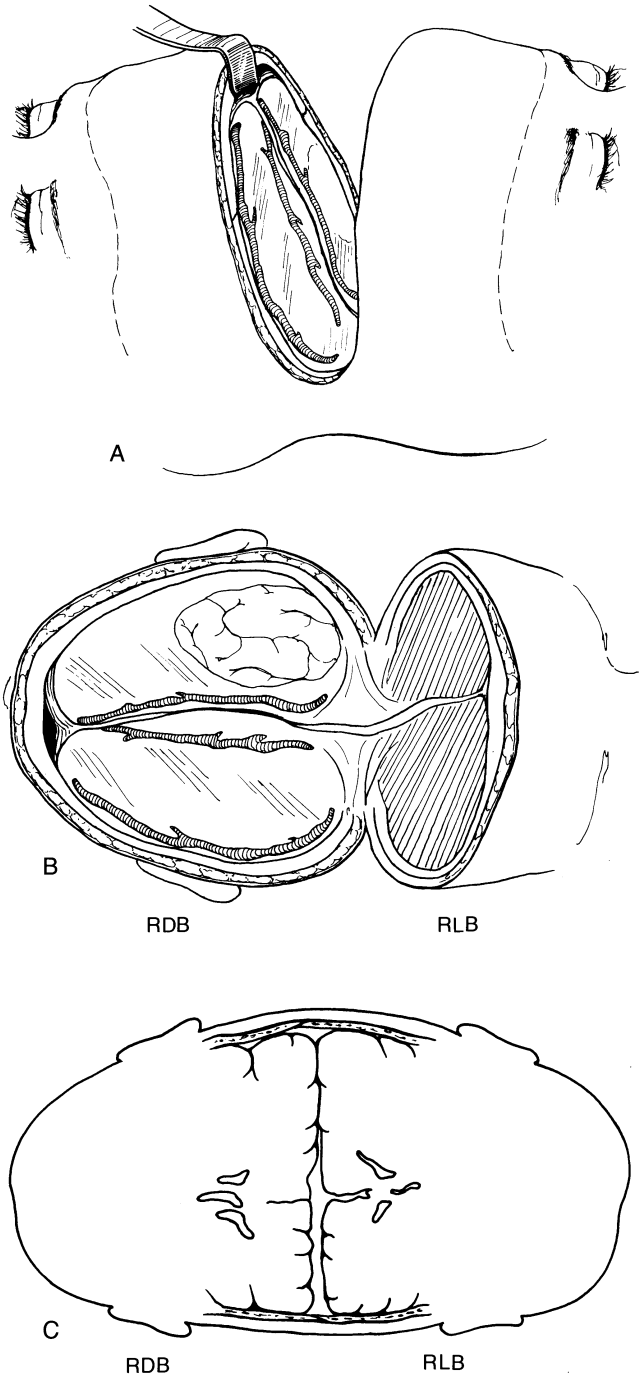


Figure 14.65. (A) Artist's drawing of findings during final separation (age 15 months). (B) Diagram of arrangement of abnormal dural venous sinuses as disclosed by operation. Note the dural defect over the right hemisphere of R.D.B. (C) Reconstruction of arrangement of dural venous sinuses, seen in coronal section. Note the apparent absence of the falx in R.D.B.

times, could eat at separate times, made separate movements, had independent bowel and bladder functions, and had individual body temperatures. In all, there was clinical evidence of two autonomous nervous systems. There was a mirror-imaged moderate asymmetry of the head (including the face), the right side of R.L.B. and the left side of R.D.B. being more prominent.

Both babies had systolic murmurs, which were loudest to the left of the sternum at the level of the 3rd interspace. It was considerably louder in intensity in R.D.B.

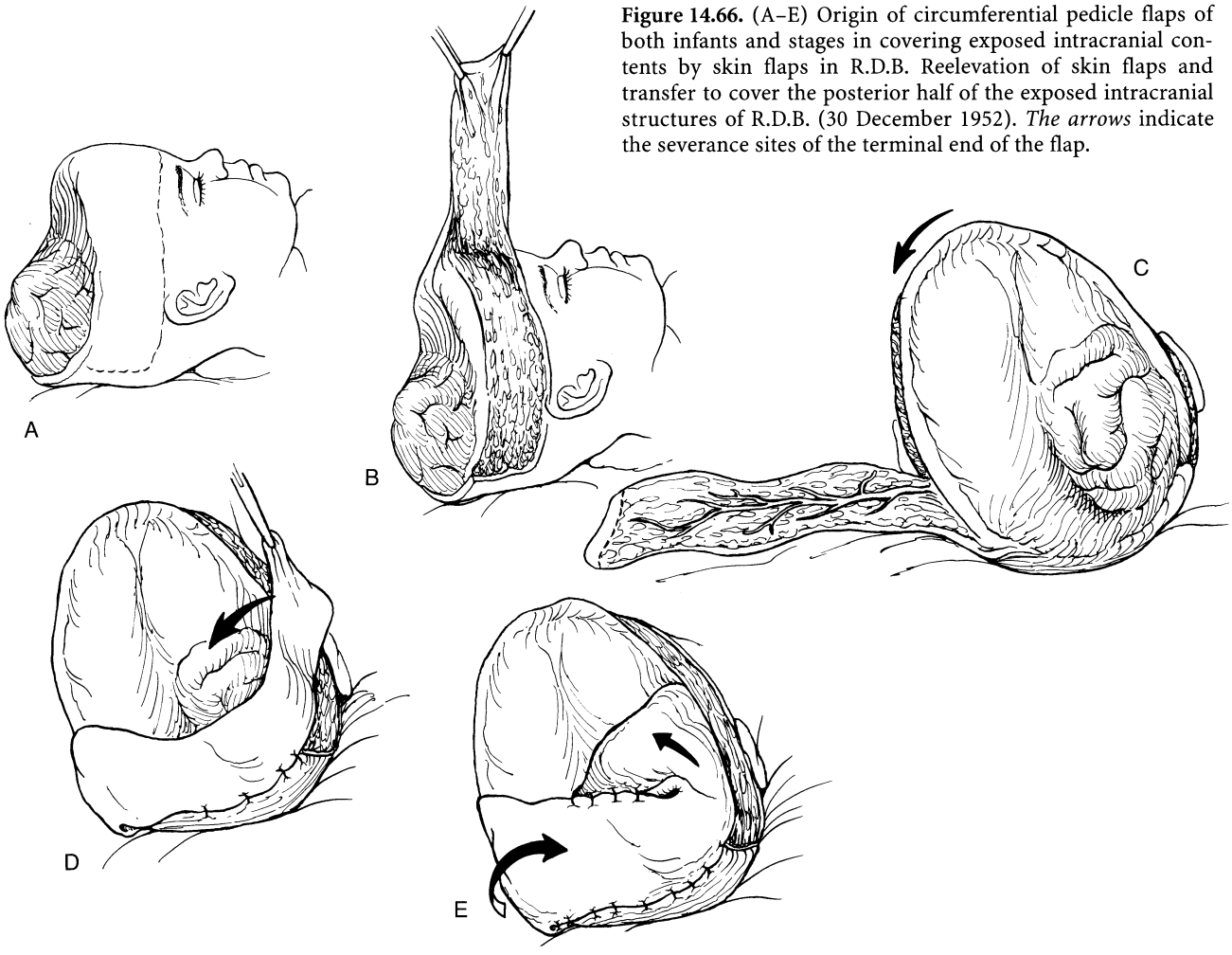


Figure 14.66. (A–E) Origin of circumferential pedicle flaps of both infants and stages in covering exposed intracranial contents by skin flaps in R.D.B. Reelevation of skin flaps and transfer to cover the posterior half of the exposed intracranial structures of R.D.B. (30 December 1952). *The arrows* indicate the severance sites of the terminal end of the flap.

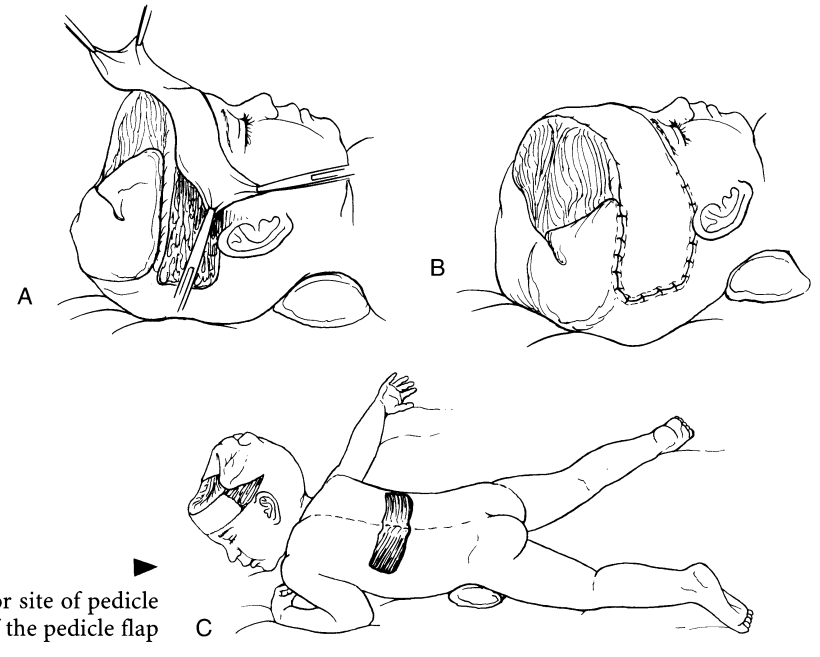


Figure 14.67. (A–C) Covering of exposed donor site of pedicle flap with split-thickness graft from the back of the pedicle flap (6 January 1953).

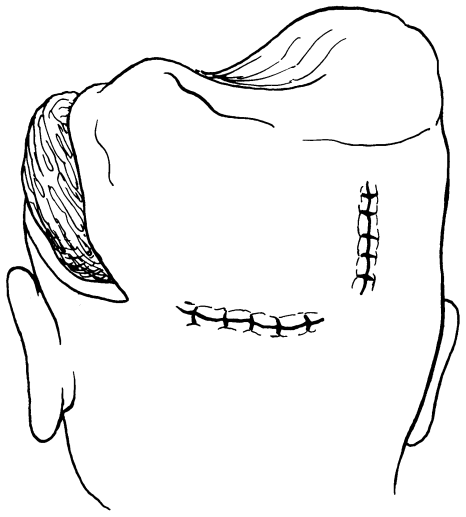


Figure 14.68. Extension of the proximal end of the full-thickness flap by delay of circulation to the base (17 January 1953).

There was no evidence of impaired cardiac function. The left side of the chest of R.D.B. was larger – corresponding to asymmetry of the faces. This was mirrored in R.L.B. The heart sizes were normal. The murmurs were believed to represent an intraventricular septal defect.

The problem at hand was whether or not a surgical separation could be attempted. Unfortunately there was little material in the literature that was of any pertinent value. The few previous attempts at separation of other craniopagus twins had failed.

Before an actual separation could be attempted, the following basic questions had to be answered:

1. Could the babies withstand numerous diagnostic and operative procedures?
2. Were the brains separate?
3. Was there a dural septum?
4. Were the cerebral arteries separate?
5. Were the venous sinuses separate?
6. Could the anticipated defects in the scalp and skull be closed?

Except for the obvious malformation of a conjoined state, results of all studies, including roentgen examination of the chests and long bones, blood studies, and urinalyses, were within normal limits. The systolic murmurs of neither child appeared to be of any functional significance; their general physical condition was good.

Clinical observation of independent activity indicated the probability that the brains were functioning independently. Electroencephalography disclosed similar but independent patterns of electrical activity normal for their age (2 months). Adequate placement of electrodes over the entire cerebrum was impossible because of the juxtaposition of the heads. Pneumoencephalography by the lumbar route disclosed separate subarachnoid spaces and indicated a dural septum between the two brains.

Carotid angiography was carried out after operative exposure of the common carotid artery (the right carotid artery of R.D.B. and left carotid artery of R.L.B.). Direct intra-arterial injection of iodopyracet (Diodrast) permitted visualization of the intracranial circulation of R.D.B.; owing to technical difficulties, adequate films were not obtained for R.L.B. (Fig. 14.64 shows independent arterial circulation): the delayed phase showed dye in the venous return of only the baby into whom injection had been made. There was no way of accurately predicting from these films what the venous drainage might be. In one attempt at dural sinography through the anterior fontanelle at age 6 weeks, the sinus could not be found. Even through a midline anterior bur hole, the sinus could not be located (age 9 months). At 11 months of age, the junction of two occiputs was explored, and one parasagittal venous lake was found. Attempts to obtain contrast visualization of this sinus failed.

Preparation of Skin Flaps

If separation were accomplished, adequate covering for the exposed intracranial structures would have to be provided. It is well known that any free skin graft will grow on exposed brain or dura. It will adhere tightly, however, and such a covering would not be satisfactory, for it would later become necessary to insert some structural support between the skin covering and dura in order to protect the brain. Transplanted skin and subcutaneous fat, but not a free graft, could be reelevated in the future (much as one might do with a normal covering), the mechanical support inserted, and the soft tissue covering then replaced. Since skin with its underlying fat will not grow as free graft, some type of pedicle flap arrangement had to be planned in order to maintain its own viability.

Although specific information concerning the venous drainage systems was inadequate, it was agreed that separation should be attempted. The skin flaps were prepared first. Many different places were considered as the source of the flaps. The most logical solution seemed to be to use direct circumferential flaps taken from around the head. They could be transferred very simply, without discomfort and without the necessity of immobilization in awkward positions. The base of the flaps, in the occipital area, could be brought to the forward part of the skull opening and thus provide some hair where it was most needed. The donor areas from which the flaps were taken could be covered readily with free skin grafts, with which an acceptable cosmetic result might be anticipated.

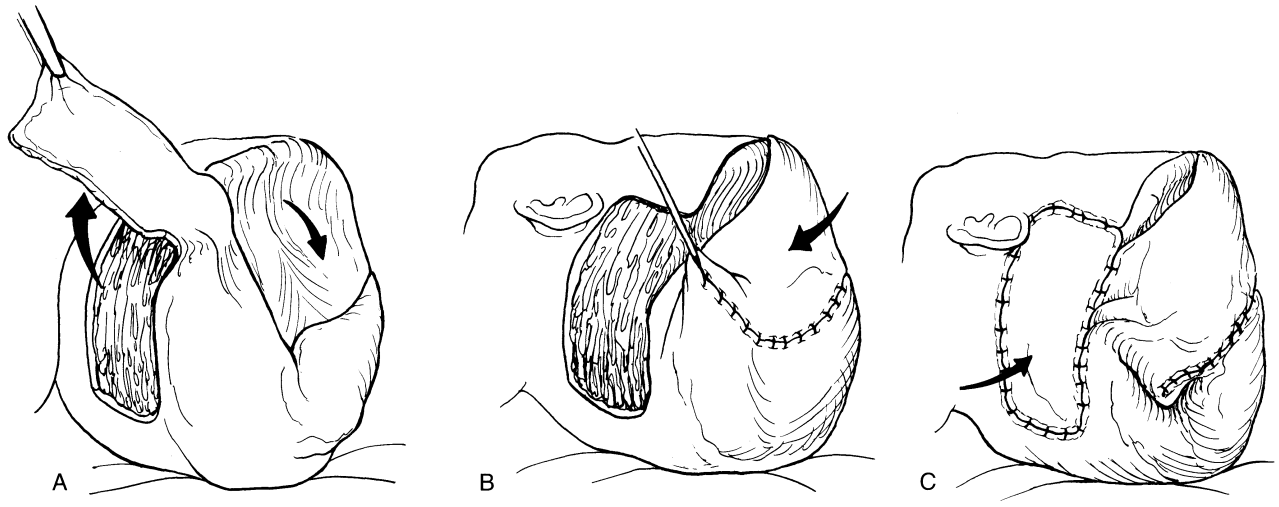


Figure 14.69. (A–C) Completion of the rotation of the full-thickness graft to cover the remaining exposed intracranial structures (10 February 1953). The donor site is covered by a split-thickness graft (1 March 1953).

When the use of scalp flap had been decided on, it was found by actual measurement that each flap would be approximately 14 in. (35 cm) long by 3 in. (7.5 cm) in width. This is very long compared with the width, especially when the normal blood supply coming in from the vertical axis would be cut off and thus make maintenance of nutrition of these narrow flaps still more hazardous. All of this could be done successfully, however, if sufficient patience and time were used. Repeated "delaying" operations would give adequate circulation if enough time elapsed between the various stages. Each stage would cut off more and more blood supply coming in through the main arteries, namely the frontales, supraorbital, and superficial temporal arteries of the two sides. As this source of nourishment was gradually diminished, it would simultaneously stimulate increased vascularity rising from the base of each flap in the occipital area and also produce a new additional blood supply in the transverse axis, i.e., parallel to the linear direction of the flap.

The foregoing plan was started on 14 May 1952. After three stages, the scalp flaps were completely divided around their peripheries. During the next 5 months (occupied by several neurosurgical diagnostic procedures) large transverse veins appeared in each flap. These proved to be of intestimable value in preventing venous congestion at the time of transfer. During the first attempt at separation on 26 November 1952, the scalp flaps were elevated completely from their beds. Their blood supply was excellent even though both were 14 in. (35 cm) long by barely 3 in. (7.5 cm) wide.

Very early in the course of our observations of the babies, it was evident that there would be numerous diagnostic and surgical procedures, some of which would require an anesthetic agent. During all procedures there was an anesthesiologist with a separate set of instruments (anesthesia machines, laryngoscopes, endotracheal tubes, etc.) with each infant. In each anesthetic procedure, the infants were given only enough of some basal anesthetic agent to produce a state of sleep. This was supplemented by inhalation anesthesia when indicated. Local anesthesia was used as the primary technique where applicable, utilizing basal anesthesia by the rectal route with tribromoethanol (Avertin), thiopental (Pentothal), or paraldehyde, and the inhalation of a mixture of nitrous oxide, oxygen, and trichloroethylene as a supplement where necessary. In this way psychic trauma was avoided as the children fell asleep and knew nothing about the procedure. Intubation with soft rubber endotracheal tubes was used in all procedures in which manipulation would endanger the airway.

The respiration of each infant was supplemented or controlled as individually required. At all times, the aim was to have minimal toxicity at the time of operation and minimal residual toxicity, in order to diminish the possibility of damage from repeated anesthesia. During the operations, continuous consultation between the anesthesiologists and the pediatricians permitted proper fluid replacement. Even in operations done with a small amount of local anesthesia, there was integration of management by all concerned.

In the major procedures that required long periods of time, the problem was magnified by the blood loss and the danger of trauma to the airway from having the tube in place for as long as 12 1/2 h. The danger of trauma was accentuated by the manipulations necessary during surgery. The problem was one of maintaining the vital functions throughout. The anesthesia was maintained at a light plane to avoid cumulative damage, but yet had to be deep enough to prevent interference

from movement of the patients. The chief agent used was nitrous oxide with as high an oxygen concentration as possible. Trichloroethylene was added only for a few minutes at a time to reestablish adequate depth of anesthesia.

First Attempt at Separation

For the first attempt at separation, an incision was made at the junction of the frontal and parietal bones (right side of R.D.B., left side of R.L.B.), and a channel 1.5 cm wide from the vertex to the occiput was made. An attempt was then made to split the dura mater into its two fused portions. This proved to be impossible beyond a depth of about 1 cm, because the two leaves of dura mater were fused into one sheet; furthermore, as dissection continued in this plane, there was considerable venous bleeding, the source of which could not be determined. Accordingly, incisions were made on either side of the midline septum, exposing the subarachnoid space on either side of the septum. It could be seen that the dural septum did, in fact, separate two brains; however, because of the peculiar position of the two heads and their immobility with respect to each other, it was impossible to look into the depths on either side of the septum. The dural septum was then incised toward the midline between the two brains. This allowed visualization, at a distance of some 6 cm, of two longitudinal sagittal sinuses that did not appear to communicate with one another in the frontoparietal region, which was the limit of the exposure at this time. This exploration and the antecedent elevation of the scalp flaps had taken 6 1/4 h, and it was felt that complete separation could not be accomplished then and that the procedure should be terminated. The incised dura mater was sutured at its periphery, and the wounds closed. The scalp flaps were resutured into their original beds. The postoperative convalescence was uneventful, except for a short left focal seizure in R.D.B., involving the arm, and lasting less than a minute, that was followed by a mild transient hemiparesis for about 18 h.

Final Separation

The final separation was made on 17 December 1952. This was started at the frontal junction with an incision made parallel to the septum and few millimeters from it, to avoid the bleeding encountered when separation of the dura mater had been attempted in the earlier operation. It was believed that the dural defect created by going on R.L.B.'s side of the septum might be covered with polyethylene film. The falx of R.L.B. was found and cut while the numerous vessels were clipped as necessary. After a few centimeters of exposure, it became evident that, in order to permit visualization into the depths, it would be necessary to clip the veins running from the surface of the brain to the dural septum. This would inevitably interfere with the venous return from one of the twins. By the time this information was obtained, there was enough interference with the brain of R.L.B. that it was clear that complete separation would not markedly increase the impairment of cerebral function already anticipated (Fig. 14.65). Accordingly, the falx was cut completely, and the veins that drained the surface of R.L.B.'s cerebrum were also clipped and cut as they entered the dural sinuses. These were found to be three in number; they lay within the dural septum. One was far out in the periphery, forming a semicircle on the left side of R.D.B.'s brain, and there was one on either side of the cut falx, which had extended from R.L.B.'s brain to the dural septum. It was possible to reach the posterior part of the falx where there were huge venous lakes connecting all of the lateral sinuses and all of the sagittal sinuses, as well as the circumferential sinus posteriorly in what amounted to a conjoined torcular Herophili. With the incision being made on R.D.B.'s side, these enormous conjoined sinuses were cut through. Hemostasis was obtained with difficulty, and the babies went into shock, which was counteracted by liberal administration of blood.

Thus was accomplished the complete separation of the two infants, 10 1/2 h after the operation was begun. R.L.B.'s brain had the typical appearance of congestion with petechial hemorrhage that would be expected from a brain with an intact arterial supply but virtually no venous outflow. R.D.B.'s brain appeared to be normal except for some congestion and edema in the right parietal region where the dural septum was defective, presumably as a result of failure of healing of the incision made in the dural septum on that side in the first separation attempt.

The problem now arose as to how to cover the denuded brains, which had no dura mater. Polyethylene film was used for this purpose; it was sutured to the dural edge of R.L.B.'s brain and to the dural and/or subcutaneous tissues over R.D.B.'s brain. The blood loss had been so great and the operation had taken such a long time that it was felt unwise to consider reflecting the scalp flaps and placing them in position on top of the polyethylene film, for this would have meant more hours of operating time and more loss of blood that might not have been withstood. It had originally been planned to put polyethylene plates in place at this time, but the molding of these plates and their anchoring into the wound would again have been too onerous a burden, so this, too, was omitted and we were content to use sterile aluminum foil to cover the top of each head before putting on dry dressings and sterile bandages. The aluminum foil was easily molded, could be sewed, and was believed to be mildly bacteriostatic. The entire procedure required 12 1/2 h. R.D.B. received 2500 ml blood and R.L.B. 3500 ml during the procedure.

After separation, R.D.B. cried spontaneously; R.L.B. showed attempts at crying while the endotracheal tube was still in place. A small catheter placed in the tube revealed an obstruction. The quick removal of the tube and tracheobronchial aspiration showed that the accumulated secretions had become rubberlike, blocking the airway. After this resuscitative procedure, the baby cried weakly.

Course of R.L.B. At the time of separation when it was necessary to separate the venous plexus, there was a sudden loss of blood that resulted in a severe cardiovascular collapse in R.L.B., so that no pulse, blood pressure, or heart sounds were detectable. The very rapid administration of oxygen by pressure on the breathing bag, positive pressure on the thorax, and rapid intravenous infusion of blood restored essentially normal cardiovascular function. He did not respond to painful stimuli. About 8 h after operation, there was marked irregularity of rate and depth of respiration, with retraction of the suprasternal and infrasternal regions. Secretions from the hypopharynx were aspirated easily, with the patient in an atmosphere rich in oxygen and moisture, but signs of right upper pulmonary atelectasis appeared. Although it was considered probable that much of the respiratory difficulty might be due to central nervous system derangement, tracheotomy was done by Dr. Cecil Riggs of the department of otolaryngology, to provide as open an airway as possible. Periodic breathing of the Biot type appeared, however, indicating a central nervous system cause for the respiratory difficulty. Accordingly, the head dressings were removed, when it was seen that the swelling and engorgement of the cerebrum seen during the operation had progressed. Relief of pressure by removing the artificial coverings of the brain brought immediate improvement in breathing. The tachycardia observed during the Biot breathing became slower.

With the long exposure of the brain, contamination and possible infection were anticipated, so penicillin and streptomycin were given intramuscularly. Organisms sensitive to these antibiotic agents were suppressed, but an infection with *Pseudomonas aeruginosa* was detected on the fourth postoperative day. This was controlled by daily local applications of 2 1/2% polymyxin B to the surface of the brain and by intramuscular injections of 2 mg per kilogram body weight per day. No evidence of systemic toxicity was discovered, although the antibiotic was given until his death 34 days after separation. By this time, the surface infection of the brain was completely controlled. Staphylococcal organisms were controlled by intermittent administration of aureomycin, oxytetracycline (Terramycin), and erythromycin. Eleven days after operation, signs of diffuse bronchopneumonia appeared, with marked expiratory difficulty and very tenacious secretions. In order to liquefy these, water was nebulized over the tracheotomy tube. Since high humidity alone proved insufficient, a solution of trypsin (Tryptar) was given as an aerosol through the nebulizer (20,000–50,000 units of trypsin in 2–3 ml diluent at pH 7.4) every 6–8 h. On several occasions, Alevaire (a solution of glycerin, sodium bicarbonate, and a detergent, Triton WR-1339, in distilled water) was used in a similar manner. Marked liquefaction of the secretions resulted, permitting their aspiration and resulting in marked improvement in respiration. No evident signs of toxicity to the drug were noted. The signs of bronchopneumonia subsided. During the survival period, the infant remained comatose and exhibited signs of decorticate rigidity. Parenteral fluid therapy was given for the first few postoperative days, and thereafter nutrition was maintained by feedings through a #19 polyethylene nasogastric tube. Up to 4500 calories daily were necessary. During the last week of life, there were marked variations in rectal temperature (108°F to 94°F), the peaks not responding to antipyretic measures. Respiratory irregularities of central nervous system origin appeared again on 19 January 1953, and he died on 20 January 1953.

Course of R.D.B. Immediately after the separation R.D.B. cried and moved all extremities. He took fluids by mouth the following day and responded to familiar persons. He was kept in oxygen with supersaturated humidity to minimize the effects of the prolonged period of intubation. Supplementary parenteral fluids and antibiotics were given with R.L.B. On the fourth postoperative day, he was irritable and refused oral feedings. There was an odor from the head dressing characteristic of a *Pseudomonas aeruginosa* infection, so the dressings were removed, and the surface infection which was found was treated with polymyixin B in the same manner and dose as in R.L.B. The thick tenacious exudate typical of infection with this organism gradually subsided. It had not completely disappeared by 30 December, when the clinical status of the patient was improved sufficiently to permit proceeding with the plastic surgery. On this day, the scalp flap was reelevated and transferred to cover the posterior half of the exposed intracranial structures (Fig. 14.66). The terminal end of the flap was turned to cover part of the anterior defect. All of this flap grew well except for a short segment along the distal edge. This ultimately was trimmed off. After the flap was transferred, the donor site from which it was taken was covered with a large autogenous split skin graft taken from his back (Fig. 14.67).

On 17 January 1953, the base of the pedicle in the occipital area was divided so as to stimulate additional return circulation from the opposite end of the transplanted flap (Fig. 14.68). On 10 February, the proximal end of the flap was completely detached from the occipital area and transferred forward to cover nearly all of

the remaining open defect (Fig. 14.54). Donor sites were covered in each case by split-thickness grafts, some of which had to be revised later.

On 11 March, the "dog ear" in the flap was divided in its central portion. The anterior part of the flap was re-elevated until it could be shifted anteriorly without tension to cover the balance of the open cranium. The excess "dog ear" was trimmed off and its edges sutured. By this time, the infection was not apparent. This twin, too, showed no signs of toxicity from the antibiotics.

Several days after the separation, when there was more spontaneous activity, it became apparent that there was a left hemiparesis and left astereognosis. Although he was able to hold objects when he looked at them, he did not know what to do with them if his eyes were turned away, nor did he place his extremities properly if he did not see them. The hemiparesis disappeared within the 1st month, but the astereognosis still continued, although less marked than before. There were no hyperactive stretch reflexes on the left, nor was there any loss of common sensibility. There were intermittent Babinski signs on both sides, which were considered to be normal for his age.

Before the separation, the neuromuscular development of the twins was considered normal. This was assessed with difficulty because of the peculiar nature of the junction of the heads, which undoubtedly interfered with the development of head control, tonic neck reflexes, and the ability to sit and stand. Both had normal infant jargon. During the 3 months that followed separation, R.D.B. showed only a slow progression of development. This is attributed to the trauma of separation, debility, infection, and repeated operations that took place during this period. Since the head covering has been completed, he has shown a considerable advance in development and improvement in his general physical state. He has had a considerable amount of occupational and physical therapy, which has contributed to this improvement. He can now sit alone, but cannot pull himself into the sitting position. He has good control of his head and good coordination of all extremities, and his jargon has increased in amount and intelligibility. He is alert, responds to favored personnel, and makes attempts to feed himself. The hip flexors (which were tight, presumably owing to lack of standing) have become normal with physical therapy. It is difficult to assess the degree of development in all spheres because of the many factors, biological, traumatic, and psychological, that have affected his present state.

There is still no solid protection for the brain, and the solution of this problem is now under active consideration. We are reluctant to use any foreign material over such a large surface of dura mater and under skin flaps because of the hazards accompanying buried foreign bodies. Autogenous bone grafts appear to offer the best replacement for the calvarium that never developed, but the extensiveness of the defect make accomplishment of closure by this means difficult.

All professional and technical skills have basically one objective – to permit this baby to grow and develop as a happy, effective human being. His handicap – like other handicaps – need not prevent him from fulfilling this goal. It has been necessary throughout, therefore, to provide for him as well as his family an understanding and comfortable environment. Without such an environment – and without the help of his parents and our help to them in understanding his problems – the boy's development might not have proceeded so well. The future planning must involve most careful attention to his emotional and social development. This will consist of his parents' greater and greater participation in his care as his physical problems permit, continued medical and surgical efforts to correct his physical difficulties with as little pain as possible, and long-term planning to assure stimulating educational opportunities that are compatible with his capacities. These provisions, associated with increasing understanding of all handicapped children by the public, will provide the best assurance that this boy will have the opportunities that should come to all children – that of the fulfillment of his greatest potentialities as a human being.

My thanks to Dr. O. Sugar (from whom I learned the basic elements of cerebral angiography) for his permission to transcribe this classic case report.

Vertebrospinal Congenital Anomalies

Vertebrospinal congenital anomalies in children, the opposite of vertebrospinal injuries, are common, becoming clinically obvious either at birth, in toddlers *when the children begin to walk,* and during the enormous growth spurt which occurs during the transition from juvenile into adolescent years. In broad terms, these anomalies may be subdivided into two groups: those associated with the dysraphic state and those characterized by congenital tumors or anomalies without defects in the posterior vertebral arch.

The Dysraphic State

Surgical anomalies of the dysraphic state include myelocele, meningomyeloeele, meningocele; lipomeningocele, lipomeningomyelocele; some hamartomas (lipoma, dural fibrolipoma, leptomyelolipoma, anterior spina bifida, dermoid tumors, enterogenous cysts) extending from the intramedullary area into the subcutaneous space; and diastematomyelia. With the rarest of exceptions, these do not exist as pure, anatomical anomalies. Rather, one blends imperceptibly into another and, in

many instances such as lipomeningomyelocele and hamartomas, several may coexist.

Before undertaking an operative procedure, one asks oneself what specifically one expects to attain: closure of a defect to protect against infection, anatomical reconstruction to restore function, resection of mass to relieve compression, or sectioning of a tethering structure to eliminate stretching.

Despite some rather startling claims made many years ago (and subsequently corrected), by Sharrard and coworkers [53], *it is a well-accepted fact that closure of a myelocele, meningocele, or meningomyelocele in a newborn does not result either in improved neurological function or in increasing the possibilities of subsequent rehabilitation.* Similarly, the particular closure technique used does not affect subsequent neurologic function. The closure simply provides the central nervous system protection from infecting organisms. It does not minimize the incidence of adhesions between the dysraphic spinal cord and repaired dura (postoperative tethering). The neuroradiological observation of the latter in a child years after repair is not a sine qua non to explain motor or sensory impairment; progressive scoliosis and hydromyelia likely are much more significant. Therefore, the procedure and the timing of its performance are to be interpreted only on the basis of protecting the central nervous system from bacterial and physical damage.

When the lipomatous portion of a lipomeningomyelocele, or a medullary lipoma, compresses the medulla spinalis, removing it affords similar, but not identical, beneficent results as removing a neurinoma or meningioma. The results are similar, not identical, because it is not possible to remove the intramedullary portion of the lipoma. Consequently, the decompression is never complete and the potential for lipoma regrowth is always present. Removal of a diastematomyelic spur, when it is pathogenetic of spinal cord deficit, may be considered palliative, as may severing of the filum terminale when it is pathogenic of a conus medullaris syndrome. "Prophylactic" surgery for removal of subcutaneous lipoma, supported by some neurosurgeons but opposed by most, must be evaluated critically.

In light then of the previous comments, techniques for the surgical treatment of the individual developmental anomalies associated with the dysraphic state are described, with the realization that one anomaly may blend imperceptibly into others. *Nowhere in the field of pediatric neurosurgery do "feelings" so disrupt objective analysis of indications, treatment, and results as in the dysraphic state. What a pity!*

Any decision to operate on a patient suffering from one or more anomalies of the dysraphic state is predicated upon a detailed understanding of the developmental anatomy and pathology of the clinical entity. Precise terminology assists one neurosurgeon to communicate his observations and concepts to another. The reader is referred to the works of Duckworth, Sharrard, Lister, and Seymour (1968) [54]; Lebedeff (1881) [55]; von Recklinghausen (1886) [56]; Morgagni (1761) [57]; Patten (1953) [58]; James and Lassman (1962) [59]; Ingraham and Matson (1954) [60]; Talwalker and Dastur (1970) [61]; Emery and Lendon (1973) [62]; and Rokos (1973) [63].

Amyelia

The clinical entity known as amyelia is of significance to the neurosurgeon only from one point of view: providing evidence that muscle masses may develop and dorsal roots may be found within the spinal canal of severely deformed fetuses and newborns who have no spinal cord at all. This allows one to understand that neural elements may be present within the dysraphic area independent of a medulla spinalis, that the presence of somatosensory evoked potentials between the lower extremities and the spinal area is not indicative of functional spinal cord.

Defects in Closure of the Spinal Cord and Posterior Vertebral Arch

It is unfortunate that we generally speak indiscriminately of "M-M" as though all children born with a sack on the back had the same congenital anomaly. This confuses treatment and prognosis. With this in mind, the following classification, taken from the 1976 edition of *Greenfield's Neuropathology* [64] is recommended:

Myelocele (Figs. 14.70–14.72)

Failure of closure of the neural tube (myeloschisis), resulting in a flattening of the spinal cord, is a defect is covered by granulation tissue, which is highly vascularized and surrounded by a translucent membrane that establishes continuity between it and the peripheral skin. If the flattened cord lies normally within the ventral aspect of the spinal cord, it is referred to as *simple myeloschisis.* If it is floating on an arachnoidal cyst which separates it from the ventral aspect of the spinal canal it is *cystic myelocele.* If the vascularized granulation tissue covering the myeloschisis organizes and epithelializes, simple myeloschisis or cystic myelocele may be confused with meningomyelocele when, in fact, it is a *scarred myelocele.* The placode of meningomyelocele is quite different, nonfunctional, pathological tissue.

The developmental pathological anatomy of the myelocele results in flattening of the spinal cord, which has not closed into a tubular structure, with medial location of the ventral horns and roots and lateral location of the dorsal horns and roots.

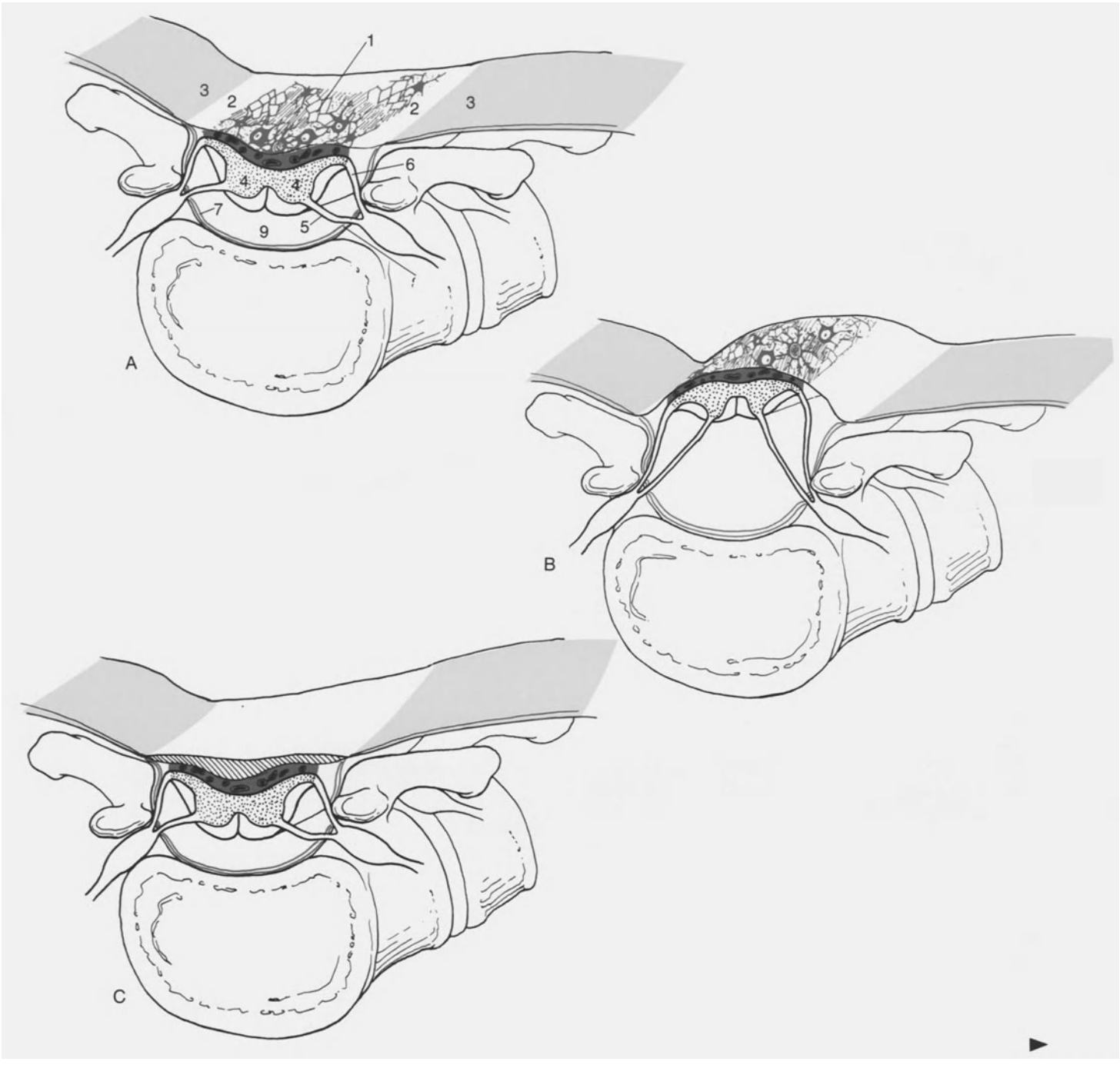
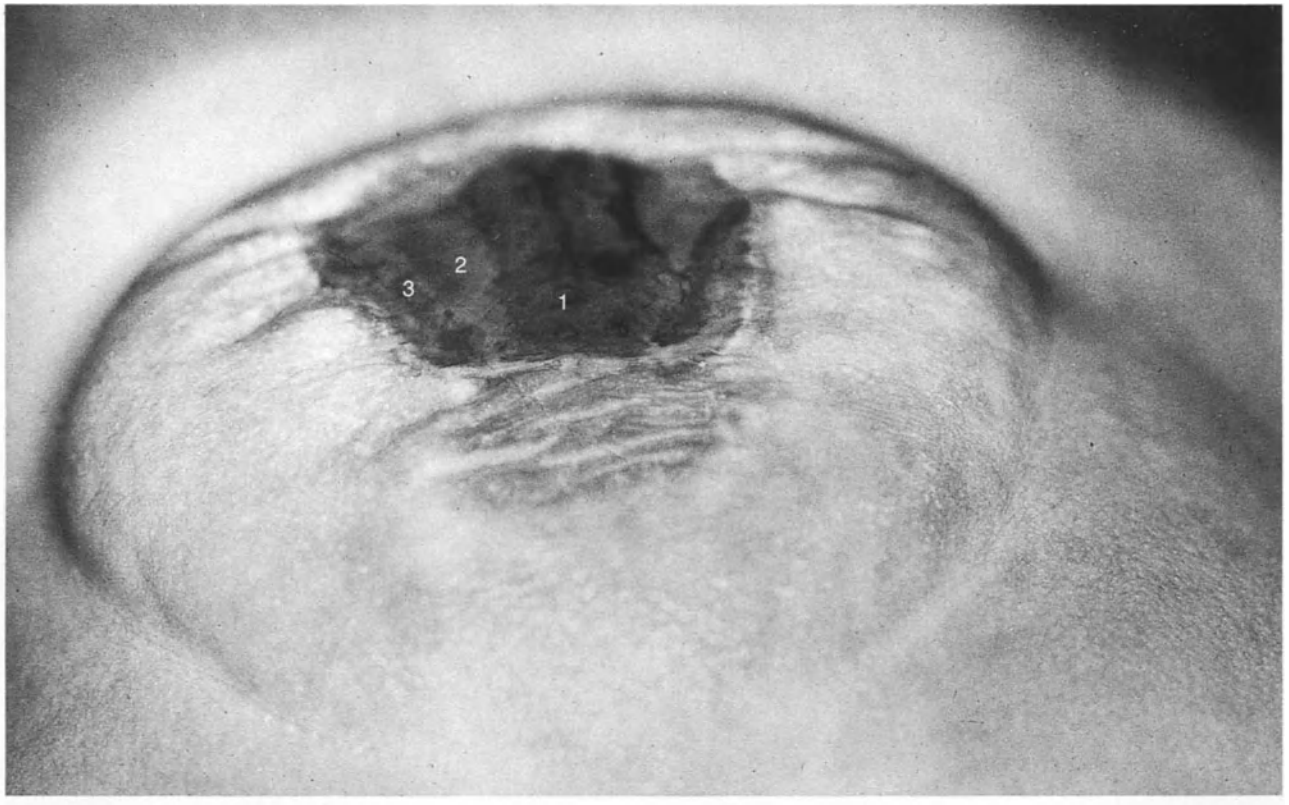


Figure 14.70. (A) Simple myeloschisis (myelocele). Its surface (*1*) is a composite of glial scar, ectopic neurons, patches of ependyma, and degenerative changes. This is the medullovasculosa. It is bordered by a zone of tissue devoid of neural or epithelial tissue (*2*) which, in turn, is surrounded by the zona epitheliosa (*3*). Note the unclosed spinal cord (*4*) with ventral (*5*) and dorsal (*6*) roots. The arachnoid (*7*) and dura (*8*) rest along the ventral surface of the spinal canal (*9*). (B) Cystic myeloschisis. This is a myelocele floating on an arachnoidal cyst, which separates it from the ventral surface of the spinal canal. (C) Scarred myelocele. The posterior surface of myeloschisis, simple or cystic, may be covered with granulation tissue, which is organized and epithelialized. This is a *scarred myelocele*, an entity easily confused with meningomyelocele.

Figure 14.71. A nonscarified myelocele (A) is illustrated with a scarified myelocele (B) to permit one to appreciate how scarification converts the appearance of a myelocele into a dysraphic anomaly which is visibly indistinguishable from the meningomyelocele. (A) The myelocele (*1*) and its zona neurovasculosa are readily identified, as is the transitional, nondescript tissue (*2*) between it and the zona epitheliosa (*3*). The bulging of the zona epitheliosa and the zona neurovasculosa (the cystic variety) results from an arachnoidal cyst located between the ventral aspect of the open, flattened cord and the dorsal aspect of the dysraphic vertebral bodies. (B) The myelocele (*arrows*) is scarified, rendering it indistinguishable from a meningomyelocele.



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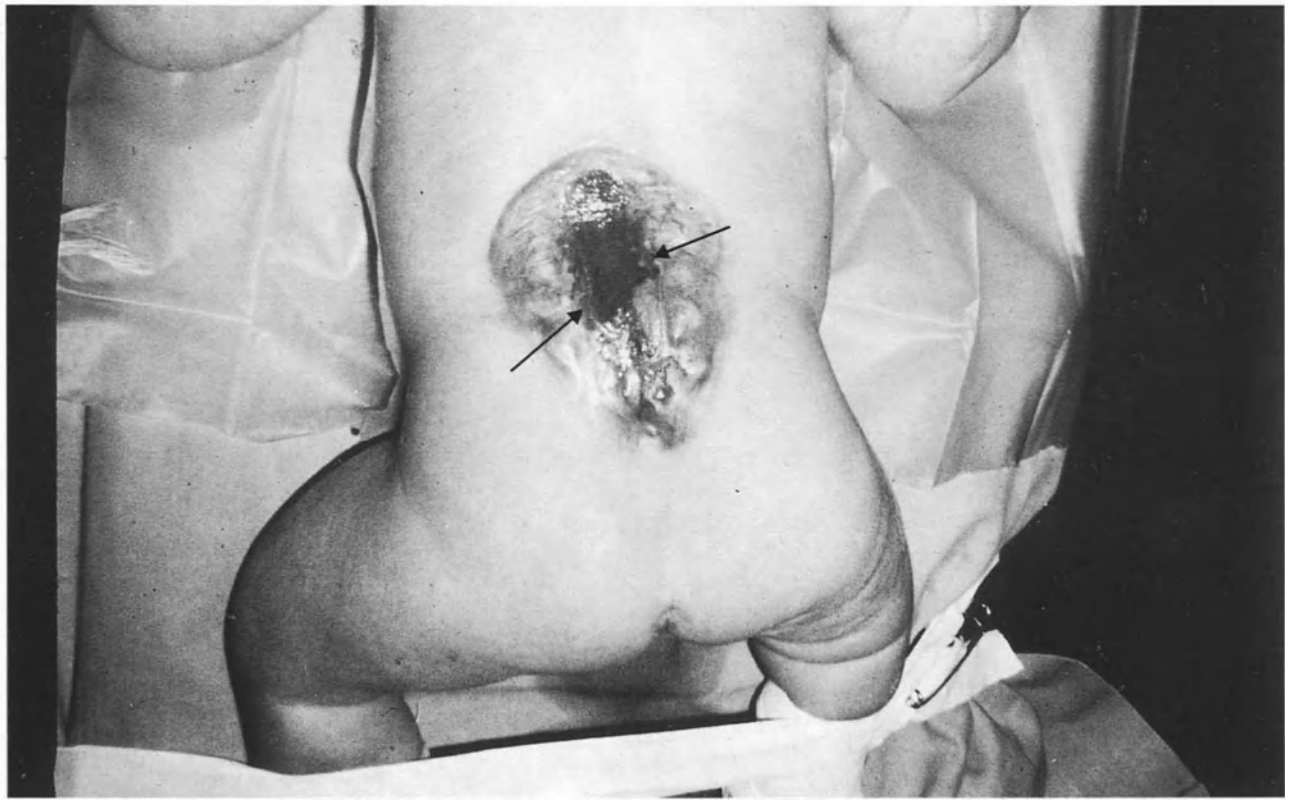
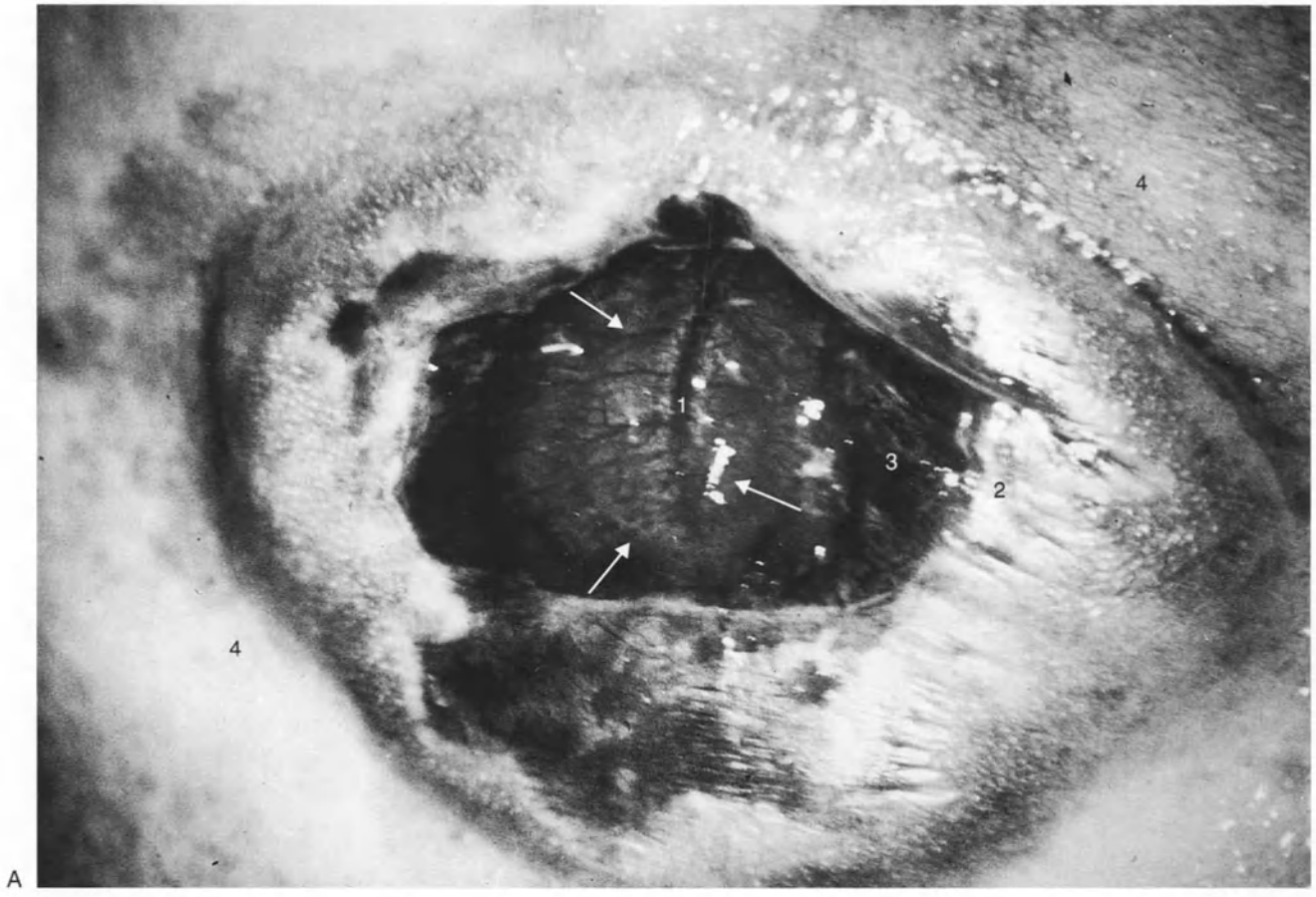


B

Figure 14.71. Legend see p. 524.



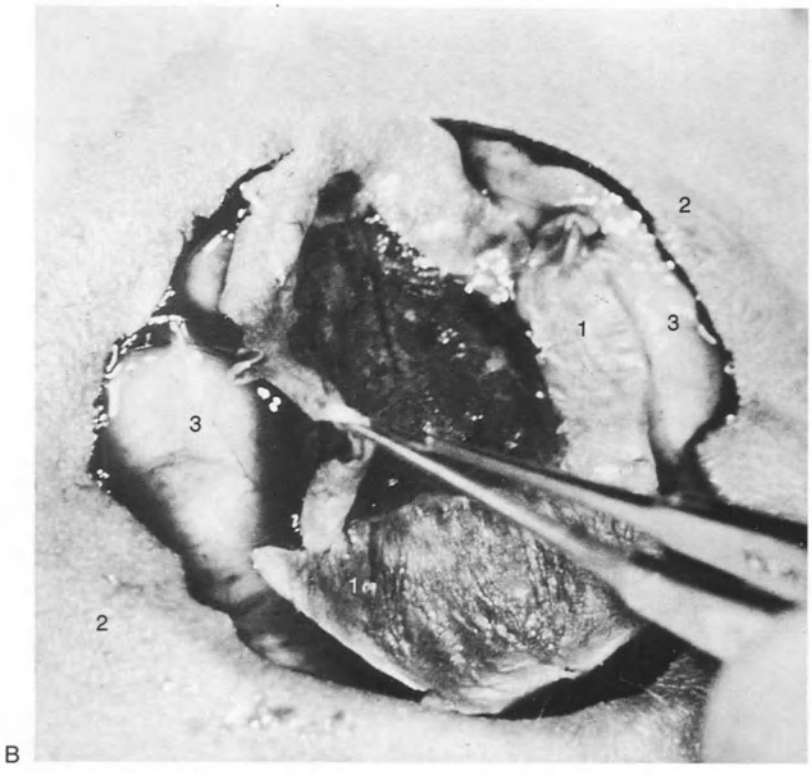


Figure 14.72. (A) Myelocele. The zona neurovasculosa (*short arrows*) consists of the flattened, opened spinal cord. One notes a groove (*1*) at the center of the open cord. Between the zona neurovasculosa and the zona epitheliosa (*2*) there is a nondescript membrane (*3*), varying in contour and extent. Normal skin (*4*) surrounds the zona epitheliosa. (B) The first stage in closure of a myelocele, as in a meningomyelocele, is to incise along the line of demarcation between the *zona epitheliosa* (*1*) and skin (*2*). This is a relatively bloodless phase of the procedure. The underlying fascia of the laterally displaced paravertebral muscle masses (*3*) is seen on either side, as the *zona neurovasculosa* sinks into the depth of the hernia cavity after cerebrospinal fluid escapes from the underlying arachnoidal cyst. At this time, one picks up the zona epitheliosa before proceeding to incise it perpendicular to the margin of the zona neurovasculosa. (C, D) see p. 527.

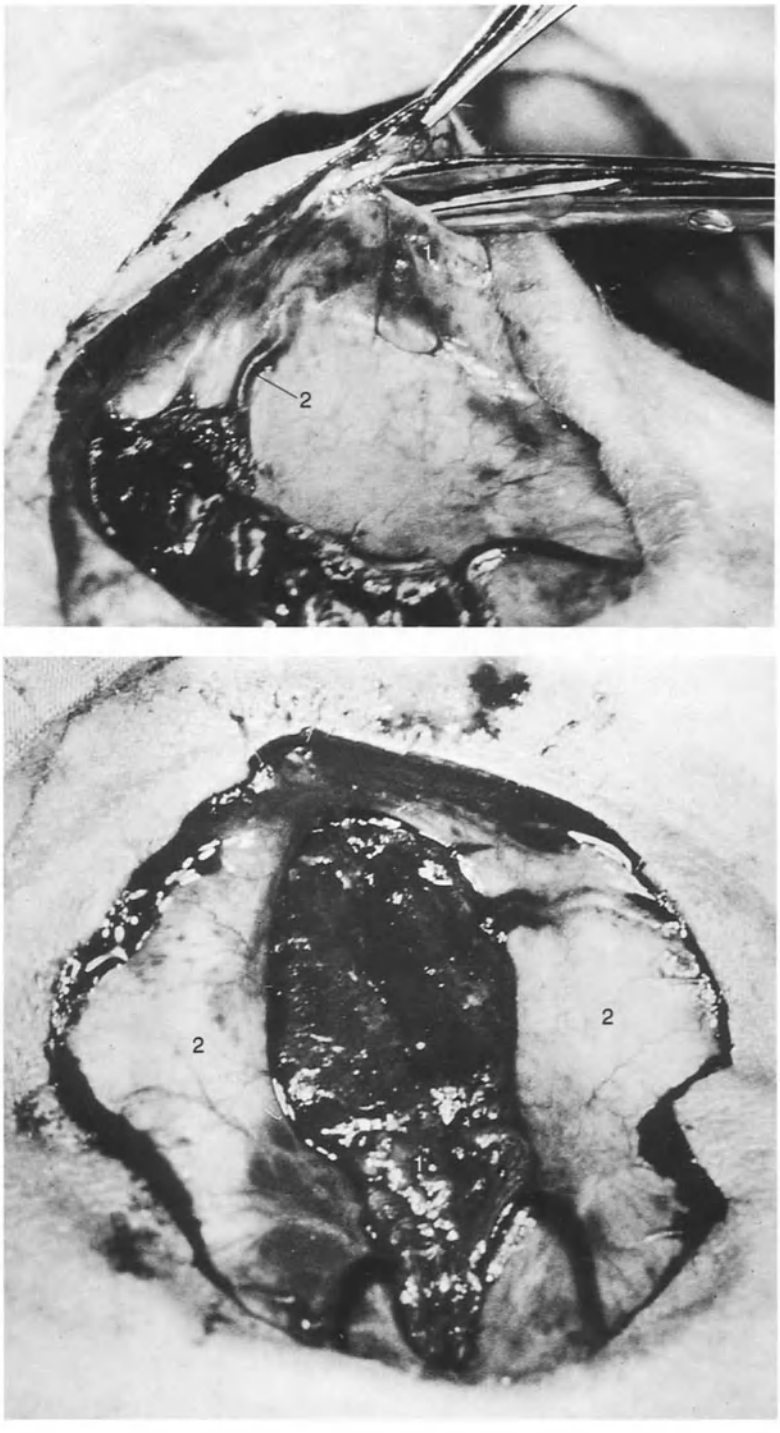


Figure 14.72. (C) Iris scissors are used to cut across the zona epitheliosa and nondescript tissue to the edge of the zona neurovasculosa. Then, the surgeon cuts the nondescript tissue from the edge of the zona neurovasculosa (*1*) so that it may be free to reduce itself into the hernia cavity. No effort is made to force reduction. Vascular structures (*2*) should be left intact. (D) After the zona neurovasculosa of the unclosed spinal cord has dropped to the bottom of the hernia cavity, one identifies first the dura mater (*1*) and frees it, preparatory to imbricating it over the myelocele. Subsequent to this, the fascia of the paravertebral muscle mass (*2*) will be separated in a similar manner, and also imbricated.

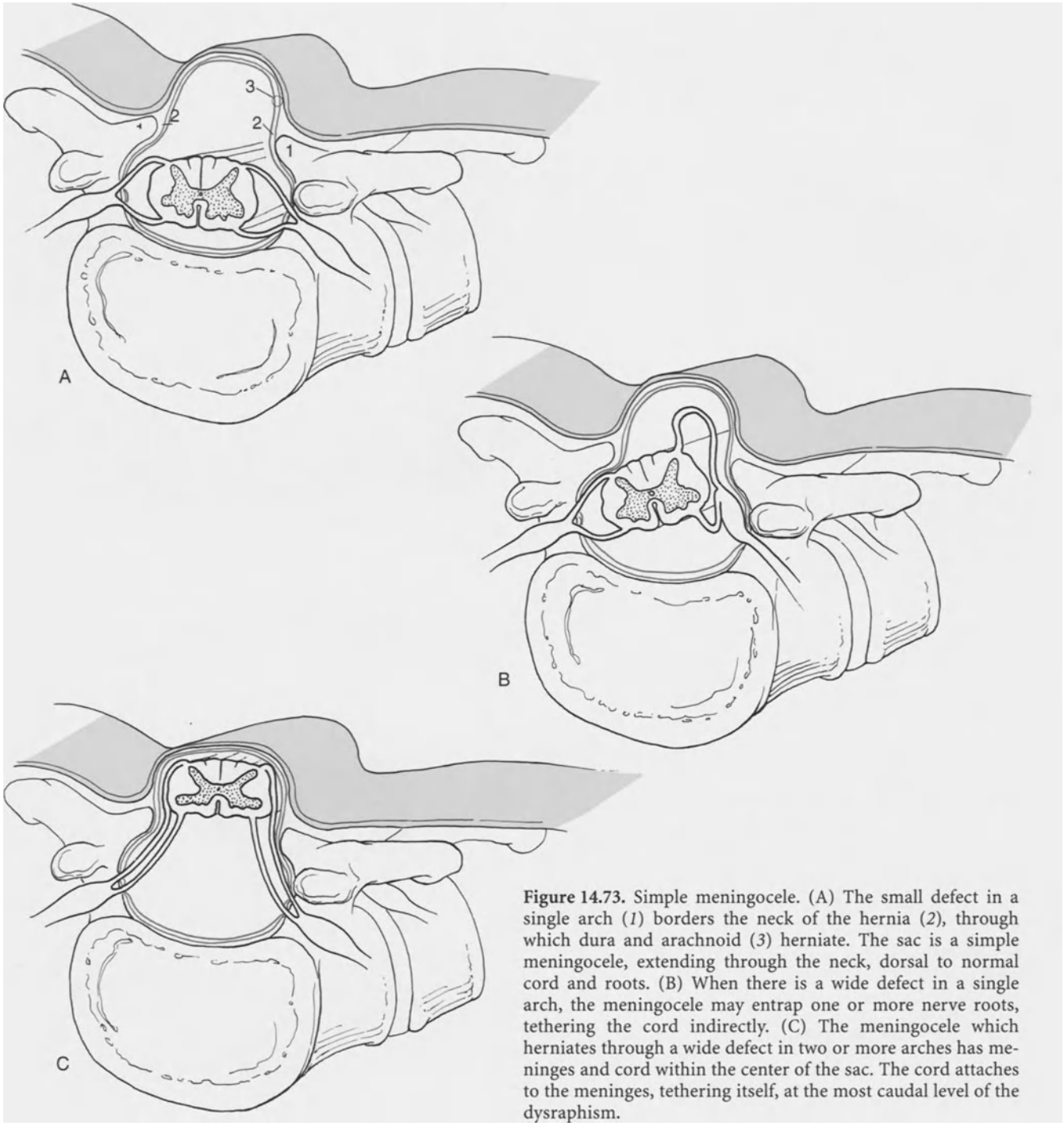


Figure 14.73. Simple meningocele. (A) The small defect in a single arch (*1*) borders the neck of the hernia (*2*), through which dura and arachnoid (*3*) herniate. The sac is a simple meningocele, extending through the neck, dorsal to normal cord and roots. (B) When there is a wide defect in a single arch, the meningocele may entrap one or more nerve roots, tethering the cord indirectly. (C) The meningocele which herniates through a wide defect in two or more arches has meninges and cord within the center of the sac. The cord attaches to the meninges, tethering itself, at the most caudal level of the dysraphism.

Consequently, all roots exit from the ventral surface of the malformed cord. On the dorsal surface there are glial scars, dysgenetic ependyma, ectopic islands of neural tissue, and extensive areas of degenerative changes. This is important to remember because closure of a myelocele necessitates separating this nonfunctional, pathological tissue from the underlying opened cord and nerve roots (dorsal and ventral), prior to dural and fascia imbrication. One must not close exposed, contaminated granulation or epithelial tissue (placode surface) into the spinal canal: infection or a dermoid tumor may result.

Meningocele (Figs. 14.73, 14.74)

The hernia sac consists only of meninges, covered by skin or epithelialized tissue, and the posterior spinal arch defect is limited to one vertebra. The spinal cord is located normally within the spinal canal, resting upon its ventral surface. Nerve roots may float into the hernia sac or be tethered to its neck. When the posterior arch defect involves two or more vertebrae the spinal cord itself may be tethered to the neck of the hernia, closely resembling the meningomyelocele.

Surgical Technique for Meningocele Closure. If the meningocele is covered with skin at the time of birth, one may profitably wait until the child is several months old to repair it. If, on the other hand, it consists of a thin layer of epidermis, it is best to repair it immediately lest it rupture and serve as a portal of entry for bacteria.

An incision, elliptical in form, is extended along the midline from superior to inferior, to the base of the meningocele, where it is bifurcated to either side of the pedicle, taking care to leave enough skin for the closure. At the inferior portion of the pedicle, the incisions from either side of the pedicle are brought into the midline from whence a single incision is extended inferiorly for a distance of about 2 cm. Only the epidermal and dermal layers of the skin should be cut, attempting to avoid incising the underlying pachy- and leptomeninges. The skin bleeding is stopped as one progresses to extend the incision superior and inferior to the meningocele. This permits the surgeon to expose the fascia, facilitates exposure of the underlying dura, and *allows inspection of its contents for neural elements.* If the meningocele is covered completely by skin, one may make the incision around the pedicle in a line of one's choosing, allowing adequate skin flap for a cosmetically desirable closure. If, however, it is covered by a thin layer of epidermis, one should make the incision along the line of transition between full-thickness skin and epidermis, postponing the planning of the skin closure until after the meningeal component of the meningocele has been repaired. In those instances in which the meningocele is covered only by a thin layer of epidermal tissue, the

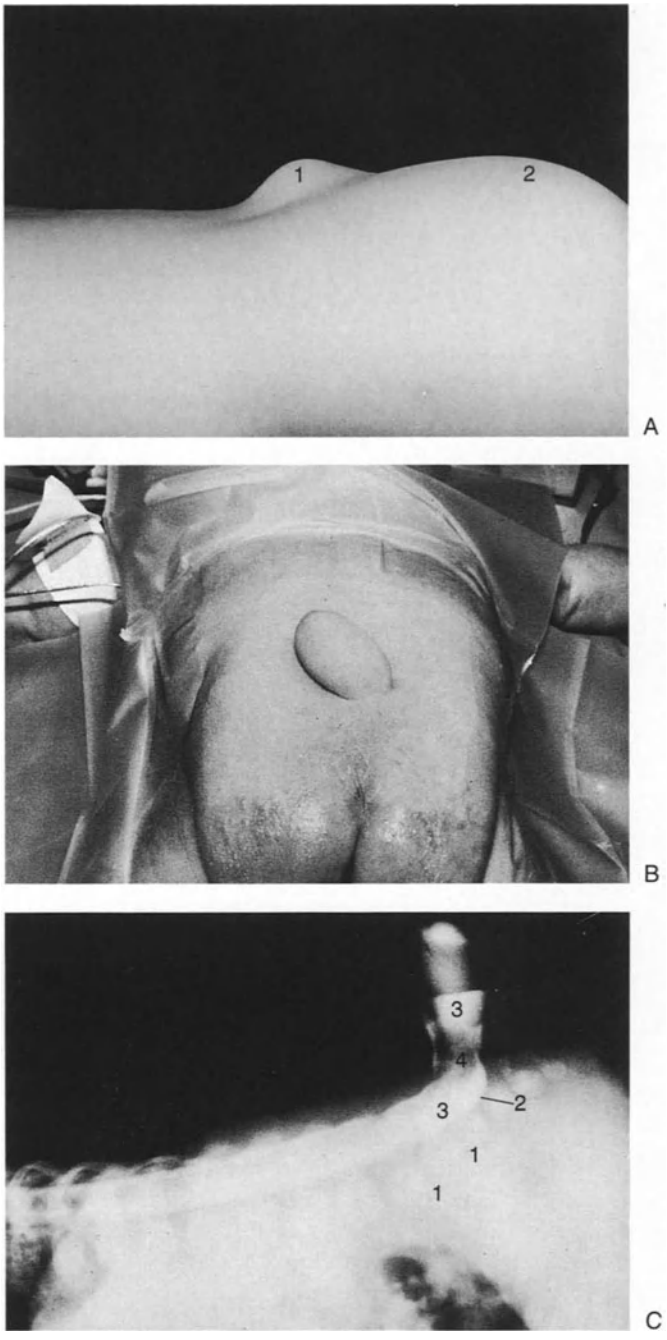


Figure 14.74. (A) This is a lateral photograph of a child with a meningocele (*1*) located at the lumbosacral area, immediately superior to the buttocks (*2*). (B) This child has a relatively large, fingerlike meningocele located at the lumbosacral junction. (C)Myelography reveals that the laminae of two vertebrae (*1*) are dysraphic, permitting exit of the neck (*2*) of the meningocele (*3*). The spinal cord (*4*) is tethered at the neck. This is the spinal cord *variety* of tethered meningocele, the type illustrated diagrammatically in Fig. 14.73B.

dura will be opened unavoidably when one cuts along the transition of full-thickness skin into epidermis.

In both instances, once the skin incision has been completed and the loose subcutaneous connective tissue separated from the underlying fascia, the exposed dura, whether intact (as in the case of meningoceles covered with skin) or dysgenetic (as in the case of meningoceles covered with epidermis) is opened. An Adson-Brown forceps is used to grasp the dura and put it under slight traction prior to cutting it with tenotomy scissors. Magnification is essential to ascertaining that no neural elements are damaged. The egress of cerebrospinal fluid occurs immediately if the meningocele is covered only by an epidermal layer, but not until the arachnoid is opened if the covering is full-thickness skin. In the former event, the surgically isolated portion of skin is separated from the underlying dura and discarded. The dura is then cut in the midsagittal plane, from the inferior to the superior extremities of the dysplastic posterior vertebral arch and then incised perpendicular to the sagittal cut on both sides, superiorly and inferiorly. This fashions two trapdoors. The arachnoid should be intentionally opened only to free and reduce the tethered roots (small hernia neck) or spinal cord (large hernia neck) in the event they are present. Whether it is intentionally or inadvertently opened, it should be closed before reconstructing the dura.

One of the two fashioned dural trapdoor openings is then laid snugly over the arachnoid and sewn to the inner surface of the dura on the opposite side, maintaining the normal dural circumference, using 4–0 or 5–0 sutures. If it is necessary to imbricate the dura at the superior and inferior durotomies in order to eliminate the pouch, this should be done with interrupted sutures. The dural trapdoor from the other side is then brought over the now closed and imbricated dura, sewing it down so as to provide additional support to the area of prolapse. Fascia from the paraspinal muscle masses is fashioned into a flap on either side, and, in turn, imbricated one over the dura first and then the other over the fascial flap, giving a four-layer closure. The skin is closed with interrupted mattress sutures.

Meningomyelocele

In the meningomyelocele the spinal cord is herniated into a sac of intact meninges which is located in the subcutaneous space. It may either be flattened out over a ventral arachnoidal cyst *(cystic meningomyelocele),* or it may be located normally within the spinal canal but have at its center an enormous hydromyelic cavity *(meningomyelohydrocele)* which distends and deforms its dorsal surface, pushing it into the hernia sac. Thus, the meningomyelohydrocele has three layers within the hernia sac: dura, arachnoid, and deformed dorsal surface of closed spinal cord.

In the cystic meningomyelocele, dysgenetic neural tissue (consisting of any combination of portions of spinal cord, conus medullaris, and nerve roots) is pathoanatomically continuous with the epidermal membrane externally and spinal cord elements centrally. There is no functional continuity, *the "placode" is not a functionally or anatomically intact structure,* nor does it hold anatomically intact neural elements. It is as much dysgenic neural tissue as are the epithelial sac and dysraphic vertebral arch, respectively, dysgenic skin and somites.

One should speak of *surgical closure, not surgical repair* of a meningomyelocele. It is impossible to repair (that is to say to restore anatomical and functional integrity). The procedure, therefore, closes the opening between the spinal canal and the environment, bringing neural and connective tissue elements, respectively, into the spinal canal and onto one another. This seals cerebrospinal fluid within the spinal canal and bacteria and foreign substances from it.

The operative procedure consists of making an incision along the point of transition between skin and epithelial membrane, stopping individual bleeders as one proceeds by applying small-toothed hemostats to the underlying connective tissue, or by using very low voltage bipolar coagulation. Once the skin has been incised, full thickness, around the entire circumference of this transitional area, it is separated from the underlying loose subcutaneous connective tissue until the paravertebral muscle fascia is identified. One then dissects one's way immediately beneath the approximately 1-mm lip of dermoepithelial membrane transition tissue, until the subarachnoid space is entered, at which time cerebrospinal fluid wells out of the field. This occurs whether the flattened cord is floating on a ventral arachnoid cyst (cystic meningomyelocele), or it has a grossly dilated central canal with the thinned posterior cord adherent to the meninges (meningomyelohydrocele).

Cystic Meningomyelocele (Fig. 14.75)

The cystic meningomyelocele is a dysraphic state with ventral arachnoid cyst and a closed, deformed cord: the placode. If one identifies a placode along the dorsal surface of the sac, and is certain that this is not a *scarred myelocele,* one should proceed to open dura and arachnoid, dissect the epithelialized placode from the underlying roots (maintaining these latter intact if possible), discard the epithelialized placode, and then reduce nerve roots and intact spinal cord into the spinal canal, if possible, before proceeding with the meningeal, fascial, and skin closure.

Tenotomy or iris scissors are used to cut the arachnoid around the circumference of the now centrally placed, totally isolated, epithelial membrane surround-

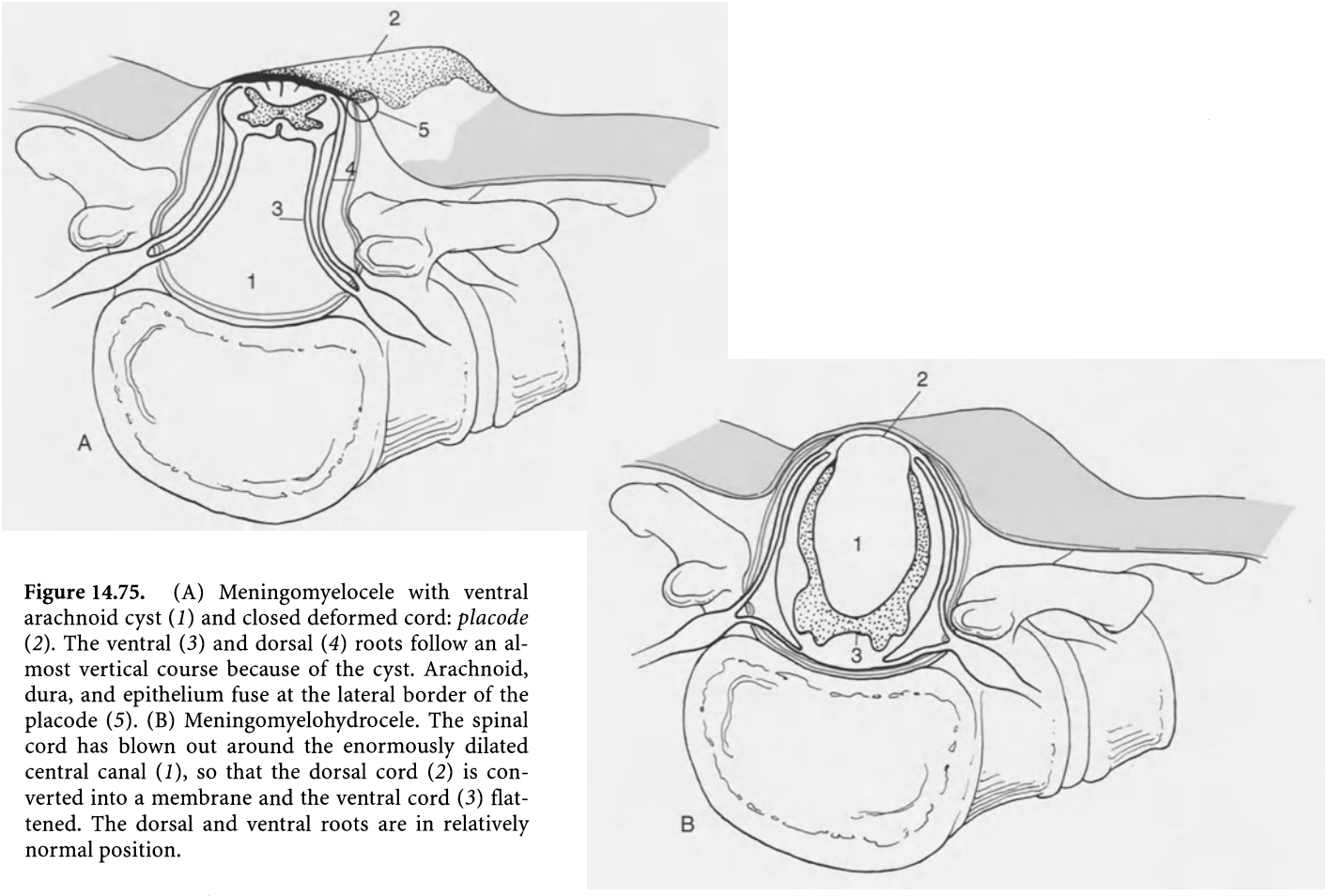


Figure 14.75. (A) Meningomyelocele with ventral arachnoid cyst (*1*) and closed deformed cord: *placode* (*2*). The ventral (*3*) and dorsal (*4*) roots follow an almost vertical course because of the cyst. Arachnoid, dura, and epithelium fuse at the lateral border of the placode (*5*). (B) Meningomyelohydrocele. The spinal cord has blown out around the enormously dilated central canal (*1*), so that the dorsal cord (*2*) is converted into a membrane and the ventral cord (*3*) flattened. The dorsal and ventral roots are in relatively normal position.

ing placode, separating this from the full-thickness skin. When the epithelial membrane and centrally placed placode are elevated, one sees dysgenetic nerve roots adherent to the undersurface of the placode. The epithelialized tissue and dysgenetic placode are then cut from the underlying posterior roots which may be adherent to it. The nerve roots are allowed to fall back into the intact inferior half of the neural arch, and the now freed placode and epithelial membrane discarded. If it is possible to identify arachnoid on either side, the two edges should be sewn to one another at this time. One should not attempt to force nerve roots or other dysgenetic neural tissue into the spinal canal, since this may result either in compression of these elements or adhesions between the neural and mesenchymal components of dysraphic tissue. Subsequently, the dura should be freed from the paraspinal muscle fascia on either side, as described above for meningocele, fashioning as generous trapdoor dural flaps as possible. These latter are brought first one over the arachnoid and underlying nerve roots, and then the other over it. The dural flaps are sewn to dura with interrupted 5–0 sutures, making every effort to attain a seal. The first trapdoor dural flap is brought across the congenital defect and sewn to dura on the opposite side, superiorly and inferiorly, taking care to avoid producing a constriction. The second dural flap is laid over the former, and sewn down similarly. Then, as described under meningocele, fascial flaps are fashioned and sewn one over the other, giving four imbricated layers of closure.

Meningomyelohydrocele (Fig. 14.76)

If the central canal is blown out into the hernia sac, its disrupted and dysgenic ependymal layer and the membranized dorsal portion of the spinal cord fuse with the arachnoid and dura to form the sac. This latter may be more or less epithelialized, though it is never covered completely by skin. At times, cloacal extrophy exists as an associated anomaly.

Its repair consists of an identification of as many individual anatomical elements of the sac as possible (though one seldom is able to identify more than a single fibroepithelial-appearing membrane), and then closure over the ventrally displaced and flattened spinal cord. If one extends the dissection into the area immediately medial to the paraspinal muscle masses, one may be able to identify arachnoid which has a glistening surface. If so, this should be closed and then the dura, in turn, identified, freed from the adjacent para-

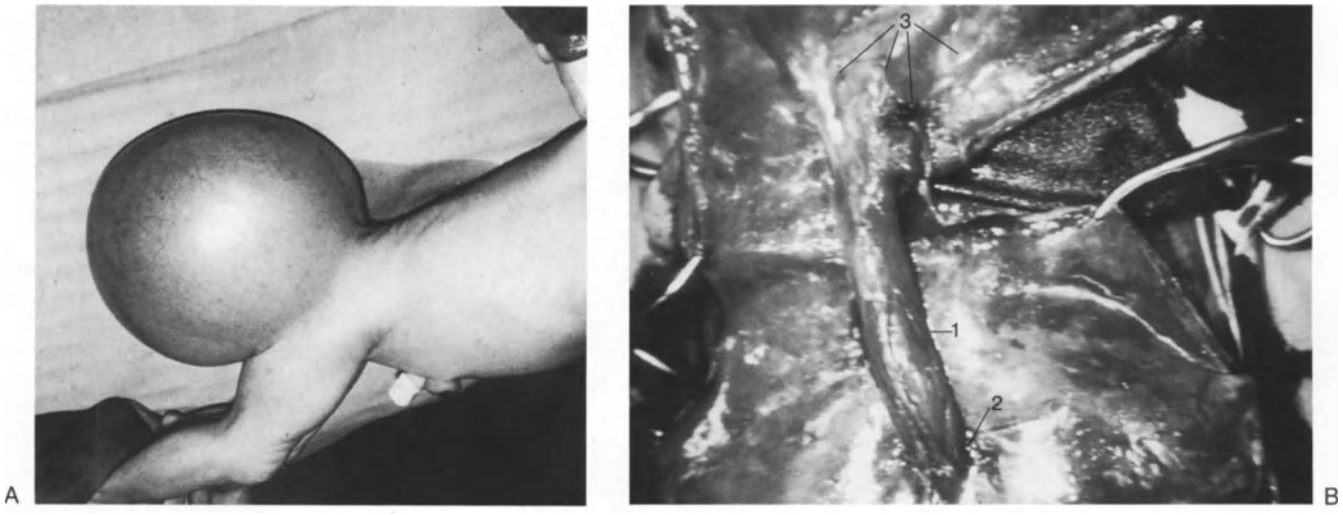


Figure 14.76. Meningomyelohydrocele. (A) The meningomyelohydrocele is located over the sacrum and buttocks. This child did not have an accompanying cloacal extrophy, something which is more obvious because of its grotesqueness than its incidence. (B) In this child, the spinal cord (*1*) is herniated through the neck (*2*), and tethered to the sac (*3*) of the hernia.

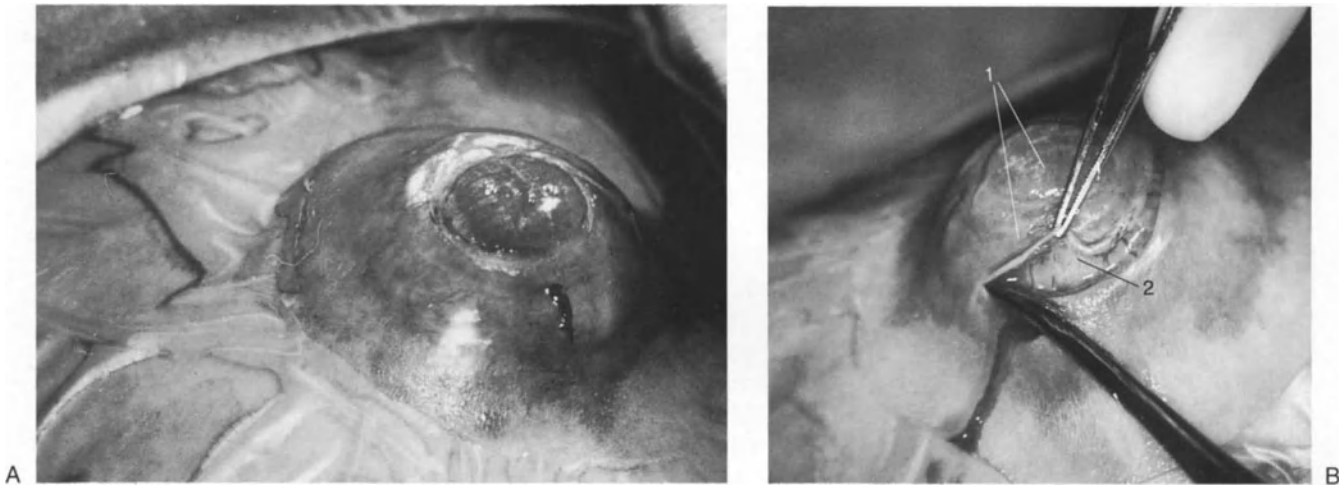


Figure 14.77. Stages in the closure of a myelocele, meningomyelocele, meningomyelohydrocele, and tethered (spinal cord or nerve root) meningocele. All four of these dysraphic states have in common herniation of meninges and neural elements into a sac composed of meninges and epithelial tissue which vary from nonrecognizable, luminescent membranes to completely developed anatomical structures. In its simplest terms, the closure consists of removing dysgenetic epithelium and meninges along with the epithelialized surface of the placode, in meningomyelocele; reducing the preserved neural elements into the ventral surface of the spinal canal without forcing them. Meninges, fascia, and skin are then closed. In the *myelocele,* the zona neurovasculosa must be preserved since it is the unclosed spinal cord. In the *meningomyelocele,* the vascular granulation and epithelial surfaces of the placode must be dissected from the underlying neural elements, discarding the former and preserving the latter. In the *spinal cord variety of tethered meningocele,* one frees the latter from the former prior to closure of the defect. In the *meningomyelohydrocele,* the redundant herniated epithelial and meningeal elements are resected; the fibrogliotic posterior rim of the cord is identified and preserved if possible (something which only rarely occurs).

The stages in closure: (A) After the surface of the hernia sac has been disinfected with surgical soap and germicidal prep, plastic draping is placed over the surrounding skin but not to the surface of the hernia sac. (B) The line of demarcation between the zona epitheliosa and nondescript tissue is identified and incised, permitting one to separate this tissue (*1*) from the underlying meninges (*2*).

spinal muscle fascia, and closed with imbricating sutures. The fascial and skin closures are the same as for a meningomyelocele and meningocele.

Concerning skin closure of all forms of the dysraphic state (Fig. 14.77), depending upon the size of the defect and the degree of posterior vertebral arch dysplasia, one may find it necessary to perform a Z-plasty in order to effect closure. Spurs of bone from the dysraphic articular and transverse processes should be rongeured to avoid compression necrosis of the skin. The skin is best closed with interrupted 4–0 mattress sutures, placing them alternatively at the superior and inferior edges, working one's way from peripheral to central, whether or not a Z-plasty is necessary. Thus, one may continuously correct for uneven segments of skin by lengthening or shortening the interval between sutures on one side or the other.

Diastematomyelia (Fig. 14.78)

Diplomyelia is invariably present, to a greater or lesser degree, when there is either a diastematomyelic spur or fibrous band. In essence, the spinal cord is cleft over a variable distance, with each independent medullary segment being covered by its own lepto- and pachymeninges, by a spur which generally has bone at its base, cartilage at its tip, and a connecting fibrous band extending from the cartilagenous tip to the dorsal segment of the vertebral arch. Occasionally, a lipoma may be present in this pathological tissue. The septum, or septum and lipoma, in its simplest terms, is the point of division between the two spinal cord segments, and is pathogenetic only in that the spinal cord presses against it as the child grows, or flexes and extends the vertebral spine. Consequently, one may have intermittent episodes of neurological deficit, or, more commonly, slowly progressive neurological deficit, referable to the spinal cord. The deficit is not radicular in nature, as one sees in the lipoma: it is medullary, upper motor. At times the bony spur is not present but, rather, the cleft between the double spinal cords contains a more or less dense fibrous band which stretches from the ventral to the dorsal surfaces of the spinal canal. The net result, when clinically obvious, is the same since the fibrous band is strong enough to compress the spinal cord.

A lipoma may be within the spur or attached to it. Hence *one could be justified in considering diastematomyelia as an intermediate congenital anomaly between the dysraphic state and hamartomous lesions.*

Irrespective of any other considerations it is recommended that the diastematomyelic spur, osteocartilagenous or fibrous, with lipoma if present, be resected immediately the diagnosis is made, even if the diagnosis is one of serendipity. The risks of surgery are so minimal and the possible damage to the thoracic spinal cord, the level at which the diastematomyelic spur most often occurs, is so great that this course of action is fully justified.

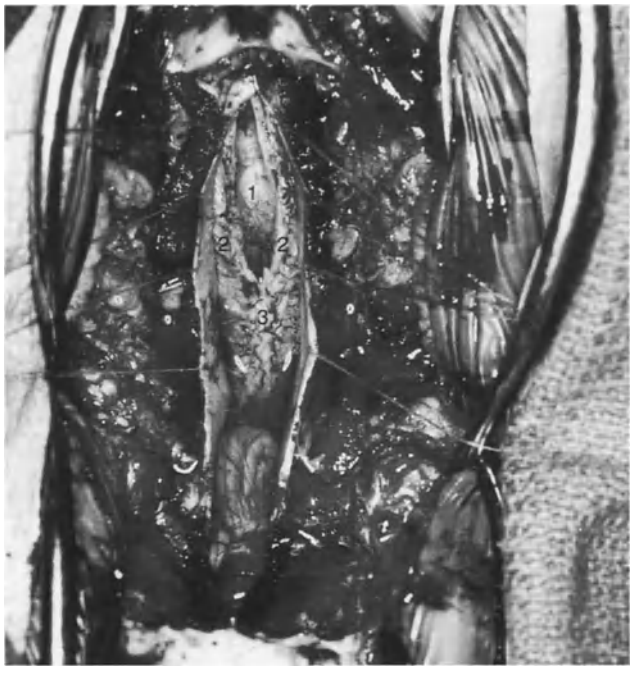


Figure 14.78. Lipoma of diastematomyelia. The diastematomyelic spur consists only of a fibrocartilaginous band. The diastematomyelic mass consists of a well-circumscribed and encapsulated lipoma whose bed (*1*) may be seen between the two spinal cords (*2*) extending caudally from the single cord (*3*). It was necessary to open the dura in order to identify and remove the lipoma. This entity is a transitional phase between the dysraphic state and hamartoma.

The operative procedure consists of performing a three-level laminotomy centered over the diastematomyelic spur. Care must be taken in removing the laminar flap, since one should avoid attempting to roll the flap away from the underlying diplomyelic deformity until one is certain that either there is no fibrous band connecting the diastematomyelic spur to the posterior surface of the spinal canal or, if one is present, that it be cut away before the laminar flap is lifted from the field. Once this is done, the surgeon has a view of the spinal cord, a single spinal cord above and below the diastematomyelic spur, with a segment of diplomyelia, each covered by intact dura mater, on either side of the spur. With few exceptions, one may state that the spur is extradural, so that its removal does not necessitate opening the dura mater. In fact, this is to be avoided. When lipoma is present, however, it is generally intradural. In these cases, one must open the dura.

If the spur consists of no more than a fibrous band, the operation will have terminated when the laminotomy flap is lifted from the field, since the surgeon already will have transected the bridging, anchoring, fibrous band running from the ventral to the dorsal sur-

faces of the spinal canal. He might choose to inspect the cleft between the diplomyelic segment of the spinal cord, to assure himself that there is not a cartilagenous or bony segment protruding from the ventral surface of the spinal canal, one which could potentially be pathogenic.

When the diplomyelic spur is an osteocartilagenous spur, the surgeon should remove it with the finest tipped rongeur available, biting away 1- or 2-mm fragments at a time. The disadvantage of using a high-speed drill, even one with a diamond bur, to do this rests in the necessity of retracting the diplomyelic segments of the spinal cord laterally, thus running the risk of compressing the spinal cord. It is preferable, therefore, to use the fine-tipped rongeur, inserting it so that the jaws open in the longitudinal (sagittal) plane, avoiding the risk of damaging the spinal cord in its narrowest, most vulnerable, horizontal plane. There is generally little or no bleeding associated with the removal of the spur. After the diastematomyelic spur has been removed, the operative procedure is considered completed. There is no need to open the dura mater.

Dysraphic Hamartomas

What are commonly referred to as lipoma or dermoid tumors in neurosurgical parlance are, in fact, heterogeneous groupings of congenital and developmental anomalies which belong in the neuropathological category of *hamartomas* in dysraphic children. *The dysraphic hamartomas may be lipomatous, dermoid, or endodermal.* The simplification of grouping all *lipomatous hamartomas* into *lipoma* and *lipomeningocele* has resulted in recommending surgery for every child with subcutaneous fatty tumor, and in very confusing interpretations of surgical results. It is recommended, therefore, that one speak of lipomatous hamartomas, and that they then be subclassified as (1) *lipoma,* (2) *leptomyelolipoma,* (3) *dural fibrolipoma,* and (4) *lipomeningocele.*

Lipomatous Hamartomas

Lipomatous hamartomas (the so-called lipomas), almost never cervical, very rarely thoracic, most commonly lumbar, present as subcutaneous masses, variable in size, which may remain in the midline or expand over one or both gluteal areas. They present either as subcutaneous masses, without neurological deficit, or with varying degrees of radiculopathy.

Lipomas (Fig. 14.79)

By far the single most common dysraphic hamartoma, lipomas are generally located within the region of the filum terminale and cauda equina. They either remain confined within the subarachnoid and subdural spaces or perforate the meninges at one discrete point to expand through the defect in the posterior vertebral arch and within the subcutaneous spaces.

The indications for surgery are either aesthetic or neurological. If the child is perfectly normal neurologically, the only reasonable indication for surgery is aesthetic. If a neurologic symptom or deficit becomes apparent, however, surgery is to be recommended.

Some neurosurgeons, especially a few years ago but always less at the present time, are recommending elective resection of the lipoma even in children without neurological symptoms or deficit. There is no evidence that the children they operate electively do not subsequently develop a neurological deficit. *In fact, quite the opposite is true.* It appears that the recommendation for elective surgery or for observing the child is one of judgment of the individual neurosurgeon, that one cannot assure a family either that the child will not subsequently suffer progressive neurologic deterioration or, if he does, that it will not be as severe as if he were not operated on "prophylactically." We prefer to observe asymptomatic children, and to operate on only those who develop clinical signs or symptoms.

In fact, with regard to the above, we call attention to the fact that the natural history of hamartomatous forms of spinal (occult) dysraphism has not been precisely determined. "According to many authors, about one-half of patients will eventually become symptomatic due to the interference with spinal cord blood flow and metabolism. This leaves the other half of them without neurological problems, and for these surgery would not offer any benefit" [65].

Surgical Technique for Lipoma

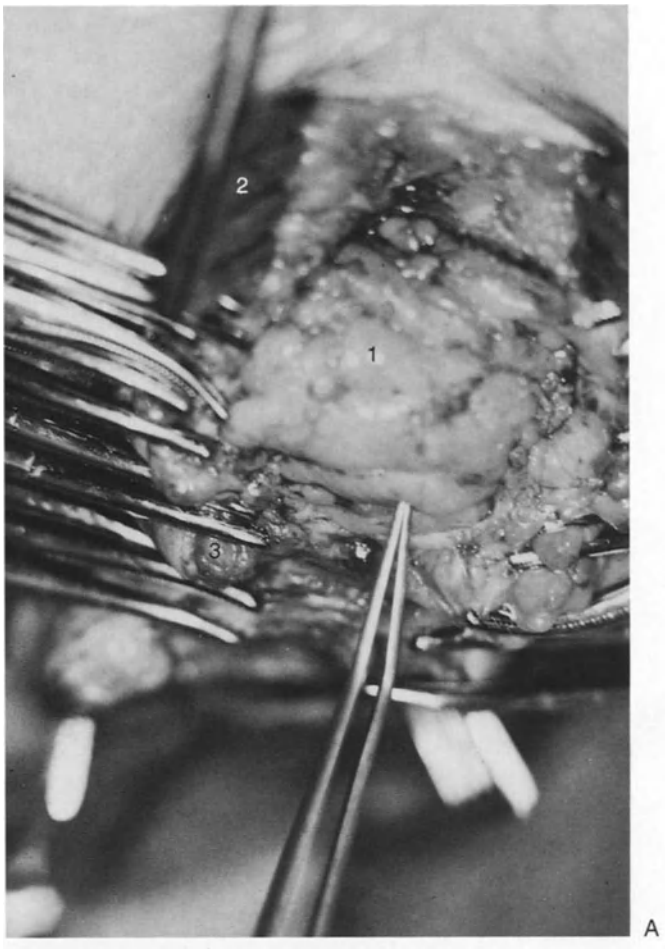
It is best to make a midline incision over the center of the subcutaneous lipoma, and then to retract the skin laterally before proceeding to resect the entire mass of subcutaneous fatty tissue. This is best done by working within the loose, subcutaneous, connective tissue surrounding the lipoma, freeing it completely from both the overlying skin and the underlying fascia of the paravertebral muscle masses. This enables the surgeon to elevate the entirety of the lipoma, so as to identify the area where it penetrates the dysraphic vertebral arches and intervertebral ligaments, prior to cutting the subcutaneous component of the lipoma from the hourglass extension into the spinal canal. One may cut this with impunity, since nerve roots are never located within this portion of the lipoma.

With the subcutaneous lipomatous mass removed, one has excellent visualization of the dysraphic and normal posterior vertebral arches, so that they may be cleared of paravertebral muscle masses by subperiosteal dissection, using periosteal elevators. It is recom-

mended that one strip muscular and periosteal tissue from at least two spinous arches beneath the hourglass extension of lipoma, and three above it. *The performance of laminectomy in order to expose the spinal canal is discouraged,* especially since these children already suffer the destabilizing effects of the dysraphic state, greatly predisposing them to scoliosis.

Most probably, the intermittent or progressive deterioration in motor and sensory function that occurs at varying periods of time, generally measured in years, after surgery is the result of scoliosis or instability secondary to laminectomy, not only or exclusively recurrent "tethering." Consequently, one is advised to perform a laminotomy, a procedure which not only contributes significantly to diminishing postoperative scoliosis, but also one which allows the surgeon to expose as extensive an area of the spinal canal as he chooses. It is recommended that the laminotomy be extended from two vertebrae below the hourglass extension of lipoma to the superior surface of the arch of the third vertebra above this transvertebral extension of lipoma.

Once the laminotomy has been performed and the epidural bleeders stopped, one may inspect the underlying dura above and below the transdural extension of tumor. Some surgeons recommend opening the dura from above downward, reasoning that they prefer to expose normal spinal cord first and then to work towards the lipoma. Others prefer just the opposite, for just the



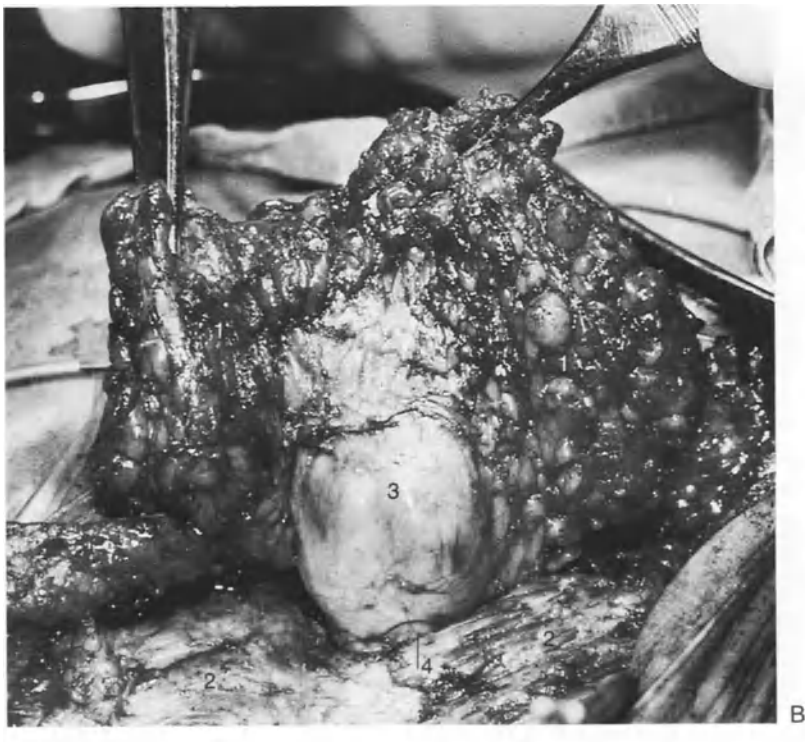
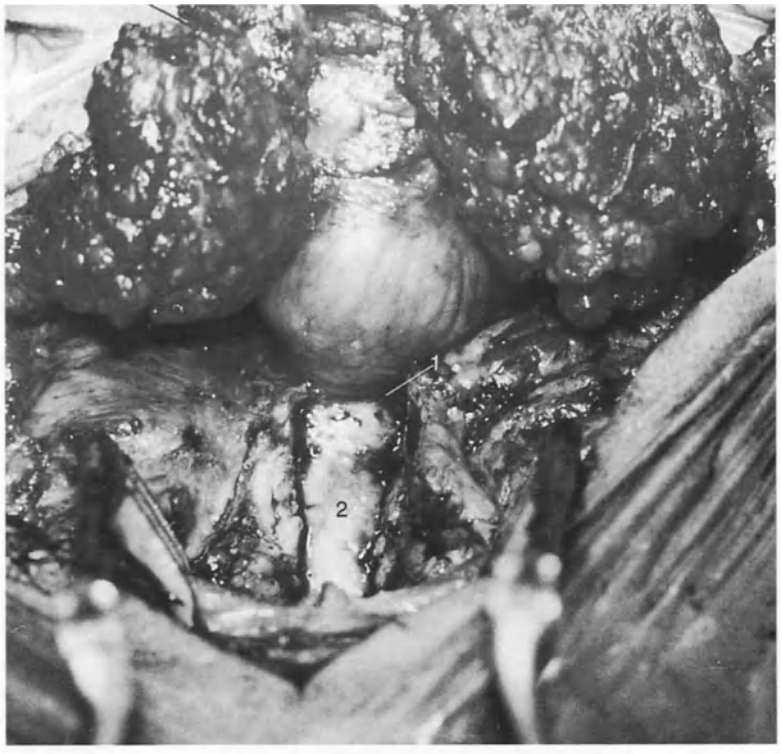


Figure 14.79. (A) Lipoma. The lipoma expands to either side of the dura, within both the epidural and subcutaneous compartments. The subcutaneous expansion may be within the midline or, as in this case, lateral to it. In fact, the lipoma (*1*) is located between the skin (*2*) and muscle mass (*3*) of the child's left buttock. (B) After the skin is incised and the subcutaneous portion (*1*) of the lipoma dissected from the underlying fascia (*2*), one may identify the extension of a lipomatous bundle (*3*) from the subcutaneous lipoma through the fascia at the neck of the hernia (*4*). (C, D) see p. 536.



C

D

Figure 14.79. (C) A laminotomy (not a laminectomy!) should be performed over two or three vertebral levels superior to the neck (*1*) of the lipoma. This uncovers the spinal canal, exposing the dural extension of this lipoma (*2*). (D) Leptomyelolipoma. In this anomaly, the lipoma (*1*), out of focus, is neither limited to the dura or remote areas, nor does it extend through the dysraphic posterior vertebral arches and dura into the subarachnoid spaces and spinal cord as a discrete mass. It does not compress, extrinsically, the neural elements. In fact, the leptomyelolipoma consists of fatty tissue and fibrous bands, neural elements and meninges, as an inseparable mass. Here, one notes the lipoma (*1*), the meninges (*2*), and lipomatous invasion of the cauda equina nerve roots (*3*).

opposite reasons. It is a matter of surgical judgment and familiarity: what works well for one surgeon is not necessarily correct for another.

In either event, whether beginning to work from the spinal cord downward, or from below the lipoma upward, individual nerve roots must be identified, so that one is obliged to work under the operating microscope and to proceed slowly in dissecting the lipoma from within the spinal canal. In essence, the entire indication for the procedure rests with successful decompression of the spinal roots coursing through or being splain over the lipoma. One does not, and cannot, resect completely all lipomas: some (the leptomyelolipomas) grow from within the center of the spinal cord, which tele-

scopes the fingerlike extension of lipoma from its center. Others, the *lipoma* and *dural fibrolipomas,* may occasionally be separated from the cord and roots. These facts, in essence, are the crux of disagreement between those neurosurgeons recommending elective removal of lipoma and those who prefer to observe the child, reserving recommendation for surgery until neurological deficits appear. It is not possible to know preoperatively whether one can remove a lipomatous hamartoma completely, since it may extend into the spinal cord, engulf the spinal cord and the nerve roots, or simply compress neural (cord and/or root) elements. The use of the laser eliminates traction on the lipoma, and, consequently, traction on the spinal cord and nerve roots. The Cavitron is equally effective for lipoma removal but the "flooded field" causes instances of blindness to tissue identification. Intraoperative somatosensory evoked potentials may be helpful, but are generally of little value.

Once one has removed all of the lipoma one is *comfortable* with removing (*this*, and only this, *should represent the end point* of the operative procedure), the dura is closed. It is only rarely possible to remove the lipoma completely, since it extends into the spinal cord! Inspection of the cul-de-sac allows one to ascertain that a tongue of fatty tumor is not left attached to neural elements at one end and dura at the other, lest the end subsequently become tethered. It is often impossible to close the dura without bringing a fascial graft into place. The laminotomy flap should be brought back into position, anchored down, and the paraspinal muscles, in turn, sewn to the interspinous ligaments. Redundant skin is resected and the remainder of the closure is effected with inverted mattress sutures. *The operative procedure itself predisposes to the production of tethering by the adhesions between cord, residual lipoma, closed dura or dural graft, and subcutaneous connective tissue dermal scar.*

Leptomyelolipoma

The simple lipoma is extremely difficult to distinguish from the leptomyclolipoma, which may expand from within the *conus medullaris* out into the *cul-de-sac,* engulfing completely the filum terminale and varying numbers of dorsal and ventral nerve root components of the *cauda equina.* These latter tumors are difficult, if not impossible, to deal with surgically, since they are composed of fatty tissue, nerve fibers, meningeal strands, and fibrous bands. One simply is not able to distinguish a fibrous band from a meningeal strand from a nerve fiber, or from an entrapped nerve root.

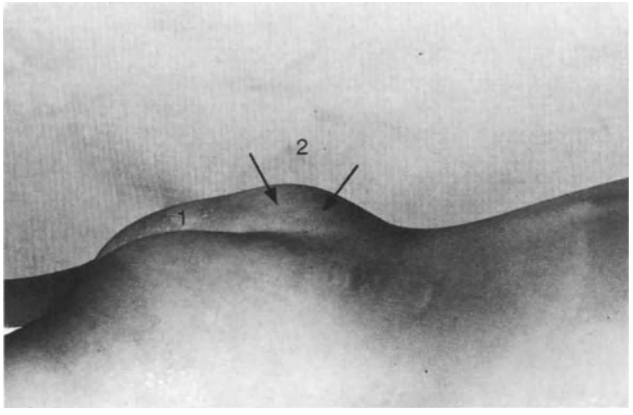


Figure 14.80. Dural fibrolipoma. The child's buttock (*1*) is to the left of the bulge from the dural fibrolipoma (*2*). After the skin has been opened, one immediately encounters the subcutaneous lipoma. Dural fibrolipomas may be adherent to the dura or, as in this case, remote to it.

Dural Fibrolipoma (Fig. 14.80)

In contrast to leptomyelolipomas, dural fibrolipomas are quite easy to identify and resect completely, since they expand as well-delimited tumor masses, dumbbell fashion, within the intra- and extradural compartments, on either side of the dysraphic posterior vertebral arch. These are similar in histology and point of origin to the lipomas which may be found within mesenchymal tissue, and dura, separating the two halves of a diastematomyelia.

Lipomeningocele

The lipomeningocele is, in essence, a lipoma which has expanded into both the extra- and intradural compartments and which has a cystic component at either end of the dumbbell.

In using classifications, the reader is urged to consider the use an author means them to serve. Lemire and colleagues [66] consider this entity as one in which the fatty mass expands on either side of dura, which it penetrates as a fibrous stalk, but state little more since their perspective is developmental pathology. We have chosen to use the anatomically more detailed, and surgically valuable, classification, which has evolved from the works of James and Lassman [59], and Karch and Urich [67].

Dermoid Hamartoma (Figs. 14.81–14.85)

Dermoid hamartomas are announced clinically by a tuft or patch of hair, abnormal pigmentation of the skin, a dimple or pit or sinus tract connected to an intraspinal dermoid tumor. There may be digitlike growth along the midsagittal plane of the body.

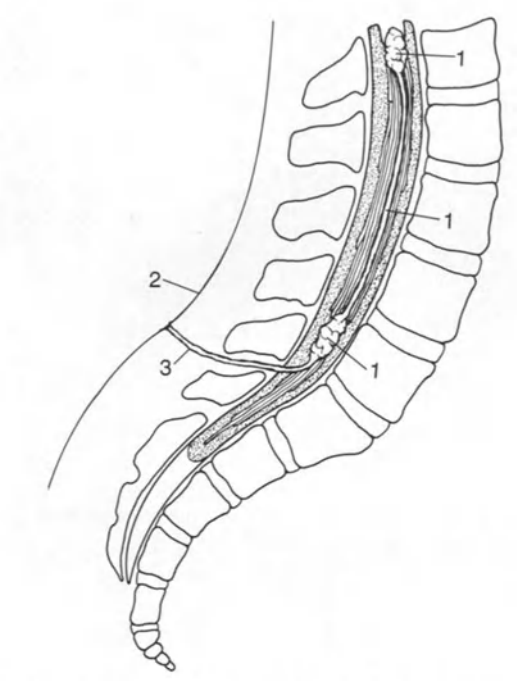


Figure 14.81. Dermal hamartomas (*1*) are connected to the skin (*2*) either by a solid or tubular stalk (*3*).

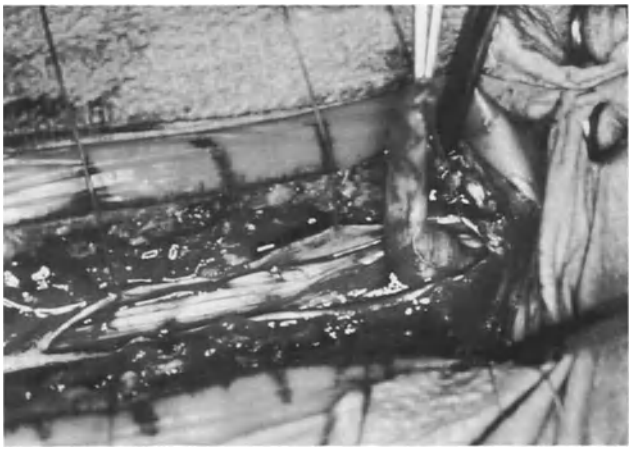


Figure 14.82. The dermal hamartoma may extend into the subarachnoid space as fibrous bands, stalks, tubes, or (as in this case) lipomatous masses.

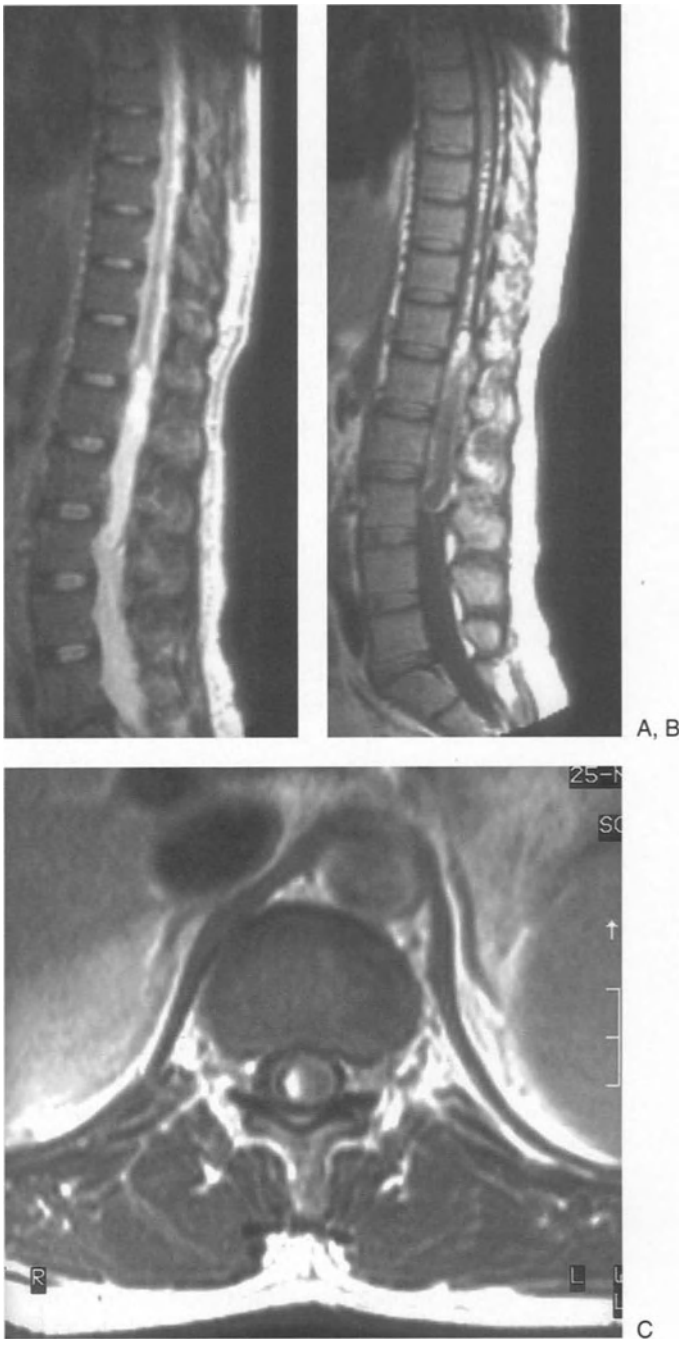


Figure 14.83. This is a spinal dermoid tumor of a 32-year-old man, which became symptomatic approximately 3-months before the time of diagnosis and subsequent surgery. At levels T12–L3, there is an intradural cystic mass which is enlarging circumferentially the conus medullaris. The lesion content is prevalently of high signal intensity on the T2 weighted images (A) and low signal on T1, secondary to cystic entrapment of fluid. There are some areas of high signal on the T1 weighted images (B,C), which represent fat tissue inside the lesion. Fatty tissue is also present as a subcutaneous lipoma extending from T8 to L3. On the T1 weighted axial image (C) the fat is identified along the wall of the cyst on the right.

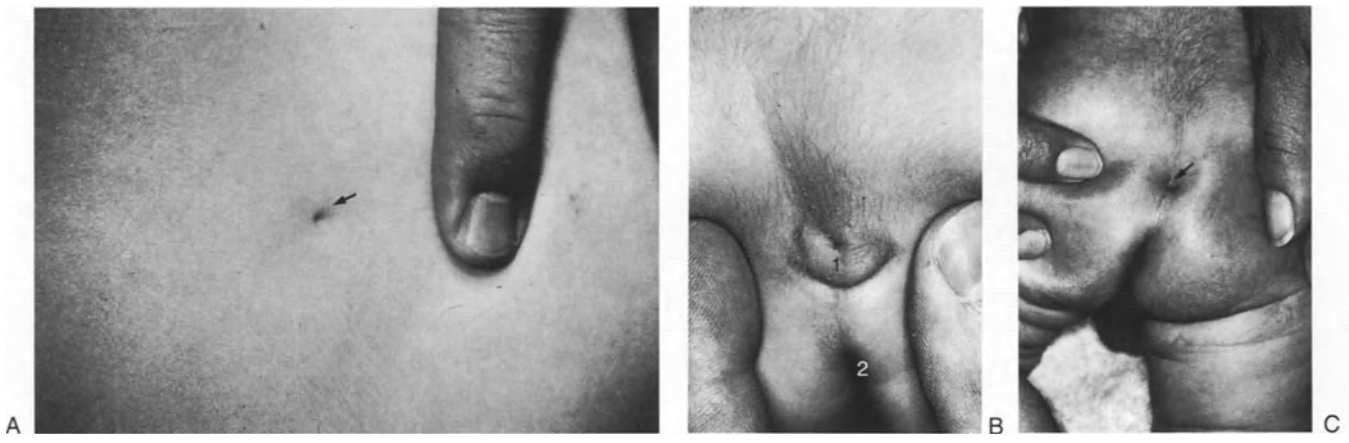


Figure 14.84. Dermoid sinus tracts, extending through the skin, muscle, posterior vertebral arch or interspinous ligaments, dura, and into either the subarachnoid space or neural elements (cord and roots), are very variable in surface appearance. They are, however, perforations of the skin and, therefore, must be distinguished from dimples. (A) A tiny pit (*arrow*) is located at the lumbosacral junction. (B) There is a relatively large fold (*1*) located immediately above the intergluteal fold (*2*). (C) The pit (*arrow*) is located at the depth of a bluish excrescence of skin.

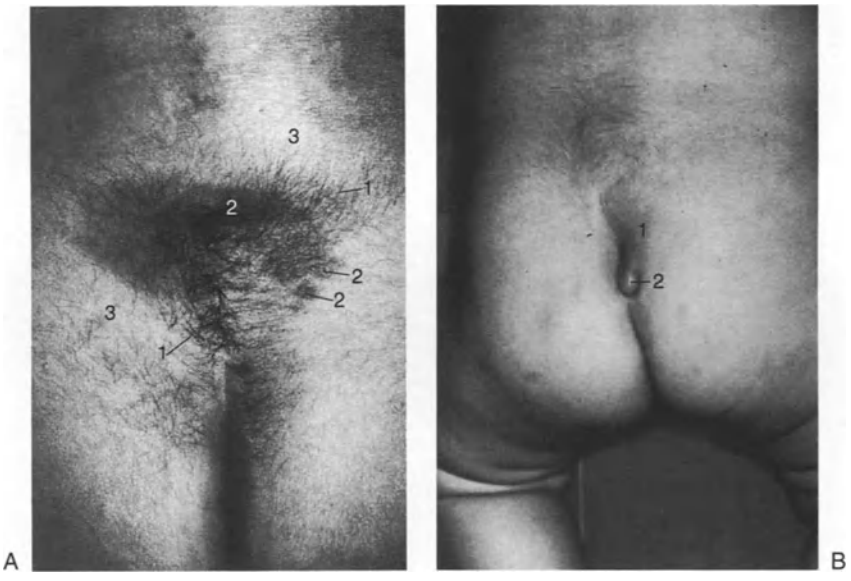


Figure 14.85. Though dermoid sinus tracts have a hole within the skin as a sine qua non for their diagnosis, dermoid tumors extending from the skin and into the spinal canal or cord may be associated with a subcutaneous mass, hypertrichosis, hemangioma, and digitlike extensions of skin. (A) Hypertrichosis (*1*), entodermal hemangioma (*2*), and redundant skin (*3*) overlie a lipoma and dermoid tumor extending from L5 into the spinal canal. (B) One notes the subcutaneous lipoma (*1*) and digitlike excrescence of skin (*2*) in this child.

The surgical technique for removing them consists of isolating the dermal sinus, if present, and performing an elliptical incision around it, taking care not to open it or empty its contents into the surgical field. Once the ellipse of skin has been cut and the incision extended into the subcutaneous area, it is best to tie a ligature around the sinus tract and cut away the portion in contact with the skin so as to remove it from the field. The dissection then extends around the sinus tract, or fibrous band, separating this latter from the surrounding tissue, and continues toward the posterior vertebral arch. If the tract extends through the vertebrae, it must be followed into the spinal canal and, subsequently, through the dura if it penetrates this structure. If one is obliged to enter the spinal canal, it is best to perform a laminotomy, and to plan to reconstruct the posterior

vertebral arches after the dermal tumor has been removed. Opening the dura allows the surgeon to evaluate the extent of intradural tumor and to remove it, if possible. Since these tumors consist of dermal elements, hair, epithelial rests, and so on, it is impossible to predict what one will find. When the dermoid tumor is associated with a lipoma, the mass may invade the cord making resection impossible. Removing dermoid elements from the *surface* of the roots and spinal cord, avoiding attempts to dissect tumor from *within* either of these neural elements, is the essence of this dissection.

Endodermal Hamartoma (Figs. 14.86–14.88)

The endodermal hamartomas may be associated with ventral meningocele, a result of failure of the two sides of the vertebral body to fuse to one another. A hernia sac extends into the retroperitoneal lumbar or sacral (very rarely retropleural) areas. At times, there is a fistulous connection to the gut. Consequently, one may encounter rudiments of intestinal tissue in either the hernia sac or spinal canal. Also, mucus-producing columnar epithelium may line the ventral surface of the spinal canal.

Surgical closure of the anterior meningocele entails identifying and opening the meningeal lining of the neck of the hernia sac, inserting a plug of bone graft into the defect in the vertebral body *(ies)*, and then using either a periosteal or *fascia lata* graft to close the dural defect. If the intra-abdominal (retroperitoneal) mass is of significant size, the general surgeon should remove it through a laparotomy.

Nondysraphic Spinal Cord Anomalies and Congenital Tumors

Hydromyelia and Hydrosyringomyelia (Fig. 14.89)

In its simplest terms hydromyelia consists of a dilation of the central canal. Though it very often complicates myelocele, meningomyelocele, and meningocele, extending the full length of the lumbar and thoracic cords, it seldom extends into the medulla oblongata. Depending, consequently, upon its severity and extent it may consist of a minimally dilated central canal or voluminous chamber which has expanded to fill the entire spinal canal, compressing the surrounding spinal cord into a membrane which is slightly thicker ventrally than dorsally.

Figure 14.86. The communication (*1*) between the anterior (intra-abdominal) meningocele and the spinal canal (*2*) is small and circular. One should use fascial grafts, sewn over inserted bone grafts, to seal the defect so as to minimize the risks of recurrence.

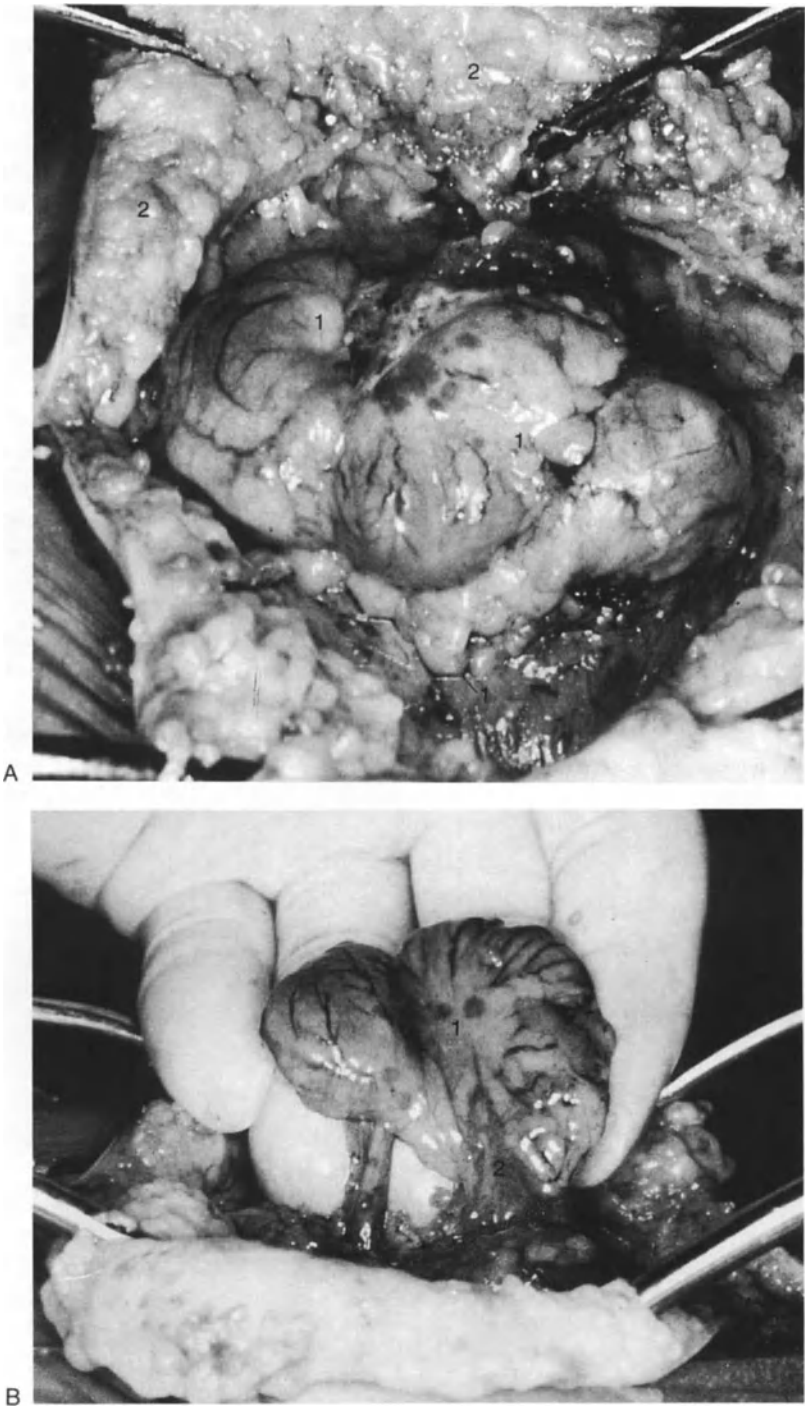


Figure 14.87. (A) Entodermal hamartomas may consist of herniation of bowel through the dysraphic vertebrae and into the subcutaneous compartment. Here, one notes vestigial bowel (*1*) occupying all the subcutaneous space. There is lipoma (*2*) adherent to the undersurface of the subcutaneous compartment. (B) One is able to identify the rudimentary bowel (*1*) and its mesentery (*2*).

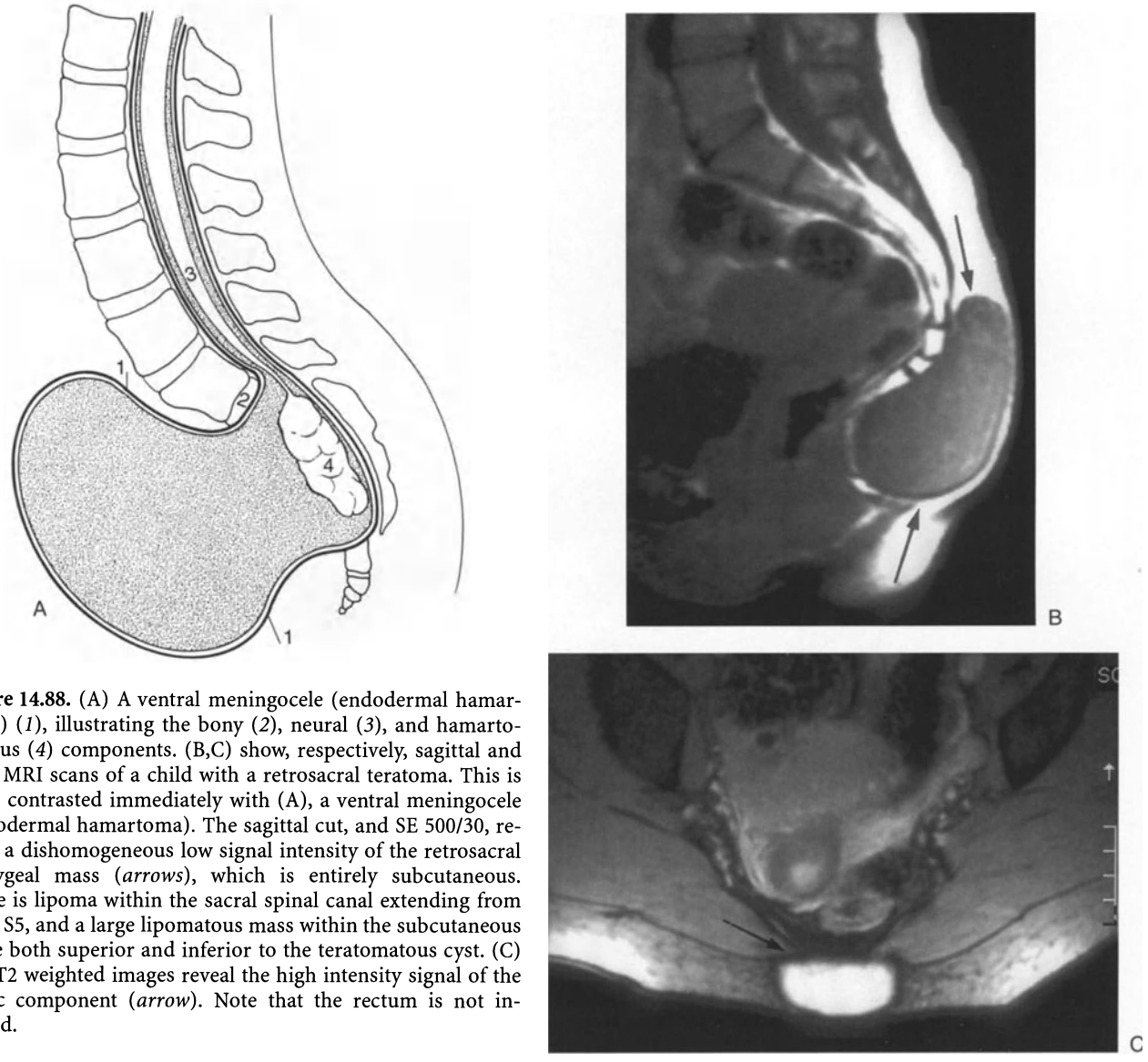


Figure 14.88. (A) A ventral meningocele (endodermal hamartoma) (*1*), illustrating the bony (*2*), neural (*3*), and hamartomatous (*4*) components. (B,C) show, respectively, sagittal and axial MRI scans of a child with a retrosacral teratoma. This is to be contrasted immediately with (A), a ventral meningocele (endodermal hamartoma). The sagittal cut, and SE 500/30, reveals a dishomogeneous low signal intensity of the retrosacral coccygeal mass (*arrows*), which is entirely subcutaneous. There is lipoma within the sacral spinal canal extending from S2 to S5, and a large lipomatous mass within the subcutaneous space both superior and inferior to the teratomatous cyst. (C) The T2 weighted images reveal the high intensity signal of the cystic component (*arrow*). Note that the rectum is not involved.

It is unfortunate mainly because of the confusion which results has resulted in the terms "hydromyelia" and "syringomyelia" being described as anatomical variations before the pathogenesis was understood. Of course, this was unavoidable, for the students of pathology who undertook the descriptions of these alterations worked with the autopsy material available to them: they had no surgical observations, no CT or MRI studies, and no long-term patient follow-ups.

We now know that the central canal dilation is both hydromyelia and the beginning of syringomyelia. We know that these occur as complications of cauda equina and medulla spinalis pathology such as ependymoma or hamartomatous lipoma, dermoid, and endodermal; *medulla spinalis* pathology such as ependymoma or glioma; *dysraphic states* such as meningomyelocele and the Chiari III malformation; and *craniovertebral anomalies* such as the Chiari I and II malformations, platybasia, and basilar impression. The common thread tying all these to dilation of the central canal and the subsequent rupture of the ependymal wall, permitting CSF to dissect its way vertically and horizontally, centrifugally along first the center then almost the full diameter of the spinal cord, is obstruction of normal spinal CSF circulation. This may begin at the foramen magnum, along the length of the spinal cord, at the conus medullaris, or within the area of the cauda equina. In essence, hydromyelia is very probably as simple, and as difficult to understand as is its cerebral counterpart, hydrocephalus. And, as hydrocephalus, very often when the causative factor has been successfully treated, e.g., meningitis or choroid plexus pupilloma, meningomyelocele or diastematomyelia, the complicating hydrosyringomyelia persists.

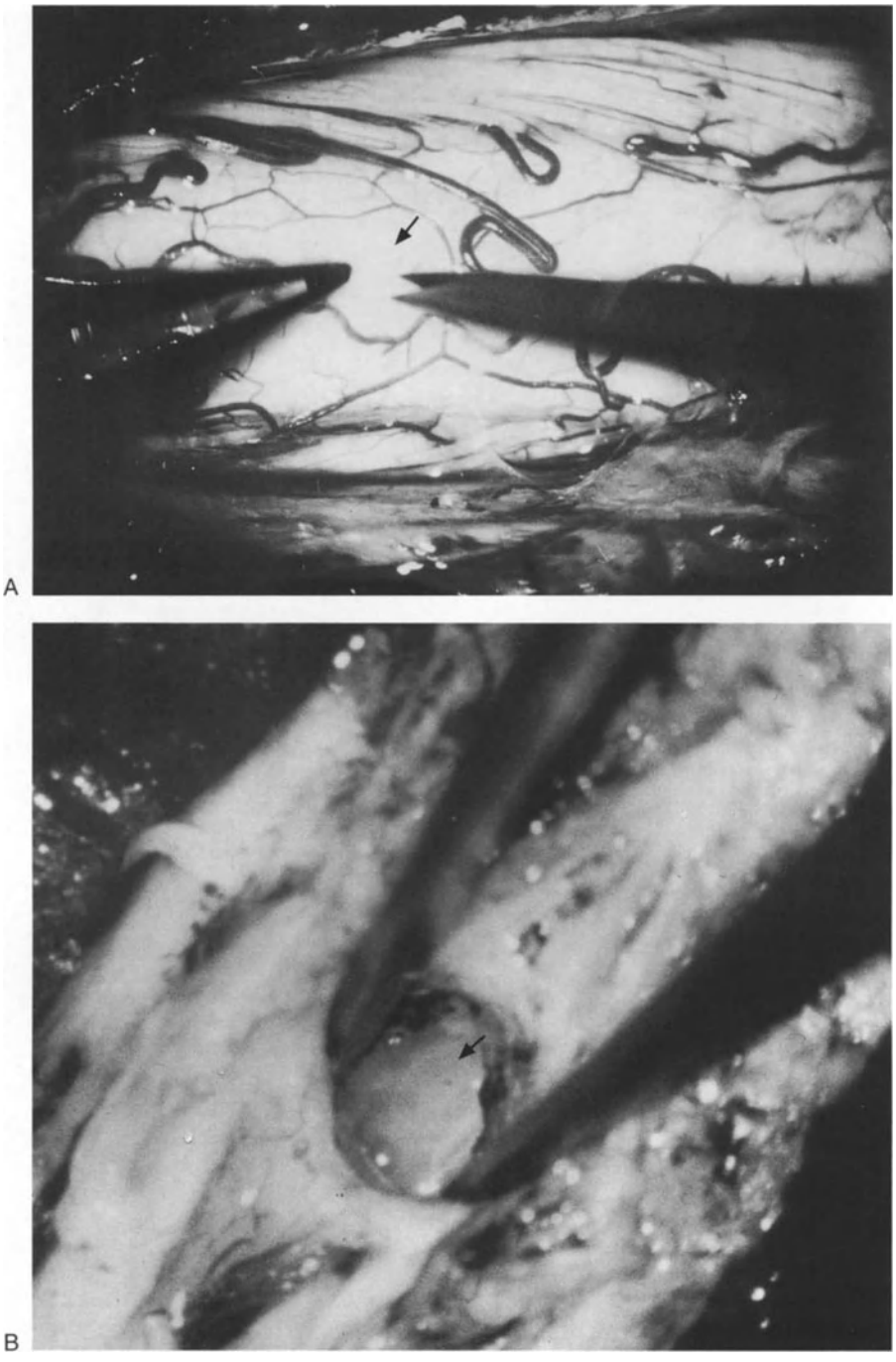


Figure 14.89. Syringomyelia. Whether idiopathic or traumatic, the surgical treatment for the non-Chiari malformation associated syringomyelia consists of marsupializing the syringomyelic cavity to the pial surface of the spinal cord and then closing the arachnoid, thus creating a fistula between the syringomyelic cavity and the subarachnoid spaces. (A) The marsupialization, if performed without a laser, is carried out by using the bipolar forceps to coagulate the dorsal surface of the spinal cord immediately above the widest portion of the syringomyelic cavity (*arrow*). The cord and wall of the syringomyelic cavity are then incised. (B) The bipolar forceps are used to open the cavity (*arrow*) and to coagulate the opening so as to impede glial scar formation and closure. This produces the marsupialization between the cavity and pial covering of the cord, thus permitting fluid from within the cavity to drain directly into the subarachnoid space. No tube is needed. (C, D) see p. 544.

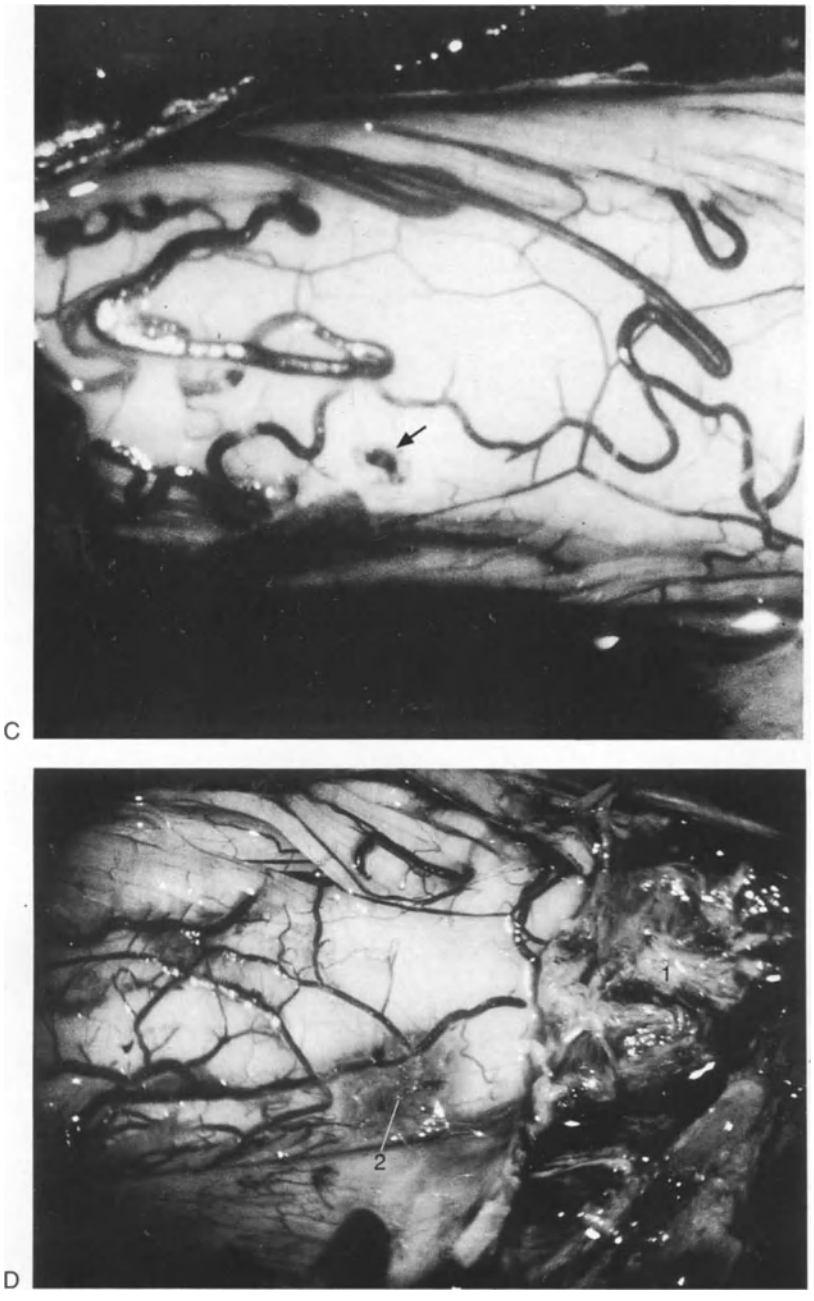


Figure 14.89. (C) If the laser is available, it is much more precise and effective in producing marsupialization between the syringomyelic cavity and the pial surface of the cord. In this illustration the laser is being used to perforate the pial surface and underlying wall of the syringomyelic cavity (*arrow*). (D) This patient suffered post-traumatic syringomyelia. The glial hypertrophy and scar formation (*1*) are seen *on the right*, and the pial wall of the post-traumatic syringomyelic cavity (*2*) *at the center*. A laser is being used to perforate the pial wall and enter the cavity.

Therefore, the treatment approaches are two: drainage of the hydrosyringomyelic cavity into another body chamber or the spinal subarachnoid spaces, and correction of the hydrodynamic dysequilibrium.

If the hydromyelia is severe, it should be treated. The recommended treatment of all forms of hydrosyringomyelia except those secondary to craniovertebral anomalies and the Chiari I and II malformations consists of laser penetration of the spinal cord, so as to establish a fistulous tract (marsupialization) between the hydromyelic central canal and the subarachnoid spaces. The arachnoid should be opened approximately 1.5–2 cm from the point of laser fenestration and then, after the fistula has been made, a watertight closure of the arachnoid performed. This permits CSF to flow from the central canal into the subarachnoid spaces. Opening the arachnoid as a trapdoor 1.5–2.0 cm away from the point of marsupialization minimizes the probabilities of arachnoid sealing the surgically created fistula.

Syringomyelia

The operative treatment for syringomyelia in association with hydromyelia has already been described. For the syringomyelia, with a dilated central canal, which is secondary to either a craniovertebral anomaly or the Chiari I malformation (see the sections above on Chiari malformations), the surgical treatment differs greatly! In fact, it is directed to the anatomical alterations to such an extent as to be considered pathological when associated with such symptoms and signs expressive of cerebellar incarceration as syncope, headache, inability to extend the head, paresthesiae, hind cranial nerve paresis, dissociated sensory loss, paresis, hyperreflexia, and ataxia. The anatomical alterations are displacement of the cerebellar tonsils into the cervical canal and their adherence to the posterior surface of the cord, absence of the cisterna magna and closure of the vallecula, diminution of the volume of the medullary and/or pontine cisterns, medulla oblongata compression, and ventriculomegaly. It is not uncommon for all of these anatomical alterations to be present, but such coexistence is not a prerequisite to the diagnosis.

The surgical treatment is directed at creating an anatomical "restructuring" which permits the reestablishment of physiological hydrodynamics at the craniovertebral junction: free flow of CSF out of the IV ventricle through the vallecula and around the cervicomedullary junction, the to-and-fro movement of CSF across the level of the foramen magnum, and the pooling of CSF within a chamber which functions as the cisterna magna along the inferomedial surfaces of the cerebellar hemispheres and tonsils.

Depending upon the severity of the anatomical changes, three variations, in degree, of decompressive surgery have proven to be very successful in reestablishing physiological CSF circulation at the craniocervical junction, eliminating the symptoms, and arresting or reversing the signs of cerebellar incarceration within the foramen magnum. These are: (1) suboccipital craniectomy and resection of the arch of C-1; (2) as in (1), plus relaxing plastic reconstruction of the dura mater at the cerebellomedullary junction posteriorly, using fascia lata graft to assure watertight closure; and (3) as in (2), with resection of the cerebellar tonsils and opening of the vallecula to permit egress of CSF from the IV ventricle.

The Tethered Cord (Fig. 14.90)

In its broadest sense, one could use the term "tethered" to indicate any bridling of the spinal cord or nerve roots, which results from a band of mesenchymal tissue extending from the spinal cord to the dura mater. Thus, a fibrotic *filum terminale,* extending from the tip of the *conus medullaris* to the dura, anchoring one to the other, may cause tethering. The cord may be anchored to the meninges of a meningocele. Similarly, a mass of lipomatous tissue extending from the center of the spinal cord through the dysraphic posterior vertebral

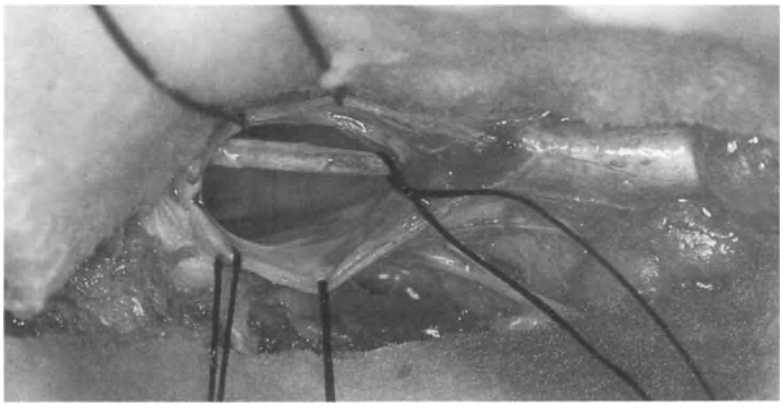


Figure 14.90. Tight thick filum at operation in child with tethered spinal cord.

arch may also cause tethering. A common cause of tethering is scar tissue binding the spinal cord and nerve roots of a previously operated meningomyelocele or lipomatous hamartoma to the repaired dura or skin.

Whether these pathoanatomical conditions are pathogenic is something which the physician must determine before deciding whether to recommend surgery. The simple identification of a continuity between cord, cauda equina, nerve roots and the dural sac is not justification for surgery. There must be a resultant neurological deficit *which one may expect to be cured or arrested* in its progress. Otherwise, what is to be gained?

The surgical implications for removing lipomatous tissue which may serve as a tethering band have already been described. Whether one is justified in reoperating on a meningomyelocele child, with a fixed neurological deficit, because myelography reveals tissue continuity between the terminal portion of the spinal cord and the area of meningomyelocele repair, is to be seriously questioned, since the tissue will scar again and, therefore, tether again. There is nothing to gain and the loss is clear: operative morbidity and further weakening of the vertebral spine.

The surgical procedure for freeing a fibrotic *filum terminale* which tethers the *conus medullaris* consists of nothing more than the performance of a laminotomy, opening the dura, identifying the tethering *filum terminale* at the tip of the *conus medullaris* (using the operating microscope so as to distinguish it with certainty from the surrounding nerve roots), and sectioning the filum. This frees the spinal cord from the potentially damaging effects of traction.

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15 Hydrocephalus

"... and so there ain't nothing more to write about, and I am rotten glad of it, because if I'd 'a' knowed what a trouble it was to make a book I wouldn't 'a' tackled it, and ain't a-going to no more. But I reckon I got to light out for the territory ahead of the rest, because Aunt Sally she's going to adopt me and sivilize me, and I can't stand it. I been there before."

MARK TWAIN, Adventures of Huckleberry Finn

Introduction (Figs. 15.1–15.9)

In his work, the seats and causes of diseases investigated by anatomy, G.B. Morgagni, in 1761, called attention to the fact that the earlier investigators of increased craniocerebral size did not have particularly clear ideas as to where beneath the skin the fluid present in hydrocephalus was located (Morgagni, cited in [1]). Specifically, pathologist, anatomist, and clinical pathologist were dubious as to whether the fluid was located beneath the skin, beneath the skull, beneath the dura, or within the brain itself. By and large, it is safe to assume that the anatomists and physicians prior to Vesalius were of the mind that the fluid accumulated beneath the scalp. Indeed, Vesalius [2] was apparently the first to give a clear description of internal hydrocephalus by stating

> Galen declared that this shape of the skull may be imagined to come from another world but not to exist in the nature which surrounds us. A boy was seen by me in Venice, one deformed and insane, with an enormous and mis-shapened head. Also, there was a beggar in Bologna with a square head which is wider than it is long. A beggarwoman in Genoa carries about a little boy as she goes from door to door, and gives him to comedians who would use him in their show to illustrate a head which is larger than the two comedians' heads together! It is my own thought that the boy suffers the same disease I first observed in Augsburg in a little girl who, at the age of seven months, had a head larger than that of any man I had ever seen. The disease I am describing was called hydrocephalus by the ancients because of the water which collected within the head but, in this child, the water did not collect outside of the skull and beneath the surrounding membranes nor did it collect within the skin – as most medical books teach – but, within the center of the brain itself, in the right and left ventricles of the brain. The depths and the breadths of these ventricles so increased, and the brain was so very swollen that they contained 9 lbs. of water, or 3 Augsburg wine measures (so help me God!). Just as the brain itself at the vertex was membrane-like in thinness indistinguishable from its own membraneous coverings so was the skull membraneous, but the base of the skull was in harmony with that of the young child before her head took on abnormal proportions. The cerebellum and the brainstem were in their natural state, so were the nerves coming from the brainstem. I found water in no other place in the ventricles of the brain; the girl was in control of all her senses until she died. When I examined her a few days before her death I noted that whenever her head was raised she coughed, her respirations became difficult, her face became red with the flow of blood, and tears dripped from her eyes.

This confirmed Vesalius's observation that the accumulation of fluid in hydrocephalus is within the ventricles, and established the basis for his conclusion that hydrocephalus in infants causes an increase in head size, but that this macrocephaly does not occur in the adult who may also suffer hydrocephalus.

Robert Whytt [1] first distinguished between internal and external hydrocephalus in 1768 by accentuating the fact that he had never encountered water between the dura mater and the brain, in the interhemispherical fissure, or over the corpus callosum, in the ten children between the ages of 2 and 15 years that he studied at necropsy. The notable absence of subdural hygroma, subdural hematoma, and subdural effusion in this study may well be due to his exclusion of children under the age of 2 years. Despite its cursory form his report definitely identified with precision the fact that in the more common forms of hydrocephalus cerebrospinal fluid accumulates within the ventricular system, dilating the ventricles and increasing head size.

Hydrocephalus has been considered simply to consist of an accumulation of cerebrospinal fluid, excessive in relation to brain volume, within the intracranial cavity. Accordingly, a distinction between external and internal hydrocephalus, depending upon whether the excessive fluid was over the surface of the hemispheres or within the ventricles, had long been made.

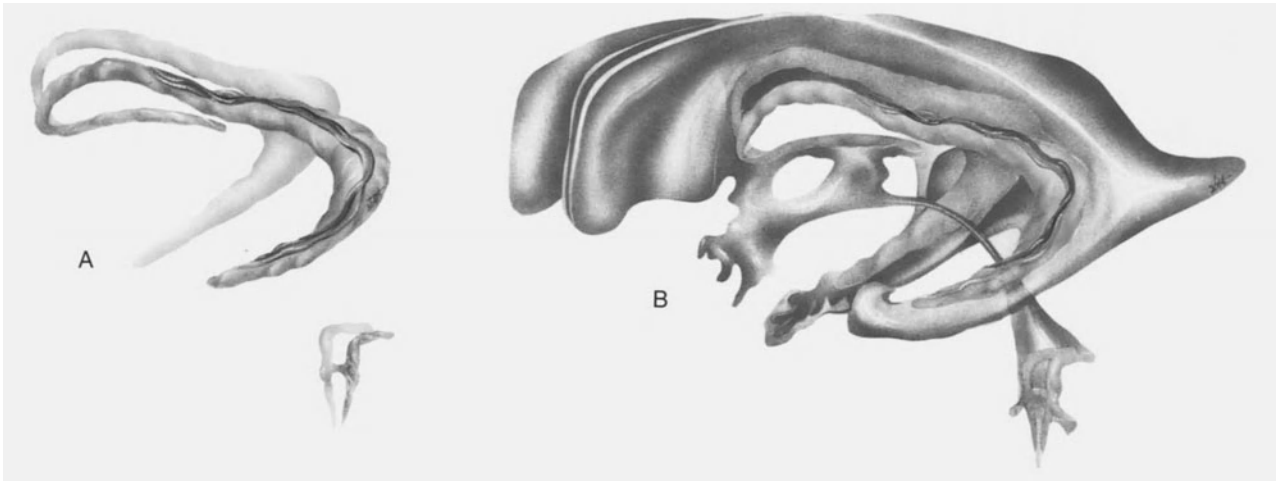


Figure 15.1. This is the first of nine sets of figures, all drawings representing diagramatically the structures producing cerebrospinal fluid, those chambers in which it is collected, the relationships between the CSF-containing chambers (ventricles, cisterns, sulci), and the cerebral parenchyma. They also show the cerebral parenchymal changes, vis-à-vis one another, which first hold back and then permit the passage of cerebrospinal fluid from the lateral ventricles into the III ventricle, and from the III ventricle into and through the aqueduct of Sylvius. (A) This is a drawing of the choroid plexus of the lateral, III, and IV ventricles. One notes that the choroid plexus "begins" within the temporal horn, and then passes posterosuperiorly to the glomus from whence it passes anteriorly to the foramen of Monro. Its passage through the foramen of Monro is continuous with its extension into the roof of the III ventricle. In the IV ventricle the choroid plexus has the appearance of two hockey sticks, one parallel to the other, connected in the form of the letter H. In (B) the ventricular system has been drawn over the choroid plexuses, permitting one to see, and readily identify, the temporal horn choroid plexus, the glomus of the choroid plexus at the trigone of the lateral ventricle, the lateral ventricle (body) choroid plexus running along the floor of the lateral ventricle to the foramen of Monro, and then the passage of the choroid plexus from each lateral ventricle through each foramen of Monro and into the roof of the III ventricle. Similarly, one may see the superimposition of the IV ventricle choroid plexus on the IV ventricle itself, extending from the fastigium, along the nodulus, and then laterally into the lateral recess. These anatomical relationships, choroid plexus to ventricular system, are extremely important to an understanding of the direction of flow of the cerebrospinal fluid. It is the pulsatile force of the choroid plexus which drives the fluid forward, in a to-and-fro manner, from posterior (dorsal) to anterior (ventral), and then from superior (cranial) to inferior (caudal) in its course through the ventricular system from lateral to III, from III into aqueduct, from aqueduct into IV, and out of the IV ventricle subsequently through the medial and lateral foramina into the cisterna magna and pontocerebellar cisterns, respectively.

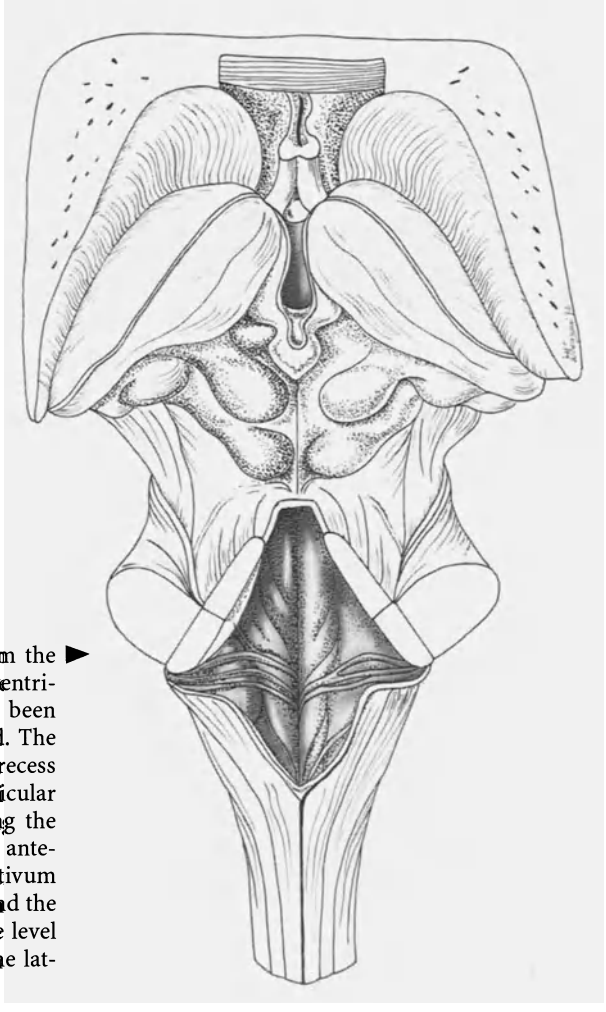


Figure 15.2. This posterior (dorsal) drawing of the brainstem, from the ► **mid-brain to the bulbomedullary junction, puts into relief the III ventricle wedged in between the thalami (the roof of the III ventricle has** been **removed). Note that the columns of the fornix have been sectioned**. The **most posterosuperior portion of the III ventricle extends as a r**ecess **above the pineal gland. Beneath this rests the pineal recess. The colli**cular **plate is the dorsal aspect of the tectum of the mid-brain, covering** the **aqueduct. The IV ventricle is particularly well visualized in that the** anterior **medullary velum has been taken away, the brachium conjunct**ivum **has been sectioned at the border of the anterior medullary velum, and** the **brachium pontis and the restiform body have been sectioned at the** level **of their entrance into the cerebellar hemispheres. One notes that th**e lateral **recesses extend inferior to the restiform body.**

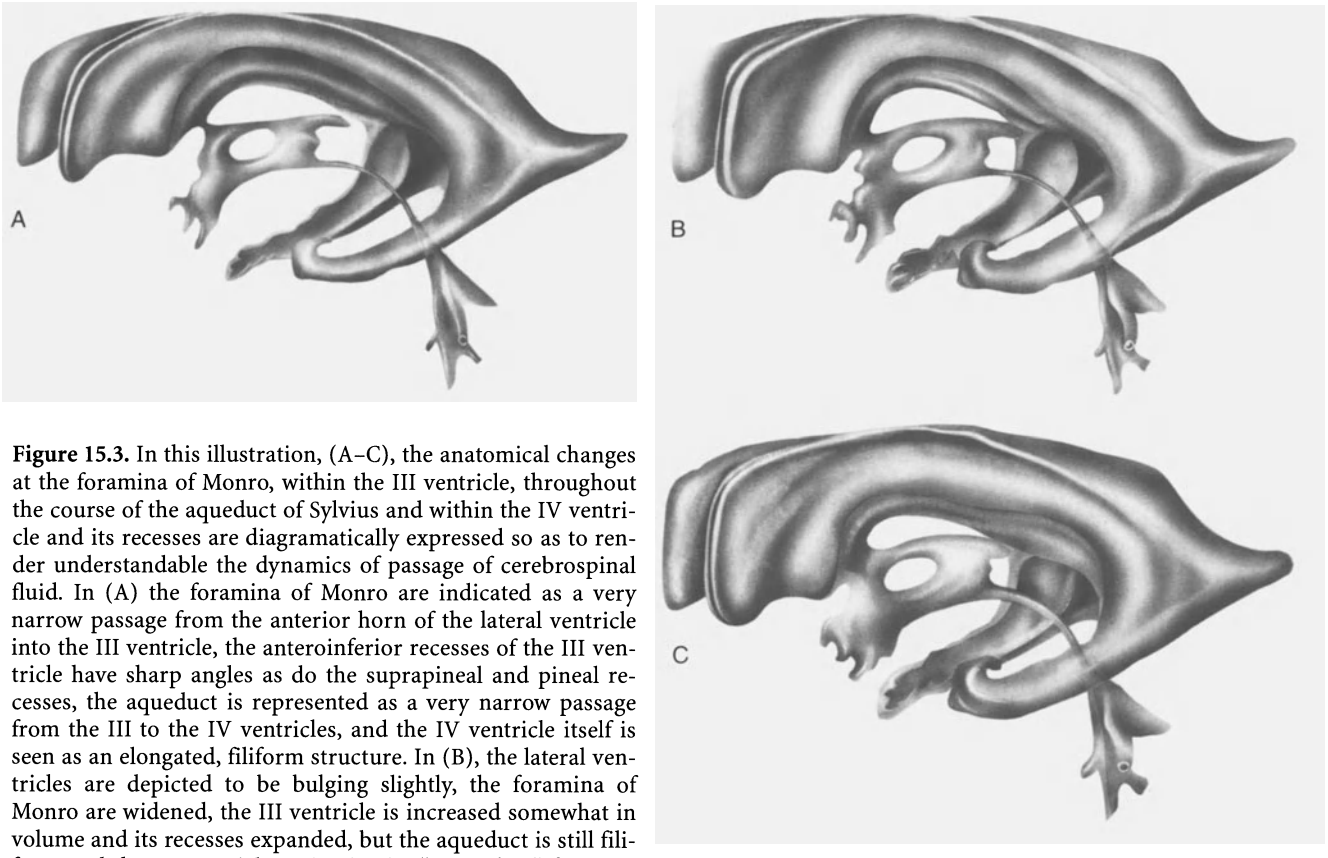


Figure 15.3. In this illustration, (A–C), the anatomical changes at the foramina of Monro, within the III ventricle, throughout the course of the aqueduct of Sylvius and within the IV ventricle and its recesses are diagramatically expressed so as to render understandable the dynamics of passage of cerebrospinal fluid. In (A) the foramina of Monro are indicated as a very narrow passage from the anterior horn of the lateral ventricle into the III ventricle, the anteroinferior recesses of the III ventricle have sharp angles as do the suprapineal and pineal recesses, the aqueduct is represented as a very narrow passage from the III to the IV ventricles, and the IV ventricle itself is seen as an elongated, filiform structure. In (B), the lateral ventricles are depicted to be bulging slightly, the foramina of Monro are widened, the III ventricle is increased somewhat in volume and its recesses expanded, but the aqueduct is still filiform and the IV ventricle maintains its "streamline" form. In (C) the lateral ventricles are further increased in volume, the foramina of Monro are maximally dilated and the III ventricle is even more distended. One notes that all of the recesses are blunted...and that the aqueduct of Sylvius is distended as are the IV ventricle and its recesses. From above, one may interpret that the flow of cerebrospinal fluid from the lateral ventricles to the III ventricle becomes possible when the lateral ventricles have so distended as to open the foramina of Monro; similarly, after the III ventricle becomes maximally distended, the cerebrospinal fluid dumps from it through the aqueduct and into the IV. In turn, distention of the IV ventricle opens the midline and lateral recesses, permitting the cerebrospinal fluid to pass respectively into the cisterna magna and the pontocerebellar cisterns.

By and large, it was assumed that external hydrocephalus and subdural effusions were identical anatomical and clinical entities, characterized by accumulation of fluid within the subdural space, a much thinner than normal cerebral mantle, markedly enlarged subarachnoid spaces, and a dilated ventricular system. Computed tomography (CT) has recently provided us with additional information: *cerebrospinal fluid may accumulate in pathological volumes within the subarachnoid spaces, in the absence of ventricular dilation, and it may pass freely across the arachnoid membrane and into the subdural spaces.* Consequently, the term external hydrocephalus was expanded to include accumulations of cerebrospinal fluid within the subdural space, and to be characterized anatomically by a much thinner than normal cerebral mantle, markedly enlarged subarachnoid spaces, and a ventricular system varying in volume from normal to dilated.

It was once thought that differentiation between external hydrocephalus and cerebral atrophy was possible only when the child demonstrated the clinical picture of increased intracranial pressure, something considered characteristic of all forms of hydrocephalus, but not present in cerebral atrophy. However, spontaneous episodic increases in intracranial pressure were detected in patients with normotensive hydrocephalus and have been correlated with REM sleep. These increases in intracranial pressure, recorded during sleep in patients with normotensive hydrocephalus, were postulated to be expressive of an inability to compensate for rapid volume variations in intracranial contents. It was observed that some hydrocephalic children may, paradoxically, continue to function normally for prolonged periods of time in spite of a patently obstructed shunting system or neuroradiologic evidence of progressive ventricular dilation. These observations were suggestive of the establishment of compensatory mechanisms for the absorption of cerebrospinal fluid (CSF).

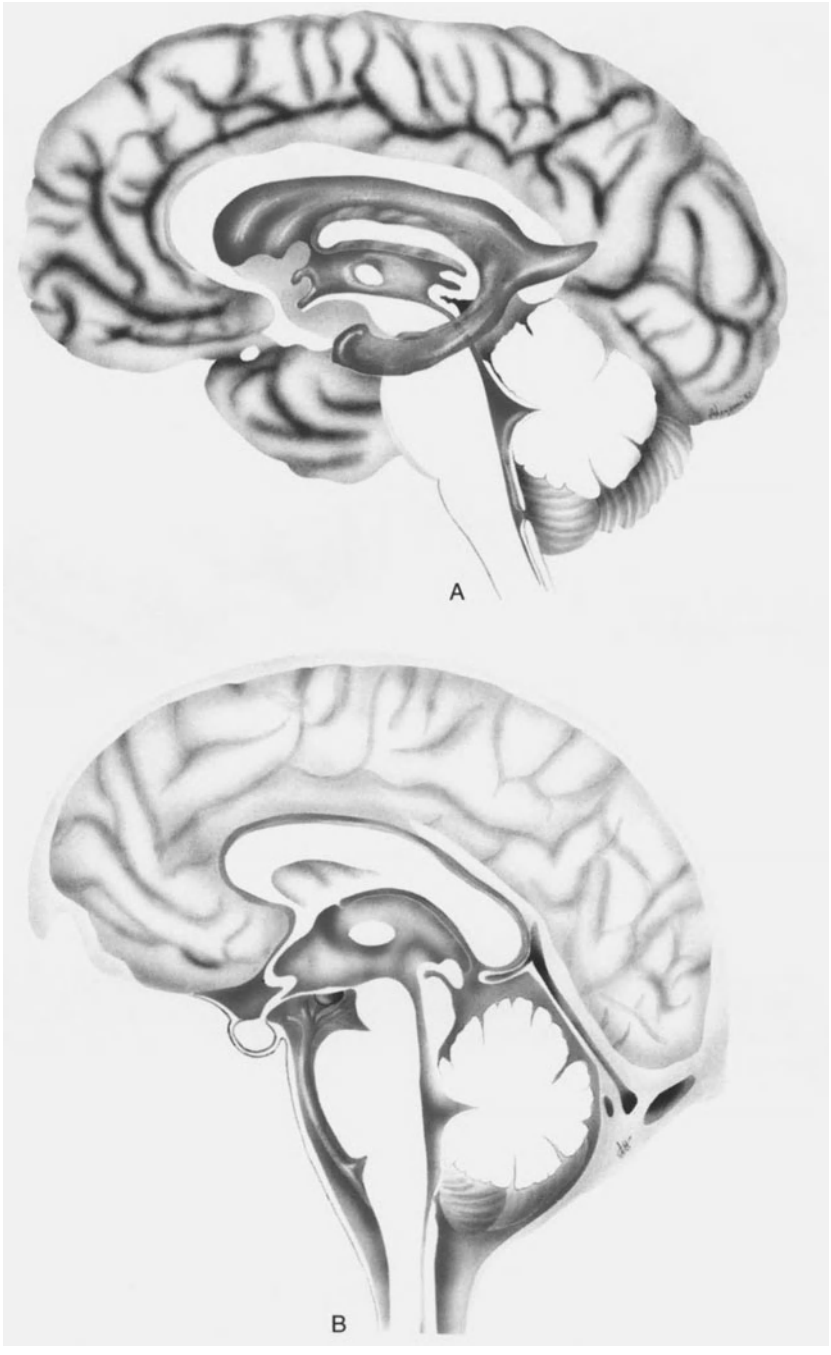


Figure 15.4. In this pair of drawings the lateral and midline ventricular systems first, and then the midline ventricular system and the basal cisterns, are projected upon the parenchymal structures with which they directly relate. In (A) one sees the lateral ventricle in the cerebral hemisphere, with its choroid plexus passing from the temporal horn through the trigone to the foramen of Monro. The III and IV ventricle choroid plexuses are projected upon their respective ventricular chambers. The midline ventricular system – III, aqueduct, IV – is projected upon the transparency image of the brainstem (midbrain, pons, bulb). In (B), the major anatomical relationships between the midline ventricular system, the ventral brainstem, the dorsal cerebellum and the cisterns surrounding the chiasm, lamina terminalis, midbrain, pons, bulb, cerebellar vermis, collicular plate, and corpus callosum may be seen as a 360° irregular circle. The basilar artery, the major driving force for the circulation of cerebrospinal fluid from the basal cisterns up over the surface of the hemispheres, and very probably a pushing force from the basilar fundus against the floor of the III ventricle to propel cerebrospinal fluid posterosuperiorly toward the aqueduct of Sylvius, is shown wedged between the rigid clivus and the massive pons Varolii.

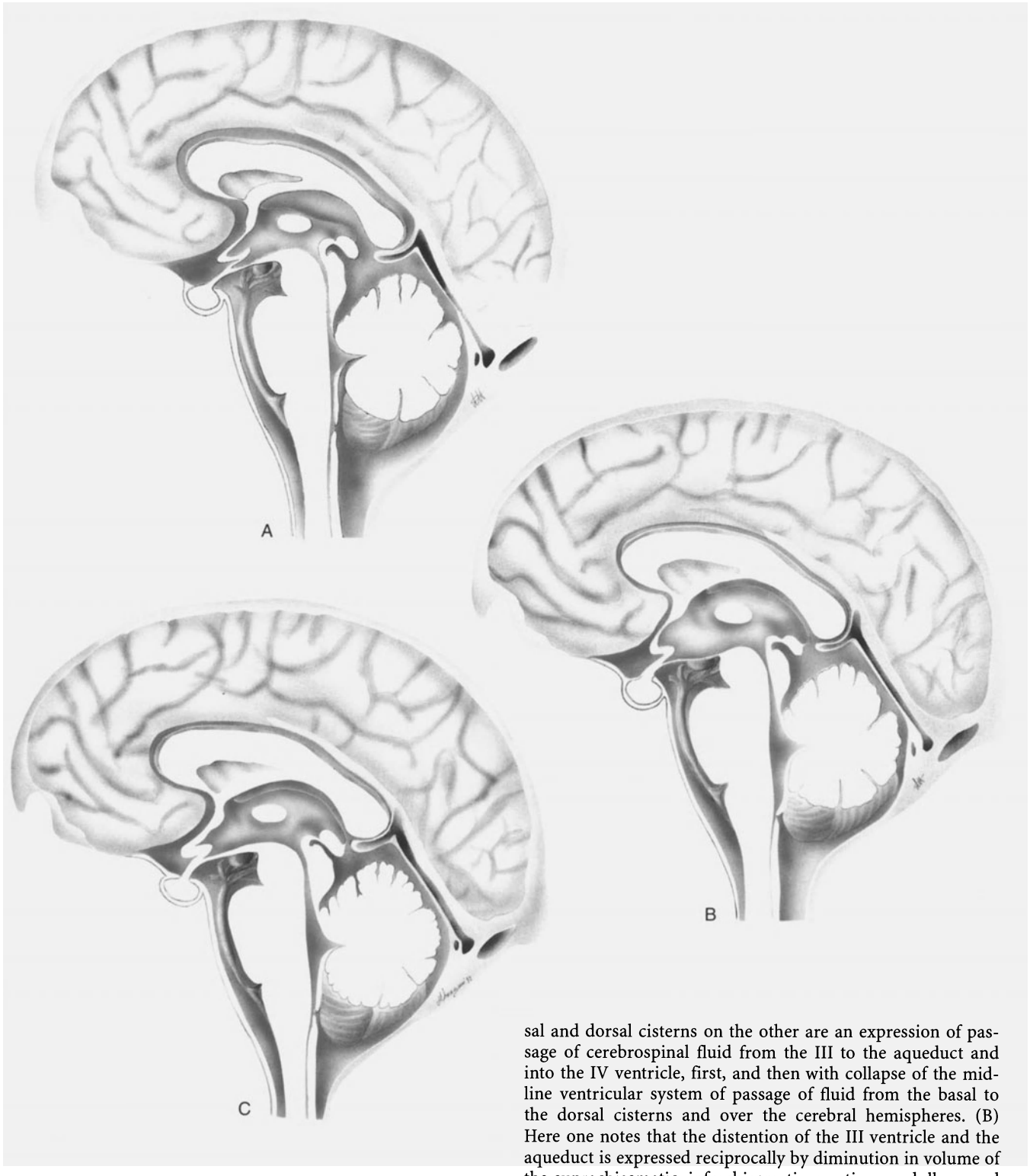


Figure 15.5. This is a series of three drawings, mid-sagittal cuts, illustrating the midline ventricular system, the basal and dorsal cisterns, and the parenchymal structures surrounding them. (A) The midline ventricular system, including the aqueduct, are seen in the "resting" state. Note that the cisterns, *depicted in blue*, are relatively larger than in the subsequent illustrations: the reciprocal relationships in CSF volume between the midline ventricular system on one hand and the basal and dorsal cisterns on the other are an expression of passage of cerebrospinal fluid from the III to the aqueduct and into the IV ventricle, first, and then with collapse of the midline ventricular system of passage of fluid from the basal to the dorsal cisterns and over the cerebral hemispheres. (B) Here one notes that the distention of the III ventricle and the aqueduct is expressed reciprocally by diminution in volume of the suprachiasmatic, infrachiasmatic, pontine, medullary, and quadrigeminal cisterns. There is also a relative diminution in volume of the supra- and infracerebellar cisterns. (C) The III ventricle is returning to its original form, but most prominent is the distention of the pericallosal, infracallosal, and suprachiasmatic cisterns: the cerebrospinal fluid has completed its cycle through the midline ventricles, into the basal cisterns, and over the pericallosal and suprachiasmatic cisterns to pass subsequently along the surface of the hemispheres.

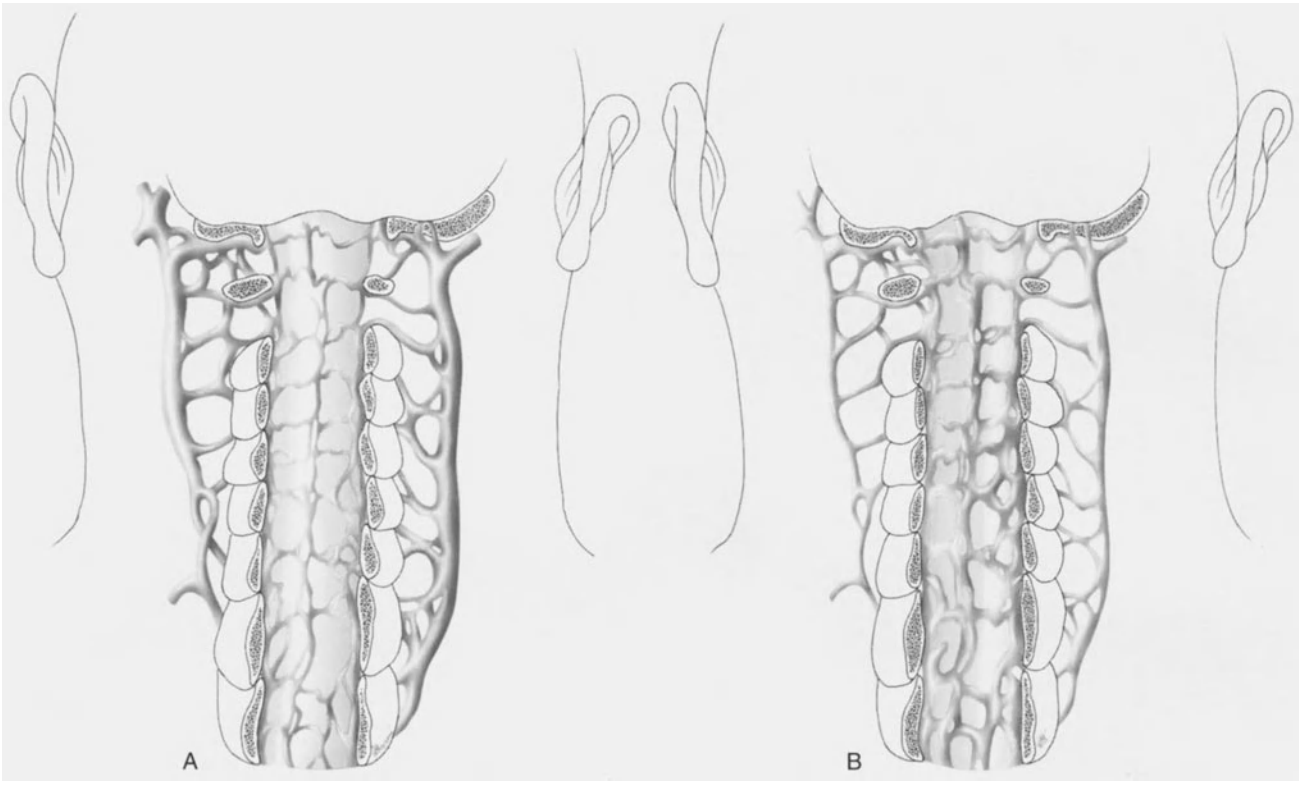


Figure 15.6. It is now well known, given the many studies on cine magnetic resonance of the cerebrospinal fluid flow, that there is a thrust of CSF from the cisterna magna dorsally and the pontomedullary systems ventrally to and across the line of the foramen magnum. It is equally well known that at this line the cerebrospinal fluid does not pass massively and in an unimpeded manner into the cervical subarachnoid spaces. There is, to be sure, a flow across the line of the foramen magnum, but this is stopped at the level of C1, where one notes a circumferential flow of CSF around the upper cervical cord, and then back up into the intracranial compartment.
Why? (A) Batson's plexus ends at C1. We know that Batson's plexus is a labyrinthine communication of epidural veins extending the full length of the epidural space from C1 to S5. We also know that the spinal dura mater is composed of only one layer, so that there are no dural venous sinuses. Lastly, we know that Batson's plexus communicates freely with the retroperitoneal venous system of the pelvis and the abdomen, with the retropleural system of the thorax, and with the cervical plexus of veins. With increases in intra-abdominal pressure, venous blood is shunted into Batson's plexus, and expansion of the cerebrospinal fluid volume is buffered. Similarly, increases in intrathoracic pressure cause the same. Increases in cerebrospinal fluid pressure and volume may compress the epidural veins, impeding flow of venous blood from the retroperitoneal and retropleural spaces. In (A) one has a diagrammatic representation of distended cervical veins but compressed Batson's plexus around the cervical cord: increase in cervical subarachnoid fluid volume. In (B) there is a remarkable extension of Batson's plexus, a diminution in volume of the cervical veins: decrease in volume of cerebrospinal fluid. These fluxes in cerebrospinal fluid volume, reciprocals of venous blood within Batson's plexus, buffer volumetric (pressure) changes, within the intracranial compartment in general but specifically in the posterior fossa.

Changes in intracranial pressure (ICP) occur during the early stages of sleep and during REM sleep, they are episodic and rapid in onset and cessation, and are therefore expressive of the fact that alterations in production or absorption of cerebrospinal fluid are not the causative factors. Rather, alterations in cerebral flood flow (CBF), mediated by cerebral vasodilation and induced by neuronal or humoral mechanisms, resulting from cerebral vasodilation, cause a greater increase in intracranial pressure (because of the exponential relationship between intracranial volume and pressure) in children with defective CSF absorptive mechanisms. These factors were identified as explanatory of how one may repeatedly record normal pressure in patients who actually have progressive hydrocephalus. This observation of significant episodic variation in ICP suggested the necessity for substituting the concept of "time-related pressure variations" for the older one of "level of pressure" in patients with defective ICP control mechanism.

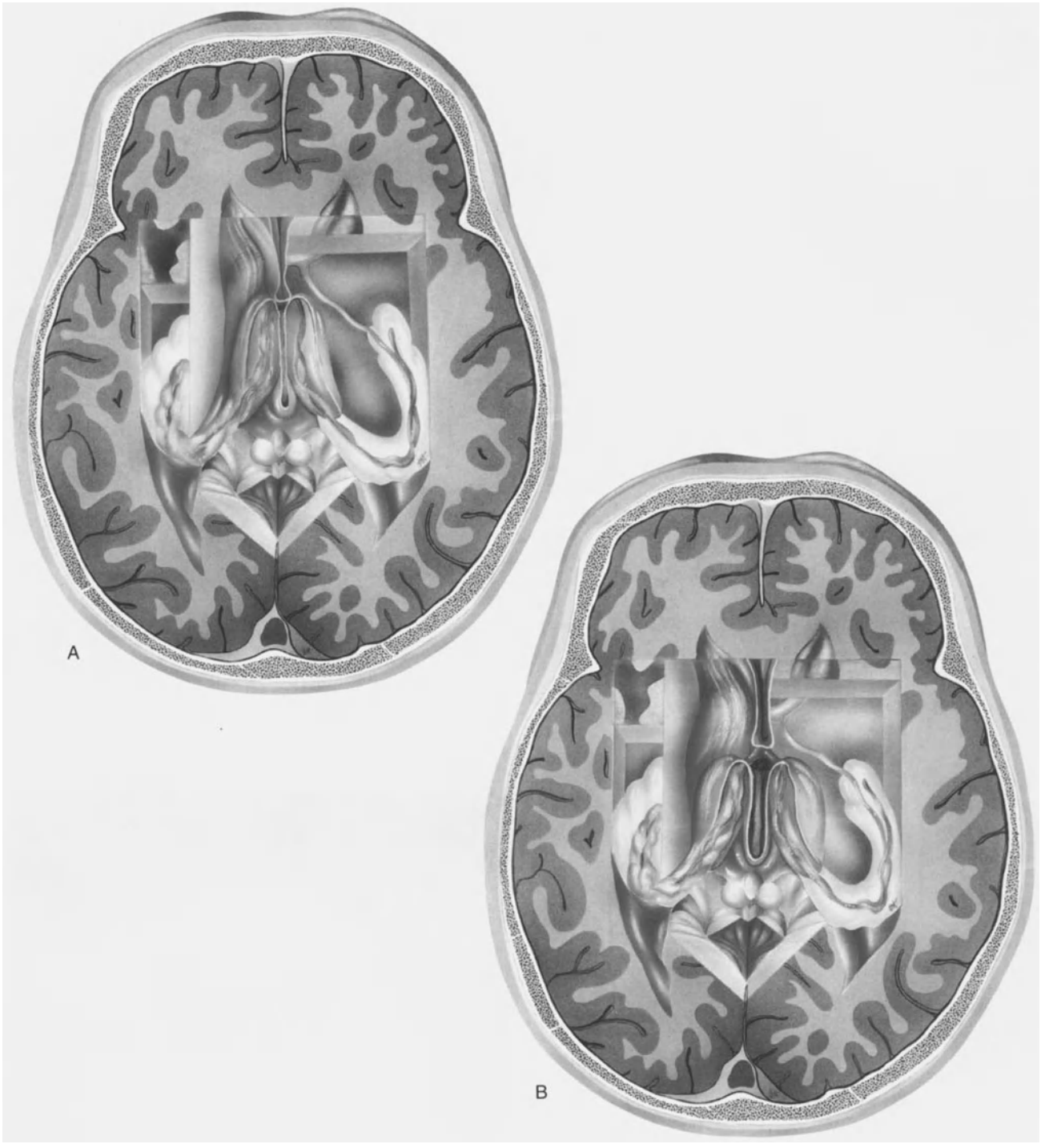
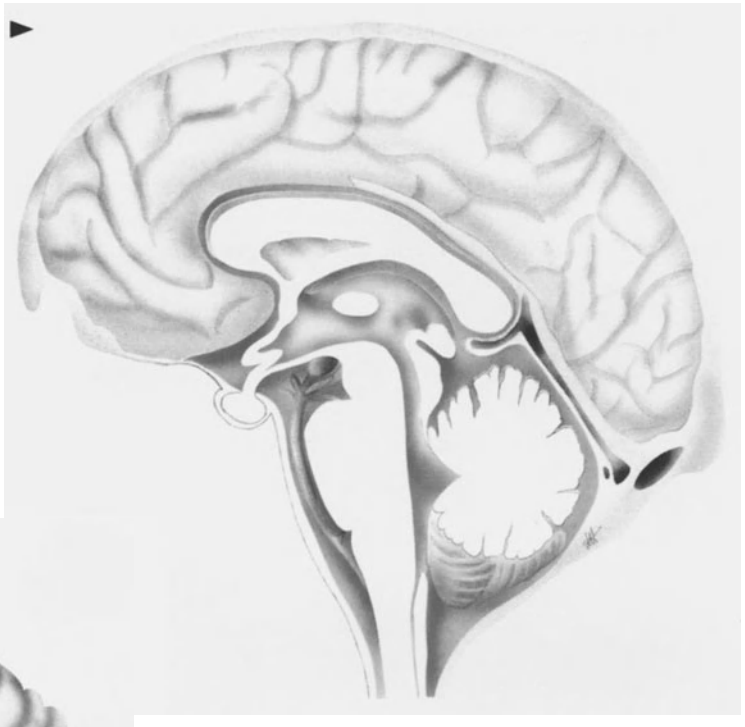
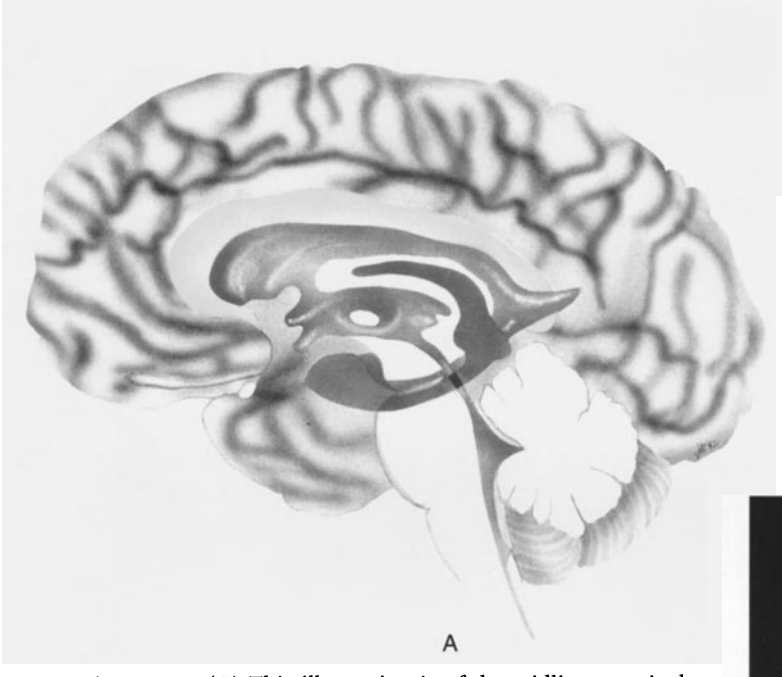


Figure 15.7. In this illustration, (A) and (B), axial sections of the hemispheres through the level of the III ventricle at the foramina of Monro are represented. There is a "cut-through" exposure of the IV ventricle. The purpose of these two illustrations is to put into relief the mechanism by which the foramina of Monro open: distention of the lateral ventricle displaces the columns of the fornix anteriorly and the anterior tubercules of the thalami relatively posterolaterally. In (A) one sees the columns of the fornix juxtaposed upon the almost touching thalami. It is also possible to see the closed III ventricle, to identify the projection of the choroid plexus from the temporal horn along the sulcus terminalis to the foramen of Monro and, *on the left*, the pes hippocampus and hippocampal formation with the anterior choroidal artery running to the pes and the choroid plexus along the surface of the hippocampal formation. (B) The lateral ventricle, *represented in green*, is dilated, and the fornices have moved forward as the thalami are displaced posterolaterally by both the distending lateral ventricle and the distending III ventricle. The opening of this "vaginal" valvular mechanism permits passage of cerebrospinal fluid from the lateral ventricles into the III ventricle.

Figure 15.8. This sagittal cut, illustrating an opened aqueduct of Sylvius, permits one to follow the passage of cerebrospinal fluid from the III ventricle into the aqueduct, as a consequence of what was described in Fig. 15.7.





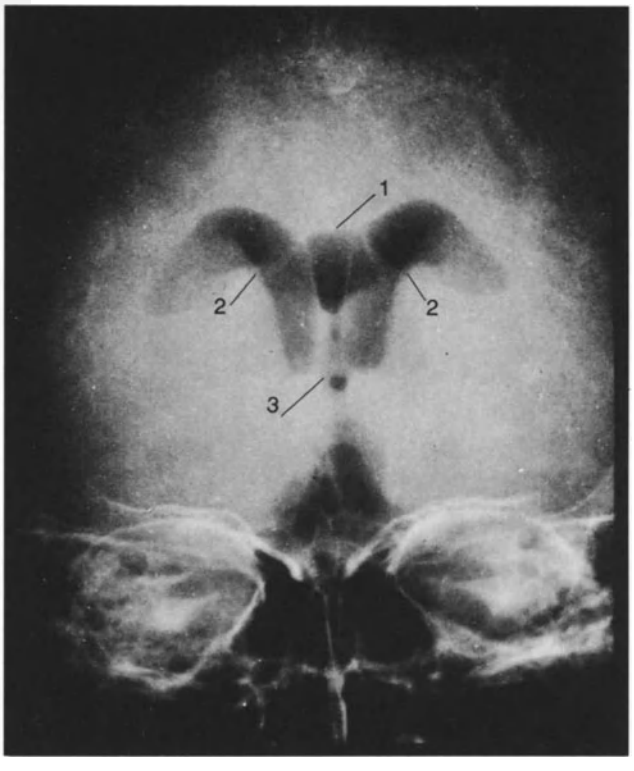


Figure 15.9. (A) This illustration is of the midline ventricular system and the lateral ventricles, but was prepared to demonstrate flow of cerebrospinal fluid from the interpeduncular cistern around the brainstem within the ambient cistern to the collicular cisterns: normal flow. However, in some children the space between the roof of the III ventricle and the inferior surface of the corpus callosum is virtual, permitting cerebrospinal fluid to pass into this space. There are cases of ventriculomegaly, and clinical hydrocephalus, which apparently result from this abnormal flow into the cavum veli interpositi, probably a result of disturbed volumetric equilibrium between intracisternal fluid and intra midline ventricular fluid. (B) Gas has filled the ventricular system directly, not the subarachnoid spaces. There is a cavum septi pellucidi *(1)* between the lateral *(2)* and III ventricles *(3)*.

Clinical and experimental evidence has been interpreted to indicate that the CSF is absorbed through the arachnoid villi along the superior longitudinal sinus *...though these villi are microscopically absent in the newborn and infant.* Irrespective of this observation, it has been presumed that the subarachnoid channels adjacent to the arachnoid villi represent the first cerebrospinal fluid compartment to dilate, reducing the CSF pressure, thereby establishing an equilibrium. When the equilibrium shifts to the left, with progressive dilation of the subarachnoid channels, the increase in cerebrospinal fluid pressure is transmitted to the ventricular system, resulting in its dilation. Progressive ventricular dilation obliterates the subarachnoid spaces as the hemispheres are compressed against the dura, resulting in apparent internal hydrocephalus in the absence of external hydrocephalus.

Subarachnoid space or ventricular dilation occur as a result of intermittent increases in extraparenchymal cerebrospinal fluid volume: the primary pressure force emanating from the subarachnoid and subdural spaces in external hydrocephalus and from the intraventricular compartment in internal hydrocephalus. The hydrocephalic child suffers both direct pressure on the brain and a progressive increase in head size. The early forms of both external and internal hydrocephalus are more characterized by changes resulting from increases in fluid volume and pressure within the subdural or intraventricular compartments, respectively, than by marked dilation of these compartments or increased head size. Internal hydrocephalus, therefore, may be present in a child who as yet does not have dilated ventricles but in whom both CSF volume and pressure are increased. *Thus, it becomes obvious that the term internal hydrocephalus is of little significance, since increases in intraparenchymal fluid – cerebral edema – cause the same volumetric changes as increases in intraventricular fluid volume. The head will increase in size whether there is an increase in intraparenchymal or intraventricular fluid.*

Definition and Classification (Figs. 15.10–15.38)

It has, therefore, become necessary to consider critically whether hydrocephalus is characterized by: (1) an increase in intraventricular pressure; (2) an increase in intraventricular cerebrospinal fluid volume; and (3) increased, or increasing, ventricular size. With the observations which the CT scanner has provided us, namely that subdural, subarachnoid, or periventricular areas may hold pathological increases in fluid volume, one *may expand the classification of hydrocephalus to include cerebral edema*, of either the vasogenic or cytotoxic varieties. This permits the inclusion of metabolic disorders, arteriovenous fistulae, and hormonal or humoral causes for alterations in cerebral flood flow.

Until now we have *not* had a definition of hydrocephalus which is both implicit and explicit, which includes the physiological condition of degree of cerebral hydration or " bathing," as well as the anatomopathological state of inverse relationships between CSF volume and parenchymal bulk. CSF has been looked upon as a cushion, not a system for transporting cells, hormones, and metabolites; not, as we suggest, a neurophysiologic ion transfer medium.

In order to define hydrocephalus, we must first define "brain fluid." It is not reasonable to think in terms of a multitude of intracranial fluids:

1. CSF: sulcal, cisternal, ventricular, perineural, central canal, spinal
2. Extracellular: white matter, gray matter, areas without blood-brain barrier
3. Intracellular: ganglionic, glial, perivascular glial cell
4. Axomyelinic: intra-axonal, myelogenous
5. Lymphatic: perivascular spaces, basement membrane

We ignore, at least in experimental and clinical approaches to the problem of hydrocephalus, the neurohumoral and cellular (ganglionic) transport functions of CSF, as well as its role in ion transfer and pH buffering activities. Therefore... *if the CSF is considered to be all fluid (liquid), other than blood or the derivatives of its breakdown, normally contained within the brain, its cavities, and its spaces... independent of its cellular or electrolyte composition, one may then consider "brain fluid" in its most elemental form.*

Such a definition of CSF permits thinking in terms of it as an ion transfer and liquid medium which permits:

1. Transmembranous passage
2. Direct and reverse pinocytotic transport
3. Bulk flow across cellular layers or through extracellular spaces
4. Secretion and absorption
5. Changes in cellular and electrolyte concentrations and gas partial pressure both over time and in different anatomical locations
6. Pulsatile intraventricular flow from one chamber to another
7. Percolation through the aqueduct and foramina
8. Pooling within basal and medial cisterns
9. Antigravitational rise through subarachnoid spaces via capillary pressure

"*Pathological increases in intracranial CSF volume, independent of hydrostatic or barometric pressure", then, could be considered a definition of hydrocephalus* which would be acceptable to clinicians and basic scientists. It would permit the basic scientist to establish criteria for determining whether the increase is a primary or sec-

ondary event, to study the permutations and variations of each. It would also permit the clinician to evaluate volume increases as individual pathogenetic events, which could then lead him to identify the specific anatomopathologic entity against which he could direct his treatment: (vasogenic or cytotoxic) cerebral edema, aqueductal occlusion or obstruction of a foramen of Monro, high flow arteriovenous shunt into the transverse sinuses, etc.

Such a classification obliges one to distinguish between CSF which has passed into the subdural spaces, through the arachnoid membrane, in the very early (neonatal) phases of hydrocephalus and immediately after a shunt has been inserted, from subdural bleeding, and, in turn, subdural blood breakdown products from liquefaction of a pyogenic exudate and blood reactive elements.

By hydrocephalus, one means a *pathological increase in intracranial cerebrospinal fluid volume, independent of hydrostatic or barometric pressure.* It may be classified as:

I. Intraparenchymal (cerebral edema)
   A. Intracellular
   B. Extracellular
II. Extraparenchymal
   A. Subarachnoid
      1. Transient, self limiting
      2. Early stages of "communicating hydrocephalus"
      3. Transformation into regional or loculated arachnoid cysts.
   B. Cisternal
      1. Cyst of cisterna magna
      2. Cysts of basal or sagittal cisterns
      3. Cysts of sylvian fissure with or without parenchymal dysplasia
   C. Intraventricular
      1. Monoventricular (lateral)
      2. Biventricular (both lateral)
      3. Triventricular (III and both lateral)
      4. Tetraventricular (IV, III and both lateral)

The anatomical natural history of hydrocephalus *begins with an accumulation of fluid within the parenchyma,* as cerebral edema. Subsequently there are disproportionate accumulations (shifts) of fluid into the subarachnoid spaces, the subdural spaces, the cisterns, and, lastly, the ventricles. This latter, resulting in structural changes of the ependymal cells and the junctions which hold them together, permits pathological amounts of fluid to pass into the cerebral parenchyma, first compounding cerebral edema and, ultimately, causing parenchymal destruction and porencephaly. Occasionally, the parenchymal destruction is so massive as to result in fistulization between the ventricular system and the subarachnoid spaces.

Hydrocephalus, then, is not a single disease entity, nor is it a syndrome. The various etiologic factors range from congenital malformations through neoplasms to meningitis. For well over 65 years now, we have thought of hydrocephalus in a purely mechanistic sense, following Dandy's development upon and dissemination of Whytt's [1] classification of external and internal hydrocephalus. Dandy subdivided the latter into communicating and obstructive subgroups. During the past 15 years, the additional category of constrictive (as in the Chiari II malformation) hydrocephalus was added [3]. The observations that toxic substances, vitamins, nutritional disturbances, etc., could result in hydrocephalus were made but were not integrated into a new classification.

The most commonly followed classification of hydrocephalus was put forth when it was assumed that there were neither compensatory pathways nor transependymal flow, when it was thought that cerebrospinal fluid could not cross the arachnoidal membranes to enter the subdural spaces, when the interrelationships between cerebral blood flow and CSF pressure were not understood, and when it was assumed that hydrocephalus necessarily entailed increased intraventricular pressure. At that time, in the face of considerable controversy concerning the exact site of formation of CSF, whether by the ependymal cells or the choroid plexus of the lateral ventricle, there was unanimity regarding the fact that fluid was formed within the lateral ventricles. Also, most authors agreed that the choroid plexus of the lateral, III, and IV ventricles served as the driving force to propel the cerebrospinal fluid through the ventricular system and into the basal cisterns. The water-hammer effect created by the pulsatile choroid plexus gives a to-and-fro movement to the cerebrospinal fluid within the ventricular system, allowing it to pass from one ventricular chamber to another, and from the IV ventricle into the cisterns.

►

Figure 15.10. (A) The normal cisterns. (B) The normal midline ventricles. Splenium (*1*) and genu (*2*) of corpus callosum, as well as quadrigeminal cistern (*3*) are missing. In (C) and (D) are demonstrated respectively the CT observations of a child with a cavum veli interpositi. In (C), one identifies very well the cavum velum interpositum situated medial to the trigones of the lateral ventricles and superior to the III ventricle. The borders of the anterior horns are blunted. (D) The same child, a higher cut, illustrating the remarkable ventriculomegaly and the accumulation of fluid in the subarachnoid spaces: grade 2 hydrocephalus. This is another example of a cavum veli interpositi. In (E), *on the left*, one identifies very well the cavum veli interpositi and the moderate ventriculomegaly. In (F), *on the right*, one also identifies a cavum septi pellucidi and lateral displacement of the bodies of the fornices.

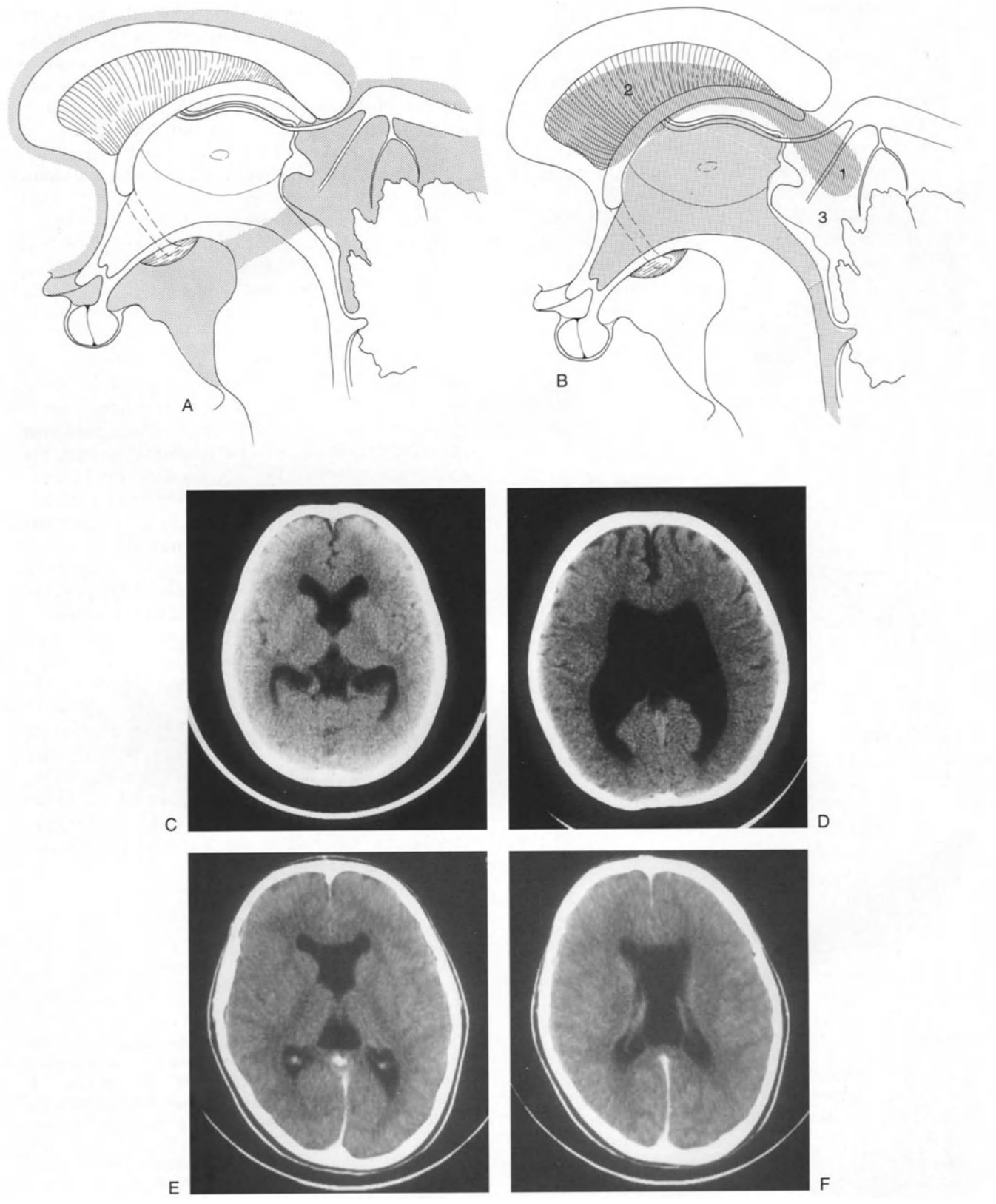


Figure 15.10. Legend see p. 558.

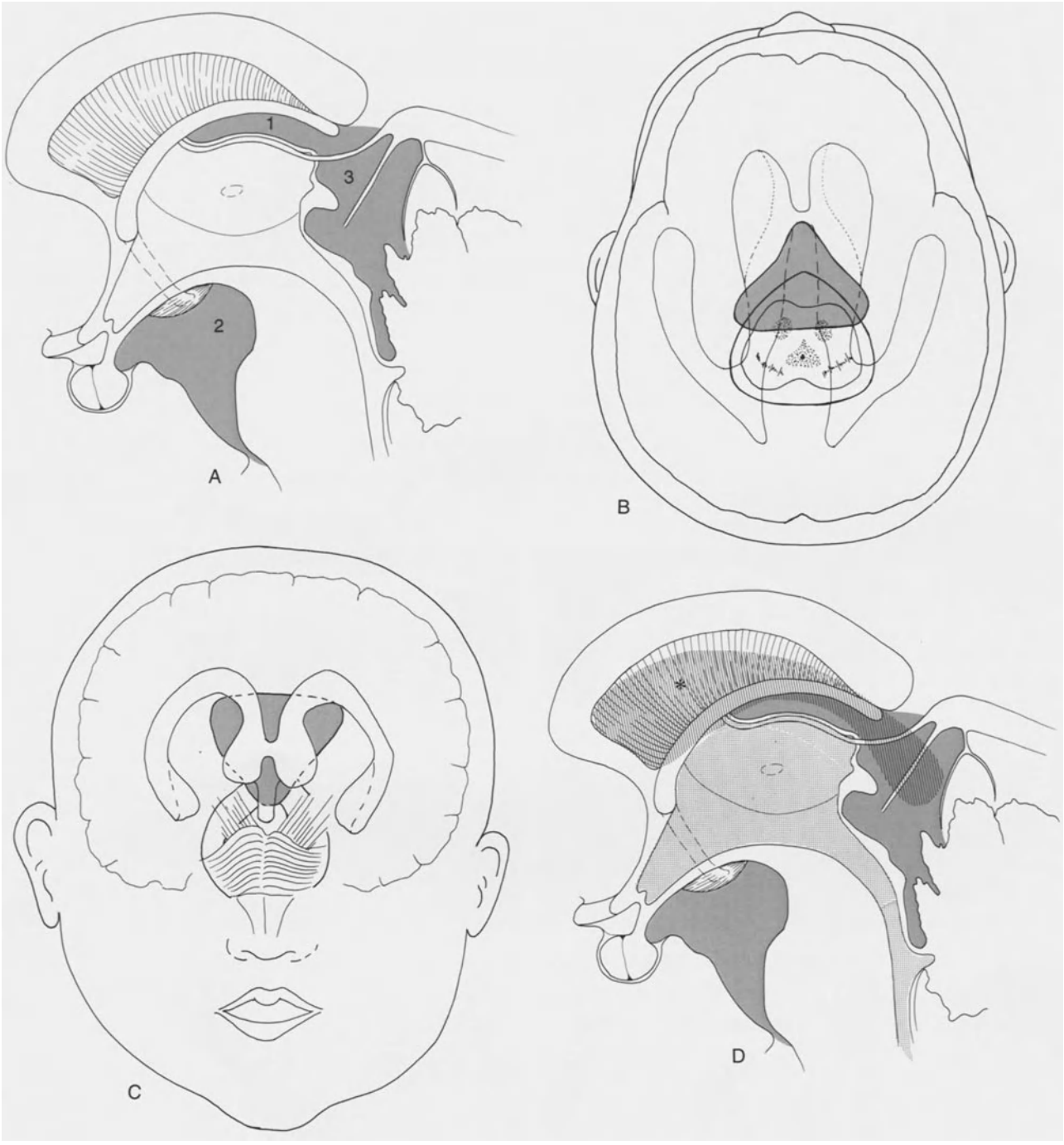
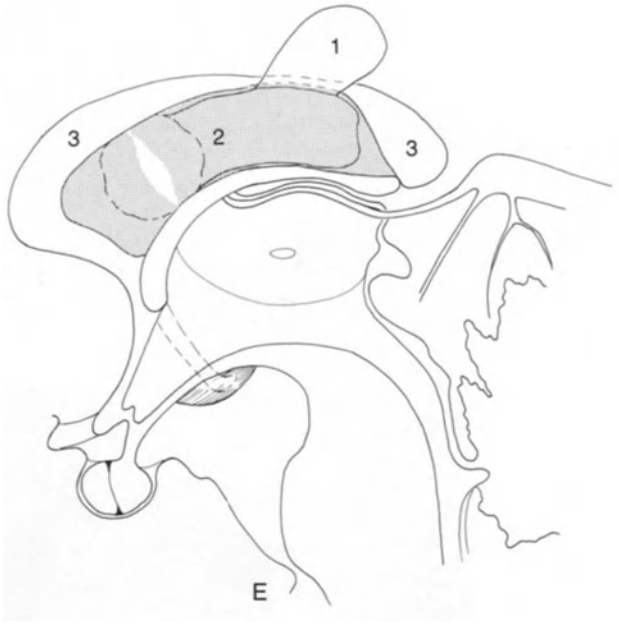


Figure 15.11. (A) The cavum veli interpositi (*1*) is filling from the basal cistern (*2*) through the quadrigeminal cistern (*3*). (B) In the axial projection, the cavum veli interpositi splays out beneath the lateral ventricles, extruding laterally at each choroidal fissure. Since the drawing was taken from an air study, the gas is projected over the lateral ventricles. (C) In the half-axial projection, we see the cavum veli interpositi insinuated beneath the lateral and superior to the III ventricles. (D) The cavum vergae fill from the ventricular system, through a cavum septi pellucidi (*). (*Continued* on p. 561).



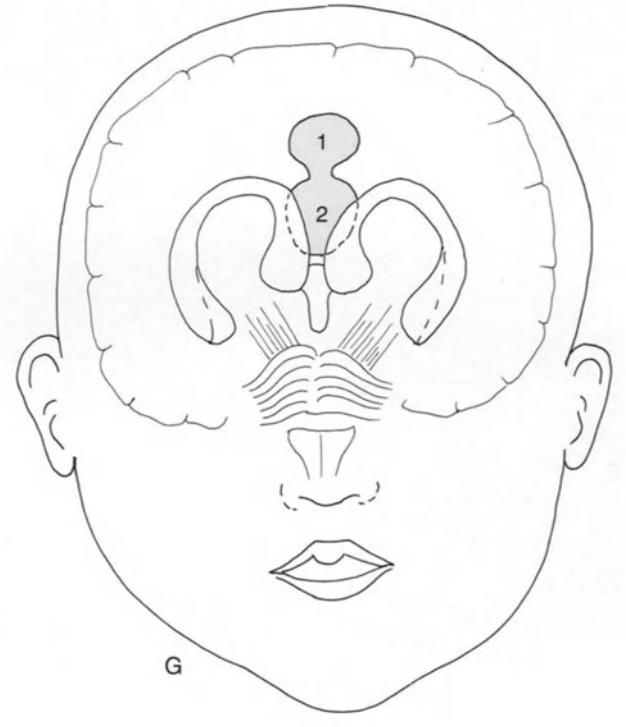
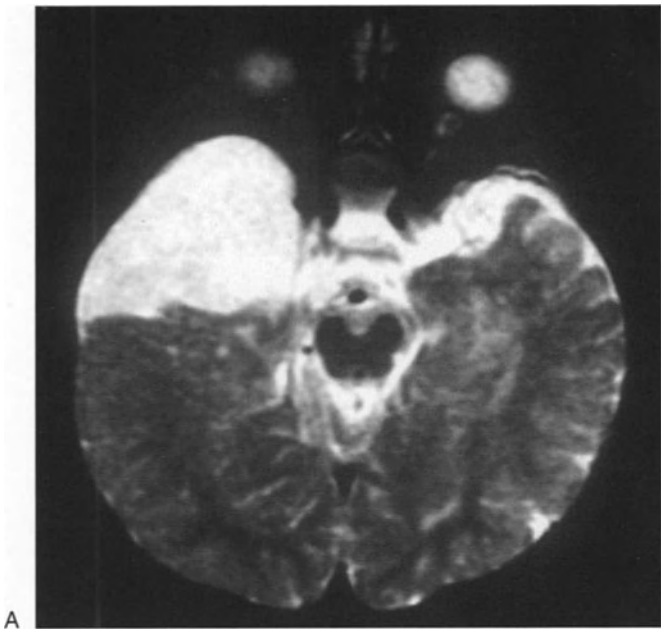


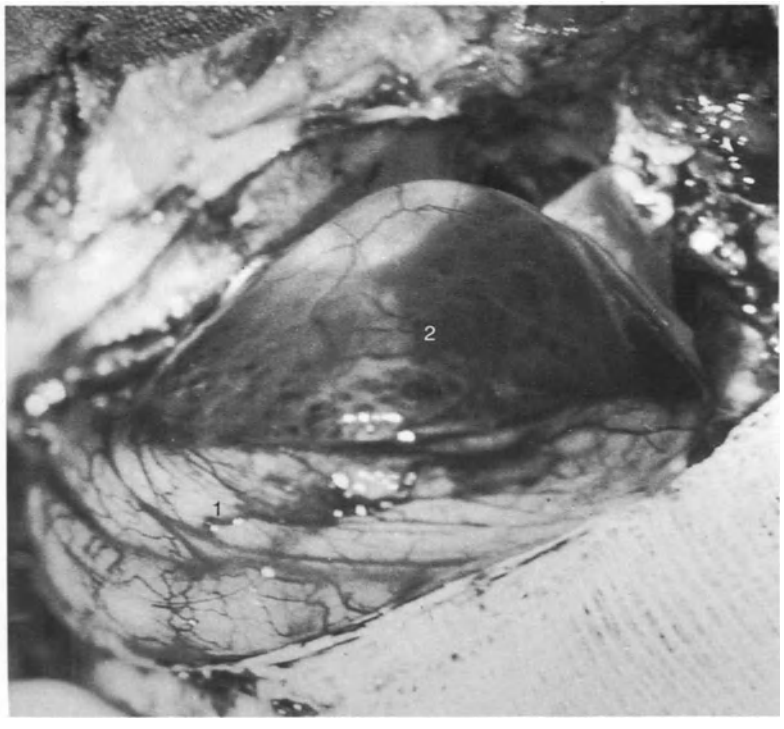
Figure 15.11. It may hang into the quadrigeminal cistern, as illustrated here, or (E) extend (*1*) superior to both the cavum septi pellucidi (*2*) and the corpus callosum (*3*). (F) The axial projection illustrates how the cavum septi pellucidi (*1*) and cavum vergae (*2*) displace the lateral ventricles. (G) The half-axial projection illustrates superior (*1*) and inferior (*2*) extension of the cavum vergae.

The circulation of cerebrospinal fluid through the foramina of Luschka and Magendie, around the brainstem, and its percolation up through the basal cisterns and over the surface of the brain offered the basis for the conclusion that: (1) the CSF may circulate freely throughout the entire ventricular system and the cisterns, but may not be adequately resorbed, in which case one spoke of communicating hydrocephalus; or (2) the CSF passage from one ventricle into another or from the ventricular system into the basal cisterns may be obstructed totally or partially, causing obstructive hydrocephalus.

Communicating hydrocephalus was considered any form of ventricular dilation in the absence of a macroscopic obstruction from the ventricles to the flow of fluid into the basal cisterns. Obstructive hydrocephalus, on the other hand, was characterized by an identifiable blockage of CSF flow somewhere along the CSF pathways, and subdivided into: (1) atresia of the foramen of Monro, (2) aqueductal stenosis, and (3) occlusion of the foramina of Luschka and Magendie. These three clinical entities varied from one another only insofar as the site of obstruction to the passage of CSF changed. The Dandy-Walker cyst (atresia of the foramina of Luschka and Magendie) was observed to be associated with dysgenesis of the corpus callosum in approximately 75% of the cases. It is true that the variation between these three obstructive forms of hydrocephalus is anatomical, but it is not true that the same pathogenesis applies to the three. *Atresia of a foramen of Monro* has, with only two exceptions published in the world literature, invariably been secondary to infectious processes of the ependyma, or of subependymal astrocytoma growing within the region of the anterior commissure or the passage of the column into the body of the fornix.



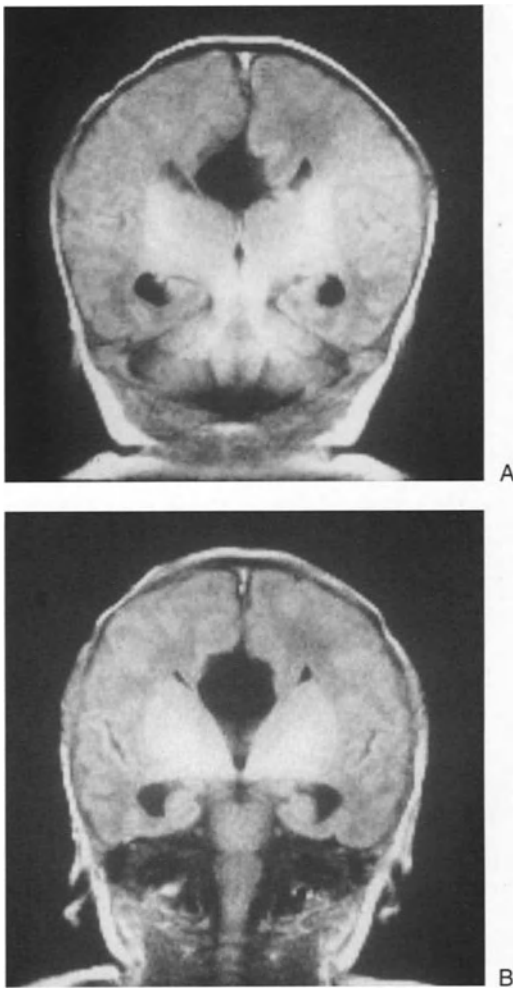
A



B

Figure 15.12. (A) This is an arachnoidal cyst of the middle fossa, *on the left*, with remarkable dilation of the sylvian cistern *on the right*. (B) Arachnoidal cyst of the sylvian fissure. A right frontotemporal craniotomy has been reflected and the dura opened. One notes the frontal lobe (*1*) flattened and draped over the expanding cyst of the sylvian fissure (*2*).
◄

Figure 15.13 (*below, right*). In (A) and (B), one sees the formation of an arachnoidal cyst (cystic transformation of the superior portion of the III ventricle?) as a result of aplasia of the corpus callosum.



A

B

Aqueductal stenosis is of such significance, historically and pathogenetically, as to merit a rather extensive discussion. It is a narrowing of the aqueduct of Sylvius which becomes clinically obvious, resulting anatomically in partial or functionally complete obstruction to the flow of CSF from the III ventricle into the IV ventricle. Complete obliteration of the aqueduct, independent of the microscopic pathoanatomical architecture, has been considered to be aqueductal occlusion. The distinction between nontumorous aqueductal occlusion and the rarely encountered membranous occlusion was previously made in the living patient only after combined neuroradiologic studies: positive contrast III ventriculography and pneumoencephalography with the patient in the sitting position. Today, magnetic resonance imaging (MRI) suffices.

Such terms as forking, atresia, and gliogenous occlusion are of purely historical value and, consequently,

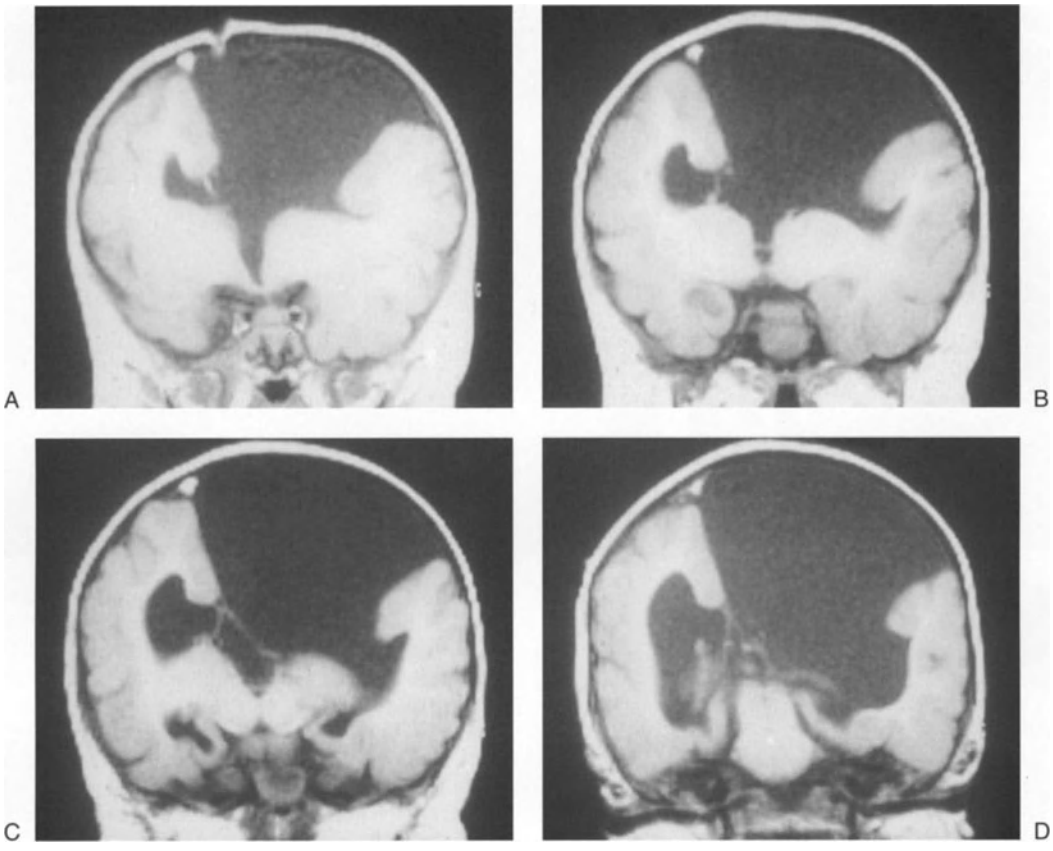


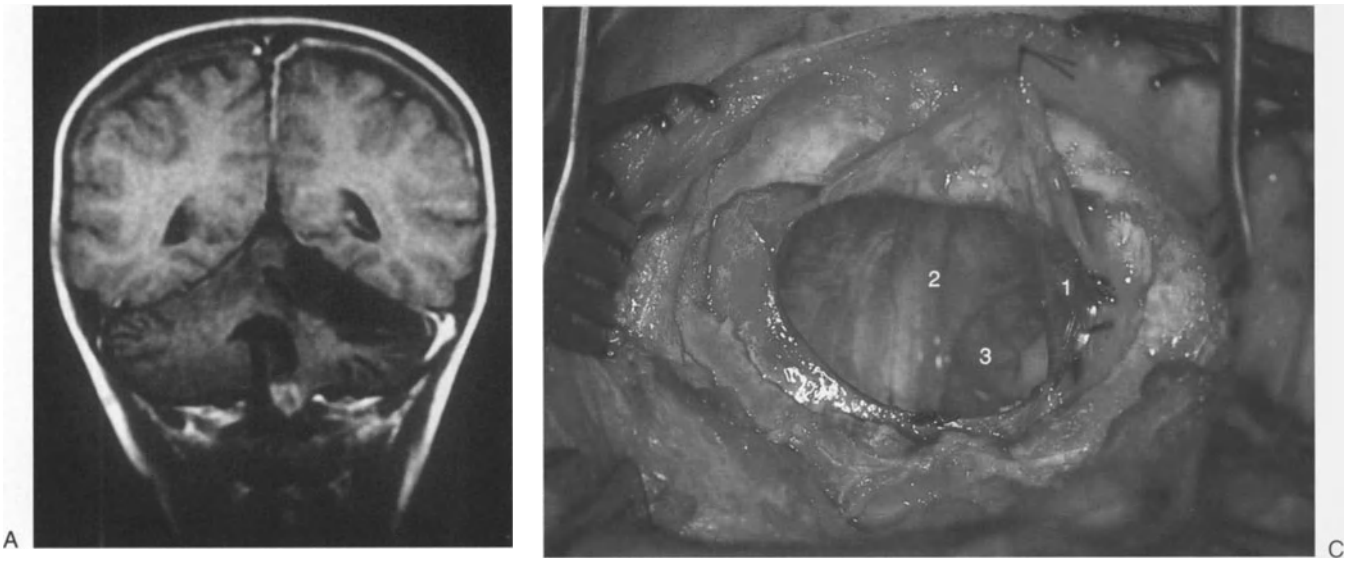
Figure 15.14. This is another example of aplasia of the corpus callosum, in this case complete, illustrating the enormous dilation into a cystic chamber superior to the III ventricle... and communicating freely with both the III and the lateral ventricles. (A–D) are different coronal slices through this cystic chamber, and all four permit the reader to understand why this, as the two previous and all subsequent examples of intracranial cerebrospinal fluid cysts, is considered a form of hydrocephalus. There is an increase in cerebrospinal fluid volume, at the expense of cerebral parenchyma.

the reader is urged not to consider them distinct etiopathoanatomical entities. Aqueductal forking, atresia, gliogenous occlusion, stenosis, and membrane formation represent a spectrum of stages or degrees of obstruction to CSF flow, and varying responses to ependymitis and cerebral edema. They are not individual, specific entities, expressive of discretely different etiological causes or maldevelopmental processes.

The history of the identification of the aqueduct of Sylvius as an anatomical structure is as complicated and, to a certain extent, controversial, not to mention speculative, as are the varying concepts and opinions concerning its size, its form, and its pathological alterations. Anatomists, neuropathologists, clinicians, and neuroradiologists have all undertaken the study of the aqueduct, each using his own investigative techniques and instrumentation, and each coming to conclusions which, at times, predictably though unfortunate, were at odds with one another.

Berengarius (cited in [4]) is given credit for the first description of the aqueduct in 1521. Jacobus Sylvius, the teacher of Vesalius, published in his *Isogoge*, in 1515 that "From the III ventricle a long and narrow meatus much larger in a living person, passes under the corpora quadrigemina into the IV ventricle" (Sylvius, cited in [4]). Twenty years after Berengarius published his observations on the aqueduct, Vesalius described the aqueduct of Sylvius in his *Fabrica* as a "meatus that extends from the III to the IV ventricle... behind and below... the testes and nates, or corpora quadrigemina." Arantius, in 1578, apparently was the first to refer to this meatus as "a canal or aqueduct" (Arantius, cited in [4]). According to Baker's review [4], another anatomist coincidentally and confusingly also named Sylvius, Franciscus – in the mid seventeenth century, wrote "From this III ventricle formed between the conjoined roots of the spinal cord (crura cerebri) there is a canal or aqueduct passing toward the IV ventricle."

Interestingly, two neuroscientists, Bonnevie in 1943 [5] and Russell in 1949 [6], came to two diammetrically opposite conclusions concerning the etiology of aqueductal occlusion. Bonnevie, studying primarily the ex-



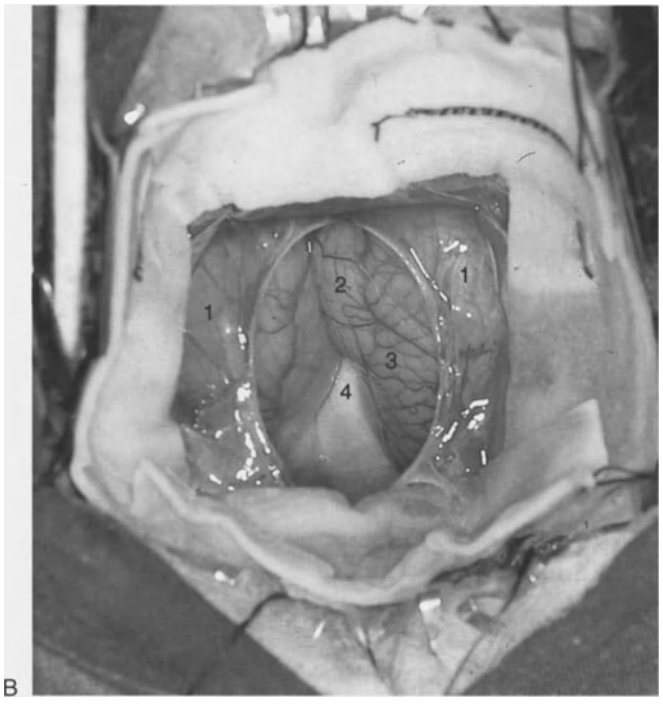


Figure 15.15. (A) This is another example of arachnoidal cyst, in which the cerebrospinal fluid accumulation is over the cerebellar vermis and left cerebellar hemisphere. (B) Arachnoidal cyst of the cisterna magna. One should distinguish between a large cisterna magna, which is not pathogenic, and cystic transformation of the cisterna magna, which is pathogenic. Here, a child with cystic transformation of the cisterna magna is illustrated. Note the very dense arachnoid (*1*) that had formed the posterior wall of the cyst, and the displacement of the vermis (*2*) and the cerebellar hemispheres (*3*). Note also the white, fibrotic, arachnoid membrane, the anterior wall of the arachnoidal cyst of the cisterna magna, densely adherent to the pyramis (*4*), and the inferior medial surfaces of the cerebellar hemispheres. This cyst could not communicate with the subarachnoid spaces. (C) Arachnoidal cyst of the cisterna magna. In this child, different from the one in the previous figure, the arachnoidal cyst occupied almost the entirety of the posterior fossa. The posterior arachnoidal membrane (*1*) has been opened and partially resected. The very dense anterior membrane (*2*) is adherent to the cerebellar vermis in the midline, and the remarkably compressed and flattened cerebellar hemispheres on either side (*3*).

perimental animal, concluded that the aqueductal occlusion was a consequence of the hydrocephalus, not its cause! Thereafter, Russell, studying children at necropsy and correlating these studies with clinical observations of her neurosurgical and neurological colleagues, concluded that "this (the interposition of some obstruction in the cerebral pathway) is responsible for at least 90% of all cases of internal hydrocephalus." Dr. Russell's manuscript refers to obstructive hydrocephalus in the broad sense as being secondary to a physical obstruction at one of the narrowings along the pathways – which she technically described as obstructive hydrocephalus – though she also allowed that defective absorptive mechanisms could be the cause for communicating hydrocephalus. Her work was probably influenced by the work of Dandy and Blackfan (1913) [7], which suggested a distinction between communicating and noncommunicating hydrocephalus on the basis of intraventricular injections of phenolsulfonphthalein, which may be recaptured normally from the urine in 10–12 min and the lumbar subarachnoid space in 2 min. In obstructive hydrocephalus this marker was found in the lumbar subarachnoid space much later, whereas in communicating hydrocephalus the dye was found in the lumbar subarachnoid space in a normal period of time, though its excretion from the urine is remarkably delayed.

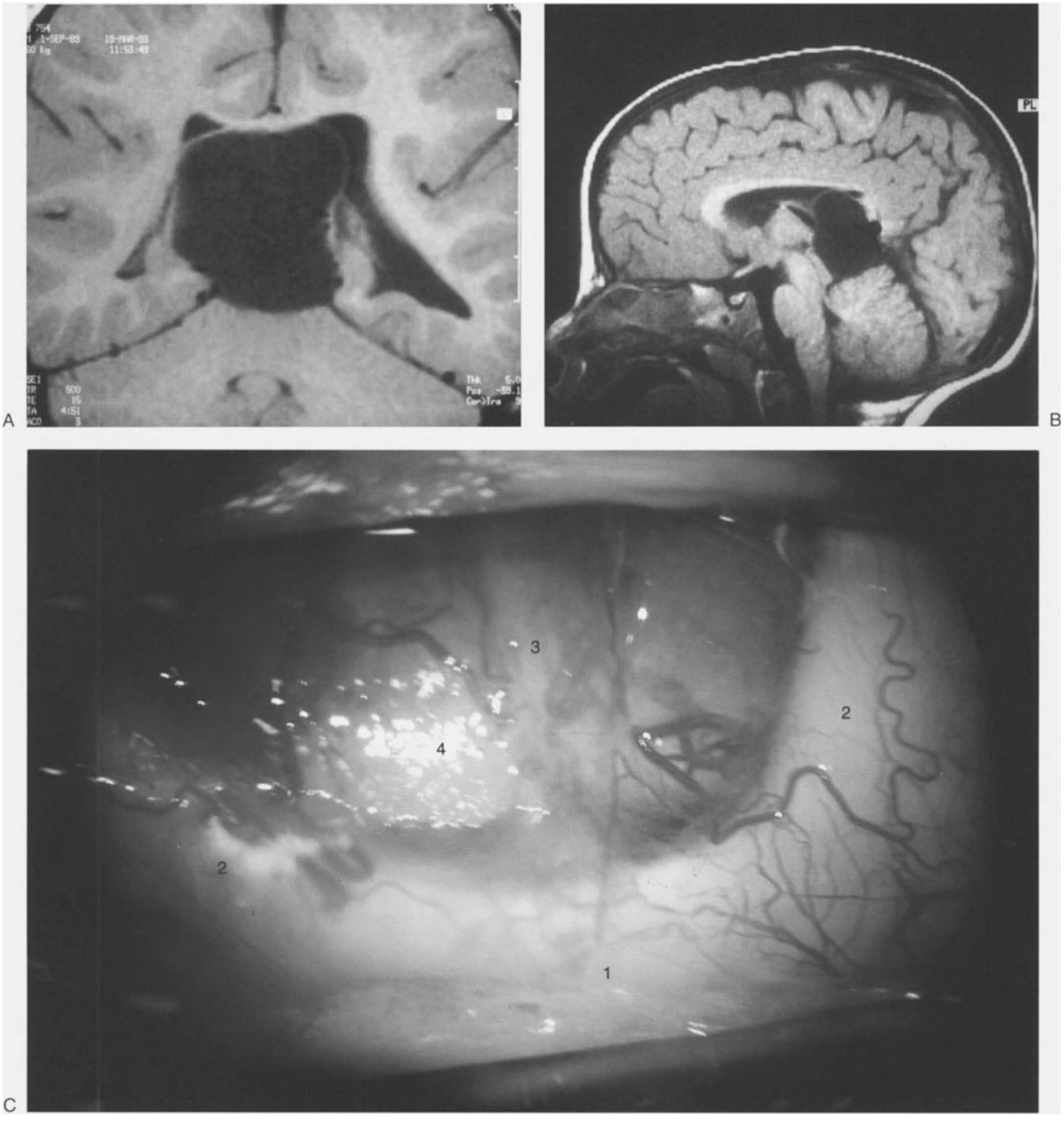


Figure 15.16. This child had an arachnoid cyst of the quadrigeminal cistern. In (A) the enormity of the cyst, its bulging into the posterior aspects of the bodies of both lateral ventricles, and its compression of the culmen monticuli are illustrated. In (B) one identifies very clearly the forward displacement of the thalamus, the remarkable compression of the collicular plate... and the very real distention of the subarachnoid spaces along the medial surface and the convexities of the hemispheres. (C) This is an arachnoid cyst involving the infrachiasmatic and interpeduncular cisterns, displacing the optic chiasm (*1*), optic nerves (*2*), and pituitary stalk (*3*) anterosuperiorly. The glistening, fibrosed arachnoid (*4*) may be seen to form the anterior superior wall of the cyst and engulf the pituitary stalk.

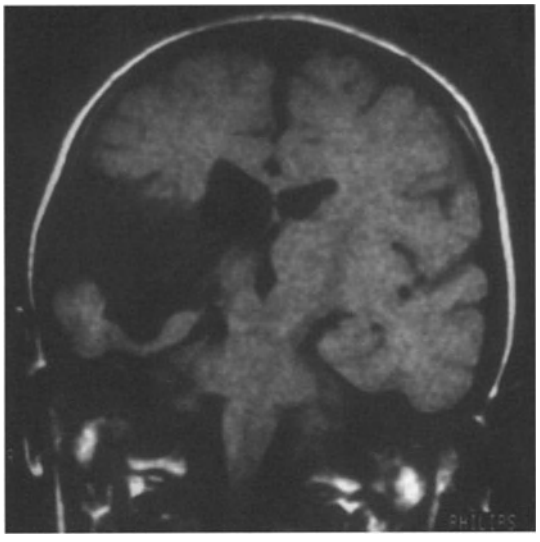
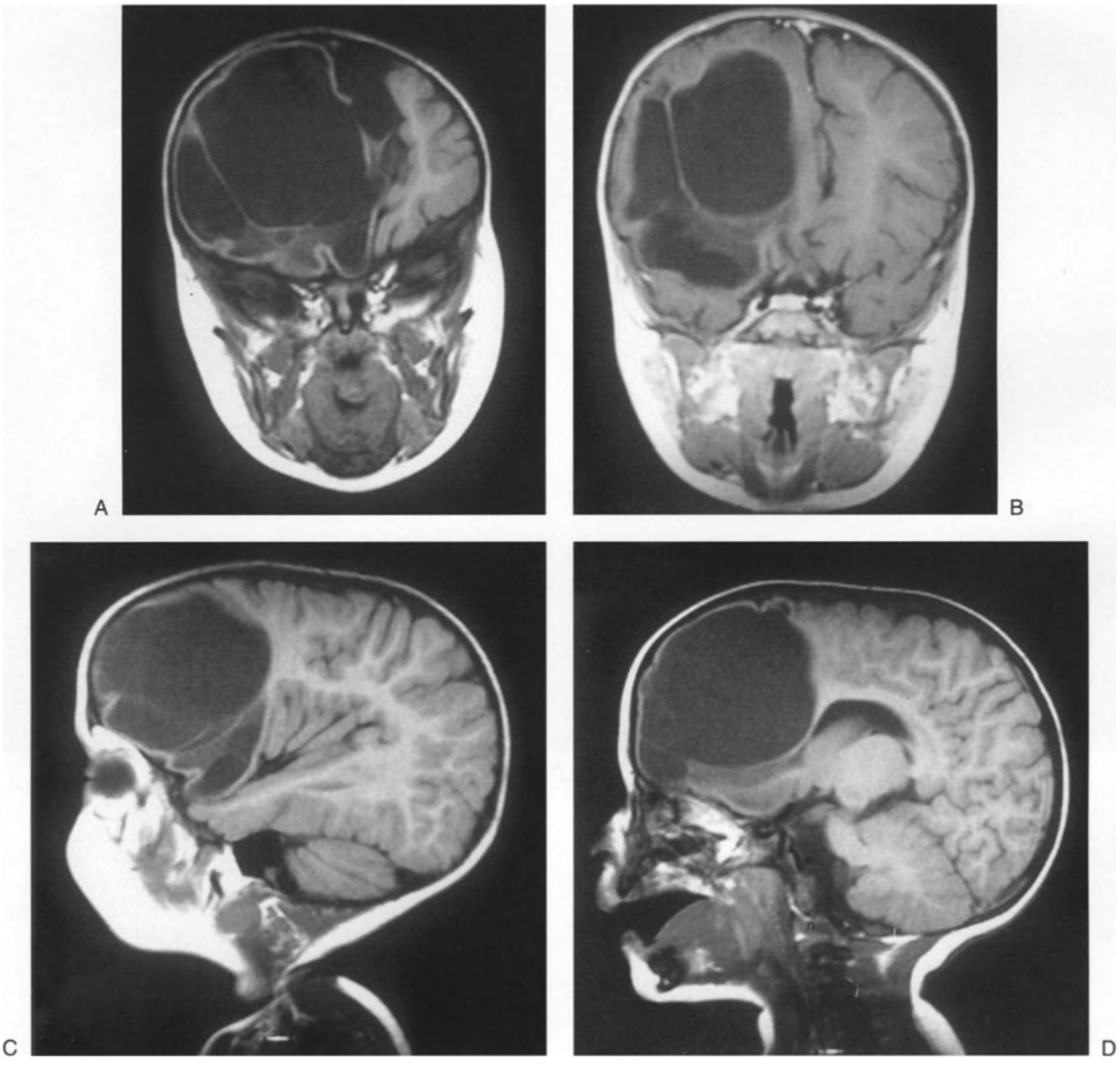


Figure 15.17. Neuroependymal cyst. The cystic cavity is in free communication with the lateral and III ventricles medially and the subarachnoid spaces laterally.
◄

Figure 15.18 (*below*). (A–D) Intraparenchymal cyst. This child had an enormous intraparenchymal, right frontal lobe, cyst which contained cerebrospinal fluid but was not in communication with either the ventricular system or the subarachnoid spaces. One may only speculate on its genesis and nature, but surgical creation of a marsupialization between the cyst cavity and the lateral ventricle resulted in a cure.



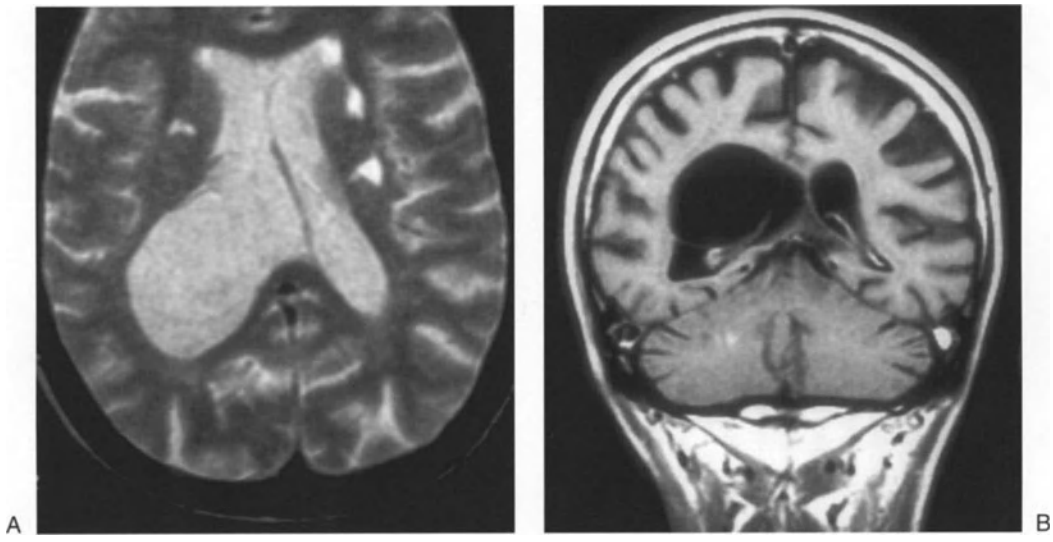


Figure 15.19. This is a neuroepithelial cyst of the lateral ventricle. In (A), an axial SE 2170/90 image shows cystic expansion of the lesion within the right lateral ventricle. The content of the cyst is cerebrospinal fluid, and there is a thin wall surrounding the cyst. That the cyst is exerting a pressure force is documented by the contralateral displacement of the septum pellucidum. In (B), a coronal SE 600/20 post-gadolinium image, one sees adhesions between the cyst and the choroid plexus.

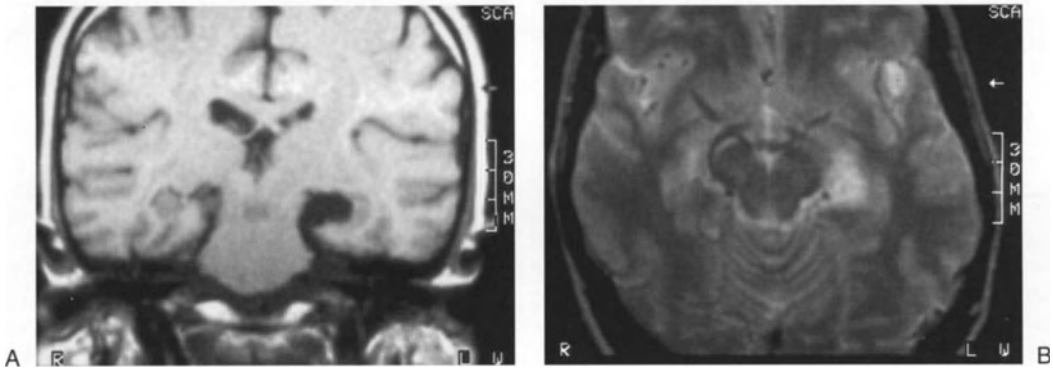


Figure 15.20. Left choroid fissure cyst. In (A), a coronal SE500/25 image, one sees an enlarged left choroid fissure which is, in essence, a cystic lesion holding fluid with a CSF signal intensity. In (B), an axial projection, SE2080/90 image, one sees the cyst displacing laterally the hippocampus.

Reovirus, adenovirus, polyomavirus, and myxovirus have all been found to be causes of aqueductal stenosis when injected intracranially or intraperitoneally. A brief period of inflammation precedes the pathoanatomical changes which result in drastic alterations of ependyma and periaqueductal glia. The aqueduct becomes occluded, without pathological evidence of gliosis. The historical significance of these studies is their demonstration that the absence of gliosis is not sufficient evidence to conclude that aqueductal occlusion (stenosis, forking, atresia) is the result of a developmental lesion. Forking of the aqueduct may be distinguished from either gliogenous stenosis or occlusion by the absence of gliotic tissue in the maldevelopmental forms and its presence in the latter forms. Aqueductal stenosis had always been looked upon mechanistically as the site of primary pathology and not the result of parenchymal changes related to cerebral edema. Raimondi et al. [8] demonstrated, in experimental animals with congenital hydrocephalus, that aqueductal stenosis and occlusion result from periaqueductal edema. Johnson and Johnson [9], in their studies of injecting myxovirus into newborn hamsters, reported a short period of inflammation in which viral particles were limited to the epen-

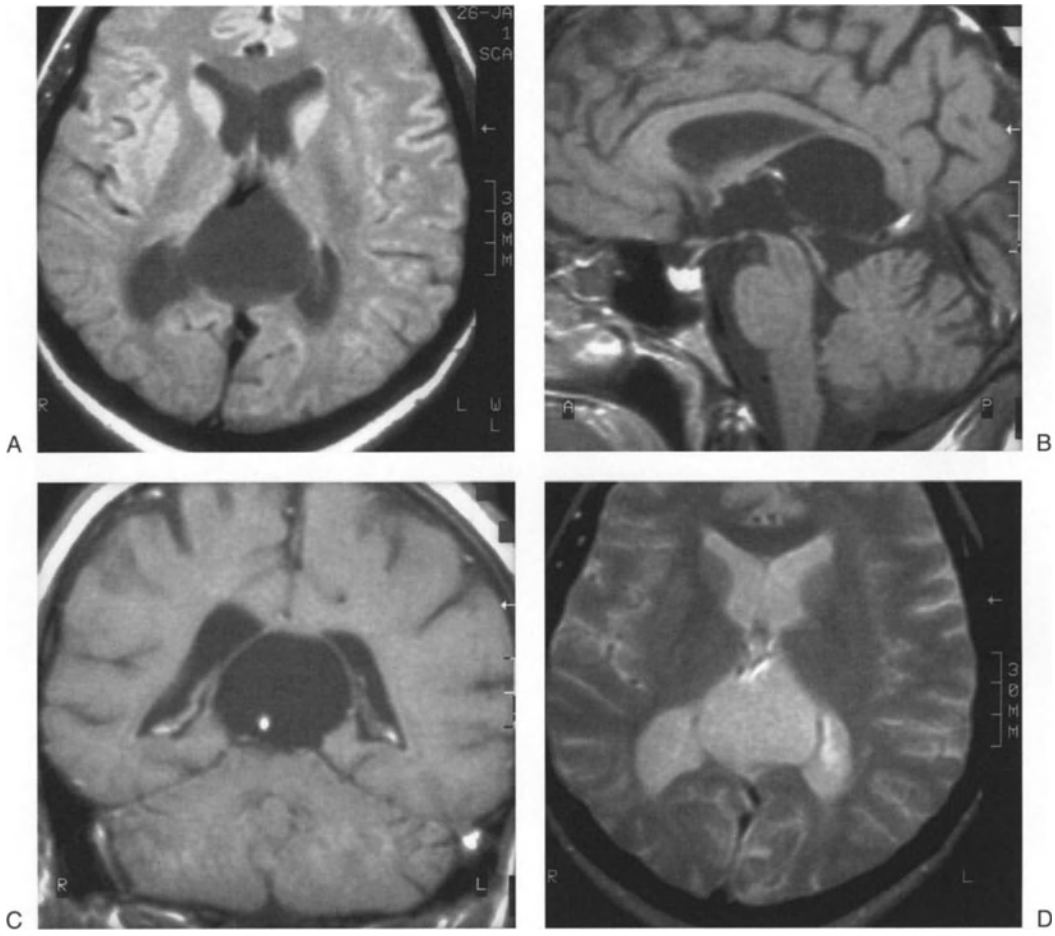


Figure 15.21. This is another case of a cyst of the velum interpositum, one which shows remarkably well the form of the cyst, the location between the roof of the III ventricle and the body of the fornix, the lateral displacement of the body of the fornix on either side and of the cerebellar vermis inferiorly. In (A), an axial SE2000/25 proton density image, one sees the cystic lesion displacing anteriorly and to the right the internal cerebral veins and symmetrically and laterally the thalami. The lesion has the same intensity as CSF. (B) A sagittal SE620/25 T1 weighted post-gadolinium image shows no enhancement of the cyst wall. The fornix is displaced superiorly along with the posterior third of the corpus callosum, the roof of the III ventricle is displaced inferiorly and anteriorly, but there is no compression of the aqueduct of Sylvius. In (C), a postgadolinium 630/25 T1 weighted image, one may appreciate to full advantage the lateral displacement of the fornices... understanding memory impairment, which may occur in these children. (D) This axial T2 weighted image SE2002/90 reveals further the lateral displacement of the bodies of the fornix and the identity of cyst contents with cerebrospinal fluid.

dyma and choroid plexus, followed by the development of hydrocephalus. When the inflammatory process subsided, extensive ependymal loss remained and the stenosed, occluded, or forked aqueduct without gliosis was found.

Russell found it difficult to reconcile the absence of pyogenic exudate in the periaqueductal subependymal glia – her evidence for the maldevelopmental, noninflammatory, origin – with the fact that there is hypertrophy of this tissue, *in the absence of cellular inflammatory exudate, in some cases of postmeningitic hydrocephalus.*

Independent of von Recklinghausen's disease, or such other neoplastic or quasi-neoplastic entities as tuberous sclerosis or subependymal astrocytoma, isolated heterotopic islands of gray matter have been identified as causative factors in obstructing or narrowing the aqueduct of Sylvius. Fulminant leptomeningitis with ventriculitis may result in the silting of pyogenic exudate within the aqueduct, thus occluding the CSF channel with thickened pus. The ependymitis of the aqueduct invariably results in its denuding and secondary periaqueductal gliosis, a more common occurrence in infants and young children than adolescents. Such forms of meningitis as diphtheroid, bacterial, sarcoidosis, and *Candida albicans* have all been reported to

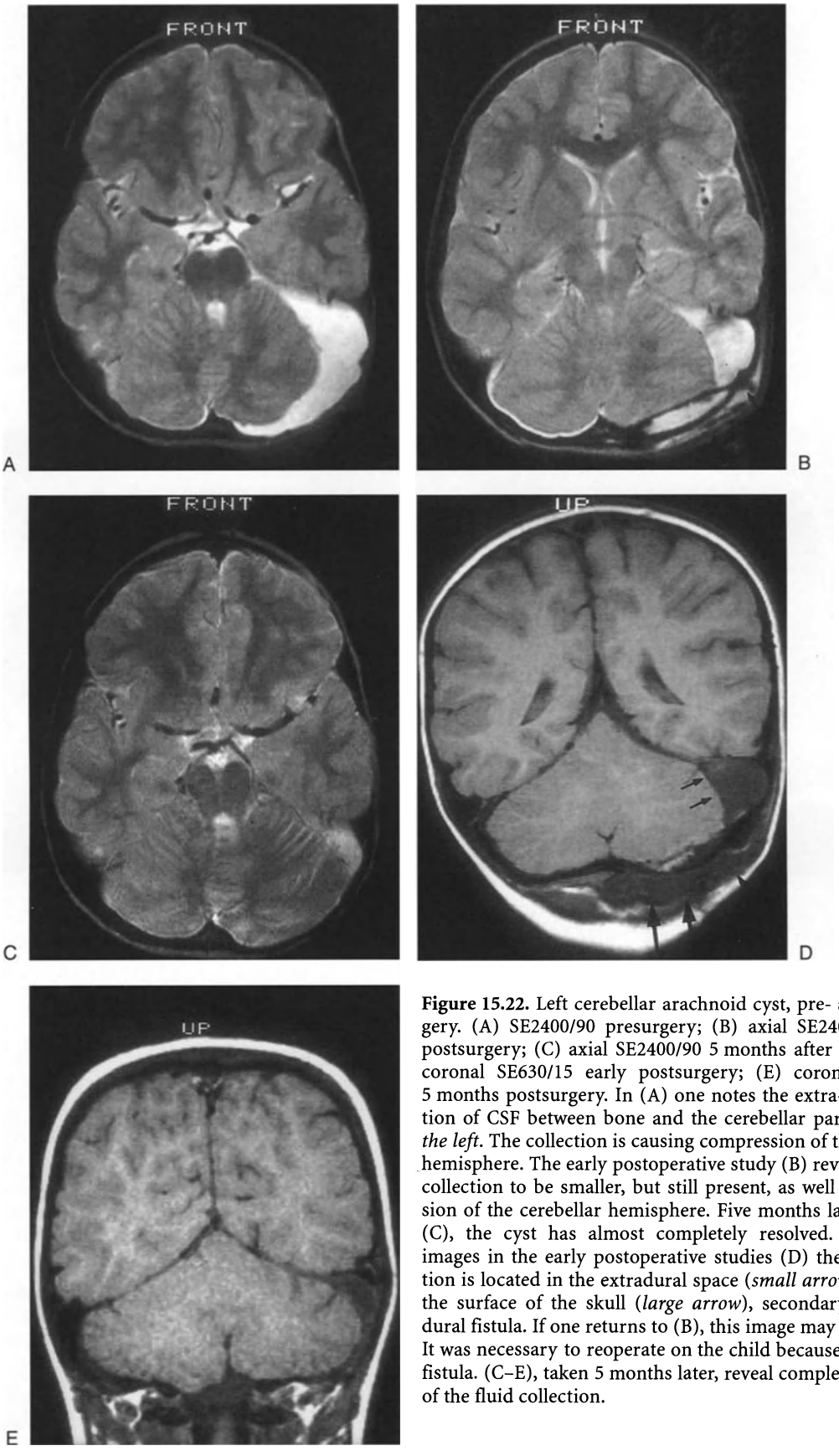


Figure 15.22. Left cerebellar arachnoid cyst, pre- and postsurgery. (A) SE2400/90 presurgery; (B) axial SE2400/90 4 days postsurgery; (C) axial SE2400/90 5 months after surgery; (D) coronal SE630/15 early postsurgery; (E) coronal SE600/22 5 months postsurgery. In (A) one notes the extra-axial collection of CSF between bone and the cerebellar parenchyma *on the left*. The collection is causing compression of the cerebellar hemisphere. The early postoperative study (B) reveals the fluid collection to be smaller, but still present, as well as compression of the cerebellar hemisphere. Five months later, however (C), the cyst has almost completely resolved. On coronal images in the early postoperative studies (D) the cyst collection is located in the extradural space (*small arrows*) and over the surface of the skull (*large arrow*), secondary to a small dural fistula. If one returns to (B), this image may also be seen. It was necessary to reoperate on the child because of the dural fistula. (C–E), taken 5 months later, reveal complete resolution of the fluid collection.

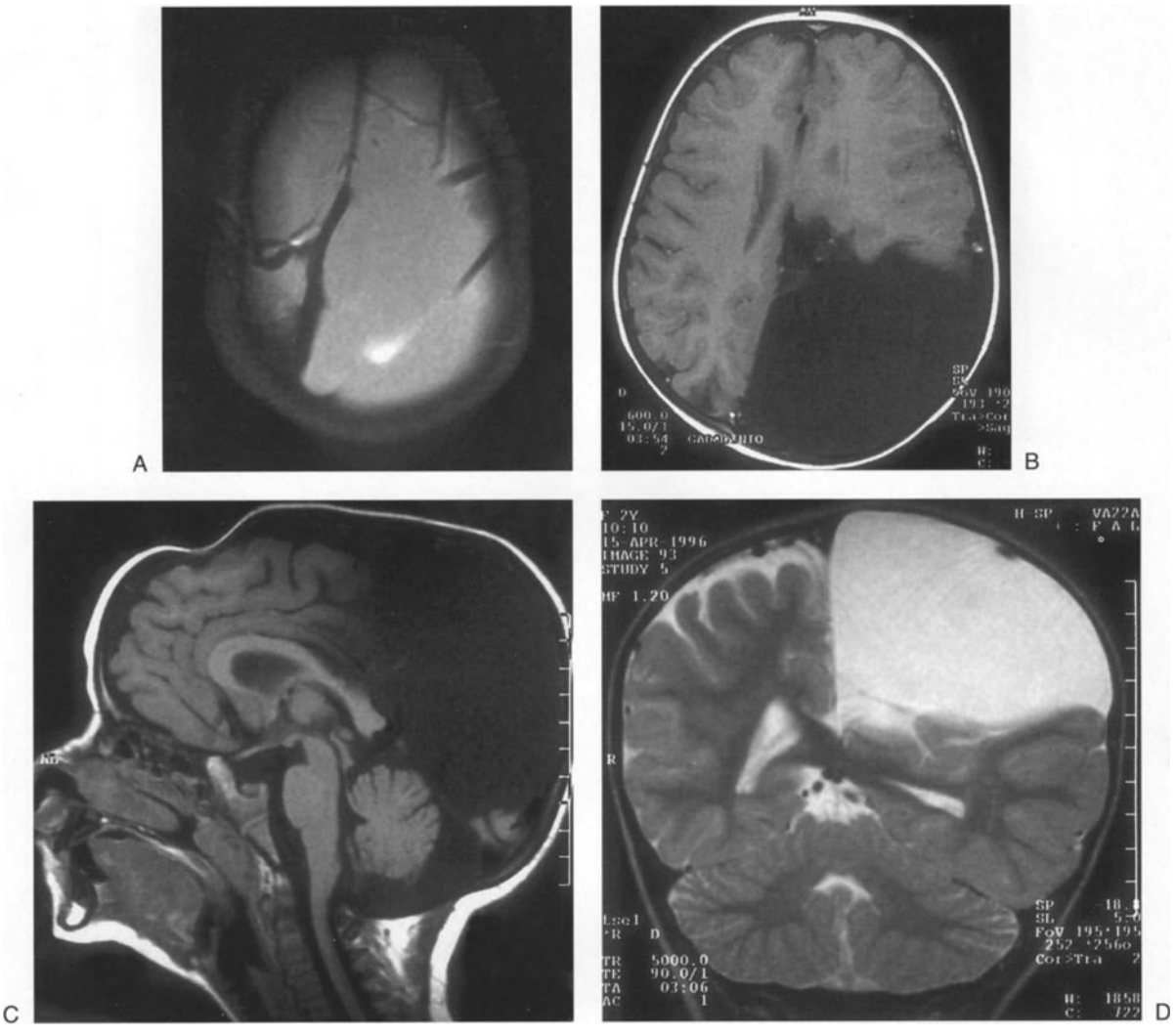
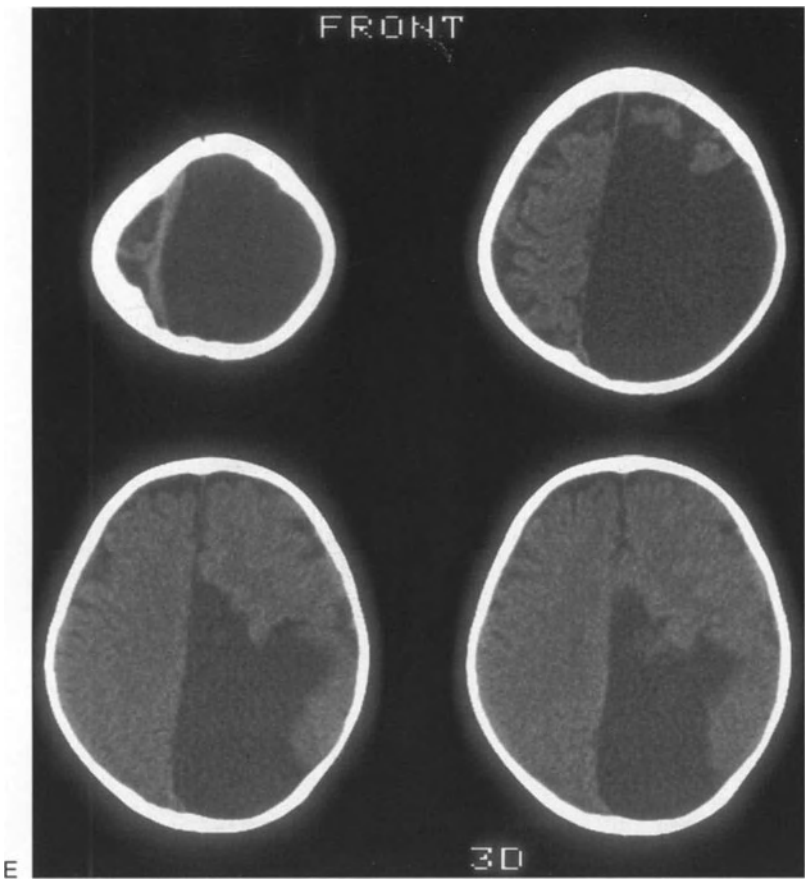


Figure 15.23. Arachnoidal cysts causing displacement of cerebral parenchyma and cortical veins in addition to deformity of the skull. (A) MR axial SE2400/90; (B) axial SE600/15 post-gadolinium; (C) sagittal SE600/15; (D) coronal TSE 500/90. (E–G) see p. 571.

cause aqueductal stenosis and hydrocephalus with pathological changes in the ependyma, and proliferation of glial cushions resulting in gliogenous stenosis. Ventriculitis, as a concomitant infection of neonatal meningitis, has been considered a rather frequent cause for aqueductal obstruction, which may either precede or be a result of the hydrocephalus.

Hereditary forms of aqueductal stenosis in laboratory mice have been known since 1943 when Bonnevie first suggested that the aqueductal stenosis and/or occlusion are probably a consequence of hydrocephalus, not the primary cause. Gruneberg [10] described hereditary forms of aqueductal stenosis in laboratory mice, and Millen demonstrated that this stenosis could be produced in newborn rabbits if the doe were deprived of vitamin A. Interestingly, in a subsequent work the same author showed that, by withholding vitamin A from the doe for prolonged periods of time, the newborn rabbit developed hydrocephalus *but did not show evidence of aqueductal stenosis until late in the disease process*. Millen et al. [11] postulated that hydrocephalus was secondary to oversecretion of CSF, but did not consider that cerebral edema could be a factor in the development of aqueductal stenosis. Dorothy Russell was aware of the fact that in 1933 Clark reported an inherited form of hydrocephalus in mice which was transmitted as a simple mendelian recessive, a form of hydrocephalus characterized histologically by narrowing of the aqueduct. She observed that malformation of the aqueduct is commonly seen in congenital hydrocephalus and that it may occur either simply, as a stenosis, or as atresia.



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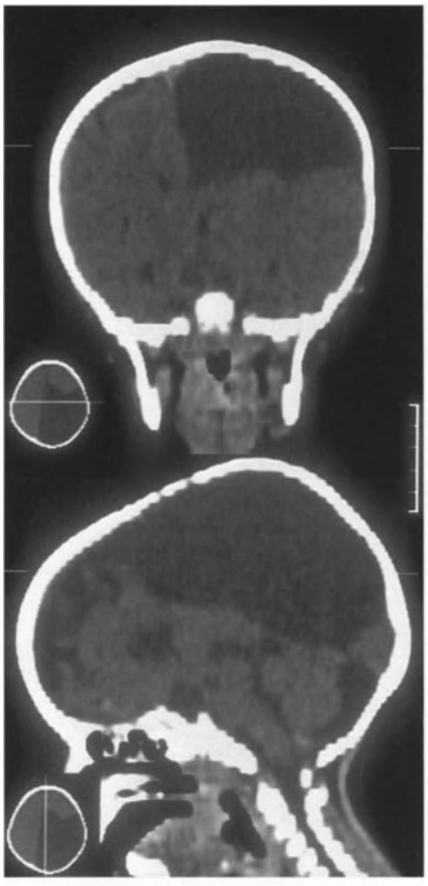

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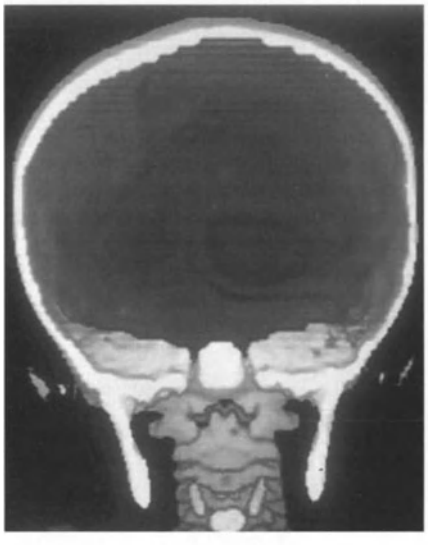

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Figure 15.23. (E) CT scan, multiple views; (F) CT reconstruction in sagittal and coronal planes; (G) three-dimensional CT reconstruction of the coronal view. These examinations show a left extra-axial fluid collection which does not enhance. The lesion has a small nodular component which at surgery was found to communicate with the subarachnoid spaces and to receive cerebrospinal fluid from them. This was coagulated in the hope that it was indeed the point of leakage of cerebrospinal fluid. The lesion caused remarkable displacement of the cortical veins and calcification of the skull. Follow-up studies at 2 months showed no change in the lesion volume, but at 4 months showed a remarkable diminution in volume of the cysts.

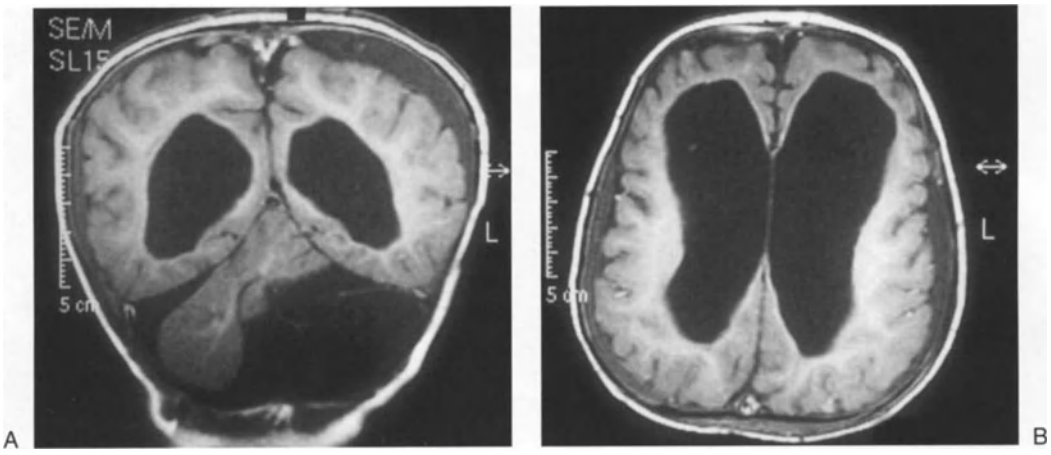


Figure 15.24. This is another example of an arachnoidal cyst of the posterior fossa, displacing the contralateral cerebellar hemisphere, with apparent partial cerebellar hemisphere and vermis dysplasia. In addition, one sees (A) a subdural collection over the hemisphere homolateral to the cyst and (B) remarkable ventriculomegaly.

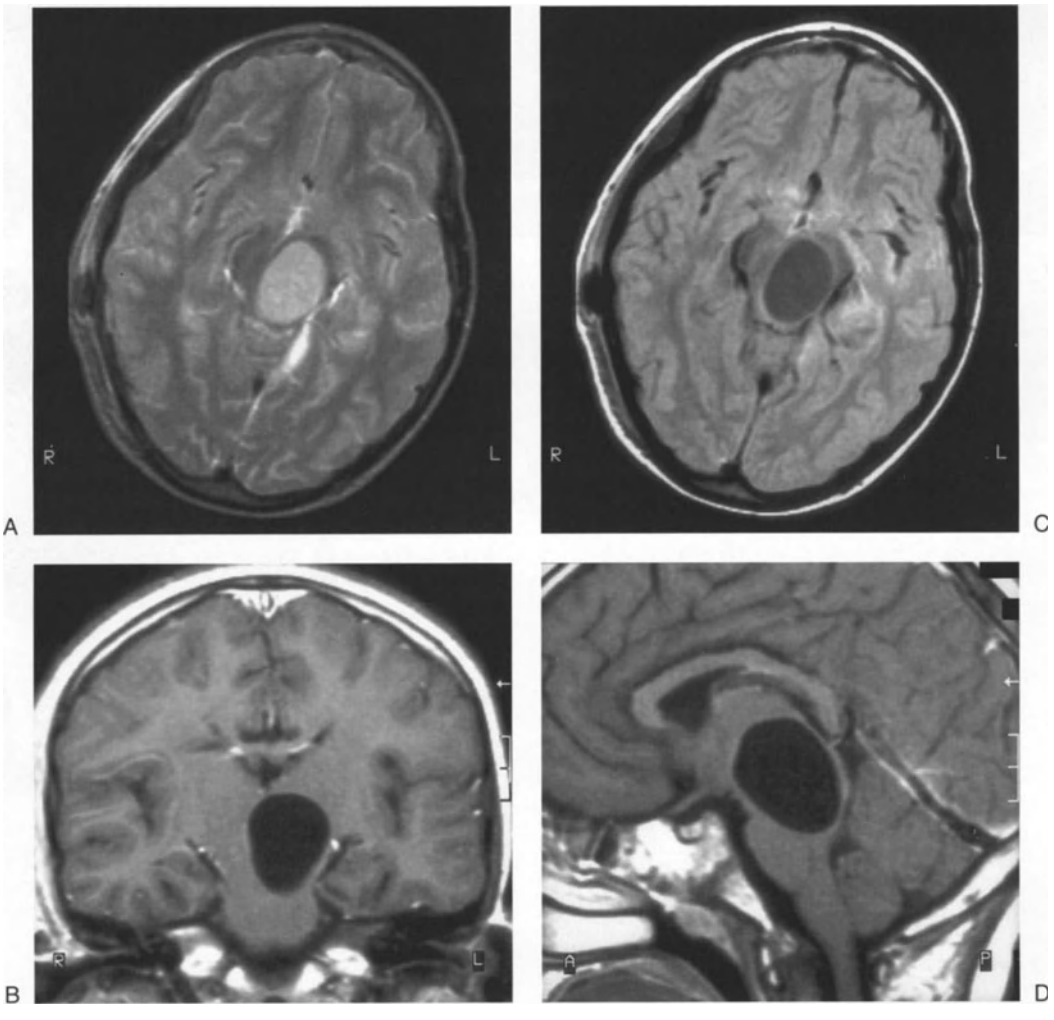


Figure 15.25. Legend see p. 573.

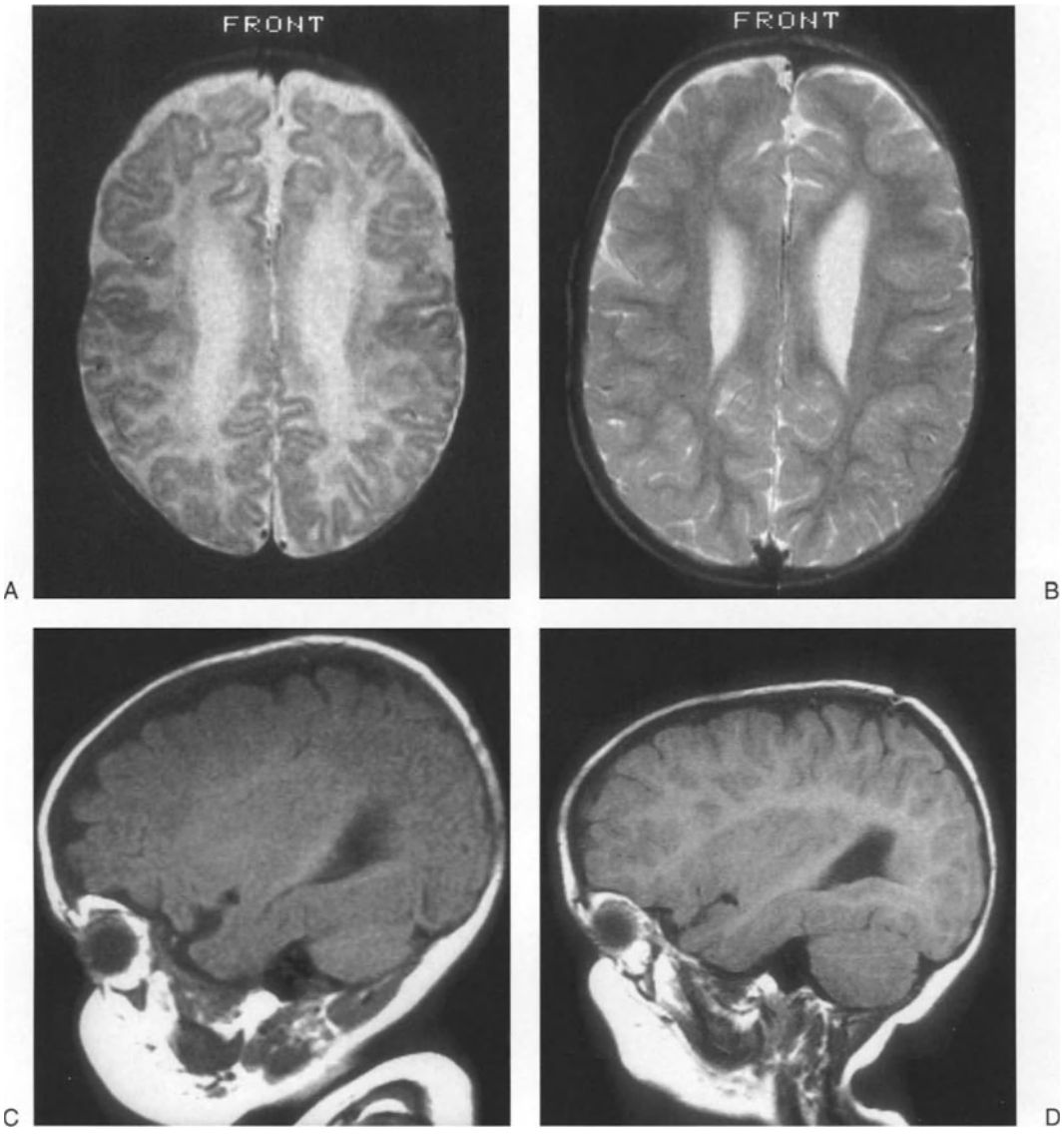


Figure 15.26. Macrocrania secondary to hydrocephalus. (A) and (B) are axial SE2400/90 images at 3 months and 2 years, respectively, without any surgery being performed. They reveal the presence of dilated subarachnoid spaces which subsequently diminished (B). (C) and (D) are sagittal SE600/15 images at 3 months and 2 years, respectively, revealing the reduction in volume of the subarachnoid spaces along the convexity of the hemisphere.

◄

Figure 15.25. Cerebrospinal fluid cyst of the left cerebral peduncle. (A) An axial SE2170/90 image reveals that the lesion has the same CSF signal intensity as cerebrospinal fluid, and (B) the coronal post-gadolinium study reveals that there is no enhancement associated with this lesion. In (C) one may appreciate that the cystic chamber extends from the tegmentun across the substantia nigra and into the peduncle, and in (D) one sees that the lesion extends from the thalamic area to the pons. This was an incidental finding, no symptoms or signs being associated with the lesion, and no surgery was recommended.

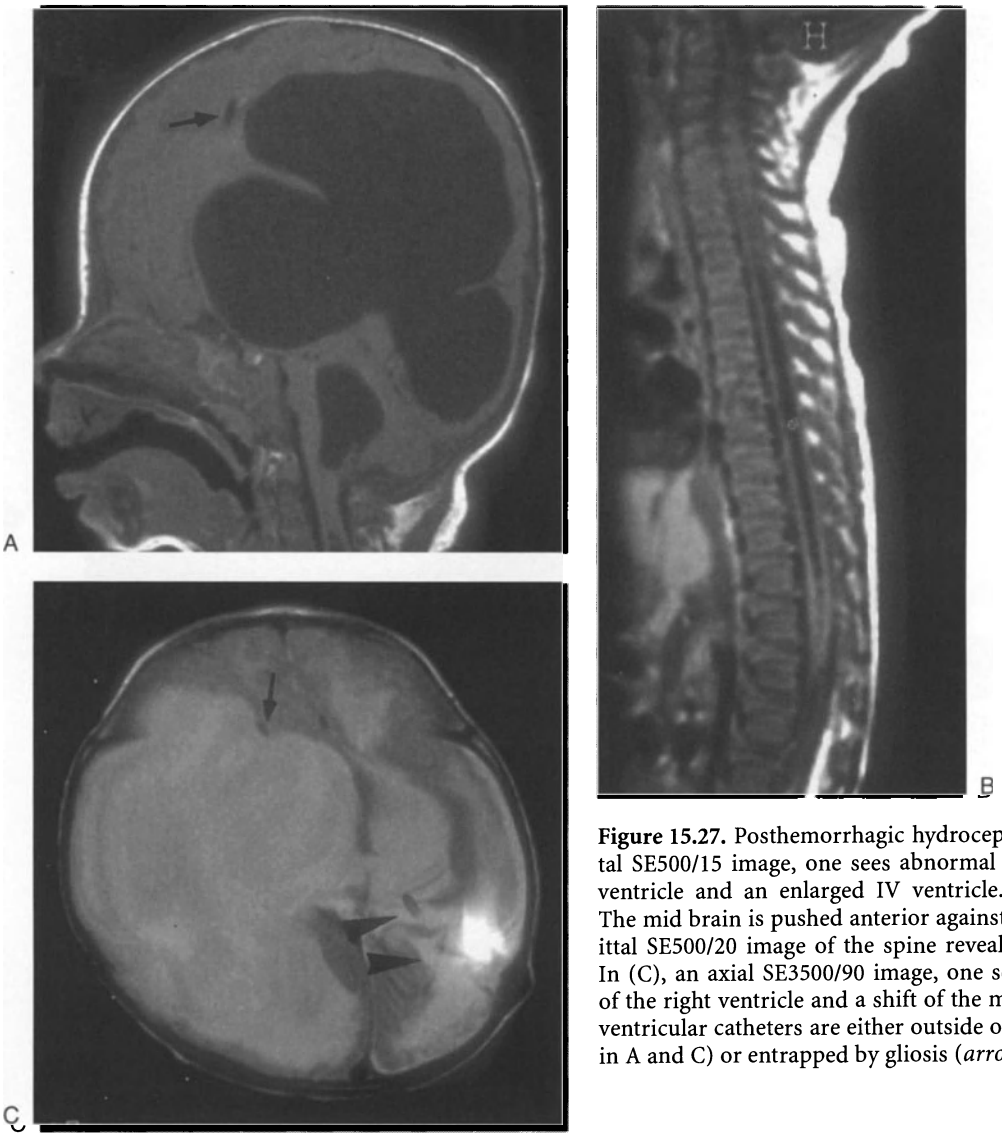


Figure 15.27. Posthemorrhagic hydrocephalus. In (A), a sagittal SE500/15 image, one sees abnormal dilation of the lateral ventricle and an enlarged IV ventricle...which is entrapped. The mid brain is pushed anterior against the clivus. (B) A sagittal SE500/20 image of the spine reveals a medullary syrinx. In (C), an axial SE3500/90 image, one sees abnormal dilation of the right ventricle and a shift of the midline. The two intraventricular catheters are either outside of the ventricle (*arrows* in A and C) or entrapped by gliosis (*arrowhead* in C).

Kesterson and Carlton [12] induced aqueductal stenosis in weaning mice by dietary substitution of hymadroziene to tupraziene with selective mesencephalic encephalitis and periaqueductal edema occurring before loss of ependymal cells, gliosis, astrocytosis, demyelination, and then severe status spongiosus. Despite the fact that they observed evidence of cerebral edema and destruction of the lateral ventricular ependymal cells prior to occlusion of the aqueduct, they considered the aqueductal stenosis to be primary. During the 1970s the causal relationship between viral infection of the brain, the induction of hydrocephalus in experimental animals, and aqueductal stenosis was postulated, with Johnson, Johnson, and Edmunds [13] inducing hydrocephalus by injecting mumps virus directly into hamster brain. They concluded that the aqueductal stenosis resulted from a selective ependymitis involving the entire ventricular system, but producing obstructive changes only in the aqueduct. Pathological changes consisted of perivascular inflammatory reactions, selective ependymitis, and proliferation of subependymal microglial cells. It is remarkable that there is no gliosis underlying the area of ependymal destruction. From this, and the observation that oral-nasal portals of entry could be used to introduce pathogenetic virus into hamster brain, one came to learn that a common virus (natural strain of mumps virus) may produce hydrocephalus in the fetus or newborn. In order for this to occur there must be a low frequency of transplacental passage and some fetal systemic disease of benign character which allow survival within the major extraneural residua of infection.

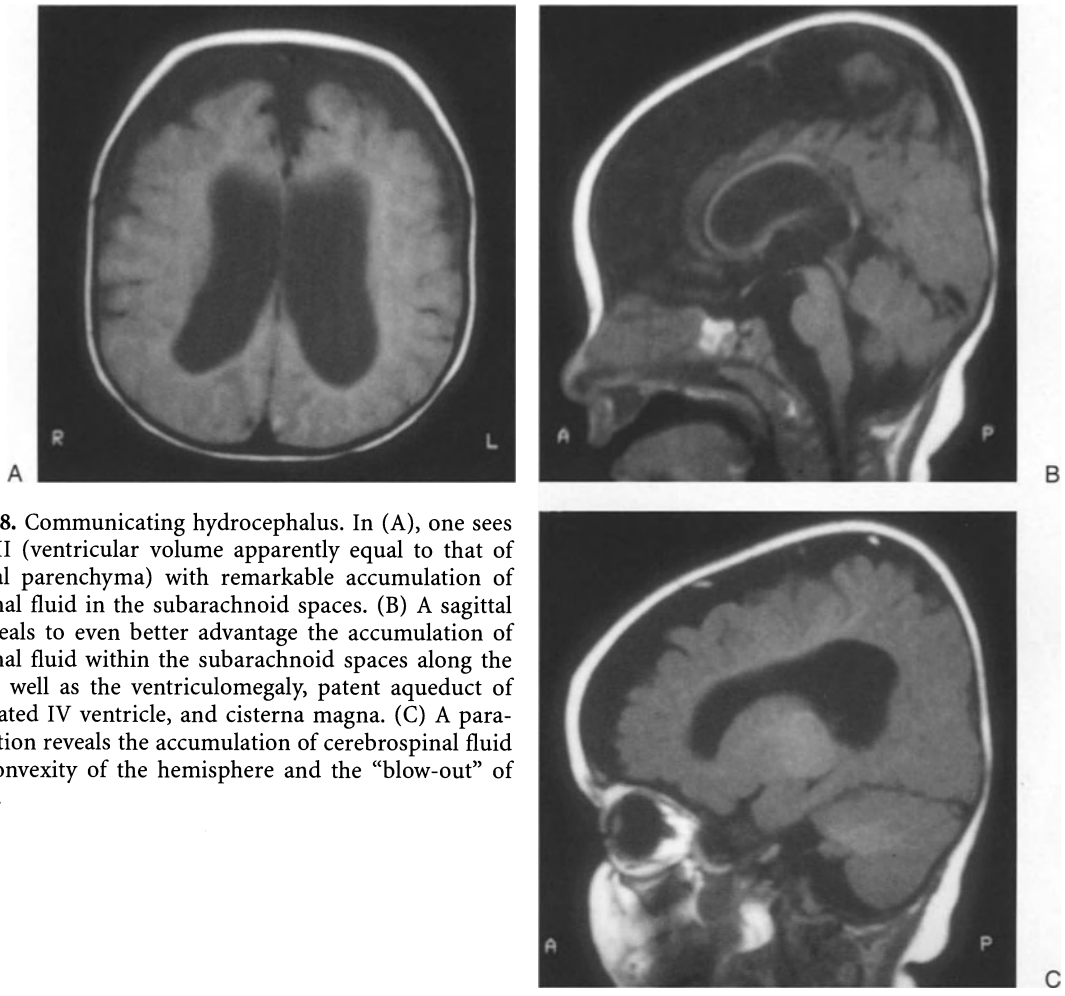


Figure 15.28. Communicating hydrocephalus. In (A), one sees the grade II (ventricular volume apparently equal to that of the cerebral parenchyma) with remarkable accumulation of cerebrospinal fluid in the subarachnoid spaces. (B) A sagittal section reveals to even better advantage the accumulation of cerebrospinal fluid within the subarachnoid spaces along the midline, as well as the ventriculomegaly, patent aqueduct of Sylvius, dilated IV ventricle, and cisterna magna. (C) A parasagittal section reveals the accumulation of cerebrospinal fluid over the convexity of the hemisphere and the "blow-out" of the trigone.

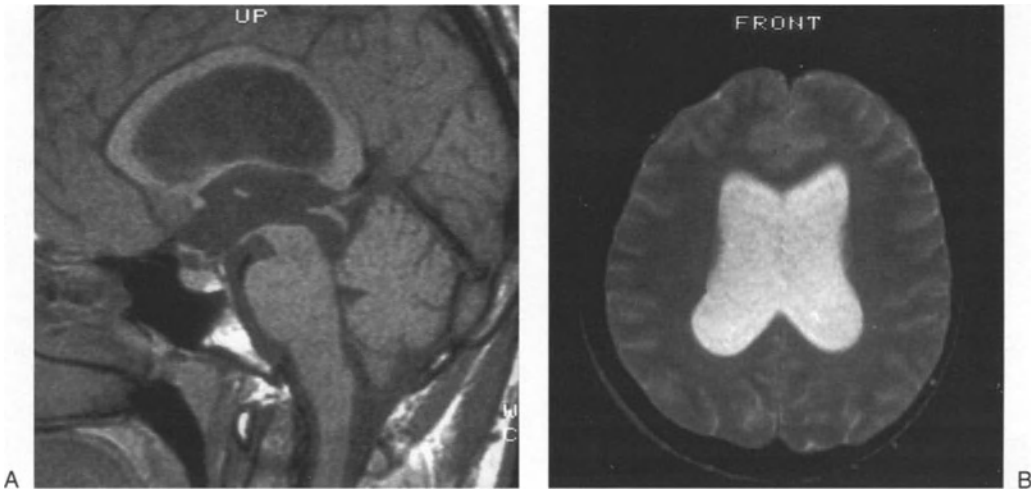
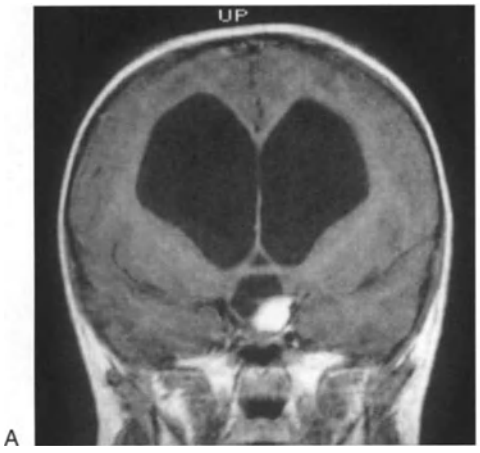


Figure 15.29. This is a most interesting case: idiopathic triventricular hydrocephalus *without* occlusion of the aqueduct. In fact, the aqueduct is remarkably dilated. (A) A sagittal SE600/15 image and (B) an axial SE2400/90 image are MRI studies performed electively after the ventriculomegaly was revealed on a routine CT scan following head injury. The MRI studies reveal enlarged lateral ventricles, an enlarged III ventricle, and a remarkably dilated aqueduct. No transependymal edema is noted. The IV ventricle is small.



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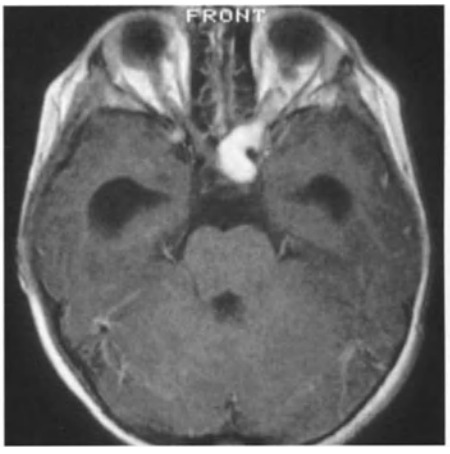

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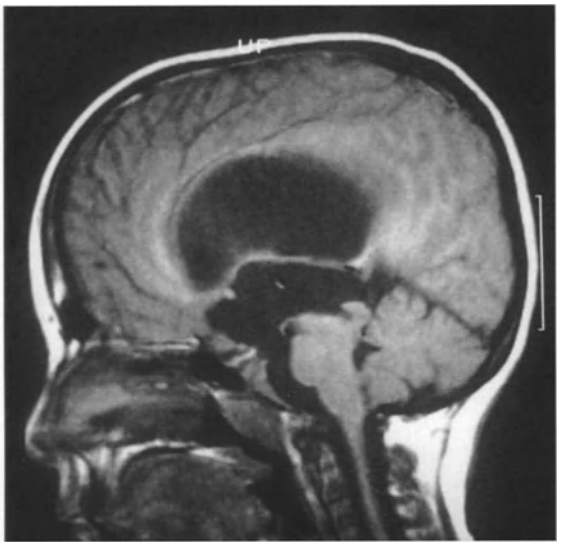
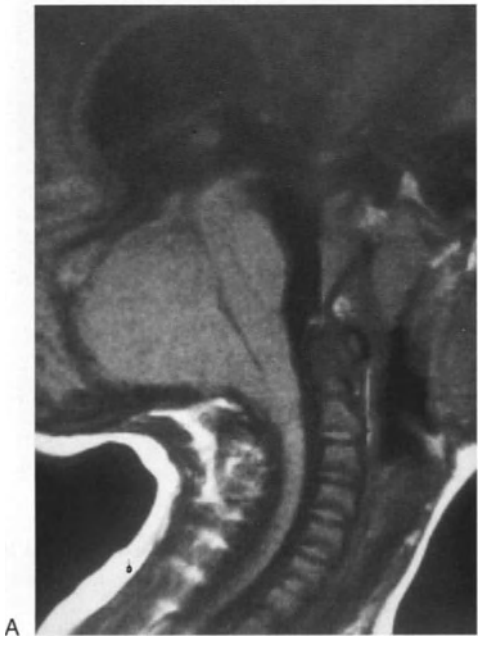


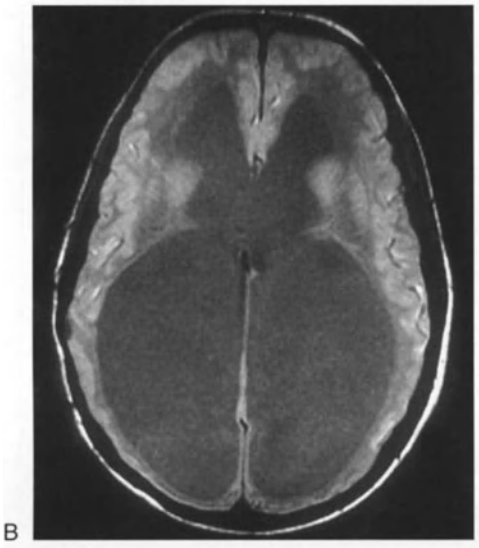
B

Figure 15.30. Hydrocephalus secondary to aqueductal stenosis in a child with von Recklinghausen's disease and a glioma involving the optic nerve and chiasm. (A) Long-standing evidence of triventriculomegaly with an enhancing mass of the left optic nerve and chiasm. (B) This sagittal projection reveals the remarkable dilation of the III ventricle, occlusion of the aqueduct of Sylvius, small IV ventricle, and cisterna magna. (C) This axial projection, post-gadolinium, reveals very well the glioma growing from the optic nerve, into the chiasm, and backward along the optic tract.



A

Figure 15.31. Chiari II malformation with hydrocephalus. Though the Chiari II malformation is discussed extensively in Chap. 14, "Congenital Anomalies," it is necessary to address this subject in this chapter since bi or triventricular hydrocephalus is present in slightly less than 80% of meningomyelocele children who have a true Chiari II malformation. (A) This lateral MRI study reveals a moderate Chiari II malformation. There is tonsillar herniation, the bulb is located below the foramen magnum, the aqueduct of Sylvius is lengthened and flattened as is the IV ventricle, the culmen monticuli of the cerebellum is flattened, and the straight sinus and vein of Galen form a right angle. There is marked dilation of the III ventricle and the mammillary bodies are displaced anteriorly. (B, C) see p. 577.



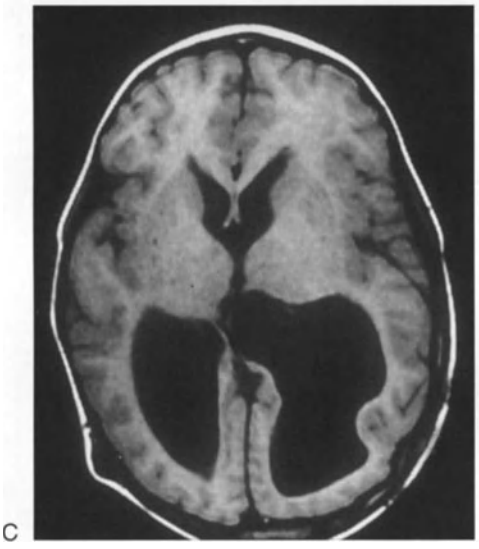
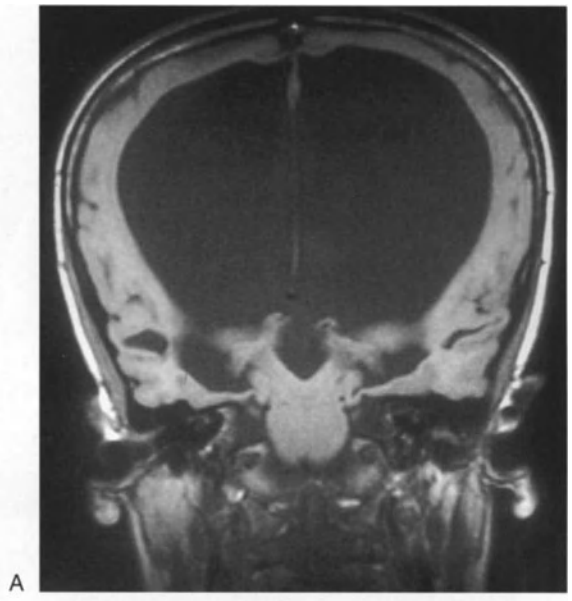


Figure 15.31. (B) This axial projection reveals the remarkable dilation of the lateral ventricles, particularly the region of the trigone and the occipital horns. This is most typical of the Chiari II malformation. (C) This is another child with meningomyelocele and the Chiari II malformation, and with the same, but much less remarkable, pattern of ventricular dilation: trigones and occipital horns much more dilated than the anterior horns. In this child, one notes that the III ventricle is almost normal in size.



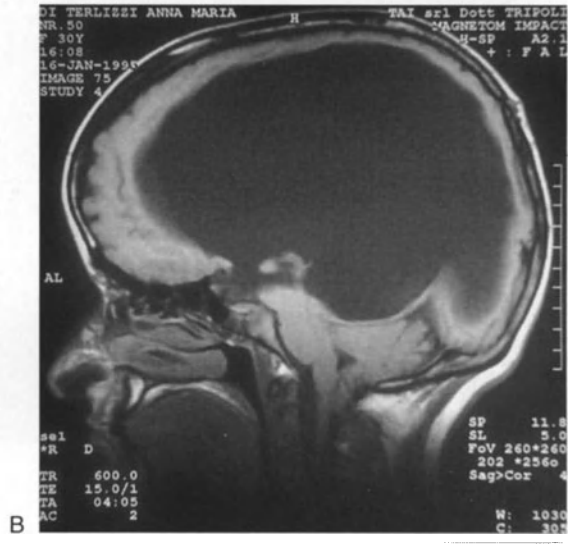


Figure 15.32. Chiari II malformation. This is another most instructive example of the bizarre ventricular alterations in Chiari II patients. This 30-year-old female functions positively in her world. In (A) one notices the biventricular hydrocephalus and only minimally dilated III ventricle which actually has the form in the coronal projection of the... IV ventricle. In (B), one is able to identify immediately that the major pressure force emanated originally from the region of the trigone, the cerebellum is flattened superiorly and herniated to the level of C1 inferiorly, the IV ventricle is at the foramen magnum as is the inferior portion of the pons: *constrictive hydrocephalus. What is most important to observe in this study is the anteroinferior displacement of the III ventricle, with it coming to rest upon the diaphragma sellae, and its shallow size. At a time when III ventriculostomies are being performed for almost all types of hydrocephalus, this image is most instructive since it advises the surgeon that Chiari II malformation patients by and large are not candidates for III ventriculosotomy: there is no interpeduncular cistern.*

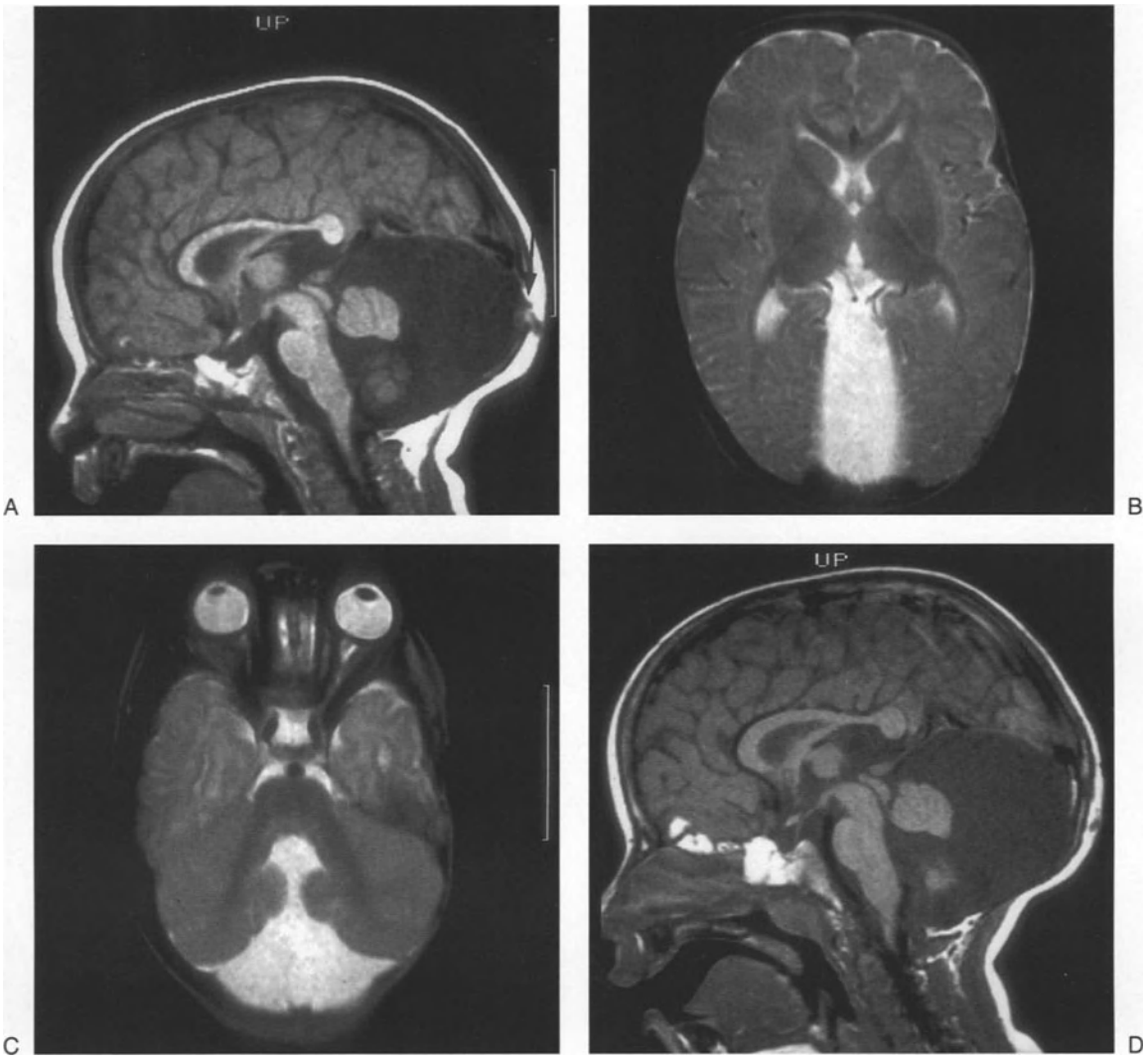


Figure 15.33. The Dandy-Walker malformation, cystic transformation of the IV ventricle, is very well known, but its variants are confusing and of dubious clinical significance. Also, cystic transformation of the IV ventricle in association with occipital encephalocele or suboccipital (Chiari III malformation) encephalocele were once very common clinical observations. Now, at least in the industrialized world, these malformations have practically disappeared from the horizon. In this illustration, a Dandy-Walker malformation in a child with an occipital meningocele is illustrated, pre- and postoperatively. (A) Sagittal E600/15 image showing a small defect in the occipital bone with herniation of the meninges (*arrow*). The vermis is absent and the IV ventricle is huge, with elevation of the straight sinus and enlarged posterior fossa. There is no supratentorial hydrocephalus! (B) and (C) show axial SE2400/90 studies with cuts at levels through the immediate infratentorial area (B) and through the IV ventricle/valeculla/Dandy-Walker cyst area (C). (D) This sagittal SE600/15 image is the postsurgical study which reveals closure of the meningocele, no alteration in the Dandy-Walker cyst, and no alteration in the hydrocephalus.

Consequently, viral invasion of the developing nervous system may cause (1) a mild form of neurologic disease which does not result in parenchymal destruction; (2) a rather specific infection of the ependyma which results in cytolysis; (3) glial-vascular repair; (4) aqueductal stenosis, and, consequently, (5) obstructive hydrocephalus. Other etiologic factors for producing stenotic aqueducts in animals with hydrocephalus are hypovitaminosis A, hypervitaminosis A, delirium, cuprizone, diaszo dies, autologous antisera, salisylates, and ionizing radiation.

The most significant matter historically is the fact that the Hy-1 mice described by Clark in 1934 [14] to have hydrocephalus which was secondary to aqueductal stenosis were subsequently reported by Bonnevie in 1943 [5] to suffer generalized hydrocephalus first and aqueductal stenosis second!

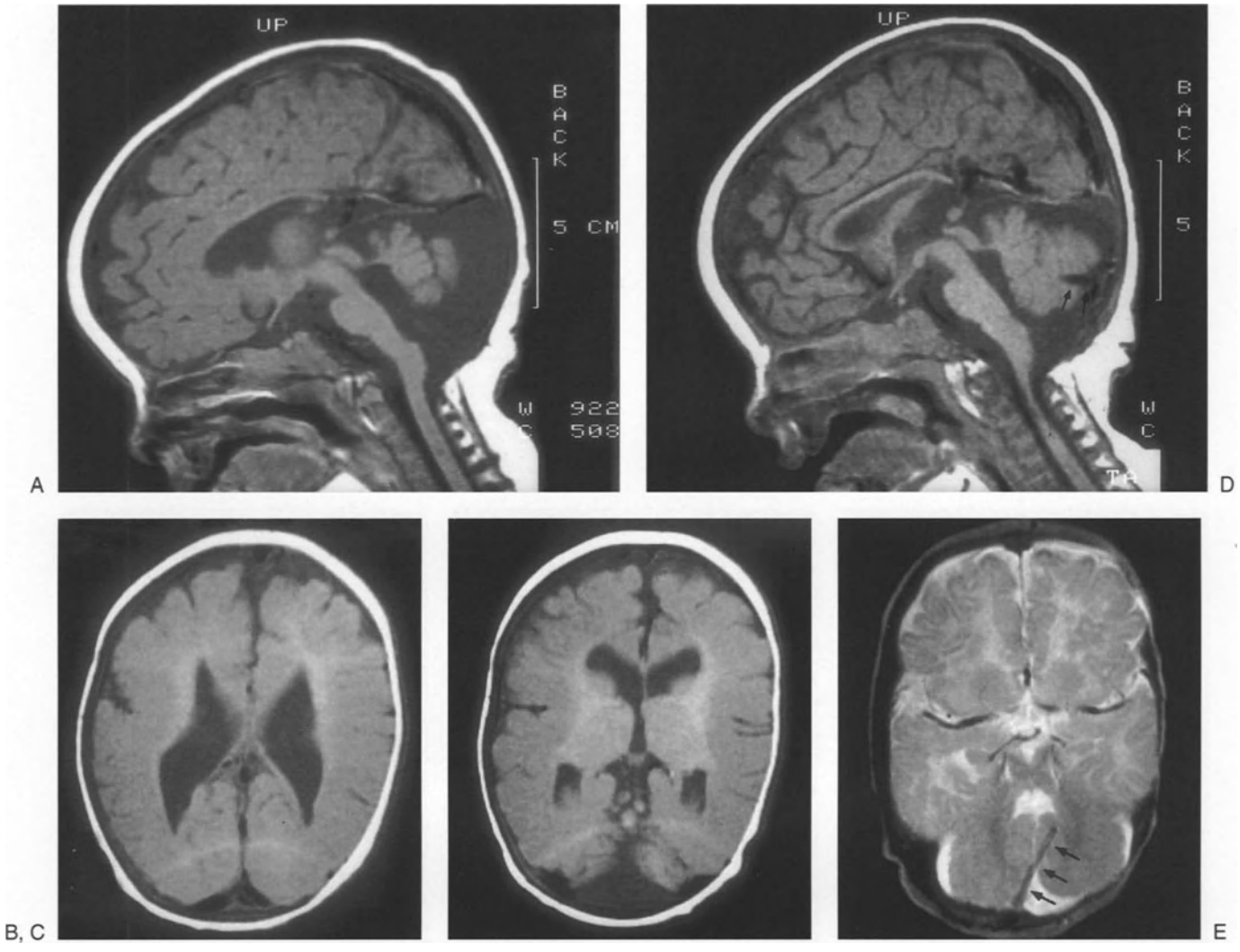
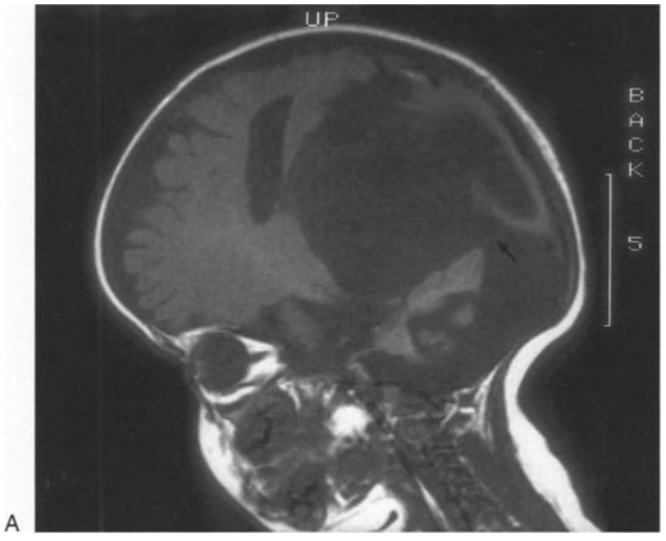


Figure 15.34. Dandy-Walker malformations pre- and postshunt. In (A), a sagittal SE600/15 image, and (B) and (C), axial SE600/15 images, one sees an enlarged posterior fossa with a straight sinus which is elevated. The vermis is absent, and there is a huge IV ventricle. The axial images also demonstrate enlarged lateral ventricles. Remarkable dilation of the subarachnoid spaces over the hemispheres is shown in (A), (B), and (C). (D) and (E) are, respectively, sagittal SE600/15 and axial SE2400/90 postshunt images. What is most remarkable is the diminution in volume of the cystic transformation of the "IV ventricle" with an almost normal return to form of the IV ventricle and presence of the cisterna magna. *The arrows* indicate the shunt within the cystic chamber.

Very likely, then, the same pathogenetic factors are active in the production of hydrocephalus whether in intrauterine or extrauterine life. Also, it seems quite artificial to attempt to identify certain pathologic agents which are capable of selectively dåmaging the ependymal cells in the periaqueductal glia of the aqueduct of Sylvius, leaving intact lateral, III, and IV ventricle ependyma. It is intriguing to contemplate in dynamic pathological terms how a given pathogenetic agent may cause edema, gliosis, axonal and dendritic degeneration, neuronal changes, spongiosis, etc., in one portion of the brain but leave the other quite intact. Very likely, what we are observing are single events in selective examples, in which the most severe changes observed occurred in one or another portion of the brain and, consequently, it is impossible to conclude that selective pathological processes may occur within the area of the aqueduct of Sylvius and not be the expressions of overall cerebral or ependymal involvement.

Functional aqueductal occlusion may result from expansile forces within both the supra- and infratentorial compartments with remarkable dilation of the III and IV ventricles. The foreshortened aqueduct produces a plication between the III and IV ventricles which occludes completely communication between these chambers when they are maximally dilated. This was de-



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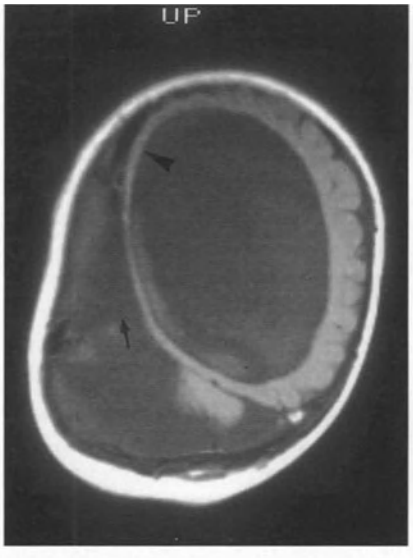

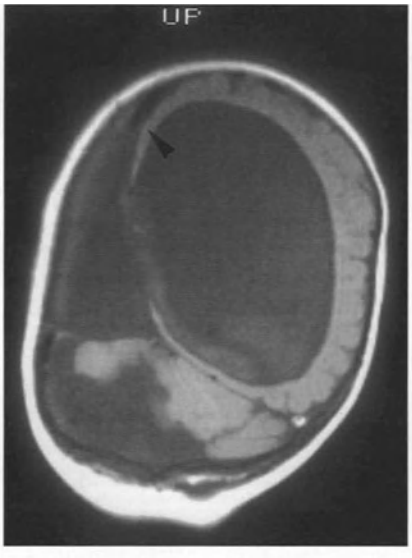

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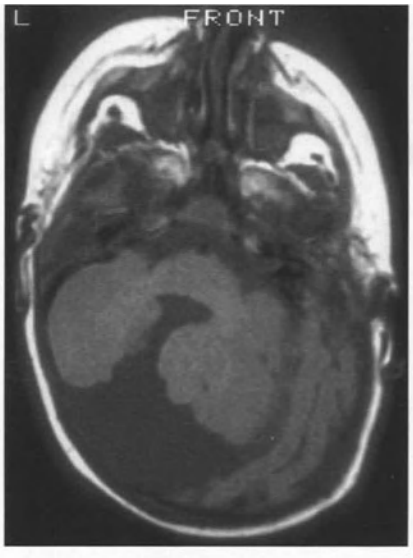

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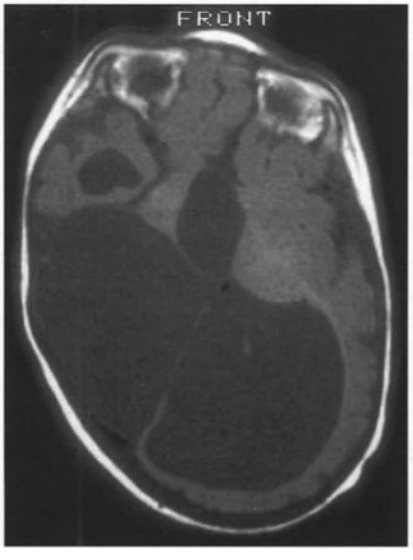
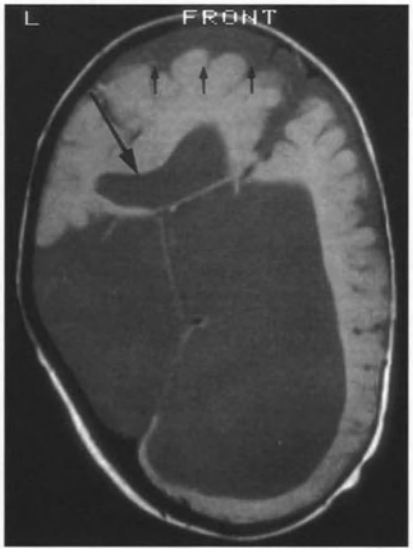
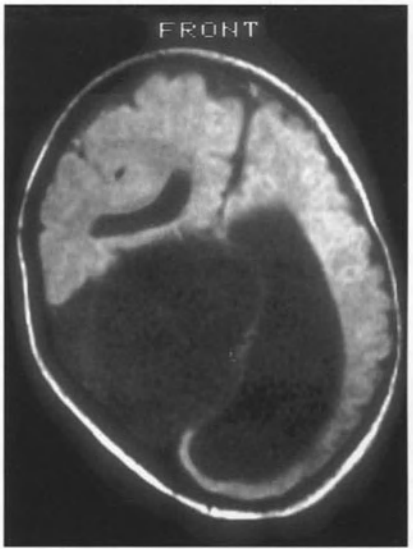






E, F

G

Figure 15.35. Dandy-Walker variant. (A) Sagittal SE600/20, (B) and (C) coronal SE750/22, and (D) and (E) axial SE750/22 images. (A–E) reveal the cyst in the posterior fossa, with upward extension into the supratentorial compartment through a defect in the tentorium *on the right* (*arrows in* A and B). This lesion causes compression of the aqueduct with supratentorial hydrocephalus. There is upward displacement of the straight sinus (*arrowhead in* B and C), and the vermis is absent (D). In (F) and (G), axial SE2400/15 images of the same patient at 8 months of age (F) and 1 month (G). One notes that in (F) there is dilation of the subarachnoid spaces (*arrows*) more than in (G), indicating that with the passage of time the hydrocephalus is progressive and more remarkably of the "communicating" variety. *Hence, one must ask oneself why this is called a Dandy-Walker... variant... since there is not occlusion of the foramina of Luschka or Magendie*

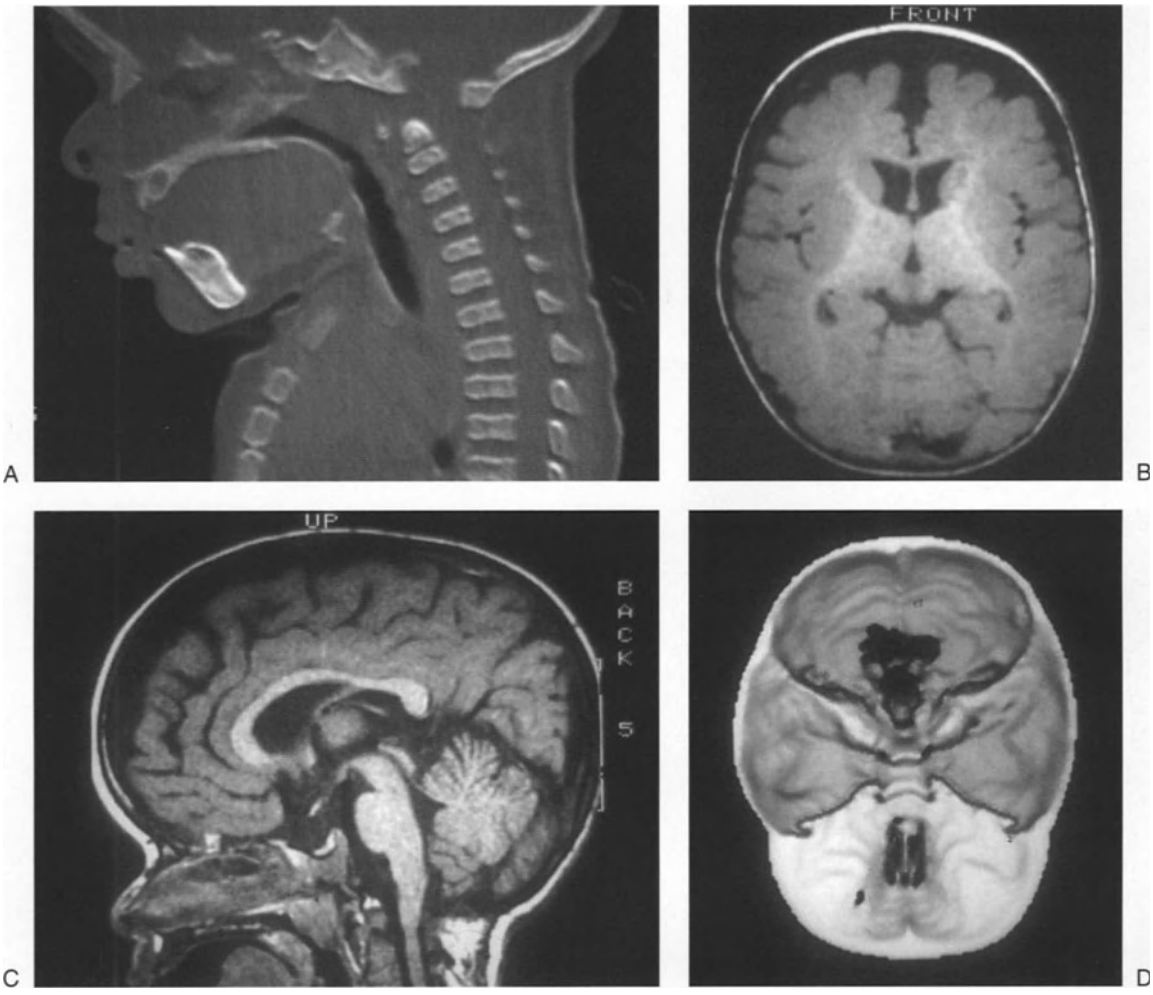


Figure 15.36. Congenital atresia of the foramen magnum. (A) The extremely small foramen magnum is well illustrated. (B) One sees only minimally dilated ventricles but remarkably dilated subarachnoid spaces over the frontal lobes anteriorly, medially, and laterally. (C) This sagittal study reveals the dilation of the subarachnoid spaces but normal aqueduct and IV ventricle. (D) This three-dimensional CT study puts into relief the extraordinarily constricted foramen magnum.

scribed by Raimondi et al. [15] in their study of patients with the Dandy-Walker cyst and confirmed by Jakurbowski and Jefferson [16].

Occlusion of the foramina of Luschka and Magendie (Dandy-Walker cyst) impairs the passage of CSF from the IV ventricle. Consequently, this chamber is transformed into a cystic structure. There is very often present an associated dysgenesis of the inferior third of the vermis, with resultant anterosuperior displacement of the remaining two-thirds of cerebellar vermis. The cerebellar hemispheres are displaced laterally, anteriorly, and superiorly, as the cisterna magna fails to develop and the IV ventricle itself extends inferiorly to the level of the second cervical vertebra. This transformation of the IV ventricle into a massive cyst is expressed anatomically by elevation of the tentorium and the torcular Herophili, and forward displacement of the brainstem.

The basic triad of Dandy-Walker syndrome (DWS) is the supratentorial hydrocephalus, the posterior fossa cyst, and dysgenesis of the cerebellar vermis.

In 1914, Dandy and Blackfan [7] reported on a 13-month-old girl with noncommunicating hydrocephalus. The disease had developed after she suffered fever, convulsions, and decerebrate rigidity at the age of 4 months, incurring progressive cranial enlargement and death. Autopsy revealed supratentorial hydrocephalus associated with aqueduct dilatation, cystic dilatation of the fourth ventricle, and atrophy of the rostral part of cerebellar vermis. As neither the foramen of Magendie nor the foramen of Luschka was recognized to be patent, the investigators concluded that obstruction of the CSF pathway had caused hydrocephalus in this patient.

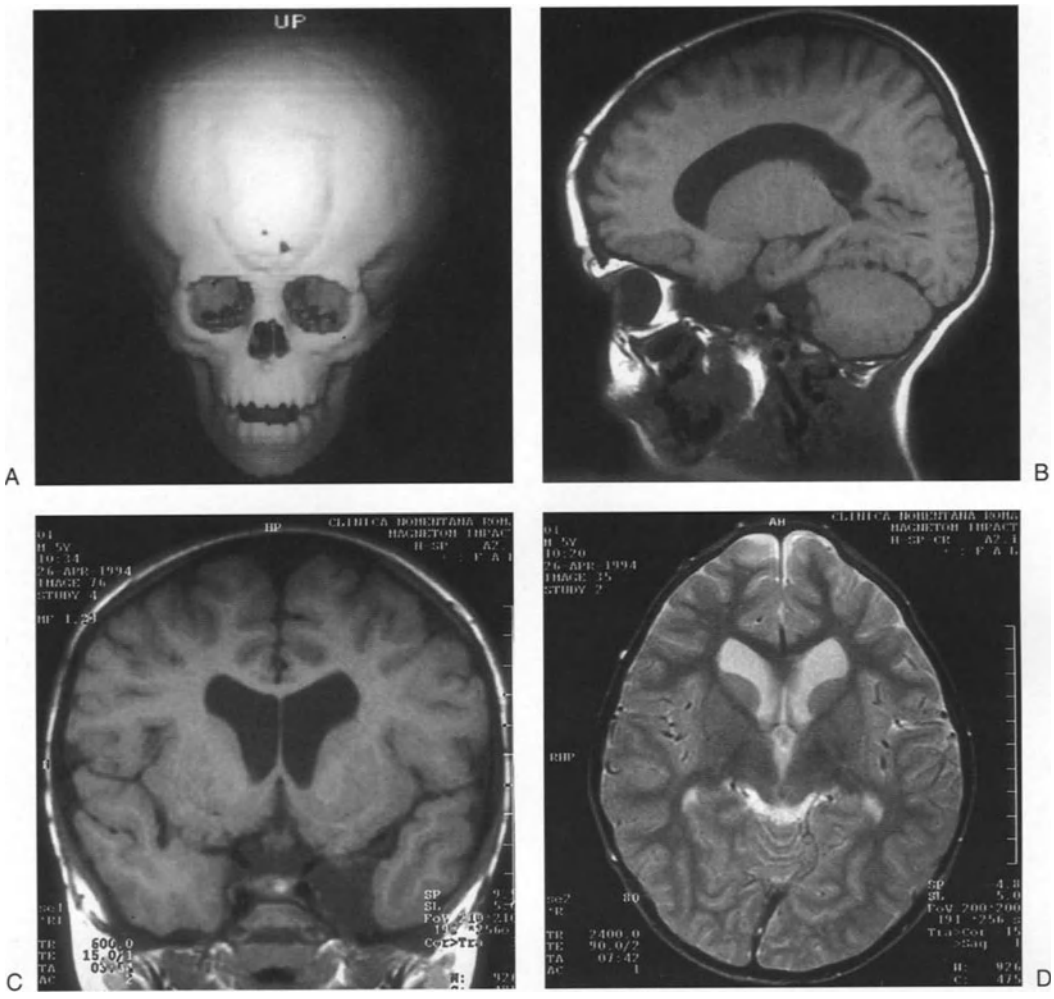


Figure 15.37. Trigonocephaly. The fact that craniosynostosis is very often the cause of hydrocephalus is now becoming recognized by most pediatric neurosurgeons working regularly with children with one form or another of this congenital anomaly. For this chapter on hydrocephalus, only a single example is illustrated: trigonencephaly. In (A) a three-dimensional CT study, one sees the remarkable keel-like deformity at the metopic suture as well as the hypotelorism. In (B) one sees dilation of the body of the lateral ventricle and accumulation of cerebrospinal fluid within the dilated subarachnoid spaces. In (C), a coronal T1 weighted image, the lateral ventricle dilation is quite obvious, as is the dilation of the subarachnoid spaces. (D) T2 weighted image which permits the reader to see clearly the cerebral deformity resulting from the trigonocephaly, the dilated subarachnoid spaces, and the ventriculomegaly.

In 1921, Dandy reported on eight infants and two adults with similar noncommunicating hydrocephalus. He classified those cases in which the foramina of Magendie and Luschka congenitally occluded as the congenital type, and those in which such occlusion secondarily resulted from meningitis as the postmeningitis type.

In 1942, Taggart and Walker [17] reported on three infants in whom hydrocephalus was assumed to result from a congenital defect in the foramina of Magendie and Luschka. Their hypothesis of etiology of hydrocephalus was interesting: if fetal development of the foramina of Magendie and Luschka is disturbed, CSF is retained in the fourth ventricle, causing it to bulge on the dorsal side and prevent the formation of vermis cerebelli, especially the posterior vermis, which extends from the cerebellar commissure to the caudal side (atresia theory).

In 1941, Sahs [18] reported a case of congenital anomaly of the cerebellar vermis that closely resembled the congenital type of hydrocephalus reported by Dandy, although the foramen of Luschka was patent.

In 1954, Benda [19] suggested, after pathological study of six hydrocephalus patients, that cystic dilatation of the fourth ventricle is not always induced by the occlusion of the foramina of Magendie and Luschka,

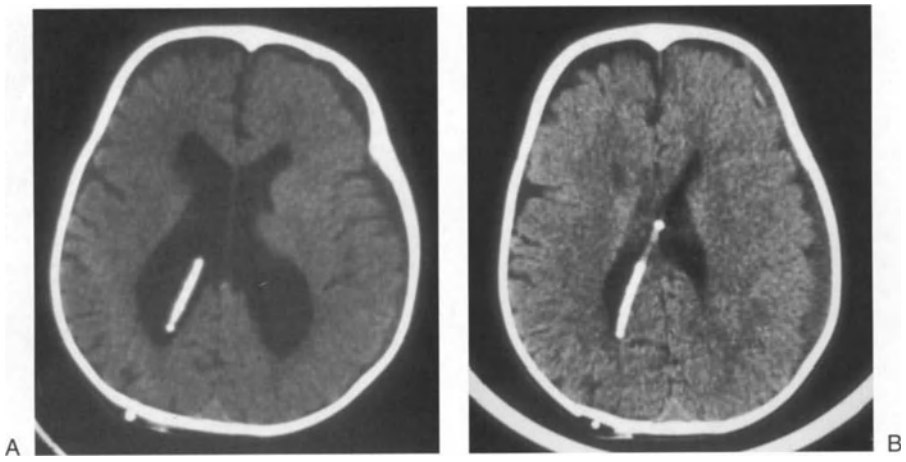


Figure 15.38. The images in this figure reveal the pre- and postoperative (ventriculoperitoneal shunt) status of the ventricles. What is of value is to study that the distribution of "brain fluid" in the preoperative state is intraparenchymal and intraventricular (A); whereas, in the postoperative state (with a functioning shunt), the subarachnoid spaces assume normal size, the brain edema has disappeared, and the ventricles have also become normal (B).

but by the congenital malformation of the cerebellar vermis itself. His theory apparently conflicted with the atresia theory proposed by Dandy and Walker. However, Benda praised their achievements and proposed naming the type of hydrocephalus associated with cerebellar vermis hypoplasia and cystic dilatation of the fourth ventricle as Dandy-Walker syndrome.

In 1954, in a report on five cases of DWS, Schreiber and Reye suggested that occlusion of the foramina of Magendie and Luschka is not always indispensable for diagnosis of DWS and its cause. In 1959, Brodal and Hauglie-Hanssen stated in a report on the pathological study of two DWS patients that the pictures of the two cases resembled the 1943 histopathological findings of Bonnevie [5] in murine hydrocephalus (Hy-1). They compared pathological findings of cystic dilatation of the fourth ventricle in Hy-1 with those in human DWS patients and assumed that this had developed before formation of the area membranacea posterior and induced dorsal bulging in the area membranacea anterior, thus arresting the development of the cerebellar vermis. They therefore concluded that occlusion of the foramina of Magendie and Luschka is not directly involved in the maldevelopment of the cerebellar vermis in DWS.

In 1956, Matson [20] outlined the plain skull film, ventriculographic, and dural sinographic characteristics of prenatal obstruction of the IV ventricle and emphasized that "intelligent and successful treatment of hydrocephalus depends upon an accurate knowledge of the location and type of obstruction to normal cerebrospinal fluid movement."

Since 1960, many other investigators have introduced new ideas on the cause of DWS. Nonetheless, it generally is agreed that the basic picture of DWS is maldevelopment of the inferior vermis associated with cystic dilatation of the fourth ventricle. However, its cause is still unknown. Because, in DWS patients, associated anomalies are found not only in the central nervous system (Table 15.1) but also in other organs (Table 15.2) and because family and hereditary backgrounds are likely to be relevant in connection with this disease, we are of the opinion that more complicated and unknown teratogenic factors are involved in DWS etiology.

Recent advances in neuroimaging, especially computed tomography (CT) and magnetic resonance imaging (MRI), have allowed us to see the anatomical architecture of the posterior cranial fossa and to obtain the pathological picture of each disease. Such investigations revealed that DWS includes a variety of similar diseases.

Table 15.1. Associated anomalies of the central nervous system in Dandy-Walker syndrome

Absence of medullary pyramid
Agyria, aqueductal stenosis
Cavum septi pellucidi
Cerebellar folial anomalies
Chiari malformation type III, cyst of choroid plexus
Diverticular cyst of third ventricle
Dysgenesis of corpus callosum
Encephalocele, fused hypothalami
Holoprosencephaly, infundibular hamartoma
Malformation of inferior olivary nuclei
Meningocele, microcephaly
Neurocutaneous melanosis
Nonspecific cerebral gyral anomalies
Polymicrogyria, specific cerebral gyral anomalies
Syringomyelia

Table 15.2. Systemic anomalies associated with Dandy-Walker syndrome

Absence of middle lobe of right lung
Bicornuate uterus, cleft palate
Coarctation of aorta
Chondrodystrophia calcificans congenita
Cornelia de Lange's syndrome
Double vagina, dysplastic and hypoplastic kidney
Ellis-van Creveld syndrome
Fetal ovaries, tracheoesophageal fistula
Fryns syndrome, hernia of accessory lobe of liver
High arched palate, hypospadia
Hypertelorism, intrahepatic biliary atresia
Klippel-Feil syndrome, laryngomalacia
Lipoma of posterior fossa, low-set ears
Macroglossia, macrogyria, macrostomia
Meckel's diverticulum, micrognathia, micro-ophthalmia
Prognathism, pulmonary stenosis
Six-lumbar vertebra, polydactylia, polycystic kidney
Rhinencephaly, sclerocornea, tuberous sclerosis
Turner's syndrome, undescended testis
Ventricular septal defect, Walker-Warburg syndrome

Table 15.3. Classification of retrocerebellar cyst

With maldevelopment of the rhomboencephalic roof
Involvement of area membranacea anterior
Dandy-Walker syndrome
Dandy-Walker variant
Involvement of area membranacea posterior
Persistent Blake's pouch
Without maldevelopment of the rhomboencephalic roof
Retrocerebellar arachnoid cyst
Cyst in the cisterna magna
Enlarged cisterna magna with communicating hydrocephalus
Mega-cisterna magna

Harwood-Nash and Fitz [21] designated a group of diseases that greatly resemble DWS; milder hypoplasia of the cerebellar vermis was the Dandy-Walker variant (DWV).

Raybaud [22] classified retrocerebellar cystic lesions roughly into true Dandy-Walker malformation, Dandy-Walker variant, and arachnoid pouches of the cisterna magna (Table 15.3). He described the first and second types as having cerebellar vermis hypoplasia, whereas the third did not. This group of diseases was therefore considered to include retrocerebellar arachnoid cysts, cysts in the cisterna magna, and persistent Blake's pouch.

Among diseases with anomaly in the rhombencephalic roof, those with apparent hypoplasia of the cerebellar vermis were placed in the category of DWS or DWV; the principle pathogenesis was malformation of the area membranacea anterior. Although the clinical concept has not been established, there probably is a disease – in which anomaly is restricted to the area membranacea posterior and in which the cerebellar vermis develops but the tela choroidea alone bulges out like a cyst – that may be called persistent Blake's pouch. On the other hand, we believe there is no anomaly in the rhombencephalic roof in patients with retrocerebellar arachnoid cysts, cysts in the cisterna magna, enlarged cisterna magna with communicating hydrocephalus, or mega-cisterna magna.

Of these patients, those with mega-cisterna magna have a basically benign clinical picture, and surgery is not indicated. Their cerebellar vermis, although small, is considered to develop undisturbed. Recently, however, Barkovich et al. [23] proposed that, since mega-cisterna magna has the same developmental anomaly as in DWS and DWV, the three diseases should be included in Dandy-Walker complex. Retrocerebellar cystic lesions, including DWS, are not systematically understood as yet and require further research.

The definition of the embryologic chronology, as well as the description of the specific pathological changes associated with this particular clinical entity, have stimulated controversy. Taggart and Walker maintained that failure in development of the foramina of Luschka and Magendie, in the 4th month of fetal life, produces hydrocephalic enlargement of the IV ventricle. This, in turn, precludes the development of the inferior vermis and prevents the normal descent of the torcular Herophili and lateral sinuses. Brodal and Hauglie-Hannsen [24] point out that the cerebellar anlagen fuse long before the fourth fetal month, and that thus the cerebellar agenesis could not be the result of later foraminal atresia. They conclude that the entire process is precipitated by a previously existing hydrocephalus of unknown cause. Benda [19] suggested that atresia of the developing IV ventricular foramina is only a part of the syndrome and not its cause, because in some cases reported the foramina were found to be patent. He stated that the main pathologic process was the "meningomyelocele" sac-like dilation in place of posterior medullary velum which results in the cleft cerebellum and the hydrocephalus. Gibson [25], through careful examination of the cystic membranes, concluded that there is no true cerebellar agenesis or aplasia and, therefore, that the whole process may well be secondary to persistent closure of the foramina of Luschka and Magendie. D'Agostino et al. [26], in a review of their pathological material, came to the same conclusion. Gardner [27] includes the Dandy-Walker cyst among many dysgeneses, all the result of persistent fetal hydrocephalus secondary to impeded flow of CSF through the attenuated roof of the IV ventricle. The foraminal atresia mentioned by others, according to him, is therefore, merely a more re-

Table 15.4. Clinical features of Dandy-Walker syndrome

	Cook County and Children's Memorial Hospitals in Chicago [28] (37 cases)	Hirsch et al. [29] (40 cases)
Enlargement of head	30	29
Enlarged posterior fossa	6	6
Headache and/or vomiting	4	6
Hypotonia	–	10
Downward-displaced eyeballs	6	6
Pyramidal syndrome and/or motor weakness	12	4
Seizures	3	6
Cranial nerve involvements	6	1
Irritability	2	–
Lethargy	2	–
Respiratory problems	3	–
Cerebellar syndrome	6	4

markable example of the impermeability of the rhombic roof.

We shall avoid this controversy and consider the basic clinical problem to be the presence of a cyst in the posterior fossa which is entirely within the confines of the IV ventricle, and which is secondary to the more or less complete occlusion of the foramina of Luschka and Magendie. We are not comfortable with "the Dandy-Walker variant" reclassifications.

Table 15.4 shows the clinical picture of 37 cases of DWS that were diagnosed and treated by us at Cook County and Children's Memorial Hospital in Chicago during the 15-year perior between 1963 and 1977 [28], and 40 DWS cases reported by Hirsch et al. [29]. Cranial enlargement is a major symptom of DWS diagnosed before the age of 12 months and is considered to be induced by the development of hydrocephalus. However, as Hirsch et al. reported, macrocephaly with occipital bulging sometimes precedes the onset of hydrocephalus. Our study of 37 cases indicated that hydrocephalus in infants less than 3 months old progressed slowly and rarely displayed intracranial pressure high enough to require emergency treatment. In fact, hydrocephalus associated with DWS has been reported to develop after birth. Normally, infants are not likely to develop serious hydrocephalus immediately after birth.

Most patients with DWS diagnosed 12 months or more after birth primarily visit hospitals with disturbance of psychomotor development or such symptoms associated with increased intracranial pressure as headaches and vomiting. The pathological site of DWS directed our attention to such signs of cerebellar dysfunction as nystagmus and ataxia, but the incidence of such signs was unexpectedly low.

Of the 37 children with DWS at Cook County and Children's Memorial Hospitals in Chicago, 89% had the complication of hydrocephalus. Only a fourth ventricle cyst was observed in the remaining 11%. However, increasing cystic dilatation in size of the fourth ventricle may cause *functional aqueductal stenosis*, which afterward induces supratentorial hydrocephalus.

Table 15.5. Associated anomalies of the central nervous system in 37 cases of Dandy-Walker syndrome

Dysgenesis of corpus callosum	14	(38%)
Aqueductal stenosis	8	(22%)
Chiari malformation type II	2	(5%)
Porencephaly	1	(3%)
Dermoid cyst	1	(3%)
None	17	(46%)

Dysgenesis of the corpus callosum was the most frequently associated central nervous system anomaly in our series (Table 15.5). Although Golden et al. [30] reported that no case with dysgenesis of the corpus callosum was found in their DWS series, it is considered that about 10%–20% of DWS cases present dysgenesis of the corpus callosum [29, 31–33].

Surgical Treatment and Prognosis (Table 15.6)

Dandy, Walker, and Matson indicated that, in surgical treatment of DWS, excision of the cyst wall and reestablishment of the CSF pathway between the fourth ventricle and the subarachnoid space were essential. However, as Raimondi et al. [15] and other investigators pointed out, reestablishment of normal CSF circulation could not, in many cases, be achieved solely by excision of the cyst wall, and additional ventriculoperitoneal (V–P) shunt operations were required for the purpose of decompressing the supratentorial hydrocephalus. Thus, in cases of DWS complicated by supratentorial hydro-

Table 15.6. Mortality rate in Dandy-Walker syndrome

	Total cases	Mortality rate
Raimondi et al. [65]	8	50
Fischer [66]	27	41
Udvarhelyi and Epstein [67]	6	0
Carmel et al. [68]	18	28
James et al. [69]	10	40
Sawaya and McLaurin	23	26
Tal et al.	12	33
Hirsch et al. [70]	40	12.5
Maria et al.	20	10

cephalus, V–P shunt operation is the first therapy selected.

Raimondi and Soare reported, on the basis of their study, that the mean full-scale IQ of DWS patients was 48.3 ± 26.1. They reported the prognosis for mental development in DWS infants to be markedly worse for those with simple hydrocephalus or Chiari malformation type II.

The outcome of treatment of patients with Dandy-Walker syndrome is to be considered generally unfavorable with regard to psychomotor development, very likely because of associated cerebral anomalies: two-thirds of the patients have an IQ below 69. The major disability before and after treatment is disturbance in gross motor function, especially difficulty in walking.

Constrictive hydrocephalus occurs in children with a craniovertebral anomaly such as basilar impression or platybasia, as well as children who have meningomyelocele associated with the Chiari II malformation. It is true that a sizable percentage of children born with meningomyelocele and who develop hydrocephalus may have aqueductal stenosis. In the Chiari II malformation, the aqueduct is invariably lengthened and passes imperceptibly into the flattened, elongated IV ventricle from a posteriorly and inferiorly displaced III ventricle. Remembering that dysplasia of the tentorium is common in this congenital malformation, and that there is also a posterior and inferior displacement of the parietal, occipital, and temporal lobes, one may imagine immediately the ease with which the inferior medial surfaces of the temporal lobes may abut upon the elongated midbrain, setting up ideal anatomopathological conditions for midbrain compression and pathological changes within it which result in occlusion of the aqueduct. Many of these children, as well as all those who develop hydrocephalus, and do not have anatomical or functional stenosis of the aqueduct, suffer constriction of both the brainstem and the inferior half of the cerebellum within the foramen magnum. This constrictive process obliterates the cisterna magna, the medullary and pontine cisterns, and the ambient cistern. The passage of CSF out of the IV ventricle is either impossible or severely impaired despite the patent foramina of Luschka and Magendie. (These children do not develop a dilated IV ventricle or aqueduct.) Actually, the posterior half of the III ventricle is relatively normal in size, and the anterior portion of the III ventricle is blown out as are the occipital horns of the lateral ventricles. The quadrigeminal and superior cerebellar cisterns may be enormous. The cerebellar tonsils and IV ventricle are displaced across the foramen magnum, which is remarkably dilated, and the cisterna magna is either nonexistent or only a potential cavity.

Intraventricular tumors such as the choroid plexus papilloma of the lateral ventricle are capable of producing increased amounts of CSF, causing communicating hydrocephalus of the hypersecretory variety. These children show a clinical picture of both the space-occupying lesion and hydrocephalus.

Milhorat et al. [34] have shown that the rate of cerebrospinal fluid formation in a 5-year-old child who had undergone bilateral choroid plexectomy for communicating hydrocephalus during infancy was 0.35 ml/min; standard deviation ± 0.02 ml/min: well within normal limits. This failure of choroid plexectomy as a treatment for hydrocephalus had been extensively documented in the world literature previously. Consequently, the cerebrospinal fluid which continues to be secreted at a normal rate is also produced in extrachoroidal sites. Therefore, the hypersecretory hydrocephalus which results from a papillary tumor of the choroid plexus may not necessarily be cured when the papilloma is resected-... probably because of irreversible changes at the sites of CSF absorption, changes which result from the extraordinarily high levels of protein secreted by the papillary tumor. Other intraventricular tumors such as subependymal astrocytoma, tubers in association with tuberous sclerosis, ependymoma, and medulloblastoma may be associated with permanent hydrocephalus ... after the tumor (whether obstructive or not) has been totally removed.

Consequently, after a diagnosis of choroid plexus papilloma of the lateral ventricle has been made in either a newborn or an infant and the tumor removed, a medium to high pressure shunt may need to be inserted so as to render the postoperative course smooth and to diminish the possibility of progressive ventricular enlargement.

Infratentorial tumors often cause hydrocephalus. Although the neoplasm itself causes symptoms and signs, the complicating secondary hydrocephalus is often responsible for the increased intracranial pressure (ICP), thus superimposing the clinical picture of "midline syndrome" (an increase in ICP without lateralizing signs) upon those signs resulting from the destructive and compressive effects of the tumor. Accordingly, the child with hydrocephalus complicating a primary brain

tumor (especially of the posterior fossa) may be considered to have two distinctly different diseases which complicate one another and contribute to the complex clinical picture of increasing ICP: (1) a neoplasm and (2) hydrocephalus. It is not always possible to ascertain with precision the specific nature of the hydrocephalus, or to predict in which patients the hydrocephalus will remain as a permanent condition even after the associated tumor, benign or malignant, is totally removed. Thus, the hydrocephalus may be temporary or permanent, obstructive, constrictive, communicating, or hypersecretory.

Intraspinal tumors as a cause of increased intracranial pressure and papilledema are now well documented. Hydrocephalus in children with spinal cord tumors may not be attributed to any single etiologic or pathogenetic factor, since there is a wide variation in location (spinal level and intramedullary or extramedullary) and nature (benign or malignant) of the tumors. One may safely exclude spinal cord block, leptomeningeal infiltration by tumor cells, and significant elevation in CSF protein level as single and definitive causative factors.

Arteriovenous fistulae may cause hydrocephalus. These may be transcranial, dural, mixed-dural-pial, or purely pial. The origin of blood supply depends, quite naturally, upon the type of arteriovenous malformation, but is of no significance regarding the genesis of hydrocephalus. Increased venous pressure, with retrograde flow of arterial blood through major draining veins and sinuses, and into the capillary bed, produces remarkable increases in venous pressure and results in communicating hydrocephalus. Very few documented cases of obstructive hydrocephalus resulting from arteriovenous fistulae have been reported. These have all been characterized by enormous intradural pools of blood, occupying the tentorium, falx cerebri, or galenic system. The same etiology – increased venous pressure at the torcular Herophili and the resultant diminished absorption of cerebrospinal fluid – has been attributed to the hydrocephalus resulting from superior vena cava thrombosis or occlusion of the transverse sinuses. Curiously enough, occlusion of the superior sagittal sinus has only rarely (one case in this book) been reported to cause communicating hydrocephalus.

Genesis of Parenchymal Destruction

Irrespective of the etiology, pathogenesis, or type of hydrocephalus, micro- and macrovascular changes are a direct cause of parenchymal destruction in infantile hydrocephalus. Ventricular enlargement causes displacement of primary cerebral arteries, followed by stretching and a decrease in the caliber of primary, secondary, and tertiary vessels (arterial and venous). Ultimately, there is a reduction in the number and caliber of the microvasculature, resulting in diminished cerebral blood flow and cerebral edema. Tissue destruction leading to ependymal rupture, parenchymal cavitation, and the formation of porencephalic cysts within the edematous parenchyma ensues. The ventricular enlargement occurs at the expense of the surrounding tissue, most notably the white matter, which becomes markedly thin. The most common end-stage changes found in the parenchyma of the hydrocephalic brain are atrophy, pallor and swelling, vacuolation and chromotolysis of nerve cells, hypertrophy of astrocytes, axonal demyelinization and degeneration, and a decrease in synapses. It had been suggested that these changes are the result of an increase in intracranial pressure which induces a diminution in cerebral blood flow in the hydrocephalic brain, in either hypertensive or normal pressure hydrocephalus.

Early observations of the brain's vasculature in human hydrocephalus by Penfield and Elvidge, in 1932 [35], were that "there may be a decrease in the intramedullary capillary bed. This decrease is only a supposition, not a proven fact, but it seems likely that vascular obliteration begins in the smallest branches of the vascular tree and that further passage of blood through these branches is prevented by their compression without thrombosis and congestion as in other forms of vascular occlusion." Hassler [36] and De [37], independently, from studies of experimental hydrocephalus, found a significant loss of smaller vessels (capillary and precapillary) around dilated ventricles, and concluded that ischemia is responsible for the associated changes in all structures along the ventricular surface.

In the hydrocephalic process it is possible to postulate a pathogenetic sequence of events which leads to irreversible brain damage.

Transependymal CSF perfusion, a compensatory mechanism which certainly occurs also in the normal state, is a response to high intraventricular hydrostatic or pulse pressure and relative ischemia of the white matter. The intraventricular cerebrospinal fluid and extracellular fluid of the cerebral parenchyma act as one liquid medium, with bulk flow proceeding freely in both directions across the ependymal barrier. The increased head of pressure both compresses and stretches the cerebral vasculature, displacing and deforming it, causing the caliber to diminish, and resulting in changes in CBF.

The *first vascular changes in the development and progression of hydrocephalus* consist of a decrease in the caliber of cortical and white matter cerebral vessels, not of the perforating branches going to the brainstem or basal ganglia. There is subsequently a decrease in the number of the secondary and tertiary vessels, which begins in the white matter but rapidly involves the vasculature of the cortex. Then, the normal "palisade" pat-

tern disappears, the edema and atrophy of the periventricular white matter following as ischemia occurs.

The end result is the following sequence of events, graded as stages, leading to irreversible brain damage: (1) ventricular dilation and intraventricular CSF accumulation under increased hydrostatic or pulse pressure occurs after the initial subarachnoid space dilation; (2) increased transependymal flow is responsible for the periventricular lucency and increased extracellular fluid, cerebral edema, and then obliteration of the subarachnoid spaces; (3) parenchymal vascular compression and ischemia, with progression of the cerebral edema from the white to the gray matter; and (4) cellular disruption and tissue destruction, aqueductal stenosis then occlusion, followed by further ventriculomegaly and transependymal flow. At times, because of the white matter ischemia and periventricular edema, porencephalic cavities extend from the ventricular system.

The onset of irreversible brain damage becomes obvious with the identification of these porencephalic cavities and the disappearance of the tertiary branches of the anterior and middle cerebral arteries. Consequently, one may conclude that decompression of the intraventricular pressure-head, by a shunting device after the second stage of hydrocephalus (when parenchymal vascular compression results in irreversible changes in cerebral blood flow), offers no hope of recovery of cerebral function. On the other hand, if one intervenes at the beginning of the second stage of hydrocephalus, when early edema of the white matter and enlargement of the extracellular spaces have only begun, and there is no more than a minimal decrease in the caliber of cerebral vessels with displacement of the primary and secondary vessels, drainage of CSF has a beneficent effect.

Diagnosis

As early as 1918, Walter Dandy [38] recognized the advantages of a water-soluble or water-missible medium for ventriculography, but subsequently abandoned the procedure because the substances he used were either too toxic or cleared the ventricular system too slowly. He concluded his studies by discovering air ventriculography. During the 1920s Thorotrast enjoyed a brief reign of ventriculographic popularity, only to be abandoned when it was found to cause serious late sequelae. In recent years, stereotaxic surgery put into relief the need for adequate III ventricular visualization without laborious repositioning of the patient's head. In 1962, various investigators found Conray 60 to have little toxic effect and to produce excellent roentgenographic contrast in animals. In the period 1962–1964, Heimberger et al. studied the dosage, relative toxicity, and radiopacity of intraventricular injections of Conray 60 in animals and, subsequently, in over 100 patients [39]. They found that the toxicity of the substance, when confined to the intraventricular ependymal surfaces, was not significant, and denoted the rapid diffusion and the clearing of this substance as it passed through the ventricular system. Subsequent to this, Conray 60 was used to perform serial ventriculography in hydrocephalic infants, so as to outline the criteria for diagnosing communicating hydrocephalus, aqueductal stenosis, the Chiari II malformation, and such ventricular and arachnoidal cysts as the Dandy-Walker malformation, III ventricle cysts, and arachnoidal cysts of the cisterna magna.

The use of small amounts of Pantopaque to identify specific sites of obstruction (foramen of Monro, aqueduct of Sylvius, outlet foramen of the IV ventricle) enjoyed temporary popularity. However, use of even small amounts of this substance required the same manipulation as the use of gas or dyes, and provided no more information. Since these complicated procedures were not always performed, shunting procedures had been carried out on infants with hydrocephalus as soon as the generic diagnosis was made.

In 1951, Tolosa [40] reported preliminary angiographic studies on hydrocephalic infants and in 1959 Fauré and Gruson [41] described the arteriographic characteristics of such specific tumors as the craniopharyngioma. Paillas et al. [42] studied the diagnosis and localization of vascular anomalies and suspected vascular tumors such as the choroid plexus papilloma with cerebral angiography. However, with few exceptions, the work published on angiography in childhood concerned itself with the juvenile and adolescent age groups. Subsequent to this, angiographic studies in the newborn and infant were undertaken to diminish the limited state of knowledge concerning normal variations, and the changes in arterial and venous angiographic characteristics which vary in accordance with birth weight and with fontanelle and suture closure. Studies on technique, normal vascular anatomy, and arterial and venous changes in congenital anomalies of the craniocerebrum, as well as in craniocerebral traumatology of the newborn, served as a basis for the systematic analysis of the angiographic characteristics of hydrocephalus. It thus became possible to make an angiographic diagnosis of hydrocephalus, to determine whether the hydrocephalus was external or internal, if internal to learn whether it was of the communicating or obstructing variety, if obstructive to learn the nature and exact point of occlusion, and to evaluate, most important of all, the status of the cortical and brainstem vascularization as an index of the reversibility of the hydrocephalic process [3].

Computed tomography (CT) simplified dramatically and added immeasurably to the means of diagnosing hydrocephalus in general, and of evaluating with precision the individual stages of the development of the hy-

drocencephalic process. Specifically, one became able to identify hydrocephalus, to distinguish between a communicating and obstructive variety, to learn that dilation of the subarachnoid spaces precedes the appearance of periventricular edema and that this latter precedes ventriculomegaly. Of more importance still, one was able to distinguish reversible from irreversible hydrocephalus and to outline the individual stages of the irreversible form, something which permits proper timing of the shunting procedure. Advances in computer technology have rendered the use of positive contrast material superfluous, whether ventricular or vascular.

Since 1983, however, MRI has been perfected to such a degree as to provide an anatomically perfect graphic representation of the entirety of the cerebral parenchyma and ventricular system as to permit absolute diagnosis of hydrocephalus and identification of the individual classification categories.

Treatment

Any form of treatment for hydrocephalus, whether internal or external, whether primary or secondary, must be predicated on one of three basic concepts: (1) diminishing the amount of cerebrospinal fluid being formed; (2) diverting the cerebrospinal fluid outside of the ventricular system and subarachnoid spaces so that it may be absorbed elsewhere in the body; and (3) increasing the amount of cerebrospinal fluid being absorbed.

Bypassing the point of obstruction does not provide an acceptable alternative, since, with the rarest of exceptions, obstructive processes (occlusion of the foramen of Monro, aqueductal stenosis, occlusion of the foramina of Luschka and Magendie) *are secondary!* Those authors who have reported successes from III ventriculostomy for aqueductal stenosis, suboccipital craniotomy and resection of arachnoid from the region of the foramina of Luschka and Magendie, cannalization of a stenotic or atretic aqueduct of Sylvius, have all performed these procedures on children who enjoyed prolonged periods of functioning ventriculojugular or ventriculoperitoneal shunts. The scientific literature still awaits an objective documentation that intracranial bypassing of the site of obstruction *in non-neoplastic hydrocephalus* is curative.

The use of hypertonic agents, carbonic anhydrase inhibitors, steroids, etc., has not been successful in diminishing the amount of cerebrospinal fluid formed. No drug has been used to increase the amount of cerebrospinal fluid absorbed.

The treatment today, as when the first shunting procedure was performed in the late nineteenth century, consists of diverting cerebrospinal fluid outside of the central nervous system to another portion within the body from whence it may be absorbed. A listing of these procedures would serve no purpose, since, in fact, every absorptive surface within the human body has been used: middle ear, the oral cavity, the venous system, the ileum, the fallopian tubes, the ureters, the peritoneum, etc., etc. Similarly, a discussion of the value and significance of valvular systems may be reduced to the basic question: "Is it possible to regulate the flow from the ventricular system with a valvular device?" The answers are as variable as the devices used.

Of significance, on the other hand, are the facts that we have been extraordinarily successful in treating (compensating!) primary and secondary hydrocephalus, and that we have managed to identify most of the complications of these operative procedures. The reported successes in our treatment range from 70% to 85% of operated hydrocephalic newborn and infants. The operative mortality has been reduced to less than 0.5% per procedure performed. The complications, ranging from transient ileus to intracerebral hematoma, average approximately 35% per operation performed.

Concerning shunting procedures, Davidoff [43] sites Hippocrates as the first physician to attempt to control hydrocephalus by decompressing the ventricles. In 1886, Zrenner implanted the first external ventricular drainage system and, in 1893, Mikulicz inserted the first internal shunt. Using gold, glass, or platinum tubes, Sutherland and Cheyne [44] developed techniques to connect the ventricles with either the subarachnoid or subdural spaces. Ballance compared the merits of gold and platinum to glass. Cerebrospinal fluid was first shunted from the lumbar subarachnoid space to the peritoneal cavity by Ferguson in 1898 [45]; from the lateral ventricle, using a small caliber rubber catheter into the peritoneal cavity by Kausch in 1908 [46]; and into the circulatory system by Payr in 1908 [47]. Haynes, in 1913, drained CSF from the cisterna magna into the cranial sinuses [48].

Following these pioneering innovations, enthusiasm for shunting procedures waned. Not until mid century did the favorable experiences of Ingraham et al. with plastic (polyethylene) shunt tubing rekindle interest [49]. Subsequently, a number of different shunting techniques were attempted. Following the initial report of the use of a silastic valve-regulated system by Nulsen and Spitz in 1952 [50], neurosurgeons began to accumulate experience with ventriculovenous and ventriculoperitoneal shunts. The multisystem complications of the ventriculovenous shunt caused many surgeons to prefer the ventriculoperitoneal shunt. Some comparative observations were reported. Matson [51] stressed the indications for use of the ureter, Ransohoff [52] for the pleural cavity, and Harsh [53] for the fallopian tube for entrance into the peritoneal cavity. These latter three authors were searching for a better absorptive site, one less fraught with complications from the blood stream and more reliable than direct insertion of an open-ended tube into the peritoneal cavity where the prob-

lems encountered were complications and difficulties of surgical opening, kinking of the subcutaneous or elastic tubing along its course, or plugging of its opened distal end by insinuating omentum.

Two of the complications of peritoneal shunting, extrusion through skin and organ perforation, had already been identified as complications of jugulocardiac shunting. The third, ascites or accumulation of CSF in cystic pockets within the abdominal cavity, may be broadly compared to the pericardial tamponade. Volvulus in ventriculoperitoneal shunting was reported by Sakoda et al. [54] and inflammatory pseudotumor of the mesentery by Keen and Weitzner [55]. Other abdominal complications consist of peritonitis, bowel obstruction, perforation of an arterial or venous structure, shunt tubing lost in the abdomen, abdominal cyst formation, inability of the peritoneal cavity to absorb CSF, gastrointestinal bleeding, low pressure syndrome, and intestinal perforation. The intracranial complications consist of shunt tubing lost in the ventricle, subdural hematoma, unsuccessful ventricular cannalization, intraventricular or intraparenchymal hemorrhage, brain abscess, and hemiparesis. The general complications consist of stitch infection, wound breakdown, electrolytic imbalance, and apnea.

It soon became apparent that the number of complications per patient were related to the number of revisions per patient, and that the number of revisions was directly, though not entirely, related to the incidence of shunt infection and disconnection. The causative organisms in most of the infections are the common flora of the skin – *Staphylococcus epidermides* and *S. aureus*. The only exceptions occur in meningomyelocele children, who tend to develop gram-negative ventriculitis. The incidence of shunt infection is much higher in those newborns, especially with meningomyelocele, who are shunted during the first weeks of life... so that it is advisable to postpone surgery to the 4th week of age if possible.

Intellectual Development and Quality of Survival

Many studies have been published concerning the intelligence of hydrocephalic children. These have included work evaluating shunted hydrocephalics and those evaluating nonoperatively treated hydrocephalics.

It is impossible to evaluate the literature regarding intellectual function in children with treated and untreated congenital hydrocephalus unless one knows the exact criteria on which the diagnosis of this disease was based. Progressive increase of head size, bulging fontanelle, split sutures, transillumination, positive Macewen sign, "setting sun" phenomenon, shiny scalp, and distended scalp veins, are all indicative of an increase in intracranial pressure, but are not diagnostic of congenital hydrocephalus. Subdural hematoma, intraventricular tumor, parasellar tumor, posterior fossa tumor, and toxoplasmosis are just a few of the diseases in the newborn that commonly cause similar signs.

In reviewing the literature, one finds that it is difficult to compare one study with another. Some of the difficulties include: (1) the lack of information regarding the shunt and the child who is shunted, (2) the use of different instruments to measure intelligence quotients, (3) differing methods of data presentation and definition of "normal," (4) inclusion of varying causes, and (5) sample bias.

The most difficult information to obtain has been that regarding the maintenance of a functioning shunt system, the age at which the shunt was first inserted, and specific information regarding the "intactness" of the compressed brain (e.g., presence of cerebral dysplasia, porencephaly).

Not all papers include the same hydrocephalic groups and their analyses. Some studies include patients with meningomyelocele but analyze the results separately, some include meningomyeloceles but do not analyze the results separately, some exclude any diagnosis but primary congenital hydrocephalus, and some exclude only meningomyelocele.

Few of these papers state whether the hydrocephalus may have been secondary to intrauterine, perinatal, or postnatal meningitis. Also, information is wanting as regards the possibility that the shunted hydrocephalic child may have suffered ventriculitis or meningitis either as an immediate or a delayed complication of the shunt. The effects of associated brain anomalies, such as agenesis of the corpus callosum, porencephaly, hydranencephaly, agenesis of the cerebellum, or temporal lobe dysplasia, on intellectual function have not been subjected to critical analysis; nor has mention been made of their exclusion from statistical cohorts.

The average IQ of patients born with congenital porencephaly is 37.2, or just barely within the range of trainable mentally handicapped. On the basis of this information, one may conclude that the presence of porencephaly prior to the first shunting procedure is adequate evidence that the child will certainly be retarded. Consequently, the rationale for shunting, namely, to provide the child with an opportunity to develop normal intellectual functions, is absent.

There seems to be no reason to believe that the newborn child with large ventricles and a thin cerebral mantle prior to initial shunt placement will ultimately be less intelligent than a child with relatively mild hydrocephalus, other factors being equal. The literature argues most persuasively in favor of surgical treatment for hydrocephalic children with even the thinnest mantle and the largest ventricles. Although no relationship has been found between IQ and severity of hydrocepha-

lus prior to initial surgery, it has been shown that the patient with meningomyelocele has, in general, less severe hydrocephalus prior to surgery than did those with internal hydrocephalus. These results may be explained by the fact that the children with meningomyelocele are watched very closely from birth to see whether hydrocephalus develops. Thus, the hydrocephalus is probably operated on before it progresses to severe stages.

Results regarding age at the time of the first shunt indicate that the earlier the shunt is placed the better the chance for normal development, but only with regard to children with internal hydrocephalus. This does not seem to apply to the group with meningomyelocele and hydrocephalus. It may be that meningomyelocele children who were shunted late did not develop hydrocephalus until late and shunted immediately upon diagnosis, whereas patients with internal hydrocephalus who were shunted late had the developing hydrocephalus over a prolonged period of time. This explanation is in accord with the observation that children with meningomyelocele have less severe hydrocephalus prior to initial shunting. Both observations indicate that the child with meningomyelocele is followed-up carefully over a period of time, and that the hydrocephalus is treated promptly if it occurs.

Certain variables definitely affect the patients' intellectual development while others do so to a lesser degree. Those variables which seem to have a large impact are shunt function, race, socioeconomic level, and age at first shunt. The less important variables are number of revisions and degree of hydrocephalus at initial shunt placement.

Surgical Management

The rational surgical management of hydrocephalus whether congenital, postmeningitic (bacterial, hemorrhagic), or secondary to neoplasia, rests entirely upon identification of whether the fluid accumulation is intraparenchymal (the cerebral edema of pseudotumor cerebri) or intraventricular. In the former, a lumbar peritoneal shunt often suffices as a compensatory procedure. In the latter, one must determine which ventricles are dilated: all four, the lateral and III, both lateral, or one lateral. Thus, one may speak of tetra-, tri-, bi-, or monoventricular hydrocephalus.

Tetraventricular hydrocephalus may be shunted either from the lumbar subarachnoid space or one of the two lateral ventricles, providing the child does not have a *Dandy-Walker cyst* (atresia of the foramina of Luschka and Magendie with cystic transformation of the IV ventricle). *In this latter event, one may attempt to shunt initially from either lateral and/or the IV ventricle(s). Within a relatively brief period of time, it almost invariably becomes necessary to insert two independent shunts*, one from the lateral ventricle and one from the IV ventricle. Triventricular hydrocephalus may be shunted from either lateral ventricle, biventricular hydrocephalus necessitates bilateral ventriculoperitoneal shunts, and monoventricular hydrocephalus necessitates a single shunt from the single dilated lateral ventricle.

Interventricular shunts (cannulation of the aqueduct of Sylvius, permitting communication between the III and IV ventricles), and *ventriculocisternal shunts* (III ventriculostomy ventriculocisterna magna), though not considered effective by a majority of pediatric neurosurgeons, have been used by some. The application of new endoscopic techniques has kindled interest in returning to the III ventriculostomy, but as of 1998 no one has reported a significant number of cases with a well-studied follow-up.

Choroid plexectomy is of no value in the treatment of hydrocephalus, though lateral ventricle choroid plexectomy is necessary (using the technique already described) when performing hemispherectomy.

Since there are so many different types of shunts used, and so many different connectors, reservoirs, flushing devices, and so on, there is no constant technique recommended for joining the proximal (ventricular) and distal (venous) portions of a ventriculoatrial shunt. Implantable device manufacturers have taken great care to describe their individual shunt elements and, in many cases, the technique for inserting them. United States government regulations concerning the production and use of these have resulted in relatively complete listings of precautions and possible complications. The descriptive circular for each system should be studied repeatedly, until one is familiar with limitations and use of the shunting system, its individual elements, and insertion and instrumentation.

In its broadest terms, *shunting may be subdivided into three operative procedures*:

1. Deviation of the cerebrospinal fluid out of the central nervous system (into either another body cavity or an external reservoir)
2. Interventricular shunting
3. Ventriculocisternal shunting

In all instances the cerebrospinal fluid must be diverted (*shunted*) from either the ventricular system or the lumbar subarachnoid spaces. Presently, the ventricular shunts used, depending upon the surgeon's preferences and the child's condition, are ventriculoatrial, ventriculoperitoneal, ventriculopleural, and ventriculogallbladder. The ventriculoureteral shunt is no longer used because of the prohibitively high incidence of electrolyte imbalance (from loss of cerebrospinal fluid through the urinary tract), and ascending infections. The same contraindications apply to shunting into the fallopian tube.

The indications and technique for external ventricular drainage have been described in Chap. 12, "Infections."

Techniques for Cannulation of the Ventricles

The lateral ventricles may be cannulated through either the occipital or frontal horns, the IV ventricle through the cisterna magna or cerebellar hemisphere.

Occipital Horn Cannulation (Figs. 15.39–15.41)

The location of the occipital horn should be plotted out on a CT or an MRI scan, through coronal and axial sections, measuring the distance from occipitoparietal cortex to the dilated occipital horn, and then identifying the coordinates for puncture of the occipital horn and threading the catheter into the body of the lateral ventricle. Points of reference should be the roof of the orbit, the transverse sinus, the superior sagittal sinus, and the base of the mastoid. (One should not use only external indicators, such as the parietal eminence, the midline, the external occipital protrusion, and the nasion.) Once the proper coordinates have been determined, the child's head should be positioned so that the surgeon may superimpose visually the predetermined coordinates onto the head (which has been placed in a plane orthogonal to the trunk). Settling for an obliquely positioned head may prove to be dangerous.

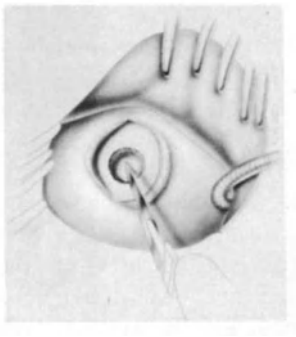


Figure 15.40. Making a cruciate incision in the dura, it is advisable to ascertain that the incision is large enough to permit the trocar free passage, for if it is small the surgeon may strip the dura from the skull as he inserts the trocar. This may result in a postoperative epidural hematoma.

Either a semilunar or, preferably, horseshoe-shaped incision is then made, taking care that it is large enough to permit placement of the anchoring clip (or reservoir) so that these will be totally covered by skin, with no portion of the incision passing immediately over the foreign body. Either a twist drill or a 10-mm bur hole opening is then made, the dura coagulated in a cruciate manner, and incised along the lines of coagulation. Any underlying vessel on the cortical surface should be coagulated and then the arachnoid cut with a #15 blade. The cannula, through which the proximal end of the shunting system will be passed, is then inserted along

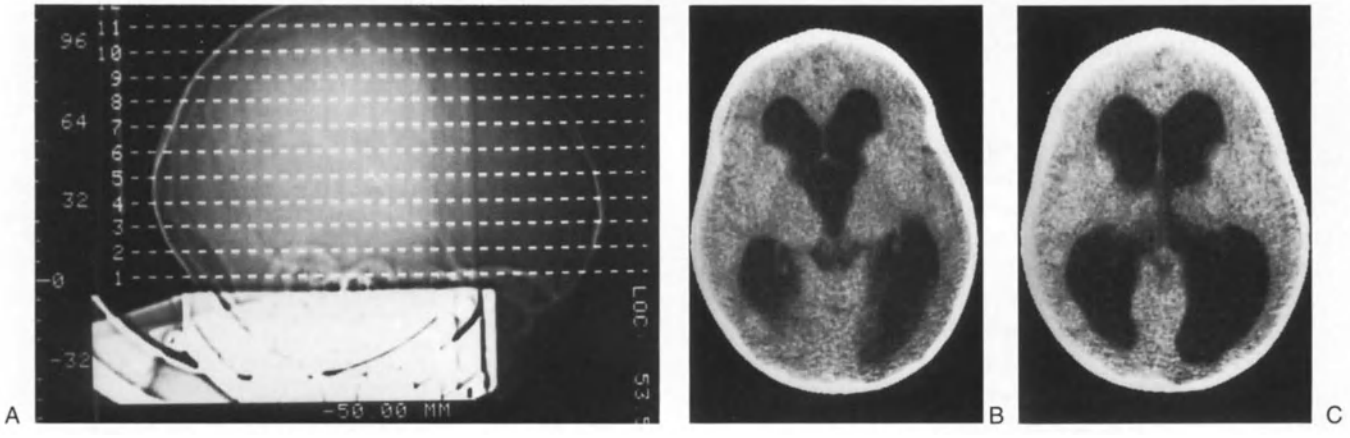


Figure 15.39. The insertion of a cannula into the lateral ventricle should be planned with the use of an MRI or a CT scan, in order to plot out coordinates for the line of insertion, rather than relying upon external landmarks such as the parietal eminence, supraorbital rim, external auditory canal, sagittal suture, etc. (A) A lateral projection, scout scan, with horizontal planes (1-cm intervals) plotted out, permit one to identify a known landmark or to cross-reference with a single CT cut in order to select the most appropriate line of insertion of the cannula into the occipital horn and body of the lateral ventricle. (B) The 4-cm line reveals the left occipital horn to extend to within approximately 2 cm of the occipital cortex surface, at a distance of 2 cm from the sagittal plane. (C) The 5-cm cut reveals both occipital horns to be well filled and the precise location of the right and left occipital horns and trigones. One may conclude that the ideal insertion would be at the 5-cm line, since the cannula may be directed immediately into the occipital horn, preferably the right, and then threaded for a distance of about 5 cm within the ventricle from occipital horn to trigone.

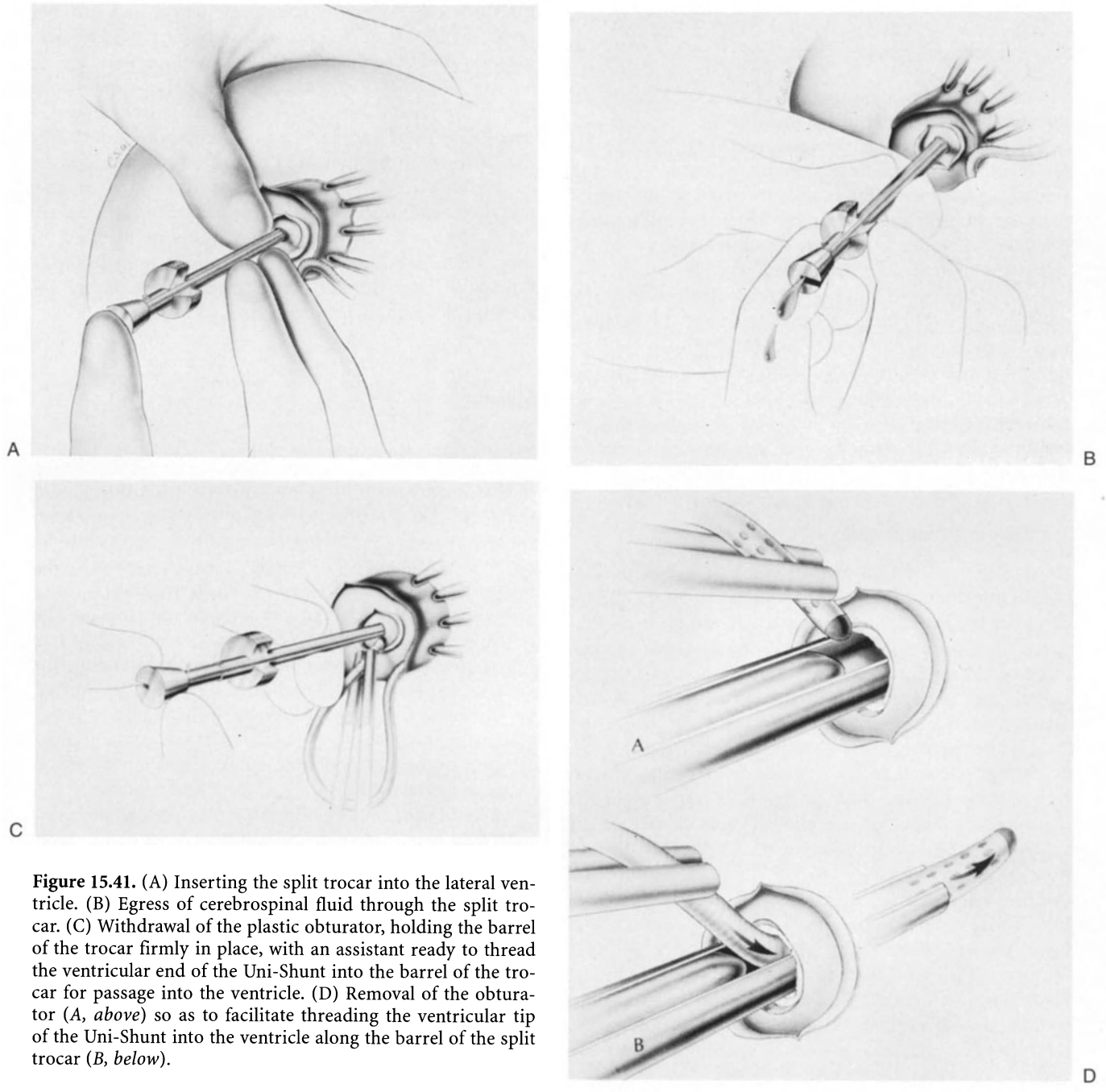


Figure 15.41. (A) Inserting the split trocar into the lateral ventricle. (B) Egress of cerebrospinal fluid through the split trocar. (C) Withdrawal of the plastic obturator, holding the barrel of the trocar firmly in place, with an assistant ready to thread the ventricular end of the Uni-Shunt into the barrel of the trocar for passage into the ventricle. (D) Removal of the obturator (*A, above*) so as to facilitate threading the ventricular tip of the Uni-Shunt into the ventricle along the barrel of the split trocar (*B, below*).

the planned trajectory. One does not often feel a distinct "pop" as the ependymal barrier is breached and the cannula enters the ventricle, so that it is best to extend the insertion no farther than 1 cm beyond the measured distance. When one sees flow of cerebrospinal fluid through the distal end of the cannula, the obturator is removed and the proximal end of the shunting system inserted through the barrel of the cannula into the ventricle. The cannula is removed and the distal end of the shunt tubing inspected to document the flow of cerebrospinal fluid. Air pockets may obstruct the flow of fluid, so that it is often necessary to irrigate with 2–3 ml saline. When free flow is observed, the proximal end of the tubing is inserted to the previously measured distance, always observing flow from the distal end so as to be certain the proximal tip is neither kinked nor penetrating cerebral tissue.

One attempts to position the tip of the proximal end of the shunting system at the foramen of Monro, theoretically to minimize insinuation of choroid plexus into

the perforations, but this is seldom possible: the ventricles diminish in size as the hydrocephalus is compensated, so that one's parameters at the time of surgery should be predicated upon the projected ventricular size and contour 6 months later.

The multiperforated, flange-tipped ventricular catheter is subject to greater adhesions of choroid plexus, rendering it potentially more prone to complications than the smooth ventricular tip which has perforation openings. The use of a rigid obturator, placed in the shunt lumen, to permit insertion of the proximal end of the shunting system into the ventricle has two very real disadvantages: (1) brain tissue may plug the perforations at the tip of the shunt, and (2) one may inadvertently insert the shunt into the thalamus, basal ganglia, and opposite hemisphere. The use of the cannula diminishes considerably the incidence of brain tissue insinuating into the catheter perforations and threading the catheter into the basal ganglia or thalamus.

Frontal Horn Cannulation

Coronal reconstruct CT or MRI scans should be used to obtain information concerning volume, contour, and location of the frontal horn, prior to planning the coordinates. The same type of skin incision and size of skull opening should be used as when one cannulates the occipital horn. Also, the dural opening and cortical incision are performed in the same way. The trajectory should be determined by a study of the CT scans, horizontal and coronal reconstruct, and the cannula (with its obturator) inserted no deeper than 5 mm from the measured distance between the superior surface of the ventricle and the inner table of the skull! This precaution minimizes risks of penetrating the thalamus and caudate nucleus. After there is egress of cerebrospinal fluid and the shunt tubing has been placed in the lateral ventricle, one should not advance the catheter more than an additional 5 mm, so as to avoid entering the III ventricle, which is undesirable only in so much as choroid plexus may engulf it, increasing the risk of tearing the internal cerebral vein when removing the shunting system electively or for revision.

Fourth Ventricle Cannulation

The IV ventricle should be cannulated with the proximal end of the shunting system only when the child suffers cystic transformation of the IV ventricle (the Dandy-Walker cyst). This congenital, or postinfectious, clinical entity results in such a remarkable dilation of the IV ventricle that the dura mater, leptomeninges, and dysplastic ependyma abut upon one another, rendering insertion into the IV ventricle easy. It is advisable to make the opening in the squamous portion of the occipital bone approximately 1.5 cm lateral to the midline, well below the transverse sinus, and just beneath the lowest nuchal line. This assures avoiding perforation of a venous sinus. It also provides an adequate amount of soft tissue to cover the shunting system, and either the anchoring clip or a reservoir (whichever is used). The bone and dura are opened in the same manner, with cerebrospinal fluid often pouring out as soon as the dura is cut: the arachnoid of the cisterna magna and the ependyma of the IV ventricle are densely adherent to the dura. The catheter tip should be inserted no more than 3 cm, making sure that cerebrospinal fluid is flowing through the shunting system throughout the insertion. No cannula is necessary.

Shunts

Ventriculoperitoneal Shunt (Figs. 15.42–15.60)

It is very important to remember that when general anesthesia is being used, one must allow the patient to become very light prior to perforating the peritoneal cavity (with the peritoneal split trocar) to avoid damage to the intra-abdominal viscera and vessels! If the anesthesiologist allows the patient to become very light, then the peritoneum will tighten, like a drum, when the split trocar is pressed firmly against it, thereby allowing the surgeon to perforate the peritoneum cleanly. *The Valsalva maneuver is not an alternative.* If the patient is deeply anesthetized, the peritoneum will be relaxed and penetration becomes impossible, increasing the risk of damage to the intra-abdominal viscera and vessels.

Surgical opening of the peritoneum adds to the morbidity by prolonging the postoperative recovery time, and increasing the risk of entering a viscus. It is also likely that the surgical opening of the peritoneal cavity predisposes the child to adhesions between the visceral and parietal peritoneum, increasing the probabilities of the shunt (a foreign body) eroding into the intestinal tract. There have been reported cases of passage of the shunt system through the gastrointestinal tract. These have all been in patients in whom the peritoneum was opened surgically, in children operated on under general anesthesia without allowing them to lighten, and in meningomyelocele children with varying degrees of abdominal flaccidity. The shunt, as all foreign bodies, may penetrate another structure or extrude through the skin at any point along its path.

In Fig. 15.43 the child is shown in the correct position for a ventriculoperitoneal shunt. Padding is placed under the thoracolumbar vertebrae, the shoulder, and the neck. The head is turned fully to the contralateral side and the neck extended, thereby placing the abdomen, the thorax, the neck, and the skull in a line so as to eliminate ridges and valleys. The broken line extending from McBurney's point to the supraclavicular space,

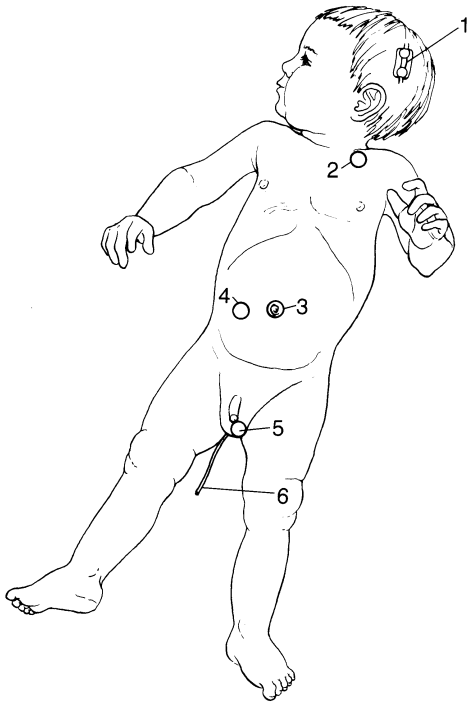


Figure 15.42. Areas of extrusion, disconnection, breakage, colonization, and occlusion along the path of the ventriculoperitoneal shunt. At the cranial end (*1*) one may observe disconnection of the shunting system if it is a three-piece unit (but not if it is a Uni-Shunt), extrusion of either a three-piece unit or a Uni-Shunt, and colonization of a reservoir if one is present. At the supraclavicular area (*2*) the shunt tubing may extrude, as it may from the umbilicus (*3*). A three-piece system may become disconnected at the abdominal end, and either the three-piece system or the Uni-Shunt may extrude at this point (*4*). It is not unusual for a child to develop clinical evidence of an inguinal hernia after a ventriculoperitoneal shunt has been inserted, since the cerebrospinal fluid dissects its way through the preexisting muscular weakness (*5*). Lastly, if the abdominal end of the shunting system penetrates the bowel, it may deliver itself through the anus (*6*). Additional points of importance concerning sites of location of shunt difficulties are the fact that the choroid plexus may grow into the perforations of the proximal end of the shunt system, binding the latter to the former. Consequently, when removing the proximal end of the system from the ventricle at the time of shunt revision one should not exert force: the choroid plexus may be torn and intraventricular hemorrhage result. The distal end of the shunting system, the intra-abdominal portion, may become coated with a proteinaceous material, which effectively seals the slit valve, obstructing drainage.

and from here to a point immediately posterior to the parietal eminence, indicates the path along which first the guide, and then the shunt, is passed. A linear incision is made at McBurney's point and a horseshoe incision posterior to the parietal eminence. It is best to make at least a 3.5- to 4-cm incision in the abdomen. Small incisions provide inadequate working space.

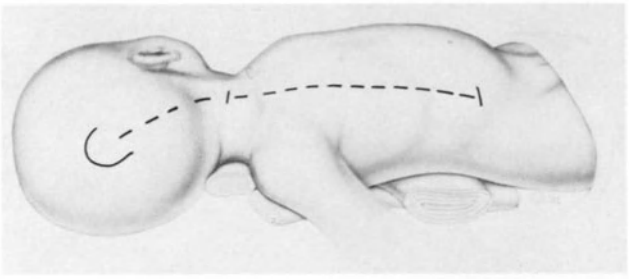


Figure 15.43. Recommended position for the patient. The tract for passage of the shunt through the subcutaneous space is *indicated by the broken line*, the skin incisions *by the intact lines.*

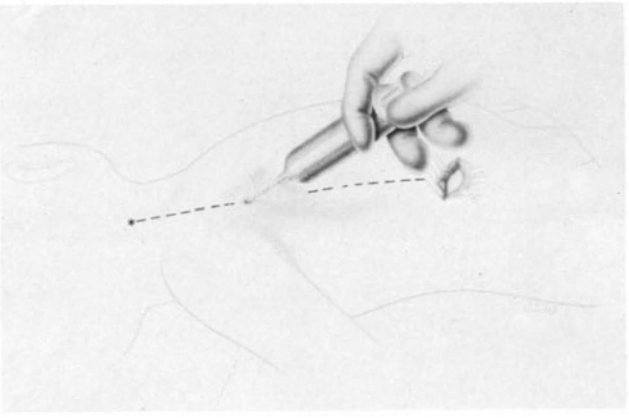


Figure 15.44. Normal saline is being injected into the subcutaneous space so as to separate the skin from the body wall. This permits an atraumatic and easy passage of the guide.

After the incisions have been made and clamps put on either the subcutaneous tissue or the galea, normal saline is injected into the subcutaneous space, along the proposed shunt path, separating the subcutaneous connective tissue from the abdominal wall, the thoracic cage, and the periosteum. The injection of saline is begun at the incision at McBurney's point. This separates the tissue planes, allowing for ready passage of the subcutaneous guide, and minimizes the risk of damaging the skin by the guide as it is passed within the subcutaneous space. This is illustrated in Fig. 15.44, where one sees that saline has been injected subcutaneously, beginning at the abdominal end and proceeding superiorly toward the clavicle. There is no need to add local anesthetic to the saline.

The subcutaneous guide shaft is malleable and may, accordingly, be molded into any contour desired. One must be cautious not to bend the tubing, or mold it quickly, since it may kink, thereby rendering impossible the passage of the leader and the shunt. Rather, gentle molding is best effected by holding the guide firmly in

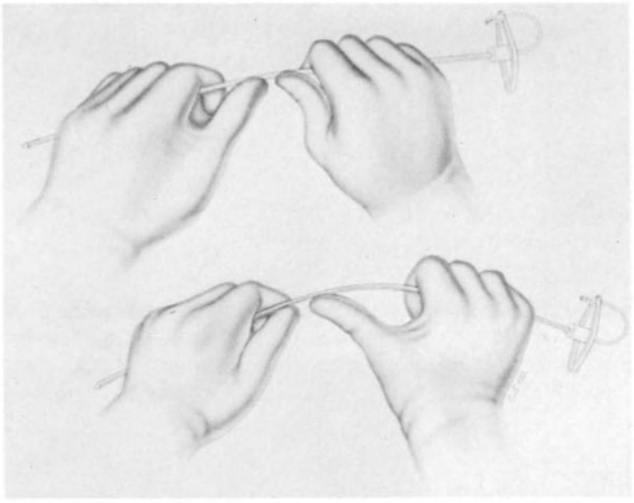


Figure 15.45. Molding of the subcutaneous guide.

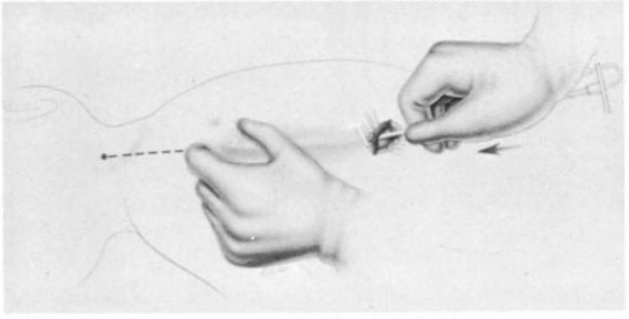


Figure 15.46. Passage of the guide through the subcutaneous space.

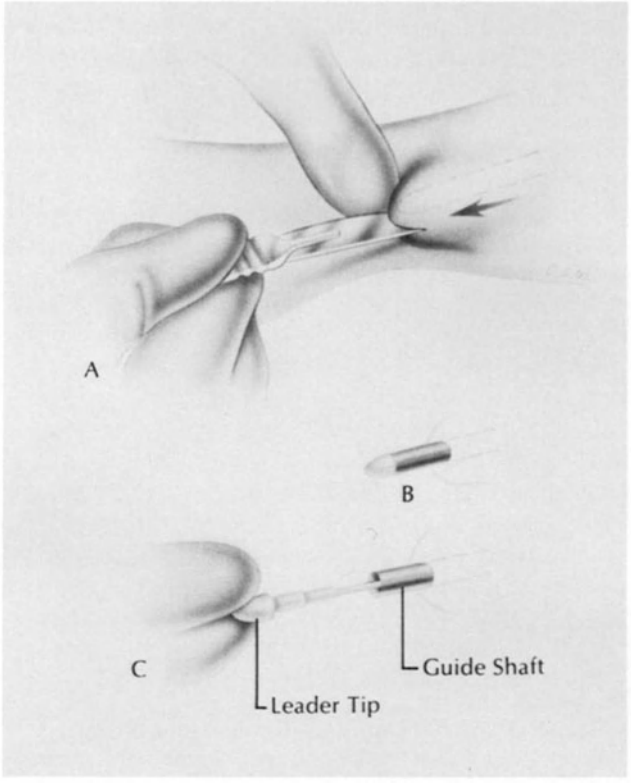


Figure 15.47. (A) The skin incision is being deepened, so as to permit exit of the leader tip and guide. (B) The leader tip and guide shaft have exited. (C) The leader tip is being pulled through the guide shaft.

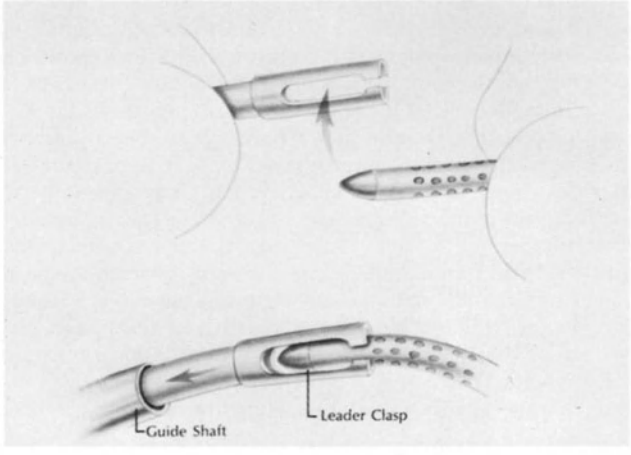


Figure 15.48. The ventricular end of the shunt is being readied for insertion into the clasp and the leader is being drawn through the guide shaft, pulling the shunt into the subcutaneous space.

both hands, and curving it to the desired form between the thumbs. This should be done before inserting the guide into the subcutaneous space: molding it thereafter may cause skin damage.

The skin and subcutaneous connective tissue at the abdominal incision may be picked up with Adson-Brown forceps. The guide is inserted directly into the subcutaneous compartment, prior to advancing it within the space distended by the injected saline. It is advanced superiorly beyond the clavicle, as indicated, in directing the tip by trapping it between the thumb and index finger of the left hand. One may hold the guide either along its shaft, or at the handle. Once the tip (which is the leader tip) has passed over the clavicle, it is pushed forward to the previously placed supraclavicular incision (placed 2.5 cm cephalad to the clavicle), permitting exit first of the tip and then the subcutaneous guide itself. No attempt should be made to pass the guide directly from the abdomen to the scalp opening; it stretches and tents the skin in the neck.

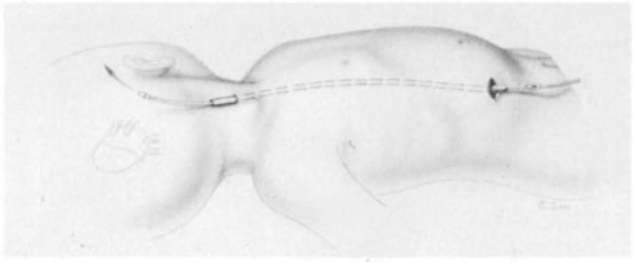


Figure 15.49. The guide rests in the subcutaneous space and the shunt is within it. When the guide shaft is withdrawn, the shunt will rest within the subcutaneous space.

Figure 15.51. Making a cruciate incision in the dura.

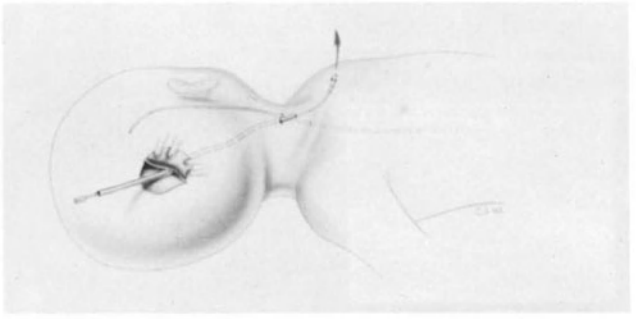


Figure 15.50. Passage of the guide and leader tip through the subcutaneous space from the parietal eminence to the supraclavicular space.

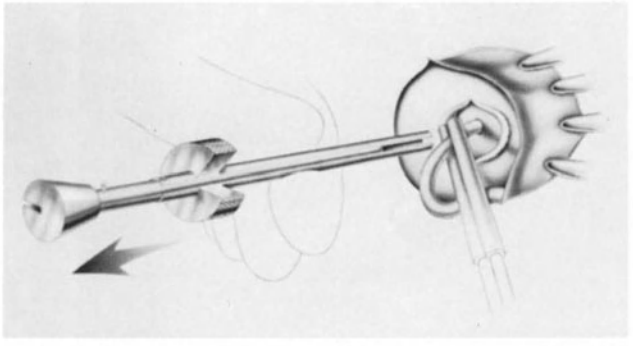


Figure 15.52. With the ventricular tip of the shunt in the ventricle, the barrel of the split trocar is withdrawn as the shunt is held securely in place.

Once the tip and guide have been advanced through the supraclavicular opening, the leader tip is withdrawn for a distance of 3–4 cm. The guide handle is removed and then the ventricular end of the shunt tubing is nestled into the clasp at the distal (abdominal) end of the leader. The leader tip is drawn through the subcutaneous guide, cephalad, and the guide shaft withdrawn caudally through the skin at the abdominal opening, thereby bringing the shunt to rest in the subcutaneous space and removing the guide.

The procedure for passage of the subcutaneous guide and the leader system from the posterior parietal area down to the supraclavicular space is performed in exactly the same manner as the passage from the abdominal end up to the supraclavicular space. However, it is helpful if the surgeon uses either a Kocher or a Peon to open a channel between subcutaneous connective tissue and the fascial layers, especially across the line of insertion of the erector capiti muscles to the occipital, highest and lowest, lines. The guide and leader tip are brought through the supraclavicular incision, and the leader tip is withdrawn entirely. Then, the clasp of the leader system is inserted through the subcutaneous guide beginning at the cephalic end and proceeding toward the supraclavicular space. Once the clasp end is advanced through the guide and exits at the supraclavicular space, the ventricular end of the shunt tubing is reinserted into the clasp, and then pulled up toward the parietal eminence. The subcutaneous guide shaft is withdrawn cephalad over the shunt tubing and leader tip. The clasp of the leader tip is then disconnected, leaving the shunt resting within the subcutaneous space from the posterior parietal area, neck, thorax, and abdomen.

At this time the periosteum between the parietal eminence and the sagittal plane is cut and stripped from the bone. A 1/4-in. twist drill opening is made in the skull. The surface of the dura is coagulated with a bipolar forceps in a cruciate manner and then incised with a #15 blade.

The ventricular split trocar is then inserted through the dural opening and compressed cerebral mantle into the dilated ventricle. This trocar permits the surgeon to obtain the egress of ventricular fluid after he has punctured the ventricle. Ventricular fluid may be removed for chemical and bacteriological analysis and then the obturator turned so as to close the system.

The white plastic obturator is withdrawn gradually from the shaft of the trocar. An assisant holds the ventricular end of the shunt in Cushing forceps, provided

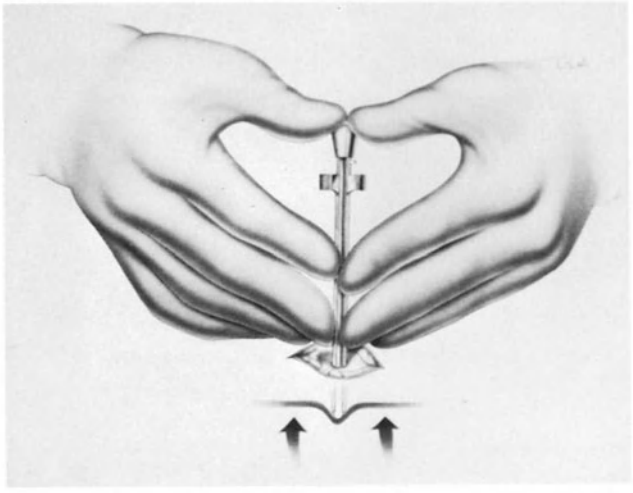


Figure 15.53. The trocar is brought against the peritoneum, exciting the guarding reflex so as to tighten the peritoneum and facilitate puncture into the peritoneal cavity. *The peritoneum must be rigid before puncturing it, lest intra-abdominal vessels and viscera be damaged!*

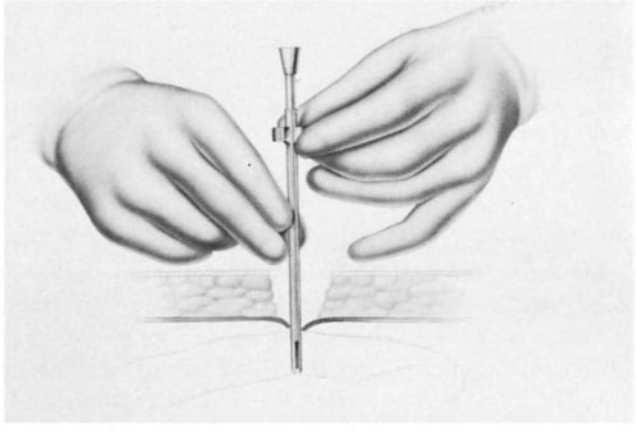


Figure 15.54. The peritoneal cavity is punctured and the obturator is being removed, leaving the barrel of the split trocar in place.

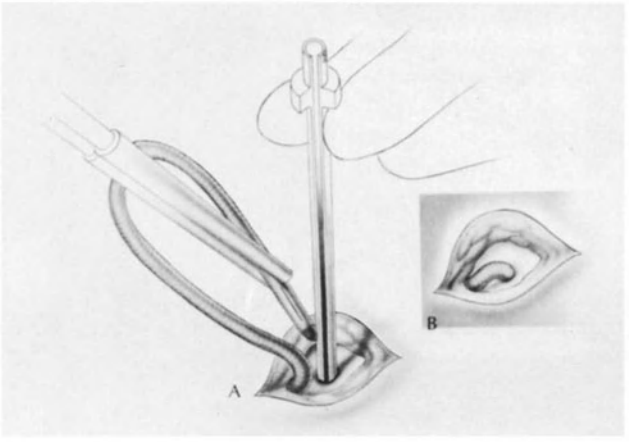


Figure 15.55. (A) The peritoneal end of the shunt is being threaded into the barrel of the trocar for passage into the peritoneal cavity; (B) then the barrel is removed.

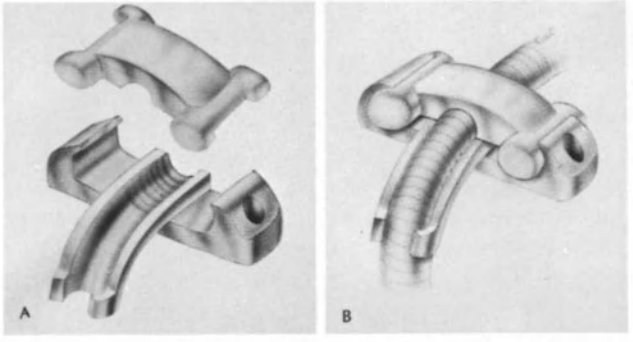


Figure 15.56. (A) The two disarticulated pieces of the clip; (B) locked into one another over the shunt.

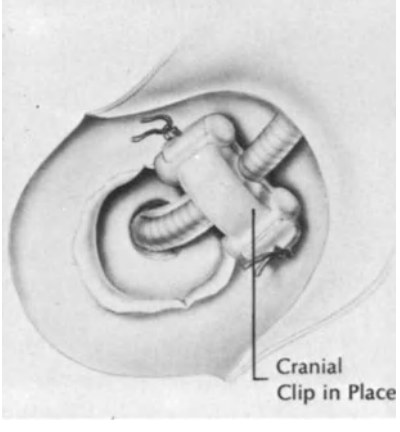


Figure 15.57. Anchored lock-clip locking shunt into place at cephalic end.

with shodded tips which diminish the possibilities of damaging the shunt tubing. As the obturator is withdrawn, the ventricular end of the shunt tubing is brought closer to the opening groove in the split trocar. The surgeon must be careful to withdraw the obturator very slowly and to insert the ventricular tip of the shunt into the opening of the split trocar quickly so as to minimize the loss of cerebrospinal fluid. A rapid withdrawal of the plastic obturator will allow cerebrospinal fluid to pour from the ventricle before the tip of the shunt is fed into the barrel of the split trocar. As the obturator is withdrawn the tube is inserted, and then advanced, first quickly and then slowly, along the trocar and into the ventricular system to the desired (previously measured) distance. Once there is a free flow of cerebrospinal fluid through the distal (abdominal) end of the shunt, the split trocar is removed. The spring catheter is anchored in the two-piece, plastic "lock-clip," which is then sutured to the periosteum.

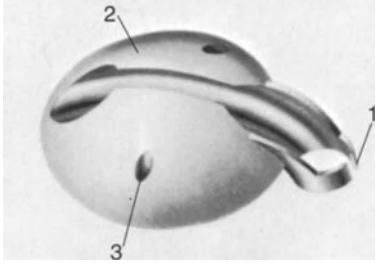


Figure 15.58. Slip-clip (abdominal end). The clasp (*1*) directs the distal end of the shunt tubing into the peritoneal cavity. The groove (*2*) is smaller than the external circumference of the Uni-Shunt, so it holds it snugly in place. The perforations on either side (*3*) are for passage of suture material for anchoring of the slip-clip to the abdominal wall.

The same split trocar as used for insertion of the ventricular end is now used to perforate the peritoneum for entry into the peritoneal cavity. It is fixed snugly between the thumbs and fingers of both hands and inserted through the abdominal musculature to the peritoneum. *Pressure is applied to the peritoneum itself until guarding is obtained, thereby assuring the surgeon that the peritoneum is rigid and may be punctured without perforating a viscus or damaging a vessel.* The puncture of the peritoneum should be performed with a quick, snapping, brisk movement, which is limited in excursion, so as to assure clean penetration of the peritoneum. Once the trocar is within the peritoneal cavity, the obturator is removed, leaving the barrel of the split trocar in place. The distal end of the shunt is then passed into the peritoneal cavity by threading it through the barrel of the split trocar. There should be no resistance to forward passage of the distal end. Once the insertion of the tubing has been completed, the spring catheter is held snugly in place by a pair of shodded Cushing forceps and the barrel withdrawn. The shunt is now in position and needs only to be anchored in place at both the cranial and abdominal ends.

The plastic "lock-clip" consists of a two-piece molded system which is curved to provide proper direction to the ventricular end. The bottom half of the clip is inserted beneath the spring catheter, which is then positioned snugly into the clip. The top portion of the clip is placed over the spring catheter, onto the bottom portion of the clip, and then snapped into place, thereby locking the spring catheter into the clip. The clip is anchored to periosteum at the cranial end.

The single piece "slip-clip" is used for anchoring the abdominal end of the catheter. It is designed to facilitate gradual slippage of the catheter out of the peritoneal cavity, thus offering the advantage of a "growing shunt." To insert the catheter into the single-piece "slip-clip,"

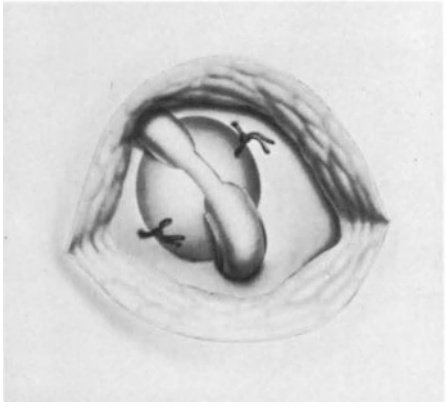


Figure 15.59. Anchored slip-clip holding shunt in place at abdominal end.

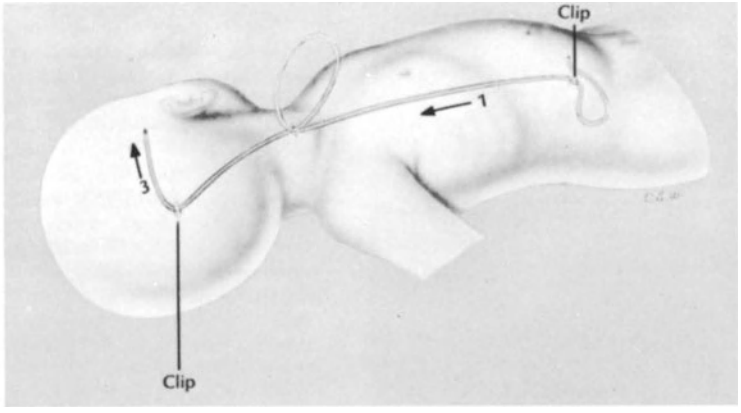


Figure 15.60. Four steps for inserting shunt if one uses split trocar for ventricular end.

gently stretch the tubing and ease the catheter into the clip. The "slip-clip" is then anchored to the abdominal musculature.

The Delta Valve

The description for the insertion of the one-piece shunts given above applied to the first fully unitized system, and to the use of cannulae, tubular (hollow) subcutaneous guides, and split trochars for shunt insertion and passage. It was also the first time clips were used for anchoring. Presently, in the author's opinion, the Delta Valve incorporates the fullest complement of technological characteristics favoring adequate CSF shunting and minimizing siphoning, as well as ease of insertion and anchoring facilities. Therefore, it was decided to reprint material from the descriptive brochure (Delta Shunt Kit, Instructions for Use, PS Medical Corp.) and the paper, "Operative Protocol for the Delta Valve in Pediatric Hydrocephalus," by Marion Walker and David Watson (PS Medical Corp., Medical Education Series).

The Delta Valve incorporates a membrane pressure valve in series with a normally closed siphon control mechanism. This combination enables the valve to maintain intraventricular pressure within a normal physiological range, regardless of the patient's cerebrospinal fluid flow requirements or body position.

The siphon control mechanism is normally closed. It opens in response to positive pressure from the ventricles, acting in concert with the membrane valve by virtually eliminating unwanted additional resistance to CSF flow. The siphon control mechanism consists of two silicon elastomer diaphragms normally closed against two outlet ports (the outlet ports communicate with the distal catheter). CSF flowing from the ventricles pushes the diaphragm surfaces away from the outlet ports, allowing CSF to flow through the ports and distal catheter.

When the patient is upright, suction on the diaphragm surfaces adjacent to the outlet ports could increase resistance to flow (i.e., positive pressure would be required to open them), due to the siphon effect in the distal catheter. The effects of this suction on the resistance to flow are minimized in accordance with the principle of hydrodynamic leverage. In the Delta Valve (Fig. 15.61), the area of the diaphragms acted on by CSF flowing from the ventricles is 20 times greater than the area of the diaphragms acted on by suction from the distal catheter. This minimizes the excessive reduction of intraventricular pressure and volume due to overdrainage of CSF. This untoward event (CSF overdrainage) may result in: (a) low intracranial pressure syndrome, (b) subdural hematoma and hygroma, or (c) slit ventricle syndrome.

The performance characteristics of this system are illustrated in Figs. 15.62 and 15.63.

Surgical Procedure

Ventricular Catheter Placement

The right angle clip assembled on the ventricular catheter may be used to bend the catheter to an approximate 90° angle at the point where the catheter exits the twist drill or burr hole. The clip may be used as a marker for planned depth of catheter insertion when slid the appropriate distance from the proximal tip of the catheter prior to insertion. This can be done with the stylet in the catheter. The stainless steel stylet is designed to facilitate introduction of the catheter into the lateral ventricle.

After the catheter is properly positioned in the ventricle, the extracranial portion of the catheter is pressed into the split tubular segment of the clip to form the right angle bend. When pressed into the clip, the catheter should not be stretched. If the catheter is to be placed in the ventricle through a tubular introducer, the

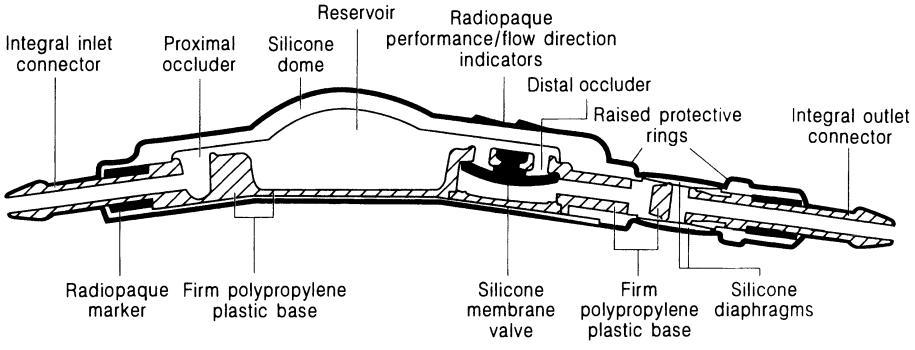


Figure 15.61. Half-sectional view of the Delta Valve. [From D.A. Watson (1994) The Delta Valve: a physiologic shunt system. Child's Nerv Syst 10:224–230.]

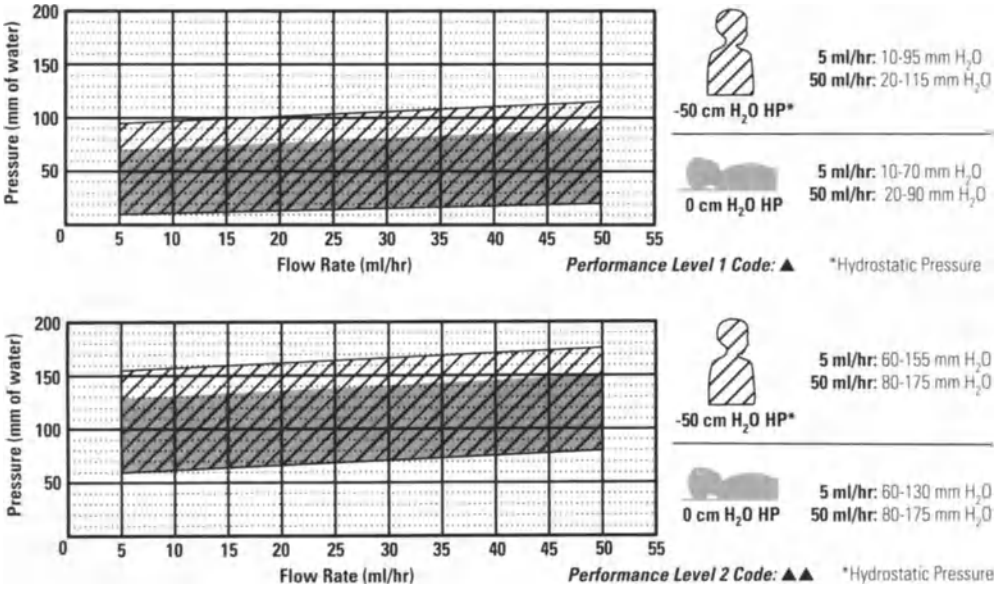


Figure 15.62. Original Delta Valve performance levels 1 and 2 pressure flow curves. [From M. Walker and D. Watson (1994) Operative protocol for the delta valve in pediatric hydrocephalus. Medical Education Series. PS Medical Corporation, Goleta, CA.]

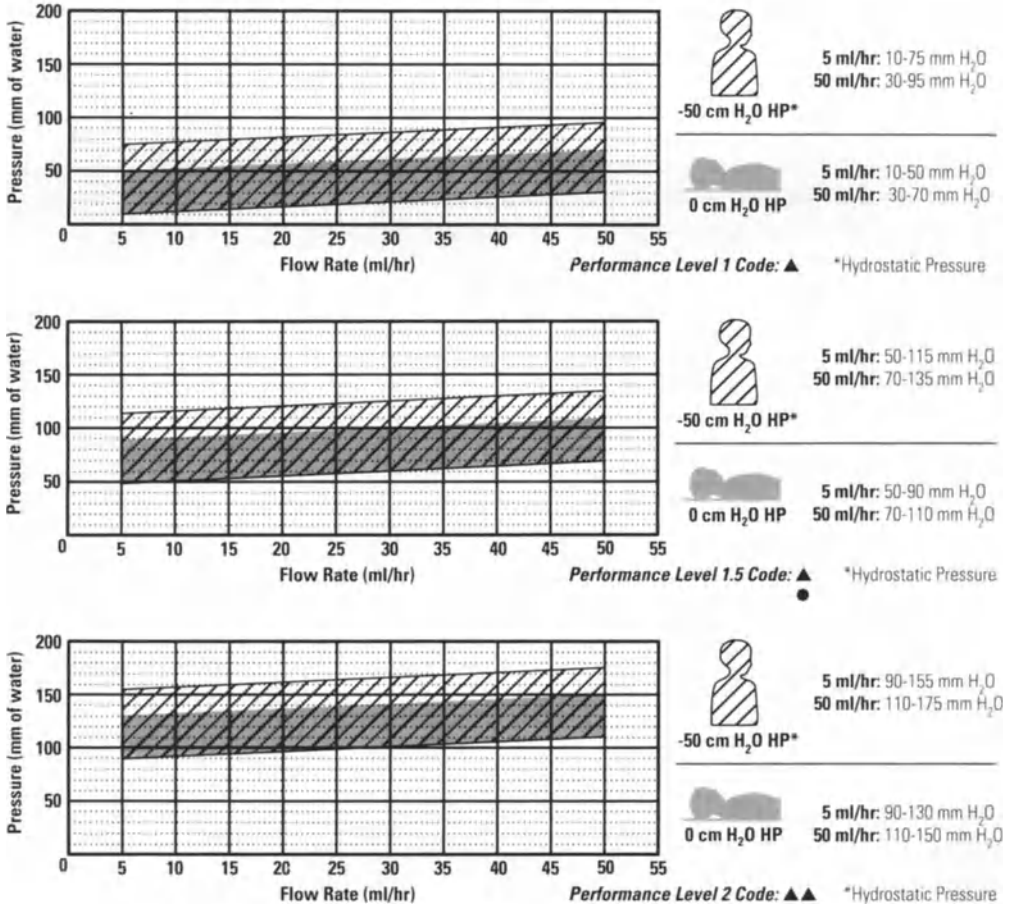


Figure 15.63. Delta Valve performance levels 1, 1.5, and 2 (1994) pressure flow curves. [Taken from reference as in Fig. 15.62.]

clip must be removed prior to insertion of the catheter through the introducer. The right angle clip should be secured to adjacent tissue by passing sutures through the two suture holes on the sides of the clip.

To connect the Delta Valve to the ventricular catheter, the integral polypropylene connector is inserted into the ventricular catheter tubing. The connector should be completely covered by tubing.

A subgaleal pocket must be formed with appropriate depth to accept the extracranial portion of the ventricular catheter and the contoured valve component of the shunt kit. Implantation of the ventricular catheter as the last step in the shunting procedure is recommended. This will minimize loss of CSF during surgery.

Peritoneal Catheter Placement

A variety of surgical techniques may be used in placing the distal catheter into the peritoneal cavity. The site of placement is at the discretion of the surgeon. The catheter length is 90 cm with wall slits at the distal end. If catheter wall slits are not desired, the distal end may be trimmed. The catheter should be checked for patency at the time of surgery.

Ventriculoatrial Shunt

Theoretically, one supposes that inserting the distal end of the shunt into the facial vein, and then threading it inferiorly into the internal jugular vein, guarantees patency of the internal jugular vein so that venous blood may continue to drain through it. In fact, unfortunately, once a foreign body (the shunting system) is within the internal jugular vein, this vascular structure closes around the tubing and thromboses upstream of its entrance into the vein. Therefore, a ventriculoatrial shunt results invariably in thrombosis of, at least, the internal jugular vein. Very often, the subclavian vein and, alas, the superior vena cava also thrombose.

It is advisable to expose the facial vein, and its entrance into the internal jugular vein, prior to cannulation of the cerebral ventricle, but one should not open the cervical venous system until the proximal end of the shunt is in the lateral ventricle, and there is a free flow of cerebrospinal fluid through the distal end of the shunting system. Prior to insertion, one should measure the distance from the facial vein to the carina, the anatomical landmark for entrance of the superior vena cava into the right atrium.

After the facial vein has been dissected from the surrounding loose connective tissue, 4–0 sutures are placed in it, parallel to one another and separated by a distance of approximately 2–3 mm. These are used as suspension sutures, which may be lightly drawn apart to put traction on the wall of the facial vein, permitting the surgeon to incise it and insert immediately the distal tip of the shunt tubing. This is threaded through the facial vein and then guided inferomedially into the internal jugular vein. The catheter is advanced gradually to the full length of the measured distance (to the point of entry of the superior vena cava into the right atrium).

An intraoperative flat plate of the chest confirms correct cannulation of the internal jugular vein and the superior vena cava, and precise placement of the tip of the shunting system at the entrance of the superior vena cava into the right atrium. It is not uncommon to estimate incorrectly the distance to the right atrium, and to cannulate the right ventricle. In fact, at times the tip of the shunting system may be threaded into the inferior vena cava.

After the shunt is placed entirely within the venous system, the two previously placed 4–0 purse-string sutures are tied to one another, thus closing the vein, which is then anchored to the surrounding muscular tissue with care being taken to avoid kinking.

The disadvantages of the ventriculoatrial shunt are many. Each insertion or revision, with few exceptions, entails the sacrifice of a major craniocerebral draining vein, superior vena cava thrombosis, vegetative endocarditis, pulmonary infarct, renal infarct, inferior vena cava thrombosis, shunt nephritis, and shunt tubing irretrievably lost in the right atrium or right cardiac ventricle. It is, consequently, recommended that the ventriculoatrial shunt be performed only when, for one reason or another, a ventriculoperitoneal shunt is impossible, never as a procedure of choice.

Ventriculopleural Shunt (Fig. 15.64)

There are times when one has no alternative other than to attempt to shunt the cerebrospinal fluid into the pleural cavity, a potential space, which may occasionally be capable of absorbing the fluid. This, fortunately, is generally not the case, since the majority of shunts into the pleural cavity result in massive collections of cerebrospinal fluid in one hemithorax, and displacement of the mediastinum to the opposite side. This pleural "effusion" impairs unacceptably the vital capacity of the patient. These inconveniences have been reported repeatedly since Ransohoff [56] first described the procedure.

After the proximal end of the shunting system has been positioned in the appropriate lateral ventricle and the tubing passed subcutaneously from the scalp to the sixth intercostal space, the intercostal muscles are separated from one another, a rib spreader is positioned so as to open a distance of approximately 1.5 cm. It is best to ask the anesthesiologist to deflate the lung at this time, to facilitate opening the parietal pleura and diminish risks of cutting the lung. An adequate amount, 6–12 cm, of distal shunt tubing is inserted into the

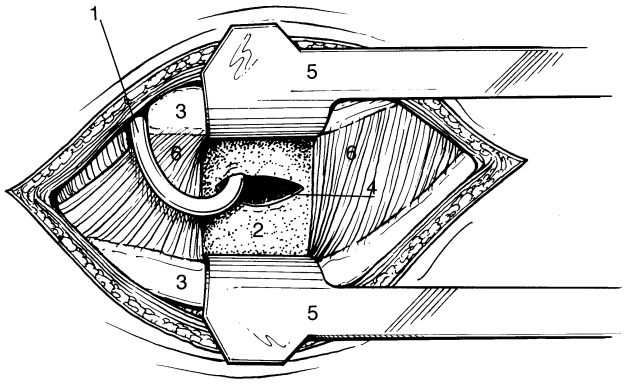


Figure 15.64. Ventriculopleural shunt insertion. At the time of insertion of the distal end of the shunt tubing (*1*) through the parietal pleura (*2*) one should spread the ribs (*3*) and ask the anesthesiologist to collapse the lung (*4*), in order to allow the shunt tubing to pass freely into the pleural cavity. At the time of closure, the rib spreaders (*5*) are removed and the ribs allowed to come back into normal position prior to closing the intercostal muscles (*6*). The lung is then inflated.

pleural cavity and then this latter is filled with saline. As the lung is reexpanded, air is forced from the pleural cavity. It is advisable to place an anchoring suture approximately 1 cm superior to the point of entrance of the shunt tubing into the pleural cavity. Care should be taken to avoid kinking of the tubing. The intercostal muscles are closed so as to produce a watertight seal. Routine chest X-rays and respiratory therapy evaluations permit one to learn whether cerebrospinal fluid is accumulating in the pleural cavity [57].

Ventriculogallbladder Shunt (Fig. 15.65)

The major activity of the gallbladder on bile composition is to remove water and inorganic electrolytes. The rate of fluid absorption by the gallbladder in vivo is approximately 16% of the total gallbladder volume per hour. Water flux rate is approximately 25 ml/h, or about 10% of the total gallbladder volume per hour. About 90% of the water is removed in the process. As a result, there is a progressive increase in the concentration of the conjugated bile acids and diminution in the concentration of chloride and bicarbonate. The ultimate product is a solution in which bile acid, sodium, potassium, and calcium concentrations are extremely high, with the latter two averaging 10–25 meq/l, respectively.

It is not possible to state with precision whether cerebrospinal fluid shunted into the gallbladder is entirely drained into the duodenum, through the common duct, or absorbed, to a greater or lesser degree, by the gallbladder epithelium. The resistance of the gallbladder epithelium to passive osmotic flow is high. Consequently, a considerable osmotic gradient must exist be-

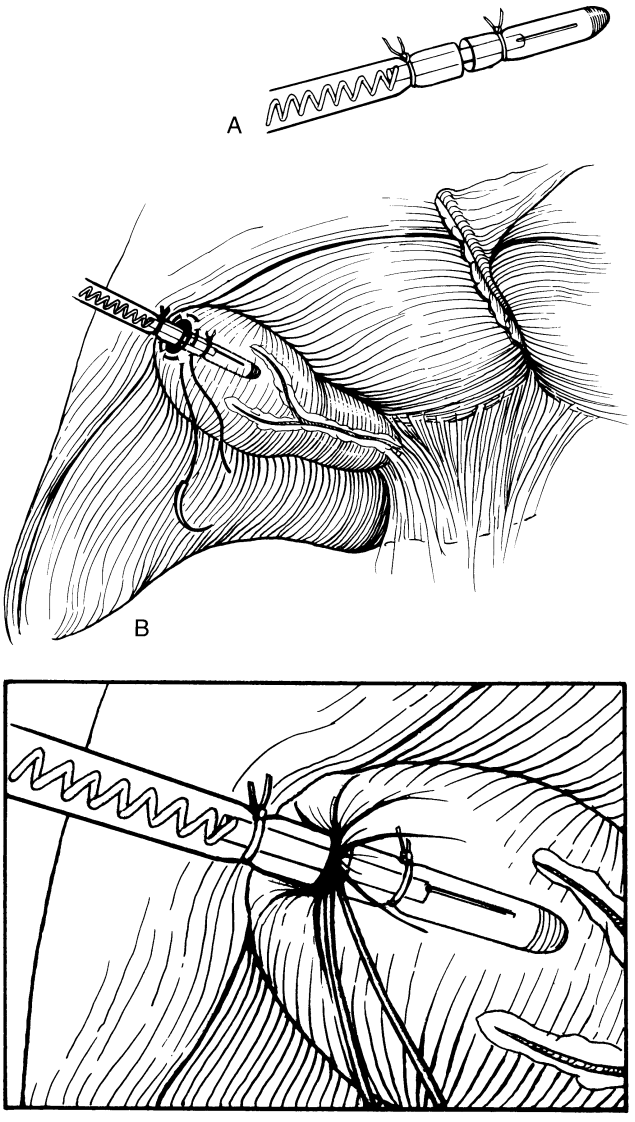


Figure 15.65. Ventriculogallbladder shunt insertion. (A) About 2 cm of the distal (slit) valve and spring catheter are prepared to receive a straight connector. After this is inserted and secured with a 4–0 suture, the proximal portion of the spring catheter is mounted over the straight connector and also secured in place. (B) Then, the distal tubing is positioned in the gallbladder so that the straight connector is centered at the opening in the gallbladder and the purse-string suture. (C) After the purse-string suture has been tied and the straight connector anchored to the now closed gallbladder wall, the serosa is sewn over the purse-string suture to anchor the gallbladder onto the straight connector.

fore water or cerebrospinal fluid may move across it at adequate absorptive volumes.

In 1958, Newman, Hoen, and Davis [58] treated a case of communicating hydrocephalus by shunting the cerebrospinal fluid into an isolated segment of ilium, demonstrating absorption over a period of 10 months. Therefore, cerebrospinal fluid shunted into the gallbladder may be absorbed by intestinal mucosa or, if there is a blockage of the cystic duct, by the epithelium of the gallbladder. In the same year, Smith, Moretz, and Pritchard [59] diverted the cerebrospinal fluid into the gallbladder in ten hydrocephalic patients, shunting from the lumbar subarachnoid space in patients with communicating hydrocephalus and from the lateral ventricle in those with obstructive hydrocephalus. Of the ten patients, three died within 3 months, and seven were alive 2 months, 5 months, 12 months, and 2 years later.

Yarzagaray (L. Yarzagaray, unpublished observations) reported a 6-year follow-up of 50 children with hydrocephalus: 35 cases of Chiari types II and III; 11 cases of aqueductal stenosis; 3 cases of Dandy-Walker; and 1 case of sarcoma of the meninges. Of this number, 32 patients had previous infections: 20 with ventriculitis, and 12 with previous peritonitis resulting in an inability to absorb CSF. Functioning shunts were reported to range from 1 to 6 years (still functioning) in this cohort. Subsequently, ten of his patients had their shunts changed from ventriculogallbladder to ventriculoperitoneal, by which time the peritoneal surface reacquired its cerebrospinal fluid absorptive capacity. At the time of conversion, laparotomy revealed the gallbladder to have a "hydrops" appearance: enlarged with thick walls, the peritoneum covering it was edematous. He reported no obstructions (intraoperative cholangiography).

Yarzagaray Technique for Ventriculogallbladder Shunting

A 5.0-cm incision is made in the right upper quadrant, two fingerbreadths below the costal margin and parallel to the ribs. The layers of the abdominal wall are incised, the peritoneum opened, and the abdominal end of the spring catheter is cut 2 cm from the tip. It is then reconnected to the spring catheter with a straight connector. The distal portion is secured with 2–0 suture material. (The only purpose of the connector is to serve as an anchoring point for the tube when it is introduced into the gallbladder lumen, permitting one to tie it in place without obliterating the shunt tube). The gallbladder, located immediately below the anterior border of the liver, is exteriorized with Babcock forceps and a full-thickness purse-string suture is placed in its fundus, leaving a 1.0-cm surface to permit puncture of the gallbladder (18-gauge needle for pressure recording and bile culture), and introduction of the distal end of the valve and the straight connector.

After the purse-string suture is tied, a serosal suture is placed to invaginate it, making certain the straight connector is anchored securely. A 25-cm loop of spring catheter is left in the peritoneal cavity, to allow for the child's growth.

The peritoneum and abdominal wall are closed, avoiding angulation or constriction of the spring catheter at its passage from the peritoneal cavity into the subcutaneous space.

Retrograde flow of bile into the ventricles may occur. Some have reported this to be fatal, others reversible after ventricular lavage. Yarzagaray has reported that 15 of his patients with "indirect clinical evidence of ventriculitis, as indicated by elevated CSF protein and low sugar," did not develop cholangitis, evidence of infection of the biliary pathways, or cholelithiasis. We have had two cases of bile ventriculitis, one fatal within less than 48 h.

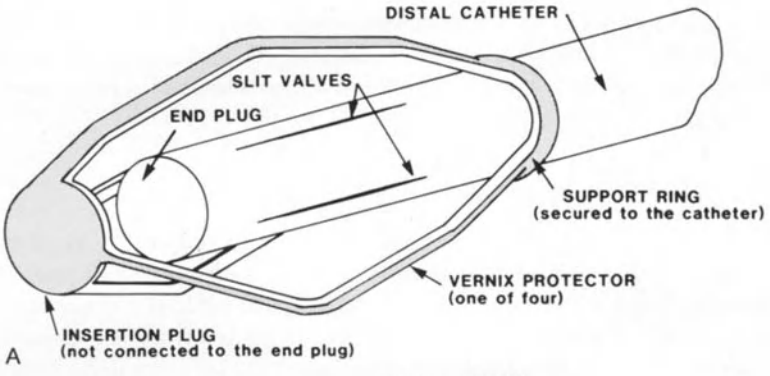
Ventriculoamniotic Shunt: J.T. Brown Technique [60] (Fig. 15.66)

The question is "can a fetal abnormality (hydrocephalus) be diagnosed in utero and, if so, should attempts be made to treat it prior to birth." If the hydrocephalus is progressive and impairs fetal development, it should be operated on "in utero." Before concluding this treatment plan, however, it is important to look thoroughly via aminocentesis and real-time ultrasonography for additional abnormalities and not just to treat what is obvious, lest a lethal, uncorrectable anomaly be overlooked.

Ventriculoamniotic shunts, placed either percutaneously or via hysterotomy, have been performed in the laboratory in hydrocephalus-induced sheep [61] and primates [62]. Most recently, percutaneous ventriculoaminotic shunts have been placed in human fetuses under sonographic direction [63, 64].

One might attempt to summarize the management options available when progressive hydrocephalus has been diagnosed in utero. If the diagnosis is made prior to 20–24 weeks gestation, then abortion may be an option for some. Clearly, this is an unacceptable option for others. If the diagnosis is made after 24 weeks then, perhaps, the fetus could be delivered early (32–34 weeks), after steroid-induced pulmonary maturity in female fetuses. One, of course, could opt for delivery after normal lung maturation around 37 weeks gestation. Delivery could also occur at term with or without a cranial destructive procedure. *This, however, as abortion, is not an option for many people.* Finally, if the diagnosis of fetal hydrocephalus is made from 20–32 weeks gestation, then a ventriculoamniotic shunt becomes an additional management option.

Figure 15.66. (A) Diagrammatic representation of vernix protector. (B) Distal (amniotic) end of the new fetal shunt with vernix protector. (C) Ventriculoamniotic shunt driven down trocar by blunt stylet. (D) Twelve-gauge trocar with sharp stylet encountering fetal cranium. (E) Proximal (ventricular) end of new fetal shunt with ventricular flange. (F) Newly proposed fetal shunt.



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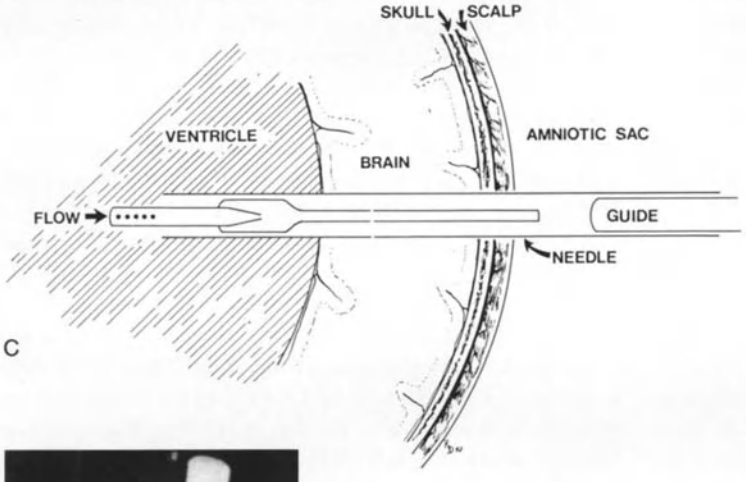

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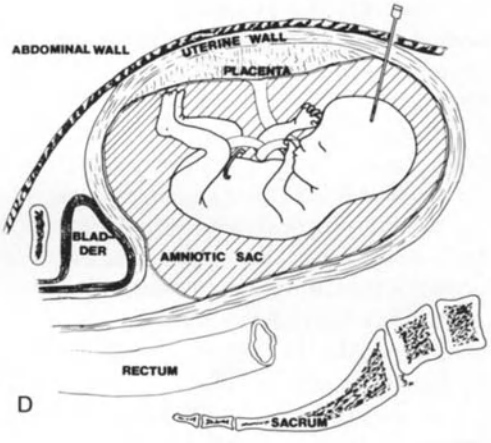

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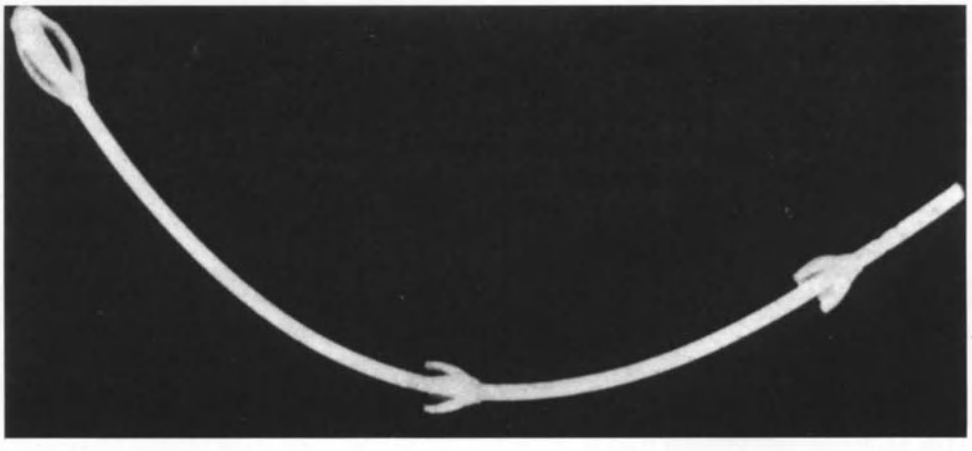


F

The surgical team consists of a specialist in maternal-fetal medicine and high-risk obstetrics, an obstetrical ultrasonographer, and a pediatric neurosurgeon.

In preparation for the procedure, the mothers are sedated with intravenous diazepam and meperidine. These medications are also given to reduce fetal movement. The same medications are intravenously supplemented, as needed, during the course of the procedure. At no time are betamimetics or other such labor inhibitory drugs needed.

After the maternal abdomen is prepped and draped in a standard fashion, a sterile (draped) sonographic probe is used to locate the precise position of the fetus' head. Not surprisingly, the fetus is often in a breech presentation. A vertex presentation, however, does not preclude placement of a ventriculoamniotic shunt.

A fetal mannequin in the operating room is useful in giving all participants a three-dimensional view of the exact fetal position. After the location of the fetal head is determined, the maternal abdominal wall in that area is infiltrated with 1% xylocaine local anesthesia and a 4- to 5-mm stab wound made with a scapel. At this point, a 12-gauge trocar with a sharp stylet is inserted into the amniotic sac and amniotic fluid is withdrawn by removing the stylet. The stylet is then reinserted and, again, under sonographic guidance the fetal cranium is encountered. It is preferable to maneuver the fetal head manually, beneath the path of the needle, rather than to direct the needle toward the fetal head. Sonographically, one can easily see the needle encounter the fetal head and then indent the skull.

The site of trocar penetration into the cerebral ventricle often varies, but usually occurs at or just in front of the coronal suture. Alternatively, one may insert the trocar just behind the parietal eminence directing it anteromedially. The right hemisphere is chosen for penetration whenever possible.

As one penetrates the thin scalp, skull, and cerebral mantle, a distinct "give" is noted. The sharp stylet is withdrawn and cerebrospinal fluid is allowed to egress. The cerebrospinal fluid is always under considerable pressure (20–25 cm H_2O), and usually slightly xanthochromic and proteinaceous. The fluid is vented until it no longer exits under pressure. This is done for fear that if the shunt is placed while the cerebrospinal fluid is still under pressure, the shunt may be extruded from the ventricle.

After cerebrospinal fluid is vented, the Newkirk ventriculoamniotic shunt (Denver Surgical Developments, Inc., 6851 Highway 73, Evergreen, Colorado) is placed into the hollow trocar. The shunt is then driven down the trocar to a predetermined depth with a blunt stylet. At this point the trocar is withdrawn over the blunt stylet, leaving the shunt in situ with one flange in the lateral ventricle (to prevent external migration of the shunt) and the other flange just beyond the scalp (to prevent internal migration). A Steri-Strip or Band-Aid is placed over the mother's skin opening.

Lumbar Peritoneal Shunt

The lumbar peritoneal shunt may be performed either by the open or closed technique, making a skin incision and exposing the laminae and interlaminar space in the former, but simply puncturing the subarachnoid space with an appropriate trocar for the latter. The closed technique is preferable.

Open Technique for Lumbar Peritoneal Shunt

For the open technique one should place the child in the lateral decubitus position and expose the III and IV lumbar laminae on either side of the L3–L4 interspace. The yellow ligament is then dissected from the laminae and hemilaminotomies performed on the inferior portion of L3 and the superior portion of L4, taking care not to perform a hemilaminectomy at either level, since this predisposes to scoliosis. The dura is exposed and opened, and then parallel 5–0 sutures are placed in the arachnoid and very gently pulled apart. The arachnoid is incised with a #11 blade, between the parallel sutures, opening an ostium large enough to permit entrance of the proximal end of the shunting system. The shunt tubing should be guided inferiorly from the L3–L4 interspace for a distance of 3–4 cm, and then the purse-string arachnoid sutures tied to one another, closing the arachnoid snugly around the shunt tubing. This diminishes the incidence of cerebrospinal fluid leakage around the tubing. The dura is then closed around the tubing, taking care to create a seal, without constricting or kinking the shunt. An anchoring stitch is used to secure the shunt to the lumbar paraspinal muscles.

A subcutaneous guide is passed from the lumbar area around the flank, to McBurney's point, where a 4.0-cm incision is made to permit exit of the subcutaneous guide and, subsequently, the shunting system passed through it. One may introduce the distal end of the shunt into the peritoneal cavity, either by using a peritoneal trocar, as recommended for ventriculoperitoneal shunting, or by using the muscle-splitting technique to open a space through the abdominal muscles and surgical opening of the peritoneum. When the parietal and visceral peritoneum have been separated from one another, tenotomy scissors are used to open the former and permit entry into the peritoneal cavity. Great care must be taken not to damage the visceral peritoneum, since this predisposes to penetration of the shunt tubing into the intestinal lumen! In the newborn and infant, distinction between parietal and visceral peritoneum is most difficult. Hemostats or toothed forceps are used to pick up the cut edges of the parietal peritoneum, opening an ostium through which the distal end

of the shunt tubing may be passed into the peritoneal cavity. The peritoneum is loosely sewn around the shunt tubing, which is, in turn, anchored to the abdominal musculature. It is essential to anchor lumbar peritoneal shunts at both the lumbar and abdominal areas.

Closed Technique for Lumbar Peritoneal Shunt

For performance of the closed technique, the patient should be put in the lateral decubitus position, both legs extended so as to permit the surgeon to expose completely the abdomen on the upper side. Following surgical scrub and draping, which should expose the left side of the abdomen, the flank, and the vertebral spine from L1 through L5, the procedure is begun.

A 1-cm incision is made over the interval between the bulges of the spinous processes of L2 and L3 and a Toughy needle inserted into the subarachnoid space, so that the curved portion of the tip of the needle is facing (caudal) toward the lumbar subarachnoid cul-de-sac. The obturator is removed to ascertain that cerebrospinal fluid is flowing freely, and then the proximal end of the special lumbar peritoneal shunting system inserted and its tip guided through the Toughy needle caudally into the lumbar subarachnoid *cul-de-sac*. The shunt tubing is firmly held in place as the Toughy needle is withdrawn from over it, leaving the tubing in position with the Toughy needle completely removed. If a one-piece lumbar subarachnoid peritoneal system is being used, a lock-clip (or anchoring suture) should be placed at the point of exit of the shunting system from the interspinous ligament. If, on the other hand, a two- or three-piece system is being used, a right-angle connector should be inserted into the proximal shunt tubing and this latter anchored over the right-angle connector, which should, in turn, have the distal end of the shunt tubing inserted over the distal end of the right-angle connector. After anchoring this distal end of the shunt to the right-angle connector, the remainder of the procedure is completed.

Whether a one-piece or multiple-piece shunt is used, the remainder of the procedure is the same. The subcutaneous guide is passed from the lumbar skin incision, around the flank, to McBurney's point on the left, where a 4-cm incision is made. Once the tip of the guide is passed through the 4-cm incision, the distal end of the shunting system is, in turn, passed through the lumen of the guide and this latter is removed from the abdominal end, taking care to avoid pulling the tubing from the lumbar subarachnoid space. If a three-piece system is used, care must be taken not to disconnect the tubing from the right-angle connector. Insertion into the peritoneal cavity is performed by using the same technique as for a ventriculoperitoneal shunt, and the tubing is anchored to the abdominal wall with a slip-clip, just as when performing a ventriculoperitoneal shunt. It is wise to get X-rays on the operating table to ascertain that the proximal end of the shunting system is in the lumbar subarachnoid cul-de-sac and that the distal end is in the peritoneal cavity before taking the child from the operating room.

Shunt Revisions (see Fig. 15.42)

Malfunctioning shunts are common. In fact, one may safely state that a child with hydrocephalus will average 2.1 shunt revisions per 3 years of life, *with the range (in my clinic) being no revisions for the first 15 years of life to 21 revisions in the 1st year of life.* The tubing may break, it may come disconnected from the anchoring devices, either the ventricular or distal end may pull from the cavity into which it had been inserted, either end may become plugged with normal tissue (choroid plexus, blood, omentum) or debris. For reasons which are not understood, the system may be intact but simply will not function in a particular child. Often, the distal (peritoneal) end is covered by a proteinaceous "glove" adherent to the parietal peritoneum. This obstructs the flow of cerebrospinal fluid into the peritoneal cavity. However, when, at the time of revision, the surgeon withdraws the tubing from the peritoneal cavity there is free flow through the distal end. In such cases, reinsertion should be through another portion of the abdominal wall. Three-piece systems have the additional failure risks of:

1. Disconnection at the proximal end with its loss into the brain and ventricle
2. Colonization of the reservoir by bacteria
3. Disconnection of the distal end with loss of the tubing into the peritoneal cavity

When the decision has been made to revise a shunting system because of malfunctioning, it is recommended that the distal end be withdrawn from the absorptive cavity first, in order to learn whether that end is occluded. If so, one should clear the obstructive material from the tubing and then observe whether there is a free flow of fluid. If so, the distal end is simply reinserted into the peritoneal cavity. If, after the occlusive material has been cleared from the distal end of the shunt system there is not an adequate flow of cerebrospinal fluid through the tubing, one proceeds to open the scalp over the proximal end of the tubing and to remove this latter from the ventricle. In the case of a lumbar subarachnoid peritoneal shunt, one should attain complete exposure of laminae and yellow ligaments before withdrawing the tubing from the lumbar subarachnoid space.

At the ventricular end, the tubing should not be slipped out of the ventricle as soon as it is freed from its anchor, since it is surrounded by leptomeninges and cerebrum, passing through these obliquely. *It is best to*

take the time to coagulate the tissues surrounding the shunt tubing so as to shrink them away from the shunt tubing, opening a wide ostium and preventing the surrounding tissue from oozing into the space left by the withdrawn catheter. This will render reinsertion of a proximal catheter less difficult. One should then grasp the proximal end of the shunt tubing with atraumatic tissue forceps and gently lift it from the underlying brain, withdrawing it from the ventricle. If the ventricular tip of the shunt tubing is tethered, necessitating more than *the most gentle traction to withdraw it from the lateral ventricle, one is urged to desist: "jamais forcé!"* When one encounters resistance to extraction of the proximal end of the shunt, it is advantageous to irrigate saline through the shunting system as tubing is withdrawn, with the hope that the irrigating solution will flush the debris from the interstices of the proximal tubing, freeing it so that it may be atraumatically removed. This is not invariably successful, so that one may be obliged to cut the tubing at the cortical surface, leave it in situ, and reinsert an entirely new shunting system. It is possible, and likely, that the proximal end of the shunt tubing is enmeshed in choroid plexus, so that forcefully withdrawing it may result in tearing a choroidal vessel and causing an intraventricular hemorrhage. Though this is not invariably a complication which results in morbidity, it may do so.

After the proximal end of the shunt has been removed from the ventricle, a fluffy cotton is immediately placed over the ostium of the shunt tract to prevent draining of the ventricle. The shunt tip is then inspected and all debris removed, prior to flushing it with abundant amounts of saline in order to clear debris from within the system and to assure oneself that it is fully patent. If there is any question about the patency or integrity of the shunt, it is removed and a new one brought into position within the subcutaneous space. The proximal end of the system is then inserted into the ostium of the previous transcerebral shunt tract and threaded to the desired depth, measuring always from the inner surface of the skull. It is best to use the same shunt tract, rather than introduce the shunt through the cerebral parenchyma, in all instances except, of course, when one has found it advisable to cut the proximal end of the shunt and leave it in situ.

Once the proximal tip has been inserted to the appropriate depth, one may inspect the distal tip to look for flow of cerebrospinal fluid. If it does not appear immediately, this does not indicate that the system is obstructed, since air bubbles may serve as airlocks, obstructing egress of cerebrospinal fluid from the ventricles. Gentle aspiration through a blunt needle inserted into the distal tip, using a 2-ml syringe with a wet barrel, will start flow, siphoning cerebrospinal fluid, which should continue once the needle has been removed from the distal tip of the shunt. If so, one is advised to wait approximately 5 min before proceeding to reinsert the distal tip into the absorptive cavity to assure oneself of steady flow and, more importantly, that there is not intraventricular bleeding. This latter may not become obvious to the surgeon for several minutes, since the blood gravitates to the bottom of the ventricle and the shunt tubing to the top.

In the event the cerebrospinal fluid is blood tinged, one should wait long enough to determine whether the fluid tends to become clear or red. It may be reinserted immediately if it remains clear, but it must not be reinserted into the absorptive cavity if it reddens: this very likely will result in plugging of the tubing with clot and, consequently, the need for another revision within a few hours.

The intraventricular bleeding generally stops within 3–5 min, though, quite naturally, intraventricular blood discolors all of the cerebrospinal fluid. One may allow the bloody cerebrospinal fluid to drip gradually through the shunt until it clears, or, preferably, occlude the distal tip with an atraumatic bulldog clamp and wait for about 15 min, opening the shunt tubing every 2 or 3 min just long enough to vent the intraventricular pressure and to evaluate the colorimetric status of the fluid. The purpose for this waiting period is to allow the bleeding to stop and the blood to form a clot. Once this has occurred, only xanthochromic cerebrospinal fluid will flow through the system, since the cellular elements of the blood will have coalesced into a clot, which will gravitate to the recumbent portion of the ventricle. The shunt tip floats to the superior portion of the ventricle, away from the forming clot. In fact, intraventricular *clots* do not obstruct a shunting system, since they dissolve slowly, over days or weeks, gradually releasing the liquefied elements of the clot into the ventricular system. *It is fresh intraventricular blood that obstructs a shunt because it clots within the tubing.* Though irrigation of the tubing with 2–4 ml saline may be advantageous in clearing blood from the tubing, repeated irrigation should not be performed because it distributes the blood uniformly throughout the ventricle. This greatly increases the chances of clot forming within the shunt tubing. It also subjects the child to uncontrollable and potentially dangerous fluctuations in intraventricular pressure. Persistent flow of fresh blood through the shunt tubing over a prolonged period of time indicates to the surgeon that the bleeding is continuing, that he should not internalize the system *but proceed to external ventricular drainage.*

If at the time of shunt revision one encounters evidence of *infection* (in the subcutaneous tissue or the peritoneal cavity, the cerebrospinal fluid flowing through the tubing), one should proceed directly to external ventricular drainage. One may opt simply to convert the shunt into an external ventricular drainage, or to remove it completely and then to insert an external

ventricular drain (EVD) on the same side, saving the opposite side for insertion of another shunting system after the infection has been cured. *One should not leave an infected shunting system in place*, settling for treating the child with intravenous antibiotics, if the infected shunt is functioning. Similarly, if the infected shunt is not functioning, one should not leave it in place, insert a fresh shunt on the opposite side, and treat the child with antibiotics. All foreign bodies (and a shunting system is a foreign body) must be removed when the child has ventriculitis, or cellulitis, or peritonitis! In the event a portion of the shunting system has extruded through the skin, through the umbilicus, or penetrated the intestinal tract, remaining coiled within the lumen of this latter or protruding through the anus, the entire system must be removed and the child put on EVD. Removal of an extruding system is performed by cutting the tubing and withdrawing it from the most contaminated portion, to avoid drawing contaminated or grossly infected tubing through a clean area.

Intracranial Shunting

There are two broad categories of intracranial shunting procedures, *ventriculoventricular* and *ventriculocisternal*. Neither, unfortunately, has proven to be particularly effective, though both have been used extensively in many centers, and repeatedly, over many years.

Ventriculoventricular Shunting

The ventriculoventricular shunt is performed when a portion of the ventricular system, because of a mechanical obstruction, is excluded from the normal cerebrospinal fluid pathways. Examples of this are occlusion of one foramen of Monro, isolating one single lateral ventricle, and aqueductal stenosis, whether anatomicopathological or "functional."

Craniotomy, cerebrotomy, and either cannulation of the obstructed foramen of Monro or marsupialization of the septum pellucidum are the interventricular "shunts" used for occlusion of one foramen of Monro. Unfortunately, for unknown reasons (but possibly because the primary problem is impaired CSF absorption and the foramen of Monro occlusion is only secondary) these procedures are generally unsatisfactory so that sooner or later one finds it necessary to proceed to a shunting operation.

Cannulation of the aqueduct of Sylvius is recommended (only after the child has had a functioning ventriculoperitoneal shunt for several years) by Lapras (C. Lapras, personal communications) for the treatment of aqueductal stenosis. The technique consists of a suboccipital craniotomy, exposure of the IV ventricle, and insertion of a double-flanged Silastic tube through the aqueduct so that the tip and superior flange rest within the III ventricle. The caudal (draining) end and the inferior flange rest in the IV ventricle. It is preferable to insert the Silastic tubing through the stenosed aqueduct without an obturator, to avoid perforating the periaqueductal tissue. Unfortunately, this is not always possible so that one may be obliged to use an obturator.

Ventriculocisternostomies

The best known of the ventriculocisternostomies is the III ventriculocisternostomy. It was used for the treatment of aqueductal stenosis before the introduction of the valve-regulated shunt. The Torkildsen procedure (placement of tubes between the lateral ventricles and the cistern magna) has been used both for the treatment of aqueductal stenosis and tumors obstructing communication between the III and IV ventricles. Fourth ventriculocisternostomy has been used unsuccessfully for the treatment of atresia of the foramina of Luschka and Magendie.

III Ventriculostomy

The III ventriculostomy may be performed either by the open or closed technique.

Open Technique for III Ventriculostomy (Fig. 15.67)

A medial frontal craniotomy and single trapdoor opening of the dura mater are followed by anterolateral retraction of the right frontal lobe, sectioning the olfactory nerve immediately posterior to the olfactory bulb, and identification of the optic nerve. The arachnoid around the optic nerve is opened and the optic chiasm fully exposed. One then passes superiorly, identifying the subcallosal gyrus and the genu of the corpus callosum.

The bulging lamina terminalis may be readily identified between the (inferior) optic chiasm and the (superior) genu of the corpus callosum, by following the subcallosal gyrus posteriorly. Bipolar forceps are used to coagulate the lamina terminalis and gain entry into the III ventricle. Once the lamina is opened, its edges should be coagulated minimally. One may now look directly into the III ventricle, and identify the supraoptic and infundibular recesses, which are separated from one another by the ledgelike bulge of the optic chiasm into the floor of the III ventricle. It is now possible to poke through the posterior floor of the III ventricle, between the infundibular recess and the mammillary bodies, to gain access to the interpeduncular cistern. This is best done with bipolar coagulation, using long-tipped, bayonet microforceps, and proceeding slowly. Do not use laser: the basilar artery fundus may rest along the cisternal surface of the floor of the III, be-

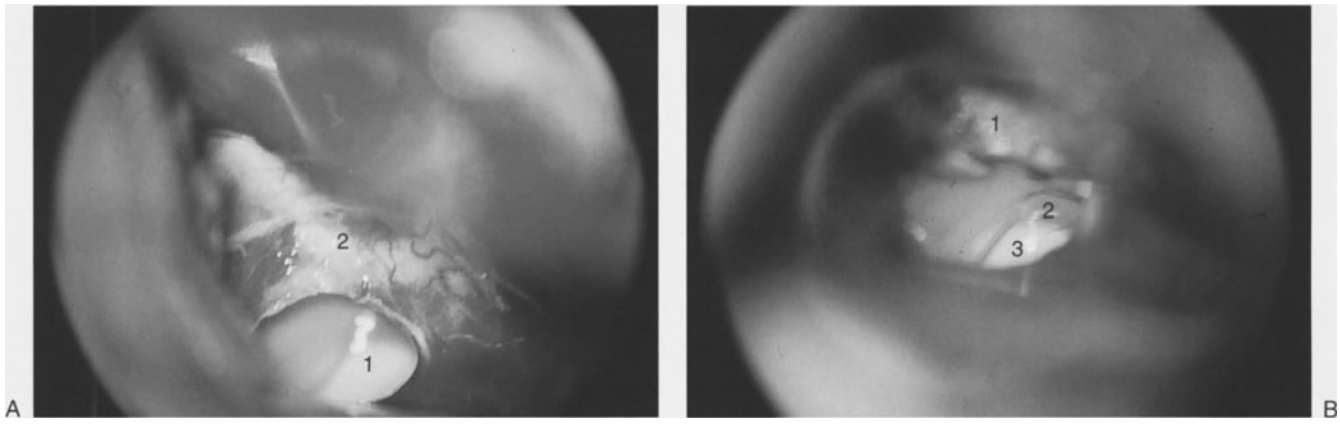


Figure 15.67. The III ventriculostomy, open technique, entails the performance of a right medial frontal craniotomy, dural opening, retraction of the frontal lobe, and sectioning of the right olfactory nerve immediately proximal to the olfactory bulb. The olfactory nerve is followed posteriorly to the optic nerve, and then the optic and supraoptic cisterns are opened. The gyrus rectus on either side is retracted lateralward. (A) After lamina terminalis is opened, one may look into the III ventricle (*1*) and at the optic chiasm (*2*). One should remember that the III ventricle has two anteroinferior recesses: the suprachiasmatic, extending over the optic chiasm, and the infundibular, extending to the region of the base of the pituitary stalk. (B) The opening in the lamina terminalis (*1*) is out of focus because the operating microscope has been adjusted to bring the infundibular recess (*2*) and the posterior floor of the III ventricle (*3*) into focus.

tween the tuber cinereum and the mammillary bodies or these latter and the peduncles.

In hydrocephalus associated with aqueductal occlusion the posterior floor of the III ventricle is extremely thin, so that one may almost see through it into the interpeduncular cistern. If, as recommended by some neurosurgeons, the III ventriculostomy is not performed until a child has had a functioning ventriculoperitoneal shunt for 1 or more years, both the lamina terminalis and the posteroinferior floor of the III ventricle will be normal in appearance, will not be bulging, and, consequently, perforating them will be considerably more difficult. It is recommended, in either event, that perforation of the lamina terminalis and the posteroinferior floor of the III ventricle be accomplished under the operating microscope. Once the posteroinferior floor of the III ventricle has been perforated, the edges of the created ostium should be coagulated to minimize the possibilities of glial overgrowth and closure of the opening.

Closed Technique for III Ventriculostomy (Fig. 15.68)

The closed technique for III ventriculostomy may be performed through a bur hole placed immediately anterior to the coronal suture and 2.5 cm to the right of the sagittal plane. The dura is opened in a cruciate manner after it has been coagulated and the underlying leptomeninges and cortical surfaces are, in turn, coagulated and incised. One may then insert a Silastic tube (with an obturator in place) into the lateral ventricle, removing the obturator when there is egress of cerebrospinal fluid. The Silastic tubing is now advanced inferomedially, passing through the dilated foramen of Monro, into the III ventricle, until the tip comes to rest upon the posteroinferior floor of this chamber. Under X-ray, or CT, control, the obturator is reinserted into the Silastic tubing and then one punctures the membranous floor of the III ventricle, passing the Silastic tubing into the interpeduncular cistern. Use a stereotaxic needle to perform this procedure [65].

Ventriculoscopic III Ventriculostomy

Perforation of the floor of the III ventricle may be effected by utilizing ventriculoscopic techniques. This has been technically very successful though the functional value remains to be documented. The III ventriculostomy by endoscopic methodology is a highly specialized procedure to be performed only by those adequately trained in its indications and intricacies.

It is our opinion that the III ventriculostomy is not adequate treatment for hydrocephalus associated with aqueductal stenosis or occlusion, and that stereotaxic perforation of the floor of the III ventricle – a *blind* procedure – is potentially dangerous.

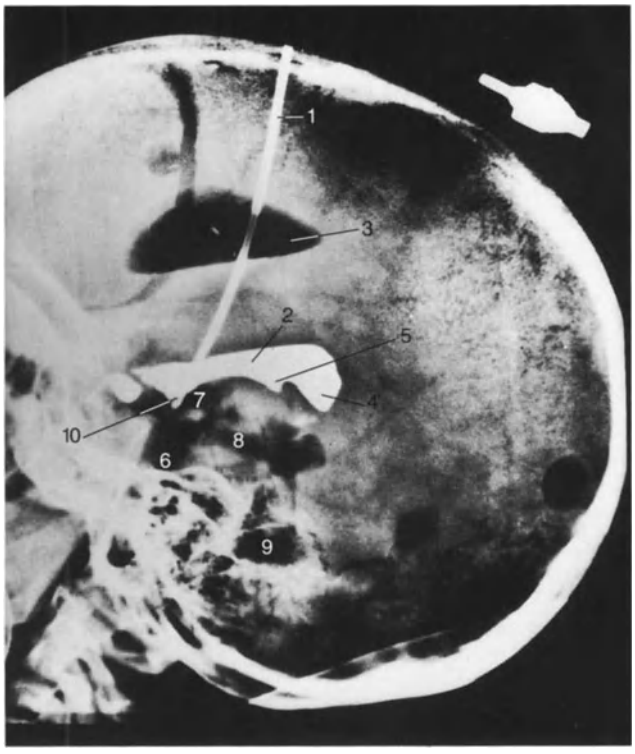


Figure 15.68. Stereotaxic, transventricular, III ventriculostomy. A catheter (*1*) has been inserted into the III ventricle (*2*) through the right lateral ventricle (*3*). One notes the positive contrast media within the inferior III ventricle and, within the chamber, the suprapineal (*4*) and pineal (*5*) recesses. Gas may be seen within the pontine (*6*), interpeduncular (*7*), and ambient (*8*) cisterns as well as within the IV ventricle (*9*). The catheter tip (*10*), under X-ray control, was poked through the floor of the III ventricle and into the interpeduncular cistern, establishing a III ventriculostomy.

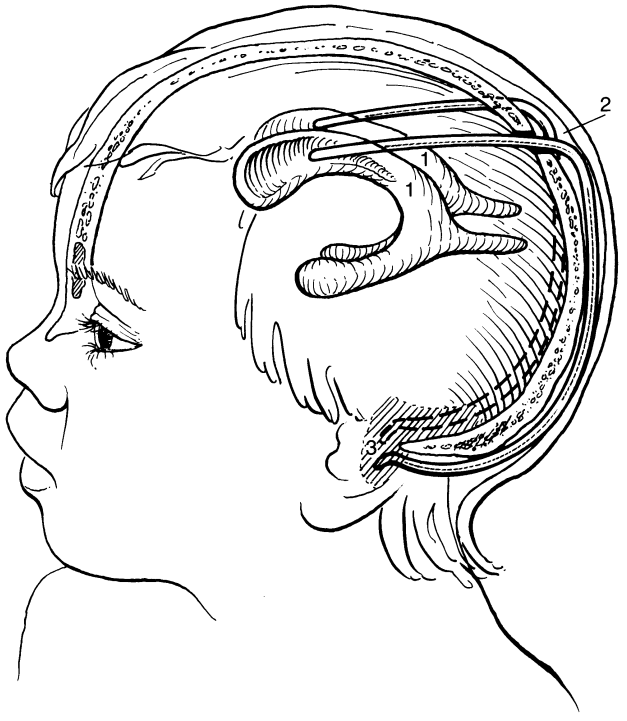


Figure 15.69. Drawing of the skull with the head placed obliquely to illustrate placement of two catheters into the lateral ventricles (*1*) and their passage through the subcutaneous space (*2*) down to the cisterna magna (*3*).

Torkildsen Procedure (Fig. 15.69)

Bypassing the obstructed, midline ventricular system by inserting tubes between the lateral ventricles and the cisterna magna (lateral ventriculocisternostomy) is a major neurosurgical procedure. It was an acceptable procedure many years ago, before shunting techniques were developed to their present state of efficiency. Presently, the author does not recommend the procedure, since it entails placement of two occipital bur holes, cannulation of both lateral ventricles, a suboccipital craniotomy and, often, section of the arch of C1, in order to insert the proximal portions of the tubes into the lateral ventricles and the distal portions into subarachnoid spaces from the cisterna magna down to the level of approximately C2. There is no simple way to revise this system if it obstructs. It is not to be considered an alternative to a ventriculoperitoneal shunt.

IV Ventriculocisternostomy (Figs. 15.70, 15.71)

The IV ventriculocisternostomy procedure has been used in vain in attempts to cure cystic transformation of the IV ventricle since this, the Dandy-Walker cyst, was first described by Dandy. It consists of a suboccipital craniotomy, resection of the ependymopiaglial cyst membrane, and establishing communications between the IV ventricle and the perimedullary and pontocerebellar cisterns. At the end of the procedure one sees the hind cranial nerves, the posterior inferior cerebellar artery, and the vertebrobasilar system. The reason for the failure of this procedure must rest in the fact that the subarachnoid spaces do not permit circulation and/or absorption of the cerebrospinal fluid: the Dandy-Walker cyst is a result of the disease process, not its cause.

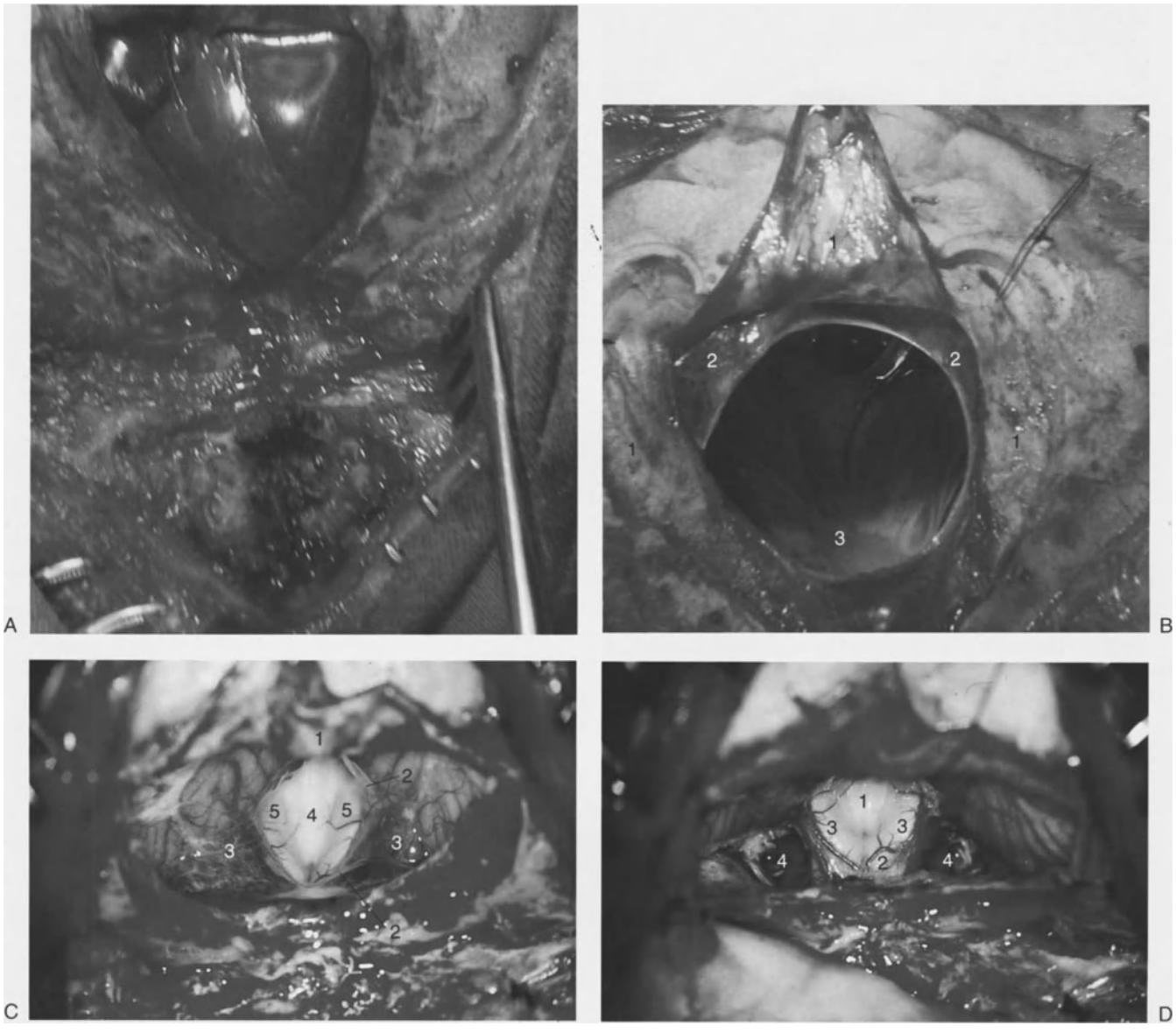


Figure 15.70. The Dandy-Walker cyst should be treated only with lateral *and* IV ventriculoperitoneal shunts. There really is no place for suboccipital craniotomy and establishment of communication between the encysted IV ventricle and basal cysterns in the management of this disease! Though this operation has been performed repeatedly since it was initially introduced by Walter Dandy, no one (including Dandy) has demonstrated its efficacy. These operative photographs are used only to illustrate the anatomical findings, not to suggest that this is an operative procedure which the author recommends. (A) The suboccipital craniotomy and reflection of the dura mater have been performed, revealing the posterior inferior aspect of the cystic IV ventricle. (B) After the dura mater (*1*) has been reflected superiorly and the posterior inferior aspect of the encysted IV ventricle (*2*) opened, one looks directly at the floor of the IV ventricle (*3*) and the enormously dilated aqueduct of Sylvius. (C) This is another child. The dura (*1*) has been reflected superiorly and the posterior inferior aspect of the encysted IV ventricle (*2*) completely resected. There are extensive arachnoidal adhesions (*3*) extending from the lateral medullary cisterns to the surface of the cerebellar tonsils. One notes the floor (*4*) and lateral walls (*5*) of the IV ventricle. (D) The colliculus faciallis (*1*) and the hypoglossal triangle (*2*) of the floor of the IV ventricle, as well as the lateral walls (*3*), are glistening white. There is no inferior vermis. Communication between the IV ventricle and the lateral medullary cisterns (*4*) has been established.

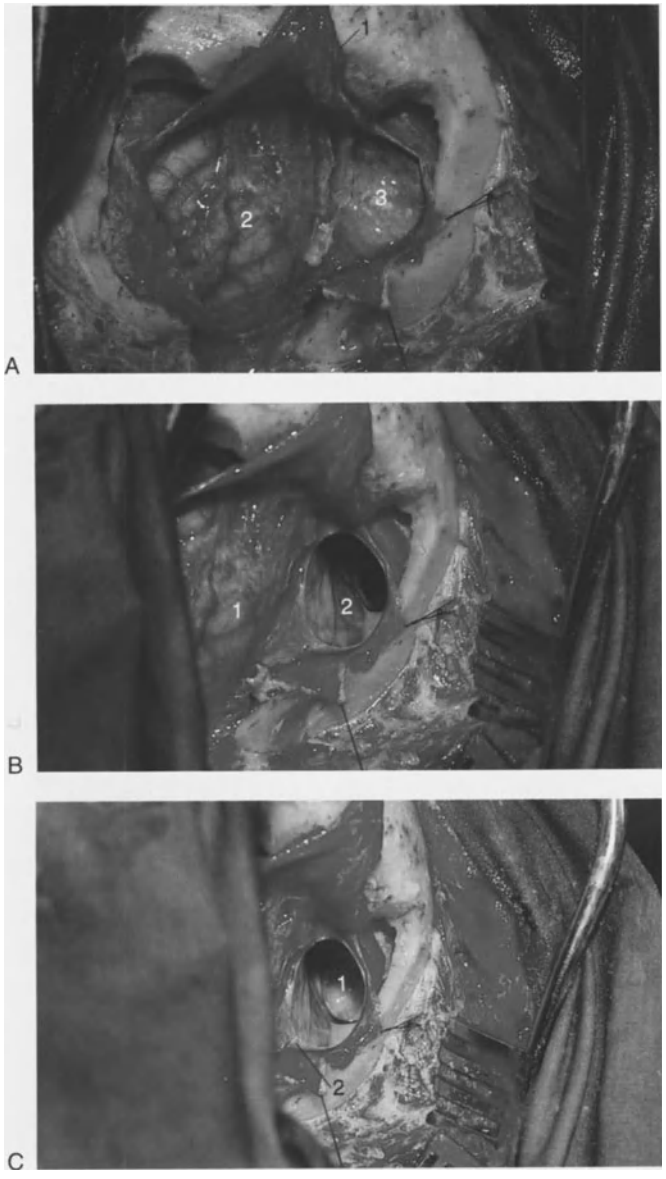


Figure 15.71. Though unusual, the encysted IV ventricle may expand lateral to one of the cerebellar hemispheres, rather than in the midline along the vallecular opening into the void left by the dysplasic inferior cerebellar vermis. When this happens, the cerebellar hemispheres are displaced to one side, with the clinical picture being that of a cystic cerebellar hemisphere tumor rather than a congenital anomaly. (A) The inferior cerebellar triangle craniotomy has been performed and the dura (*1*) sewn out of the way superiorly. The cerebellar hemispheres (*2*) are displaced from right to left by the cystic transformation of the IV ventricle. (B) The posterior wall (arachnoid-pia-glia) of the posterior lateral extension of the cystic transformation of the IV ventricle has been opened, exposing the underlying normal surface of the right cerebellar hemisphere (*1*) and the most anterior extension of the cyst wall (*2*). (C) The anterior wall of the cyst has now been resected, exposing the enormously dilated lateral recess of the IV ventricle (*1*) and the atretic foramen of Magendie (*2*).

Basic Structure of Shunt Systems

In this, the MRI era, one must not use shunt systems which have metallic components. The following devices are described because they have been in use since before the MRI era, and both to discourage the reader from using those with metallic components and to alert the reader to their presence.

The basic structure of the first valve-regulated shunt systems (Fig. 15.72) used consisted of three functional parts: (1) ventricular catheter, (2) reservoir/valve, and (3) peritoneal/atrial catheters. Subsequently, improvements evolved into two-piece functional systems, joining proximal and distal catheters at a single connector, and then into a one-piece system to provide maximum simplicity and efficiency.

1. Ventricular catheters (Fig. 15.73): For ventricular catheterization, ordinary and spring armored catheters are available. Barium loaded and spring armored catheters are useful for radiographic visualization with plain craniography. Previously described artifacts ascribable to them are not as disturbing as once considered, but they no longer have a role in that they render MRI dangerous.
   Flanged catheters, designed to be advantageous in that they are not obstructed by the choroid plexus or the ventricular walls, are liable to become *unremovable* at the time of revision. The X-ray marked, at 1-cm intervals, catheters are convenient for insertion into the ventricle.
2. Flushing devices: Theoretically flushing devices are classified into two types: i.e. one which functions as a reservoir (Fig. 15.74) and the other which, in addition to the above function, has a pressure-controlling valve (Fig. 15.75). The hoped-for objectives for placing a reservoir are the ability to evaluate shunt patency and to permit sampling of the cerebrospinal fluid. However, the reliability of the evaluation of shunt function by palpating the reservoir is as low as 60% and, though a reservoir permits CSF sampling when a shunt infection is suspected, puncturing it carries very real risks of introducing infection. Therefore, serious doubts exist concerning the placement of a reservoir for these purposes. Basing clinical decisions upon the tactile impressions received from compressing a flushing device is discouraged, puncturing it is ill-advised.
   For the flat bottom type device, the twist drill hole fits well, but, with the bur hole type device, better fixation of the reservoir can be achieved; consequently, pressure upon the skin may be reduced. In neonates and infants, since the placement of a reservoir may cause tension in the skin and eventually lead to skin necrosis, it is desirable to use a right-angle tube, connector, or, preferably, Unishunt so as to avoid the use of a reservoir. Recently, a reservoir

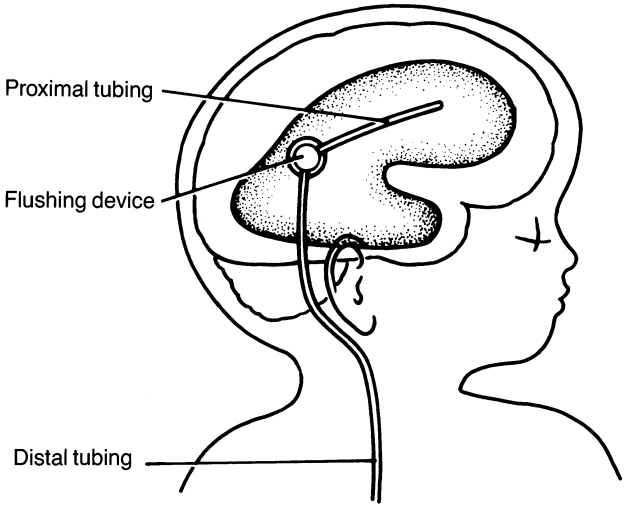


Figure 15.72. Basic structure of CSF shunt system.

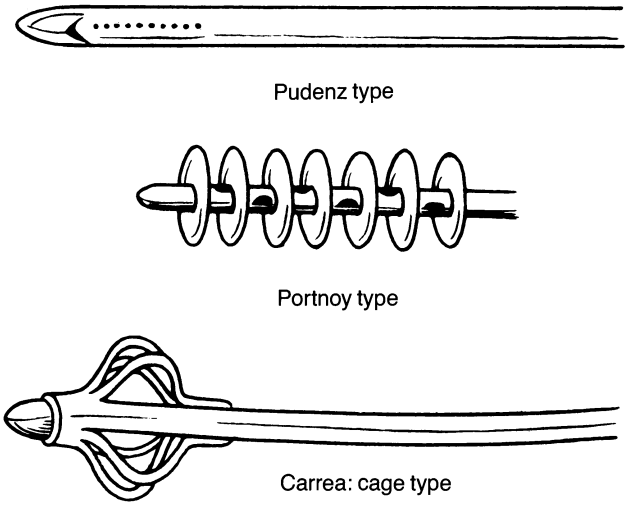


Figure 15.73. Various types of ventricular catheter.

with a smaller outside diameter so as to exert less pressure on the skin has been produced for use in infants.

3. Peritoneal catheter (Fig. 15.76): Regarding peritoneal catheters, the spring armored kink-proof and hard king-resistance types are available. The kink-proof catheters are not kinked within the abdominal wall and can be used safely.
   The tip of the catheter is equipped with a slit valve. According to the length of the slit, the valves are classified into three types: i.e., low, medium, and high pressure valves. When the reservoir has a built-in pressure control valve, a peripheral catheter with a low pressure valve is used. When a reservoir has an antireflux valve, an open-ended catheter may be used.
4. Other devices: With the hope of preventing excessive CSF drainage due to the "siphon effect" in the sitting or standing positions, antisiphon devices and on-off valves to control the intracranial pressure (Fig. 15.77) are sometimes used. Shunt filters (their efficacy is doubtful) to prevent dissemination of tumor cells into the abdominal cavity have been recommended for use in children with malignant pineal tumors.

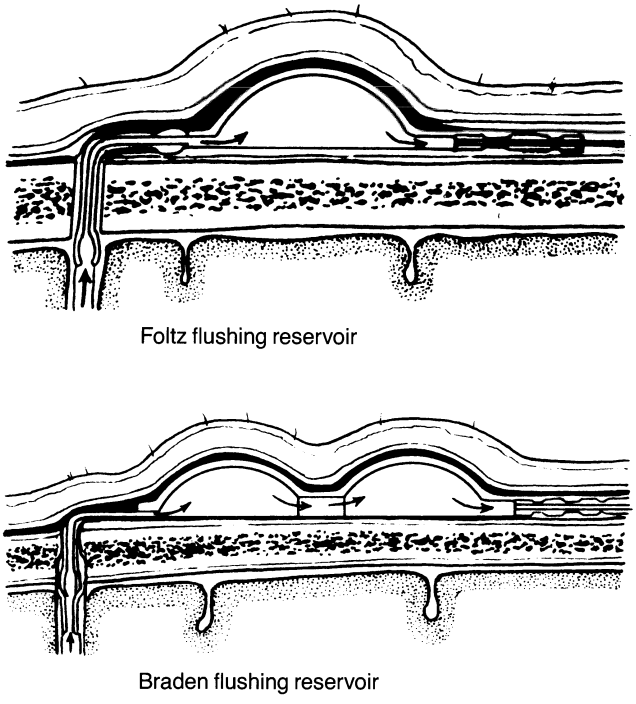


Figure 15.74. Various types of ventricular catheter.

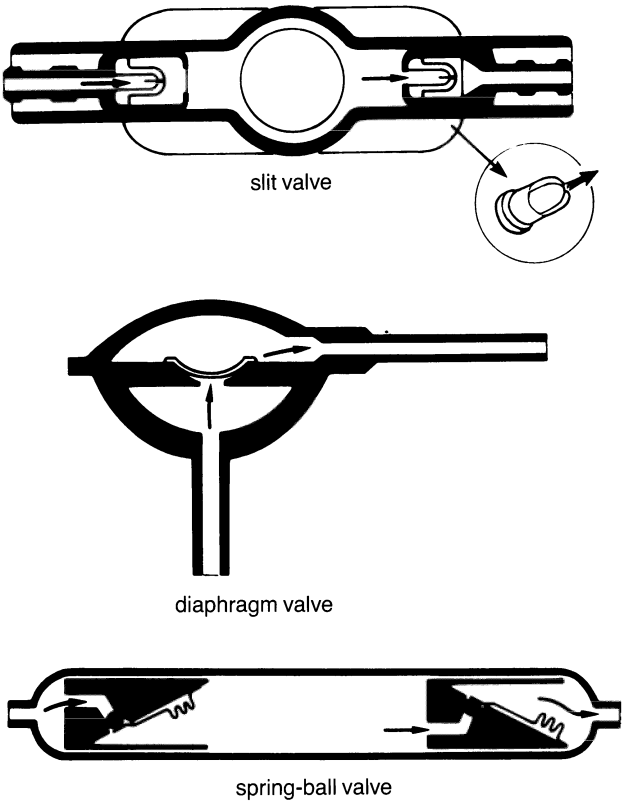


Figure 15.75. Various types of pressure-controlling valves.

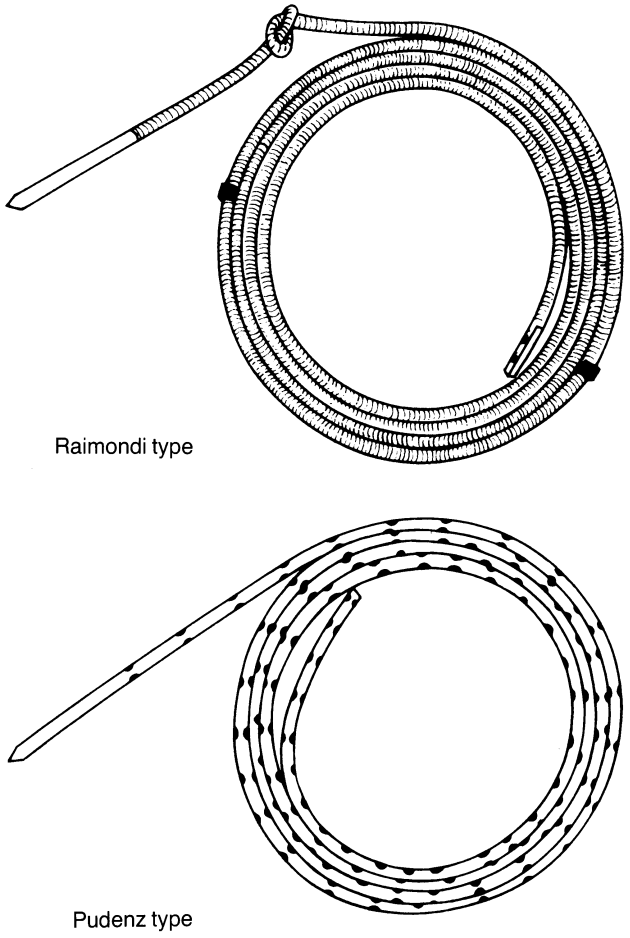


Figure 15.76. Various types of peritoneal catheters. These are entirely extracranial, so they do not present a risk to the patient. However, they do cause unacceptable artifact.

Characteristics of the Flow Rate

The drained CSF volume (ml) per minute is shown in the pressure-flow rate (resistance) curve (Fig. 15.78). In interpreting this curve, it is important to compare the pathological features of the patient reflected in this curve with the CSF production per minute (approximately 0.35 ml/min) and the CSF pressure (approximately 40–110 mmH_2O) in normal infants and children. When the CSF pressure is more or less normal, the flow rate at each pressure does not vary greatly. However, when the CSF pressure is high, in some systems the flow rate at the low pressure is greatly increased, as compared with those at medium and high pressures.

The pressure flow-rate curve is obtained by plotting the flow rates at various pressure differences examined by using two water baths (Fig. 15.79). The different curve profiles depending on various systems are ascribable to differences in the pressure control valve, and, on the basis of Poiseuilles's law (Fig. 15.80), on the diameter and length of catheters used in each system.

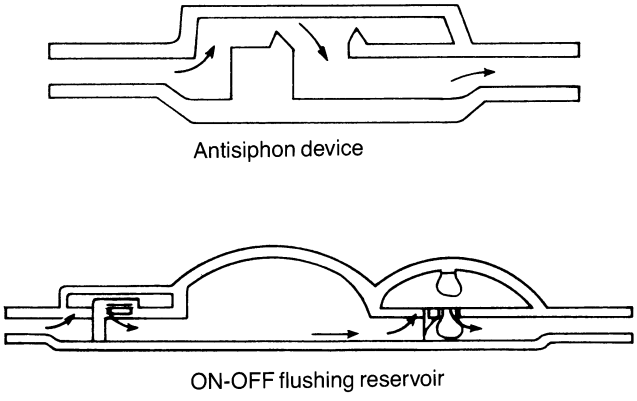


Figure 15.77. Other devices.

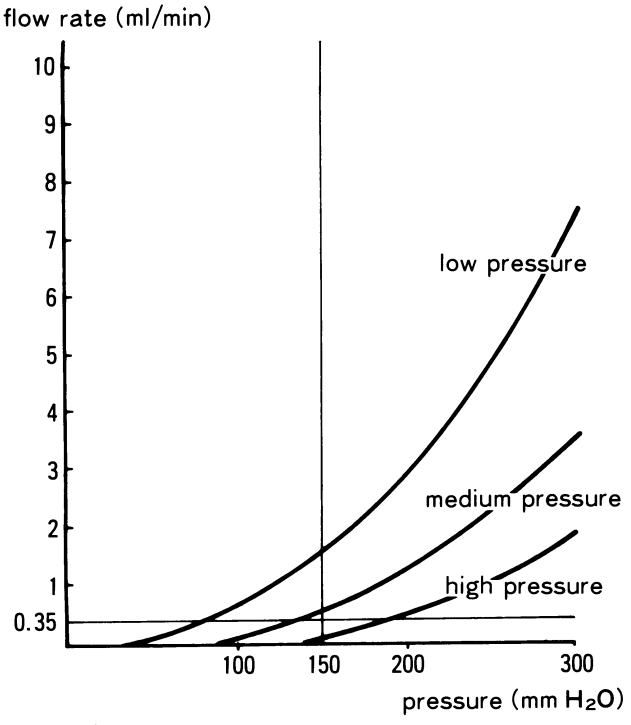


Figure 15.78. Pressure-flow rate curve.

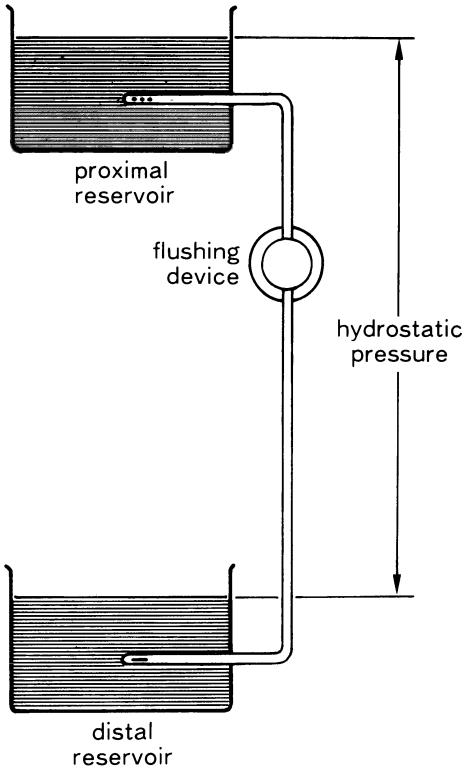


Figure 15.79. Measurement of the flow rate of shunt systems.

$$F = \Delta P \left(\frac{\pi}{8}\right)\left(\frac{1}{n}\right)\left(\frac{r^4}{l}\right)$$

F = volume passing through the tube each second
P = pressure difference between the inlet and outlet
n = viscosity of the fluid
r = radius of the tube
l = length of the tube

Figure 15.80. Poiseuille's law.

Opening Pressure and Closing Pressure

The opening and closing pressures can be measured by introducing small air bubbles into the shunt system. Having adjusted the water surface levels of the water baths at the ventricular (proximal) and peritoneal (distal) ends to be equal, the distal water surface level is gradually lowered. The opening pressure is indicated by the difference in the water surface level between the two baths at the point when the air bubbles begin to move. Similarly, the closing pressure is indicated (when the water surface level at the peripheral side is gradually elevated) by the difference in the water surface level between the two baths at the point when the air bubbles cease to move.

The ideal shunt system is one which normalizes the elevated intraventricular pressure without collapsing the ventricular system, thus facilitating CSF drainage without causing overdrainage or underdrainage.

Since the pathological features of hydrocephalus and the anatomophysiology of children vary among cases and age categories, a shunt system which functions satisfactorily in all patients has not yet been developed. Shunt system selection, therefore, is somewhat subjective and this varies from neurosurgeon to neurosurgeon. Since shunt systems are permanent, it is desirable to select one which will cause fewer complications and will be easily revised in case a shunt obstruction occurs. Therefore, the system should be simple!

Currently, three types of valves are available (Fig. 15.75). In slit valves, the flow rate is liable to change when the intraventricular pressure changes, while in spring-ball valves and diaphragm valves the low rate theoretically is maintained at a constant level, even if the intraventricular pressure and the viscosity of CSF change (Fig. 15.81). These types of shunt systems are thought to be useful, for example, in cases of hydrocephalus secondary to meningitis, where the CSF protein levels are elevated and shunt occlusion is common. However, these two types of valves have been considered by some to be subject to the so-called "siphon effect," and overdrainage has been reported to occur when patients are in the sitting and standing positions (Fig. 15.82).

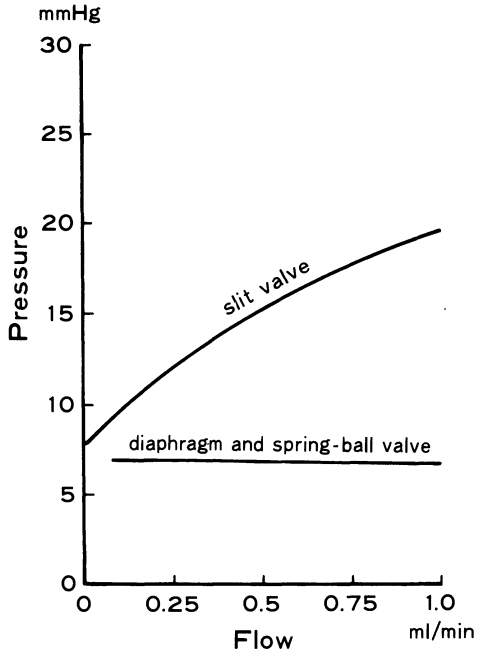
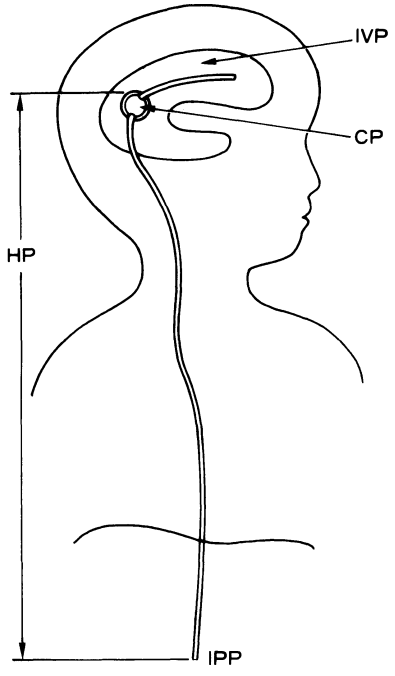


Figure 15.81. Types of valves and pressure-flow curves.



$P = IVP + HP - (IPP + CP)$

P = perfusion pressure
IVP = intraventricular pressure
CP = valve closing pressure
HP = hydrostatic pressure
IPP = intraperitoneal pressure

Figure 15.82. Factors determining perfusion pressure of CSF shunt system.

It is desirable to control the pressure of the shunt system with a proximal valve. When more devices are connected to the system, a correspondingly higher resistance results and shunt function is likely to become inadequate. Consequently, when a distal slit valve is used for the prevention of regurgitation, it is preferable to employ a low-pressure valve to reduce the resistance to the minimum level.

Concerning "programmable" valves, up to the present time there have been no reported series documenting their equally satisfactory effectiveness, much less one documenting superiority. They give the surgeon, and the patient, a false sense of security because of their "high-tech" characteristics, and they are expensive.

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