Pediatric Neurosurgery

Anthony J. Raimondi

Pediatric Neurosurgery

Theoretical Principles Art of Surgical Techniques

With Chapters on Pediatric Neuroanesthesia
and Neurologic Intensive Care by
Badr A. Ishak and Zehava L. Noah

With 1096 Illustrations, 214 in Color

Springer Science+Business Media, LLC

Library of Congress Cataloging-in-Publication Data
Raimondi, Anthony J.
Pediatric neurosurgery.
Includes bibliographies and index.
1. Nervous system—Surgery. 2. Children—Surgery.
I. Ishak, Badr A. II. Noah, Zehava L. III. Title.
[DNLM: 1. Neurosurgery—in infancy & childhood.
WL 368 R153p]
RD593.R27 1987 617'.48 87–20800

Originally published by Springer-Verlag New York Berlin Heidelberg in 1987
Softcover reprint of the hardcover 1st edition 1987

Federal Republic of Germany.

9 8 7 6 5 4 3 2 1

ISBN 978-1-4757-4204-6 ISBN 978-1-4757-4202-2 (eBook)
DOI 10.1007/978-1-4757-4202-2

To my son Paolo,
who lived his entire life in childhood

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

Contents

Chapter 10

"Better is a dinner with herbs where love is, than a stalled ox and hatred therewith."

SOLOMON
Kings– The Holy Bible

Preface

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 specialty 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 specialty 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 identified. 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 Neuroradiology: "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 cognition. 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.

Acknowledgments

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.

"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

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.

Introduction

From working exclusively with patients less than 18 years of age through performing operations to treat congenital diseases, first the pediatric surgeon and most recently the neurosurgeon identified the striking differences between adults and children, tailoring their care to special diseases and anomalies. Subsequent specialization in pediatric neurosurgery has brought the student of this discipline to defining the field in which he works, and to developing concepts and techniques specific to the problems at hand.

One approach to collecting all current aspects of pediatric neurosurgical problems into a single text, dealing with them as an holism[1], is to identify individual age categories as anatomophysiologic phases during which certain pathologic processes (congenital, developmental, neoplastic, etc.) occur, and then present these latter as integral clinical entities before proceeding to suggest detailed surgical indications and techniques. That is done in this text, which expresses the author's readings, experiences, perspectives, and spatial (I like to refer to it as the use of the parietal lobe in formulating motor) concepts, as regards disease identification and surgical technique.

It is hoped that the reader will *study* this book—holistically, even when using it as a *reference* source to obtain an answer to a single question. To facilitate the realization of two *seemingly* incompatible goals, the presentation of both a comprehensive text and a reference source, the theory of each entity is presented first, then the indications for surgery and what one may expect to accomplish by operating are described. The details of surgical technique embrace these theoretic concepts spatially, initially with general principles and later, after each pathologic entity is described, with operative details specific to the disease.

The structure of the book, designed to permit this transfer of perspective, consists of sections dedicated to the three age groups encompassing

---

[1] The tendency in nature to form wholes that are more than the sum of the parts by creative evolution. (*The Concise Oxford Dictionary of Current English,* H.W. Fowler and F.G. Fowler, Eds. Oxford University Press, 1964.)

true Pediatric Neurosurgery: newborn, infant, and toddler. I do not see significant differences between the juvenile/adolescent on one hand and the adult on the other to justify particularization of these age categories, either with regard to discrete neurosurgical pathologic entities or the execution of technical maneuvers. Of course, one may identify a factor, postulate a consideration, but there are not presently known generalities of sufficient moment to allow conclusions that apply only to the juvenile/adolescent. In fact, there are no isolated characteristics so specific as to defy effective transfer of concept and art from the realm of adult neurosurgery—other than in areas (cervical fractures or dislocations, scoliosis, etc.) universally managed by orthopedic surgeons, or those (craniofacial anomalies) managed primarily by highly specialized teams directed by plastic surgeons. The juvenile/adolescent age category is characterized by linear and volumetric body growth, and by endocrine changes that preceed and accompany such structural changes as air sinus development, musculotendinous pull on bony insertions, and refinement of psychomotor skills. The infrastructure of a specialized surgical discipline—discrete anatomophysiopathologic characteristics of a specific organ system—does not exist. Therefore, the presence of juveniles/adolescents on a pediatric or adult neurosurgical unit is one of convenience: more comfortable and less difficult for juveniles to be placed with younger children, adolescents with adults. Hence, in this work, the diagnosis and management of juvenile/adolescent neurosurgical clinical entities reflects a Janus effect, looking simultaneously in two directions: at adult pathology and the changing anatomical framework of a growing child.

Much attention is herein given to positioning children for surgery, to considering the positioning of the surgical team and its instruments, and to allowing immediate and complete access of the anesthesiologist to the child. Two separate chapters emphasize anesthesiologic and neurointensive care considerations, work for which I am extremely grateful to Drs. Badr A. Ishak and Sehava L. Noah. The state of the art with regard to medications and specific monitoring techniques may change, and rapidly, but the basics, those essential to a neurosurgeon to permit him to work with and depend on the anesthesiologist, at least in light of what is foreseeable today, should certainly apply as long as anything else in a book such as this.

Conceptualization of the target area for surgery is the essence of positioning the child *supine* for approaches to the anterior fossa, the parietal and parasagittal areas, and the middle fossa; *prone,* for occipital and some areas of suboccipital craniotomies, and for laminectomies; and *lounging,* for mid and superior cerebellar triangle lesions approached through suboccipital craniotomies. The theory and techniques for skin incisions, suturotomy, craniotomy, and laminotomy are correlated with dural openings and cerebral retraction, emphasizing utilization of cisterns and sulci, avoidance of major vessels, exploitation of gravity, and three-dimensional movement of the operating table and the surgeon's chair. Photographs and transparency drawings represent the medium for conveying these concepts. Similar iconography is used to describe cerebrotomy and cerebral resection for access to deep parenchymal and intraventricular lesions, for exposure of masses medial to the limbic lobes, for lobectomy. Scalp, bone, dural, cortical vein, and dural sinus; capillary and arterial; parenchymal; and tumor bleeding are treated individually, as are the technical aspects of using bipolar thermocautery instrumentation, topically applied protein derivatives, and synthetic fibers; the preparation and application of instantly hydrated cot-

ton "fluffies" to occlude opened vessels or to identify persistent bleeding sites for mechanical or thermocautery sealing.

I am of the firm opinion that one cannot be a master pediatric neurosurgeon unless he has already mastered adult neurosurgery. Otherwise, deep parasellar and pineal region tumors, vascular anomalies, craniovertebral junction and vertebral column pathology, complex craniocerebral and vertebrospinal neurotraumatology, all lie within the epidemiologic and diagnostic purview of the pediatric neurosurgeon—but beyond his touch.

In the text, the reader will find tumors arranged and dealt with along lines of specific age categories and anatomical location, with only indicative value or attention being given to histologic appearance. Concerning prognosis, great attention is given to the anatomical location/age factors at time of diagnosis. What, it is hoped, stands out is the spatial concept in localizing and operating the tumors, the means by which as total a resection as possible may be accomplished, and—with the sole exception of medulloblastoma—the limited, very limited, value total resection has in affecting favorably the outcome. The compounding effects that tumor expansion and complicating hydrocephalus have both on the child and on the ease of surgical resection are emphasized. Thus, the prognostic importance of separating mid-line from hemispheral—supra or infratentorial—tumors stands in relief, as do the decision to insert a preoperative shunt and the election of either unilateral or bilateral systems.

The reader will not have misunderstood me if he concludes that, within limits, total resection of craniopharyngioma is more a matter of the surgeon's perspective that a pathoanatomical reality; histologically favorable pineal region tumors are ideally managed with as complete resection as possible and Röntgen therapy; arteriovenous anomalies of the Galenic system are very different clinical entities in different age categories and their total obliteration is not essential to a very favorable outcome in children beyond six months of age.

The range of post-traumatic pathology imparts particular, one might even say intriguing, interest to this section, since so little is known of the developmental pathology of the injured brain during different stages of life. Consequently, there are many commonly used operations, all very different one from the other, to treat the chronic subdural, normal pressure hydrocephalus, post-traumatic ependymal or arachnoidal cysts. The ubiquitous CT scanner has suggested to some clinicians to withhold surgical treatment of epidural hematoma, albeit in relatively asymptomatic children, following a wait-and-see course. Increased experience has resulted in almost all clinicians giving little or no value to routine skull x-ray films. Certainly, such ill-founded, drastic, aggressive, antiphysiologic treatment protocols as barbiturate coma and prolonged artificial ventilation have proven themselves to be of no positive value and, coupled with the combination of intracranial pressure monitoring/megadose steroids/mannitol, even of negative value. What better example than this could one want to demonstrate the dangers of transferring directly adult *experimental* therapeutic regimens to the *routine care* of children? What more complicated and misleading clinical evaluation protocol could one imagine than applying the Glasgow Coma Score to children under two years of age?

The incredibly profound, imaginative, technically audacious studies of Paul Tessier (a plastic surgeon) and Jacques Rougerie (one of the earliest and best pediatric neurosurgeons) in craniofacial surgery have resulted in extremely significant, predictable, consistent contributions

to Pediatric Neurosurgery. We now have, though certainly not without some limitations, the theoretic basis for comprehending much of the dynamic pathology of the complex craniofacial dysostoses, and the understanding upon which to predicate bone reconstruction. The facial work is entirely within the realm of the specialist in plastic surgery; hence, it is dealt with schematically in this text. The cranial work is dealt with more extensively, as these dysostoses and premature suture closures are diagnosed in most of the Western world during the newborn and infancy periods. Therefore, suturectomy is sufficient for scaphocephaly, and suturectomy and bony reconstruction for plagiocephaly and trigonocephaly. Of course, if the skull deformity is not brought to the attention of the surgeon until the juvenile age, extensive procedures—totally unnecessary in infancy—may be indicated. The more interested student of this subject will not find satisfaction in this text: craniofacial dysostoses and complicated craniocerebral disproportions are an area of specialization that demand special training and total dedication.

Will the future oblige us to say the same about craniofacial encephalomeningoceles? Surely, at the present time we continue to approach most cerebral (ependymal or arachnoidal) cysts, cranial encephalomeningoceles, and cranioschisis mechanistically. The CT scanner and Magnetic Resonance Imaging have made their specific diagnosis straightforward. We must now determine whether the simple presence of an otherwise nonpathogenetic lesion can justify surgery, whether the expected results of operating severe craniofacial or craniocerebral encephalomeningoceles are ethical, and whether Society will tolerate the pediatric neurosurgeon who, in addition to acting as clinician and surgeon, also acts as moralist, ethicist, and philosopher.

Nowhere in Pediatric Neurosurgery is this more obvious, more painful, more definitely asserted and bitterly contested than in the management of children who are victims of the dysraphic state: between neurosurgeon and the community for *spina bifida aperta,* and between neurosurgeon and neurosurgeon for dysraphic hamartomas. Herein, an effort is made to present the different clinical entities in a comprehensive manner, correlating surgical technique with fundamental anatomopathology, with the hope that the reader will evaluate the clinical problem in light of what degree of damage is permanent, whether in fact damage may *ever* occur, and consider the very real advantages of following carefully a child with an anomaly that is only potentially pathogenetic. No amount of words, in whatever form or spirit presented, can become decisively influential because we are scientists *and* humanists.

The length of time—more than one hundred years—since first used, and the variety of shunts available, at present indicate both that the very best, if not the only, treatment for hydrocephalus is a shunting procedure, and no system has yet been devised that *cures* the hydrocephalus. In fact, in some children it may be necessary to shunt, revise the shunt, change the shunt, use another type of shunt or another drainage site, and then to begin all ever again. However, where else in our specialty does one obtain as good overall results, consistently, than in the management of hydrocephalus with shunting systems? Is a *valve* necessary?

Pediatric Neurosurgery

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

SAADI
The Fruit Garden

Chapter 1

Positioning

It is not realistic to specify the single most important aspect of an operative procedure, namely, diagnosis, anatomic localization, blood 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 and will always have the lesion at the center of his operative field.

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 owing to their uniform size, the constant relationship between brain and skull, and the lack of anatomical considerations such as open fontanels 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, all of which assume primary importance. Therefore, this chapter is organized to present to the reader general and specific considerations concerning age, individual body positions, relative position of surgeon vis-à-vis patient, and positioning of the head. Thereafter the recommended positions for specific operative procedures are discussed before they are described.

General Discussion

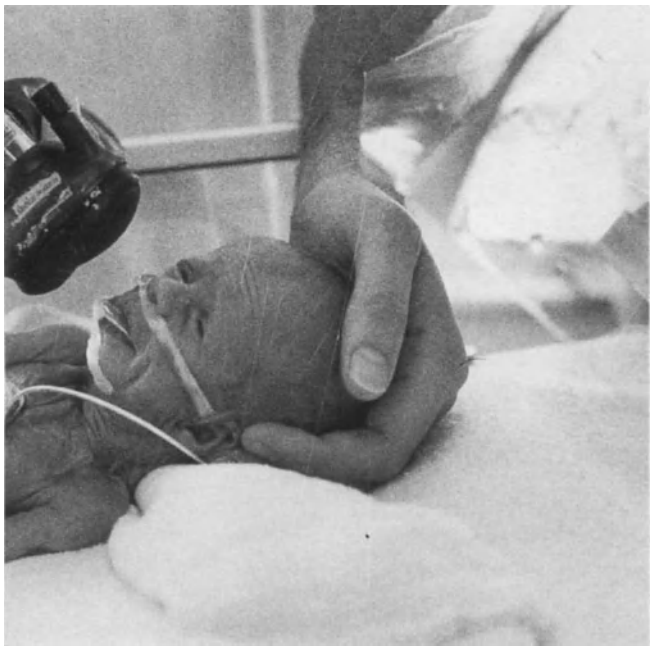
Age

The relative sizes of the surgeon's hands and the head of the newborn, infant, toddler, and adolescent put 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.

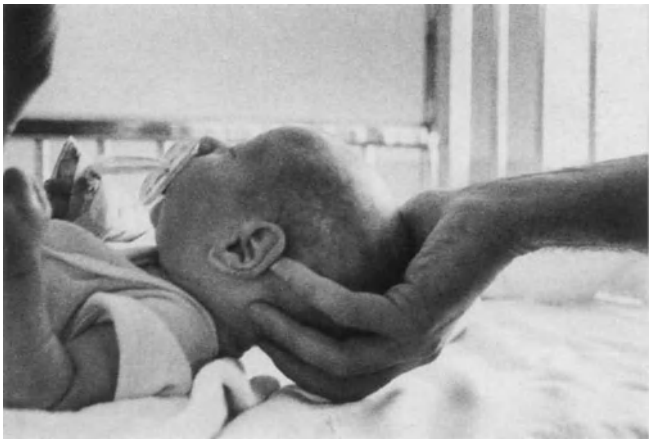
The head of a premature newborn may be so small as to fit within the palm of the surgeon's hand (Fig. 1.1 A), whereas that of a term newborn rests comfortably within the fully cupped adult hand (Fig. 1.1 B). The heads of the infant and toddler (Fig. 1.1 C, D) are proportionately larger. The same hands 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 fontanels and narrowing of the sutures.

Premature Newborn

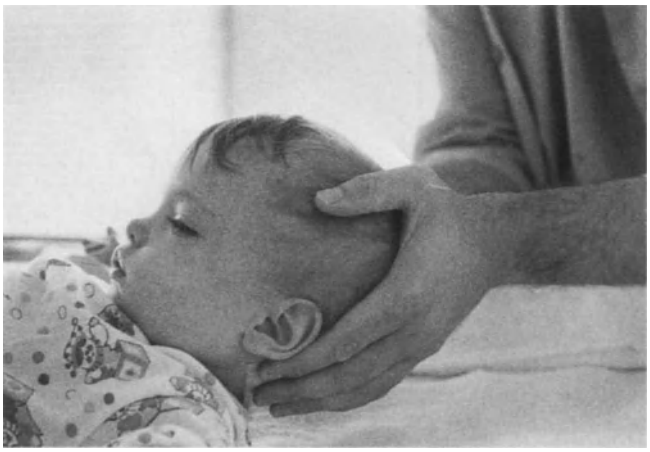
For all intents and purposes, and with only the rarest exceptions, neurosurgery on the brain of the premature newborn is limited to placing an external ventricular drain or inserting a ventriculoperitoneal shunt. Consequently, the supine position, with the head turned to



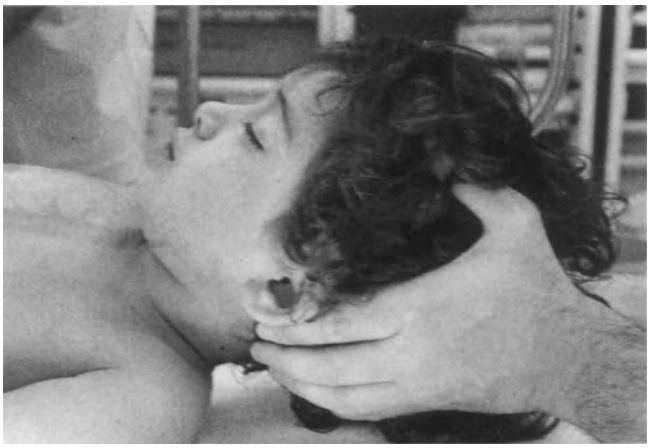
A



B



C



D

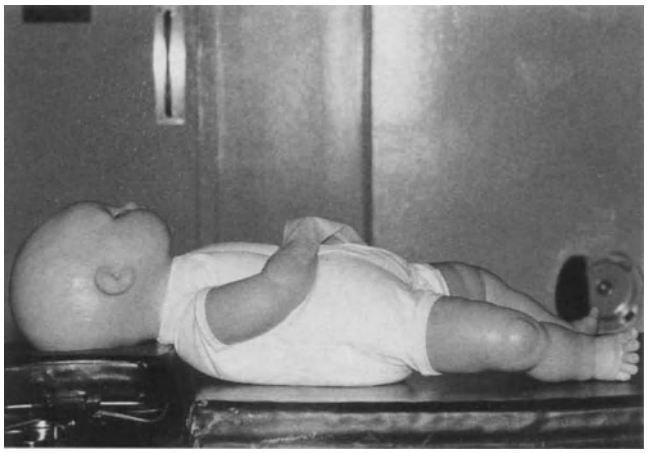
Figure 1.1. (A) Premature newborn. (B) Term newborn. (C) Infant. (D) Toddler.

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.

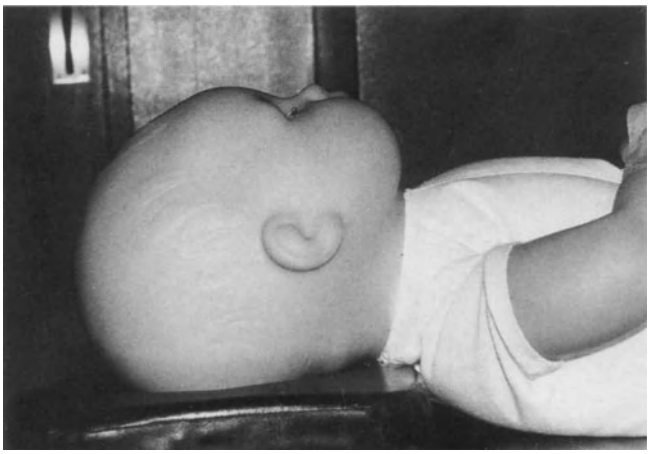
Term Newborn and Infant

The term newborn and the infant can suffer the full range of neurosurgical diseases, so that it may be necessary to operate on children in these age ranges in either the supine, the prone, or the sitting positions. Although the sitting position in the newborn is extremely difficult to maintain (he 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 if at all possible. The anterior fossa, orbits, frontal and parietal lobes, and metopic suture may all be exposed with the newborn or infant supine and his 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 hemispheral convexity (Fig. 1.3A, B, C). 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, C) 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 exposure 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

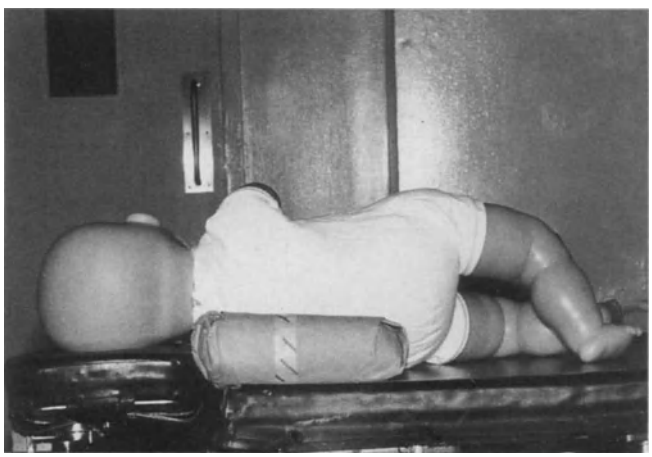


A

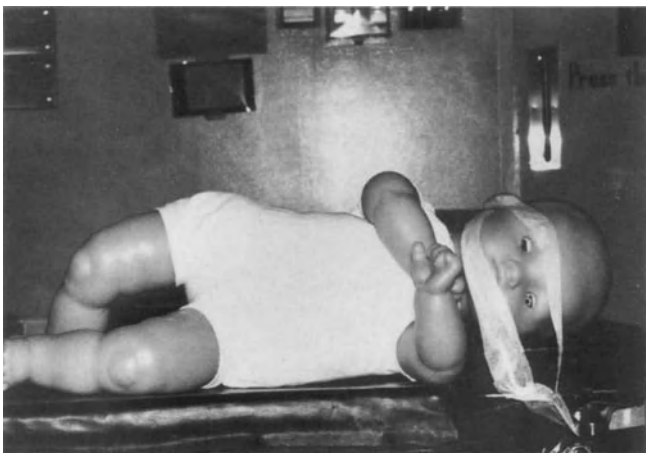


B

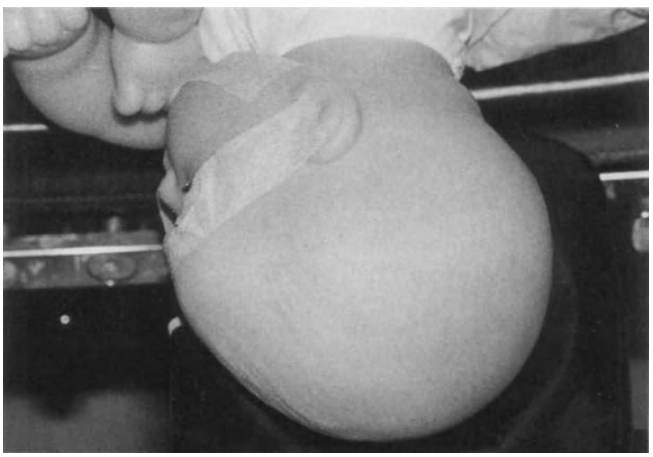
Figure 1.2. (A) The body is positioned supine, with the arms taped loosely across the thoracoabdominal junction. This permits ready access to the chest wall and avoids compression of the abdomen. (B) This is a view of the head's position when the child is supine. The head is neutral. One may either flex 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.



A

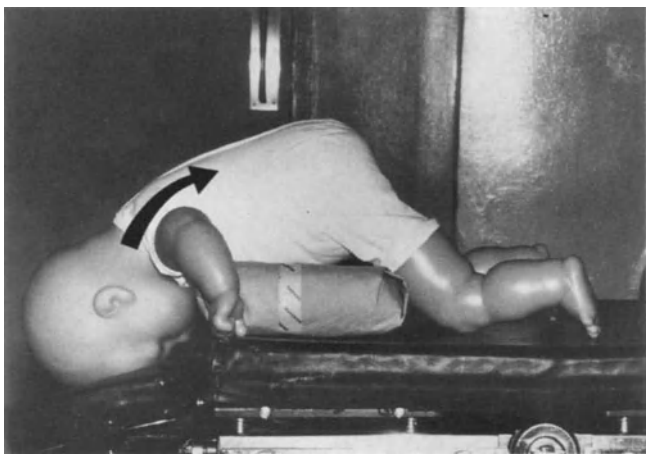


B

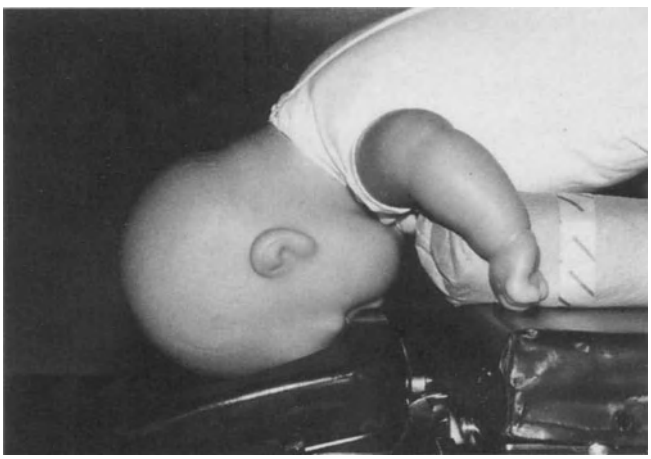


C

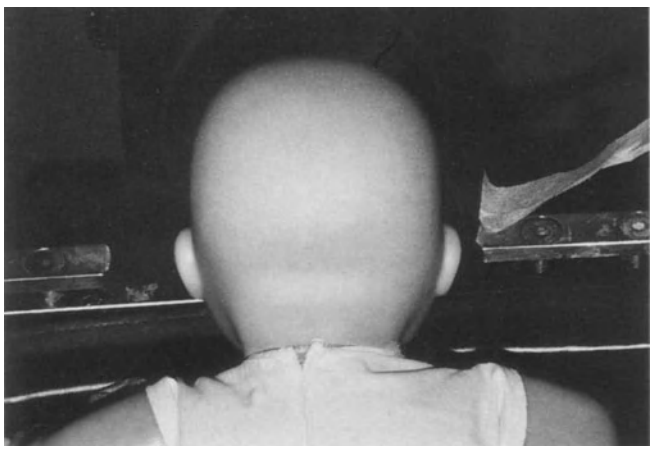
Figure 1.3. (A) The supine newborn may be rotated from his back to his side and held firmly in position with nothing more than a single sandbag, thereby bringing him into the lateral decubitus position. (B) The arms are folded across the shoulders. Tape suffices to fix the head firmly in the desired 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. (C) This is an *en face* view of the lateral surface of the neurocranium with the newborn turned partially onto his side from the supine position. The head has been angled downward so that the sagittal plane of the skull is at a 7°–10° angle to the coronal plane of the body. The head is *not* hanging, the apparent pendant position is an optical illusion resulting from the two-dimensional limitation of photography. This projection allows one to appreciate the surgeon's view of the lateral surface of the head.



A



B



C

Figure 1.4. (A) Access to the cervical spine, the craniovertebral junction, and the occipital lobes may be had 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 degree of flexion of head on neck. Note that the frontal eminences, not the face, nestle into the headrest. (C) This is the surgeons's view of the child's occipital and posterior parietal areas when positioned prone. Again, the pendant position of the head is apparent, not real, a result of the two-dimensional limitations of photography. True perspective may be appreciated by looking at the lateral view in either Figure 1.4B or Figure 1.5.

when performing a suboccipital craniectomy for decompressing the foramen magnum in children with the Chiari II malformation. *One is never 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 patient prone.* It is also difficult to gain a direct line of vision to the superior cerebellar triangle in the newborn 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 by lowering and extending his head.

Toddler

The toddler may be put safely and effectively into either the sitting or lounging positions because his trunk is long enough to sit him up, and his 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 second year of life. Such lesions as medulloblastoma, primitive neuroectodermal tumors of the superior cerebellar vermis, arteriovenous malformations of the Galenic system, pineal tumors, arachnoidal cysts of the quadrigeminal and superior cerebellar cisterns, all occur with increasing frequency in this age category. Figure 1.6 illustrates relative positions of the surgeon and patient for 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 parietooccipital lobes. Turning the head to either side (bringing the cor-

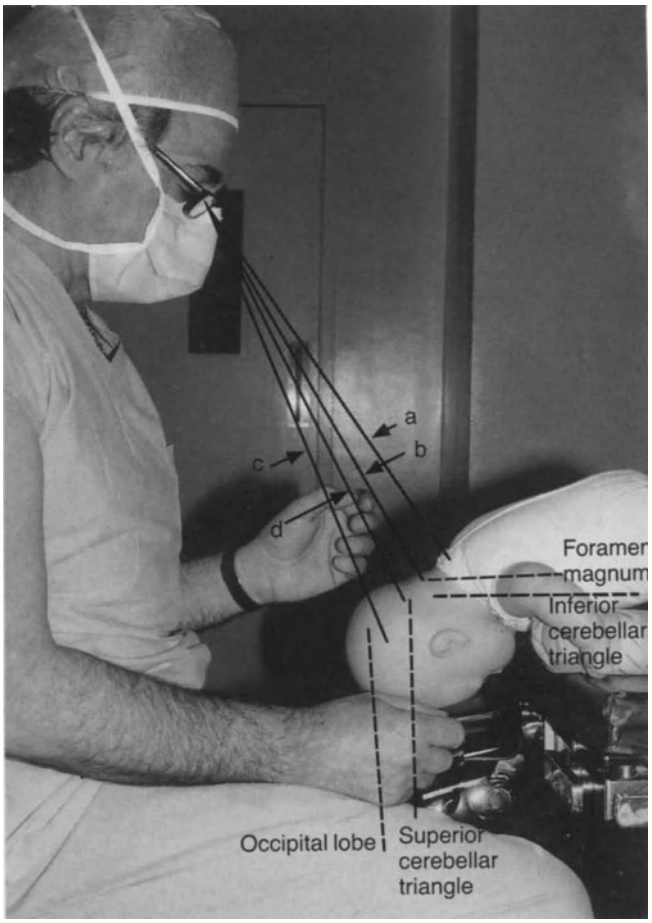


Figure 1.5. The relative positions of the surgeon and the child's head, with the newborn prone, illustrate: (a) an *excellent* line of vision to the cervical spine; (b) an *adequate* line of vision to the foramen magnum (except in Chiari II children), and the inferior cerebellar triangle; (c) an *unacceptable* line of vision to the superior cerebellar triangle because of the ledge of squamous occipital bone and the transverse sinus; (d) a good line of vision to the occipital lobes.

onal 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 into the body of the lateral ventricle through either the occipital horn and trigone or the frontal horn.

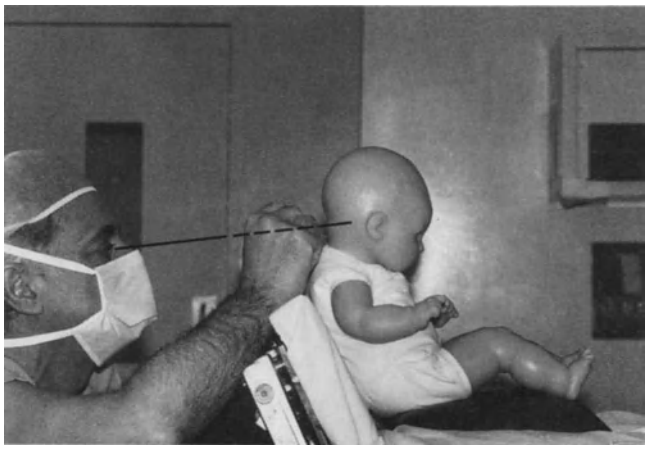
Although extension of the head around and axis running through the auditory canals does not embarrass venous drainage, flexion may cause the horizontal rami of the mandibles to compress the internal jugular veins. Distraction of the skull prior to flexion minimizes this risk of jugular compression by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

Prone Position

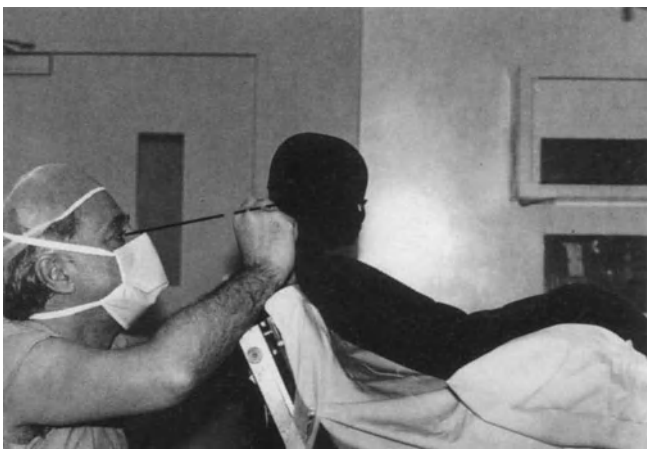
By placing the child prone with the head in the neutral position, one may expose the lambdoidal sutures (the parietooccipital region) for immediate access to the occipital lobes. Flexing the head and distracting it at the craniovertebral junction provides access to the squamous occipital, craniovertebral, 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 he is supine, particular care must be taken to 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 newborn and infants), but somewhat facilitated by the Gardner-Wells* headholder (which may be applied to toddlers and older children). Whether using the horseshoe or Gardner-Wells headholders, adequate clearance between the *symphysis 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

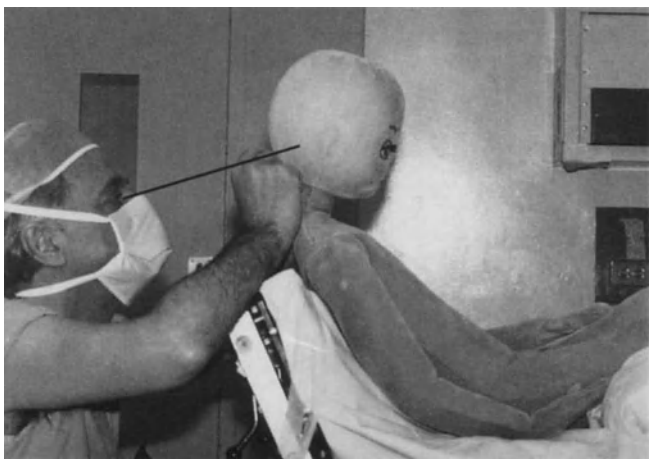
* Codman, & Shurtleff, Inc., Johnson & Johnson Company, Randolph, Massachusetts 02368.



A

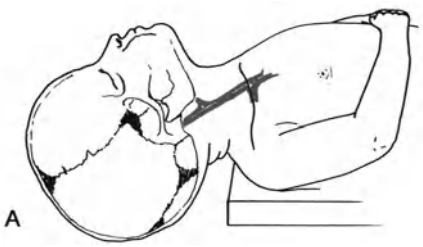


B

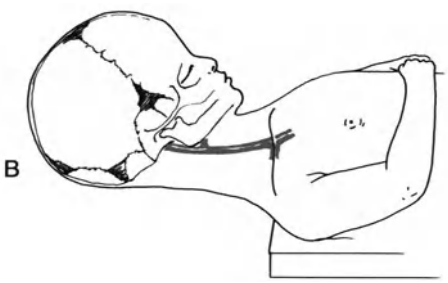


C

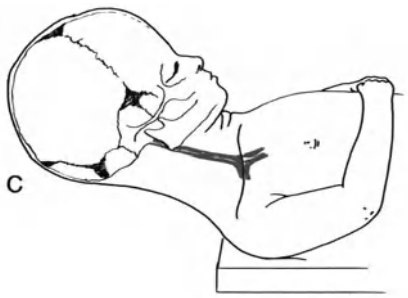
Figure 1.6. Note the relative sizes of the surgeon's hands and the child's head as shown here with (A) the term newborn: the surgeon's hands, in the neutral position, must be held apart to permit visualization of the operative field between them, since the newborn head is so small that one may not look over them and into the posterior fossa; (B) the toddler; (C) the juvenile: the head size is such as to permit 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 considerably 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 considerably the relative working space.



A

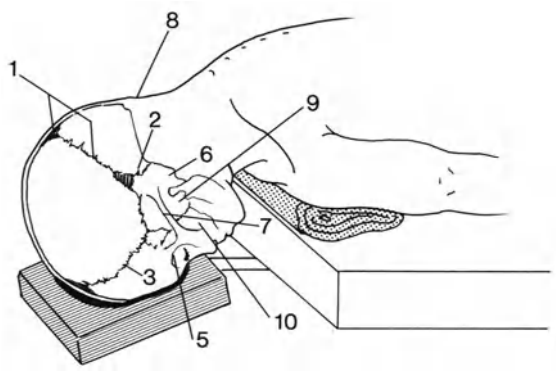


B

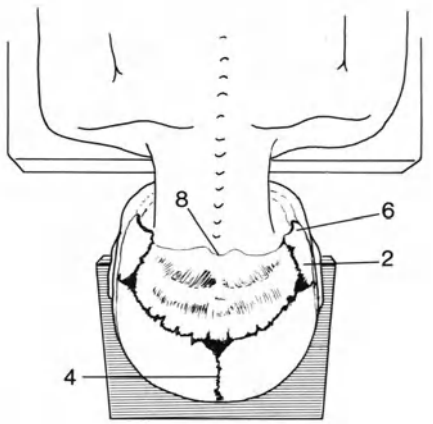


C

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 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.



A



B

Figure 1.8. 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 zygomatic arch (*7*); the rim of the foramen magnum (*8*); and the squamous temporal (*9*) and greater wing of the sphenoid (*10*) bones.

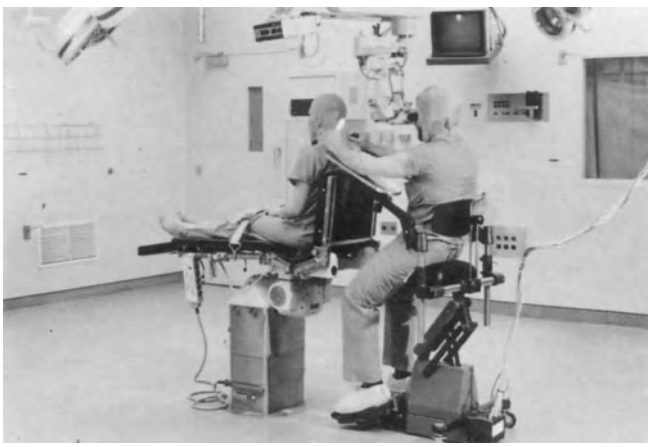
procedure, may be enough to cause a decubitus of the chin. One must leave enough room for the anesthesiologist to check and manipulate the endotracheal tubing.

Lounging (Sitting) Position

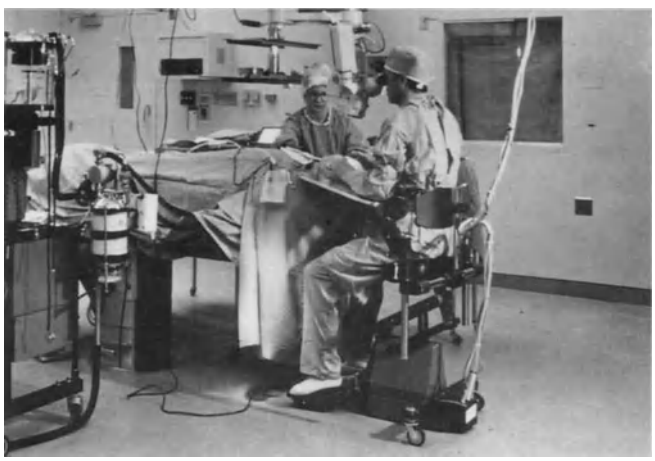
The "lounging" position is ideal for access to the posterior III ventricle, the superior cerebellar triangle, and the falx-tentorial junction. *Irrespective of the physical inconvenience to the surgeon* (Fig. 1.9A, B) *and the truly negligible risk of air embolism if appropriate anesthesiologic precautions are taken, it is the only safe way to operative lesions in the superior vermis, brachium conjunctivum, superior cerebellar hemispheres, opening of 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 a direct line of vision to the superior portion of the cerebellar vermis and the tentorial opening. Fixing it securely holds it suspended against its own gravitational force. Figure 1.10A shows this with the use of a horseshoe headholder in an infant, and Figure 1.10B with the use of the Gardner-Wells headholder, which has been scaled down for children, but which has not been designed for the thinner calvarium or relatively more voluminous diploic spaces. The risk of air emboli through the diploë remains very real.

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

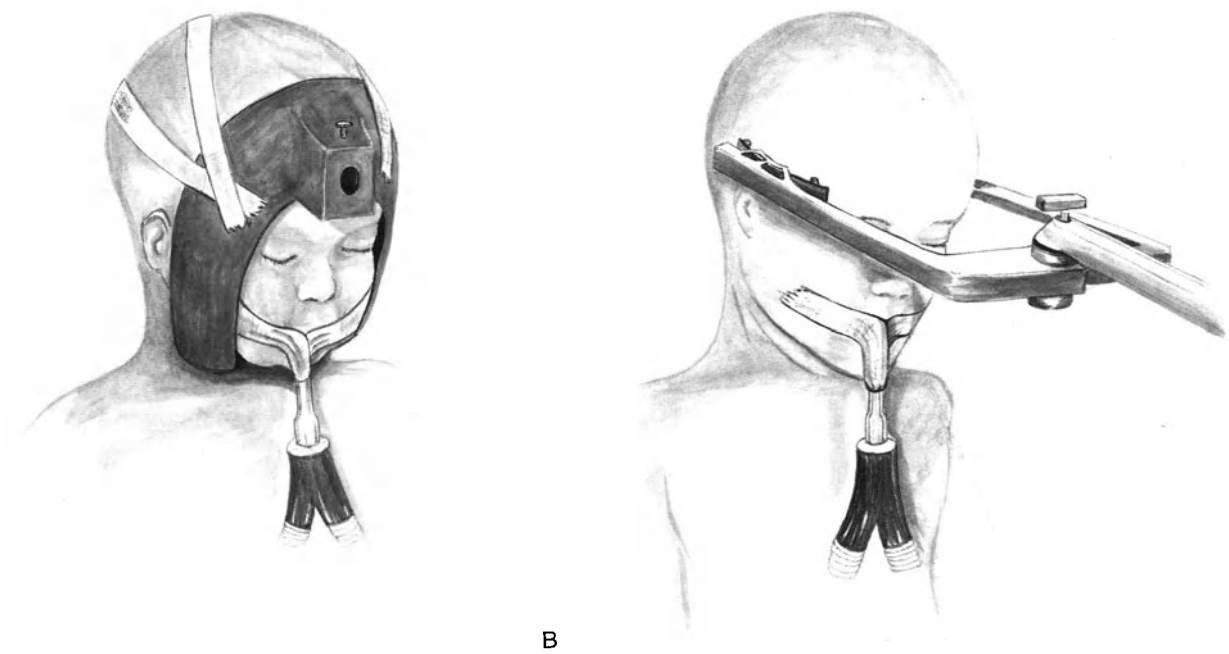


A



B

Figure 1.9. The sitting position (A) requires the surgeon to sit or stand in a more tiring and uncomfortable posture than the prone or supine positions (B).



A B

Figure 1.10. 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).

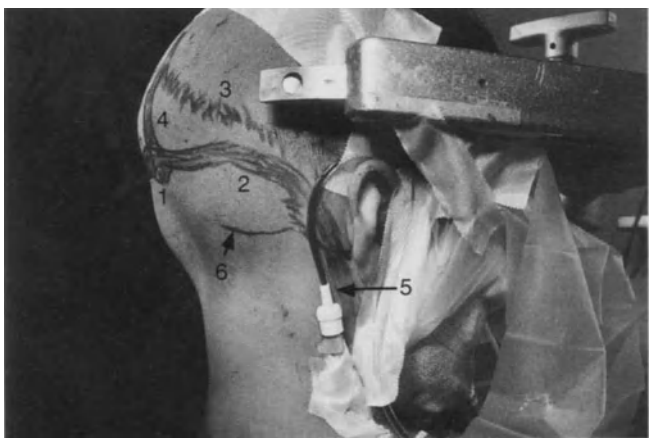


Figure 1.11. After distraction, the headholder is locked, maintaining head and neck position throughout the operation. In this child, one of the pins had to be set close to the operative field, something which at times is unavoidable, and the tubing for the central venous line had to be brought superiorly 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.

in either the sitting or lounging positions). Some form of plastic draping may be used to cover the tubing. Such a situation is illustrated in Figure 1.11, 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 the child, and in securing him 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 can be purchased almost 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.

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

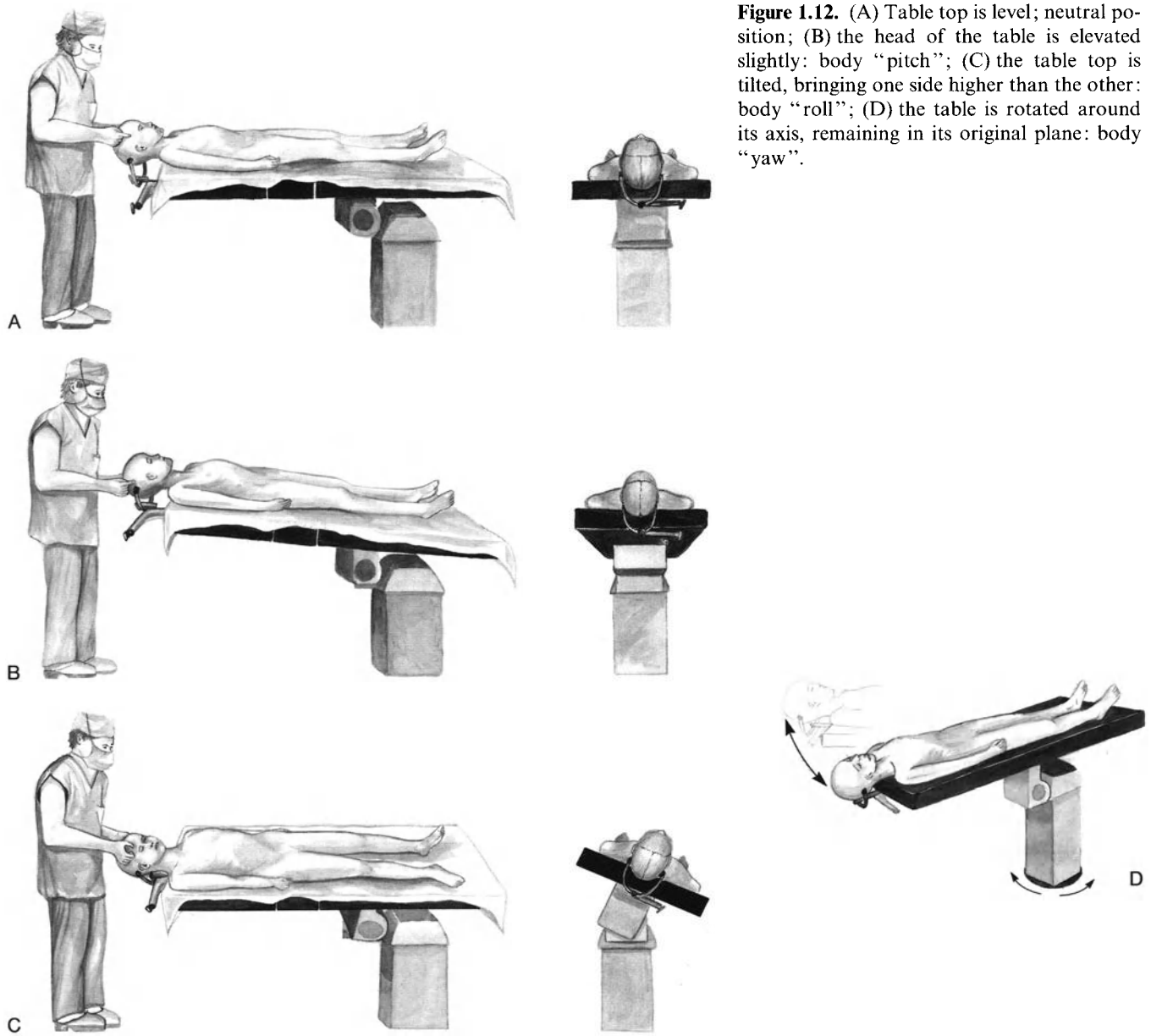


Figure 1.12. (A) Table top is level; neutral position; (B) the head of the table is elevated slightly: body "pitch"; (C) the table top is tilted, bringing one side higher than the other: body "roll"; (D) the table is rotated around its axis, remaining in its original plane: body "yaw".

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

The single most important consideration in positioning the child for surgery is not to complicate his 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 him at will, bringing him 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, 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 in 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 Figure 1.12A–D, the operating table is moved from neutral so as to provide body "pitch", "roll", and "yaw". In Figure 1.13A–C, one appreciates the mobility of the headholder from neutral to elevation

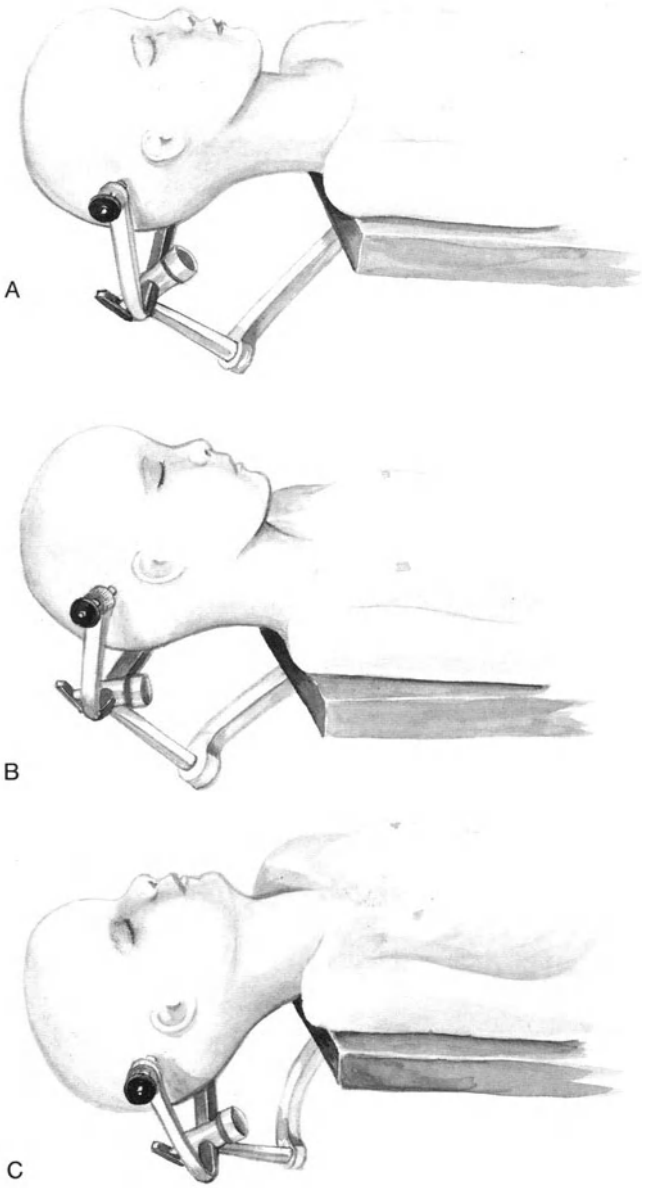


Figure 1.13. The head is shown in the neutral position (A), elevated but not flexed (B), and depressed but not extended (C).

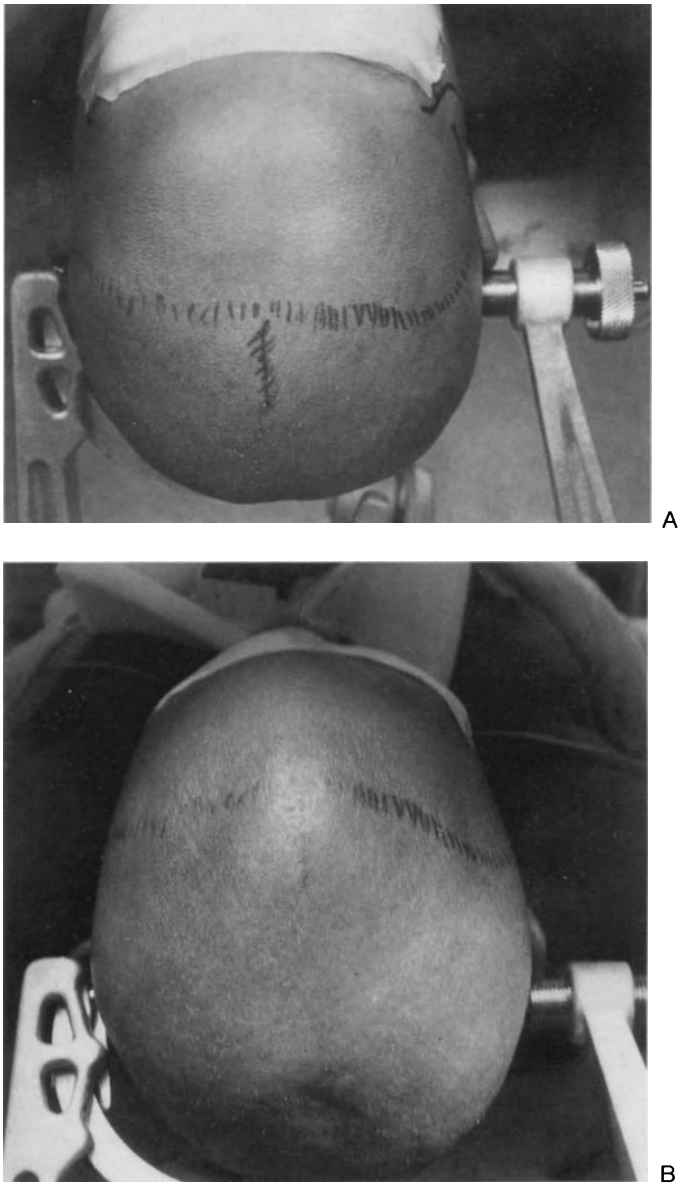
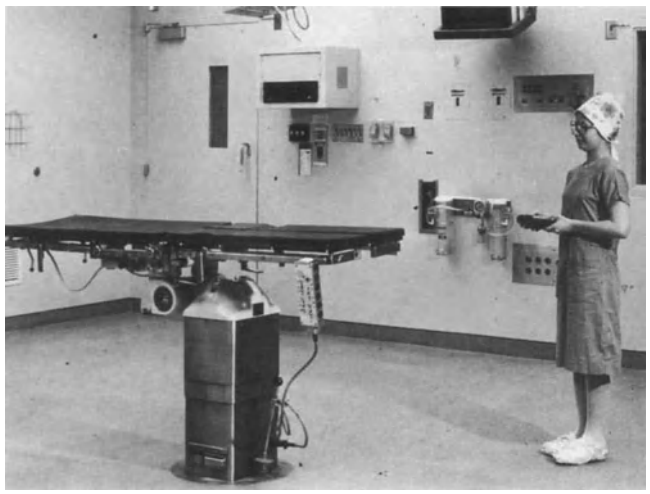
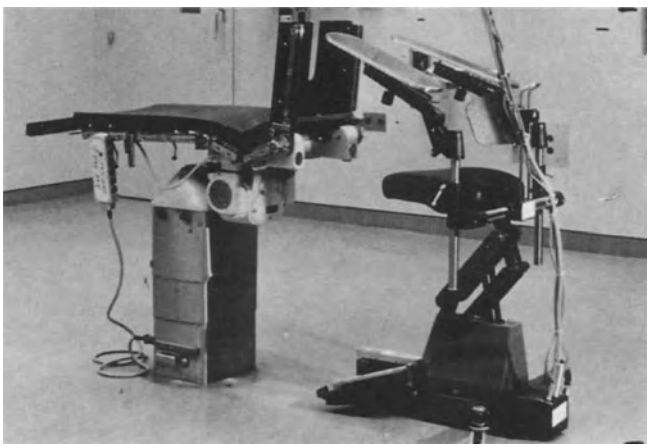


Figure 1.14. (A) The surgeon's view of the frontal bone, which is provided by using body "pitch" or head lowering with the child supine. (B) The surgeon may change his target area (shown in A) without changing his own body position, simply by altering body "pitch," lowering, or elevating the head.

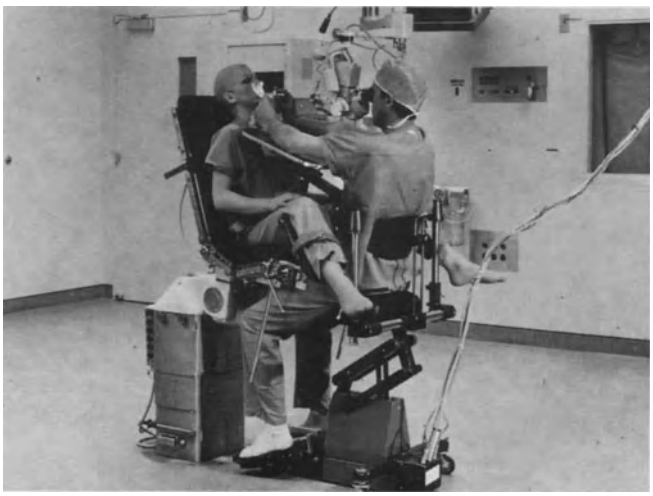
and depression of the head. All these changes in position may be obtained during the procedure, moving the operative site directly into the surgeon's line of vision or bringing a desired intracranial structure more clearly into view. This is crucial when working in the parasellar area so as to take advantage of the bifrontopterional exposure, in the pineal region, within the trigone of the lateral 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 his line of vision as the target area changes during the operative procedure is illustrated in Figure 1.14A, B.



A



B



C

Figure 1.15. (A) The Universal Operating Table* has a telemetric, hand-held device for elevation, depression, pitch, roll. (B) The Universal Surgeon's Chair** may be provided with a motor unit that permits graduated elevation and depression, as well as foot pedals to control coagulation, and shutter release, coarse and fine microscope focus magnification, and "roll" of the Universal Operating Microscope***. (C) This is the most convenient (least uncomfortable) position for transsphenoidal surgery.

Positioning of the Operating Microscope and the Laser, Multivariant Evoked Potential, and Echoencephalography Units for Intraoperative Use

The positioning of the operating microscope for intraoperative use is very variable, ranging from those neurosurgeons who always position the microscope in the same place in the operating room, and then accommodate themselves and position the child around the microscope base and arms, to those who utilize highly mobile floor- or ceiling-mounted microscopes that may be adapted, albeit with considerable effort, to the child throughout the operative procedure. Valid points may be made for either. I prefer the latter because it provides greater range of motion and superior utilization of the instrument. Different anatomical locations of surgical lesions in childhood demand a versatile, highly mobile operating microscope.

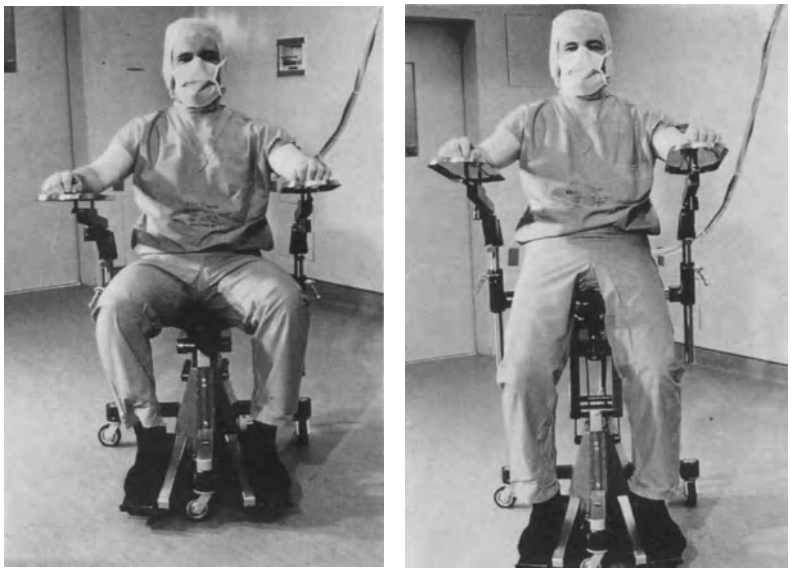
The Universal Operating Microscope***, with its operating table and chair (Fig. 1.15A–C) are so designed as to complement one another, allowing the surgeon to adjust automatically the position of either vis-à-vis the other, irrespective of the position of the child. The best-designed surgeon's chair provides a greater degree of comfort and a lesser degree of fatigability: the closer the chair position approximates normal sitting posture (Fig. 1.16A) the longer the surgeon will be able to operate without tiring. When it is necessary to extend the chair from the physiological sitting posture into one more closely resembling a high stool with armrests (Fig. 1.16B), fatigue occurs more quickly. The ceiling-mounted Universal Microscope may be easily and quickly adjusted so as to permit operating on a patient in either horizontal (Fig. 1.17A) or sitting (Fig. 1.17B) positions, without encumbering the floor space or readjusting the positions of the anesthesiologist, the neurophysiologist, and the nurses.

Both the laser and echoencephalography units are

* Universal Operating Table, Möller-Wedel, Hamburg, Federal Republic of Germany.

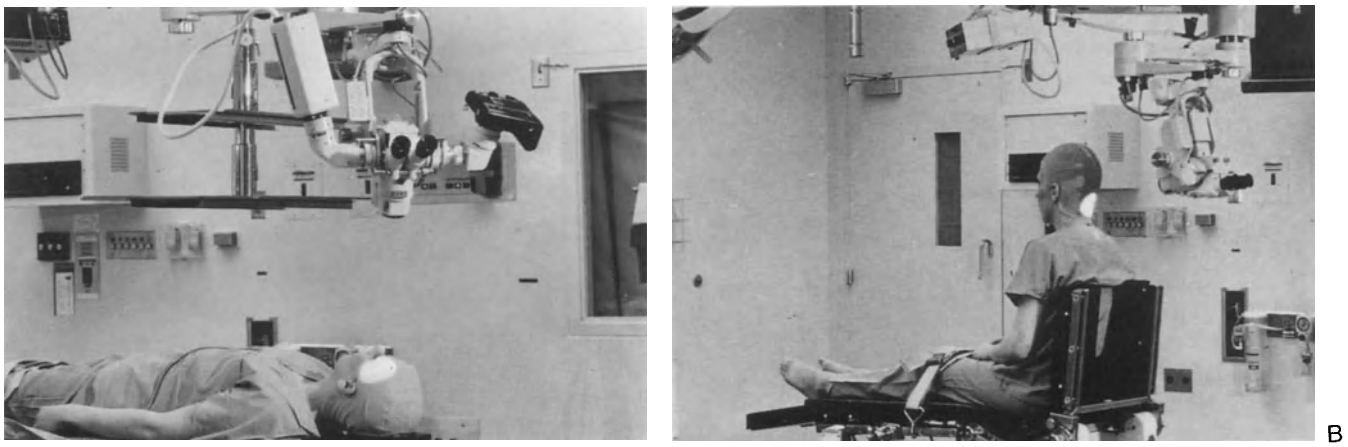
** Universal Surgeon's Chair, Möller-Wedel, Hamburg, Federal Republic of Germany.

*** Universal Microscope, Möller-Wedel, Hamburg, Federal Republic of Germany.



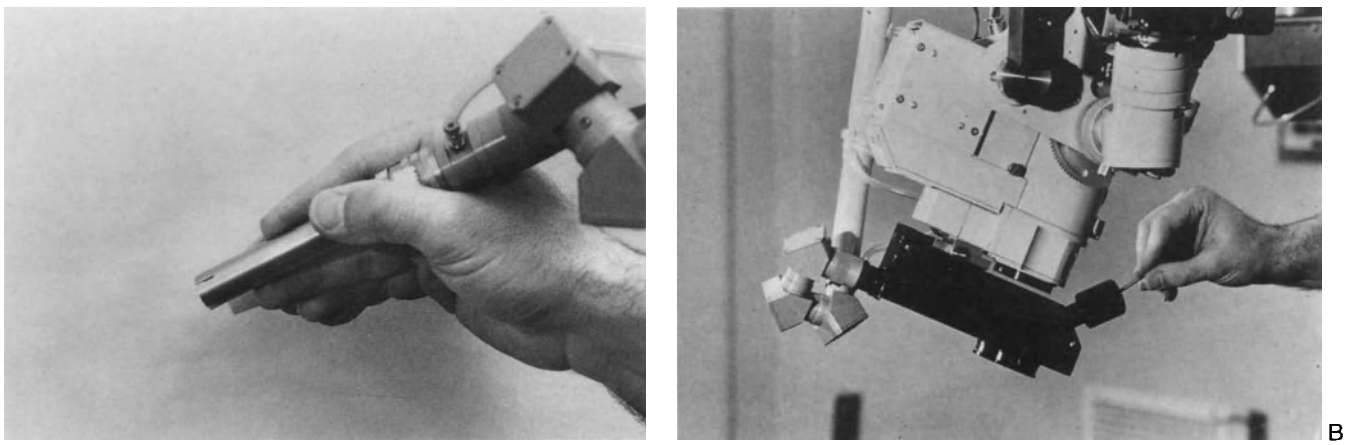
A B

Figure 1.16. (A) The surgeon sits comfortably in the operating chair when its position conforms to the normal anatomical sitting posture. Foot controls permit him complete independence for use of thermocautery, photographic, and microscope regulators. (B) The limited degree of comfort which one has when the surgeon's chair is in maximum elevation, approaching that of a high-stool, may be appreciated by studying this photograph.



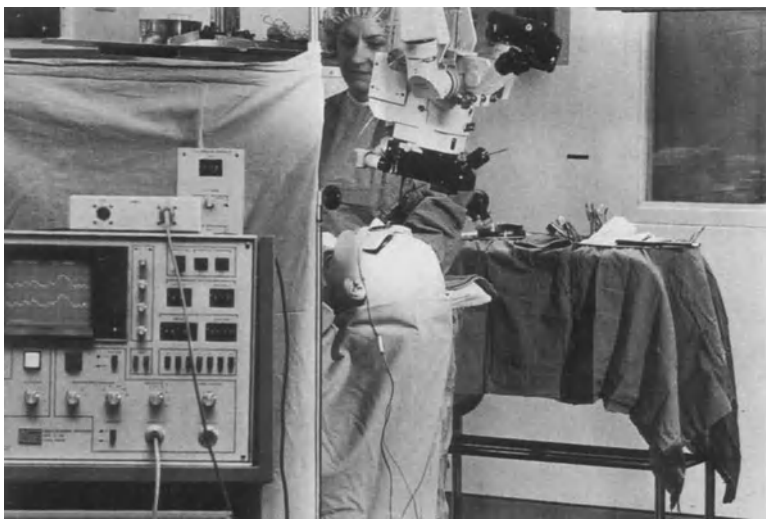
A B

Figure 1.17. The ceiling-mounted microscope may be moved about in three dimensions to bring varying portions of the operative field into view, with the patient either horizontal (A) or sitting (B).

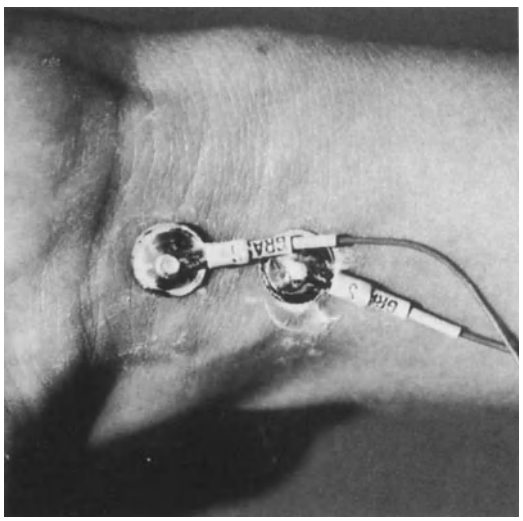


A B

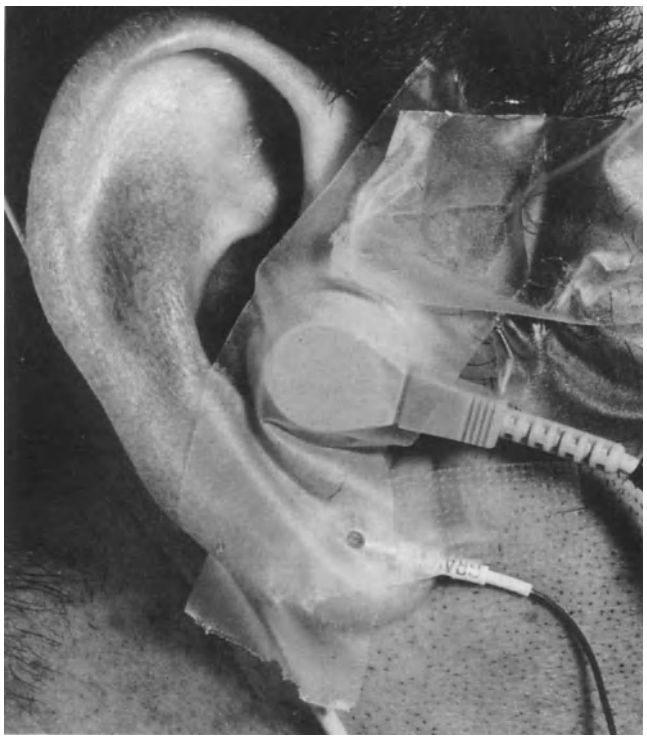
Figure 1.18. (A) is a photograph of the hand-held device for direction of the laser beam, (B) of the micromanipulator for direction of the beam through the microscope lens. Note the relatively rigid and cumbersome coupling device (system of mirrors) presently necessary in order to mount the carbon dioxide laser to the operating microscope and to the hand-held device.



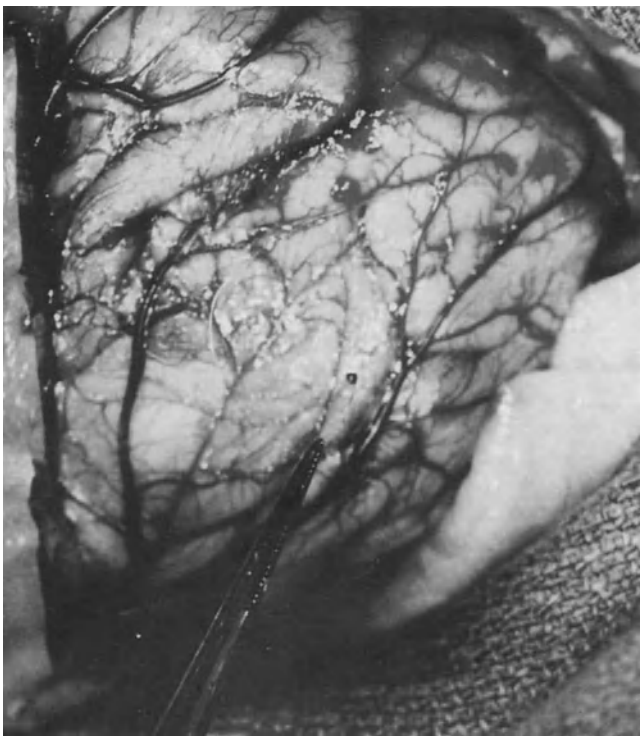
A



B



C



D

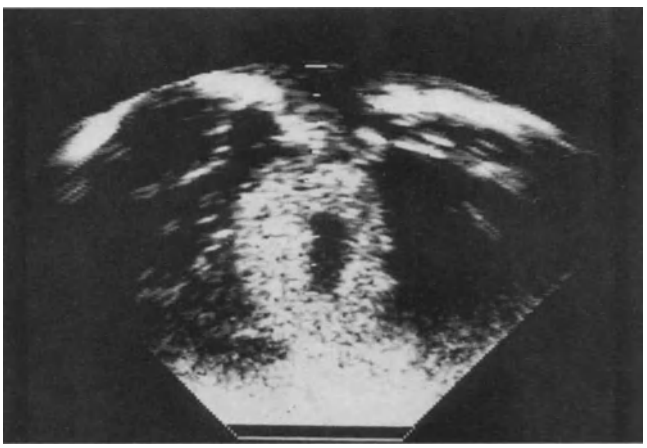
Figure 1.19. (A) Visual evoked potential stimulators have been applied to patient's closed eyelids and the multivariant evoked potential monitoring system positioned. (B) Surface electrodes have been applied over the median nerve. (C) A clicking device and needle electrode have been inserted, respectively, into the external auditory canal and lobe of the ear. (D) A 1-mm ball electrode has been applied to the cortical surface.

large, floor-mounted, awkward, and difficult to manipulate. However, the benefits each provides the surgeon are well worth the encumbrance, fatigue, and effort. Each operating room and each procedure are so very different from one another as to render it meaningless to attempt to describe detailed guidelines for the use of these instruments. Very likely, advances in technology will simplify considerably the laser, echoencephalogram, and multivariant evoked potential (MEP) recordings, allowing the laser to be used with ease through the operating microscope and resulting in the development of a much smaller "head" for effective use of the MEP. They are all very expensive, delicate, highly technical instruments that require training for their use, and the MEP requires experienced neurophysiologists.

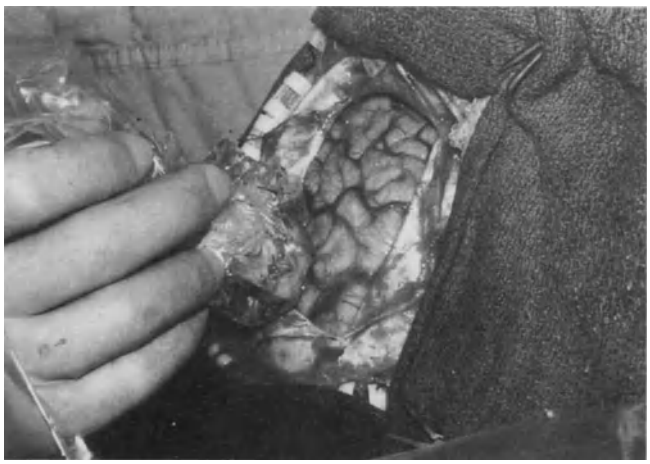
Irrespective of whether one is using a carbon dioxide, argon, neodymium YAG, or other laser, the beam may be delivered to the target area by using either a hand-held device (Fig. 1.18 A) or by mounting a micromanipulator to the operating microscope (Fig. 1.18 B).

Intraoperative MEP's provide supportive data concerning functional integrity of the visual (Fig. 1.19 A),

A



B



C

Figure 1.20. (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.

somatosensory (Fig. 1.19B), or auditory and brainstem (Fig. 1.19C) conductive pathways. Electrodes may be applied to the cortical surface (Fig. 1.19D) in order to identify more precisely the motor or sensory cortices.

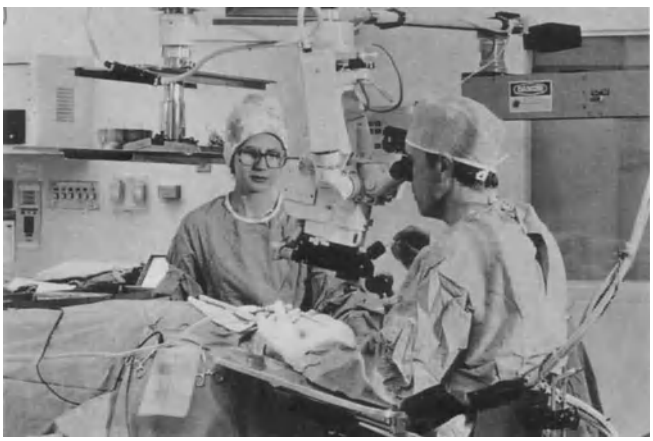
Intraoperative echoencephalography entails the application of a sonar-type emitter (head) to the dura or cortex, and computer elaboration of these signals into a visual image that is recorded on a screen (Fig. 1.20A). The head (of the echoencephalography unit) is still, unfortunately, quite large. However, when feasible and successful, intraoperative echoencephalography provides the surgeon with a remarkably precise visual image of the size, physical characteristics, and location of an intraparenchymal lesion (Fig. 1.20B, C) or ventricle. It must be realized, however, that sonography does not invariably "see" tumors or cysts: it, too, has serious limitations.

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 became 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 (Fig. 1.21A): the second assistant must stand with his back to the anesthesiologist, separated from him by a sterile drape passing from the operative field to an intravenous stand; the first assistant, normally positioned between the surgeon and the scrub nurse, makes it difficult for the nurse to pass the instruments quickly, and makes it impossible for her to see the surgery. Consequently, the scrub nurse should also serve as the first assistant, passing suction or bipolar cautery to the surgeon along with other instruments (Fig. 1.21B). 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 Universal 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.

A

B



C

Figure 1.21. (A) With two assistants (*1*, *2*) there is little body room for the surgeon (*3*), and the nurse (*4*) is at an unacceptable distance. (B) Without the assistants, the surgeon (*1*) has ample room and is immediately adjacent to the nurse (*2*). Note the draped operating microscope (*3*) and heating lamps (*4*), and fully draped frontal scalp (*5*). (C) This photograph, using a model, illustrates the surgeon, using the operating microscope, and seated in a surgeon's chair, without need of an assistant. Note that the micromanipulator for the laser has been attached to the operating microscope.

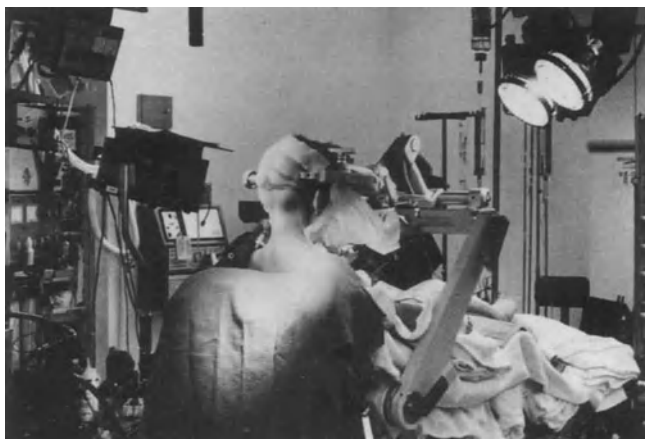
The placement of television monitors within the direct line of vision of the nurse (across the operative field) and the assistant and anesthesiologist (across the length of the patient's body) permits everyone to observe directly the details of the micro- and macrosurgical procedures. The coordination 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. The laser may be attached directly to the lens of the operating microscope, or connected to a micromanipulator, so that vaporization of tumor tissue may be effected without bringing another piece of equipment into the operative area (Fig. 1.21 C).

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.

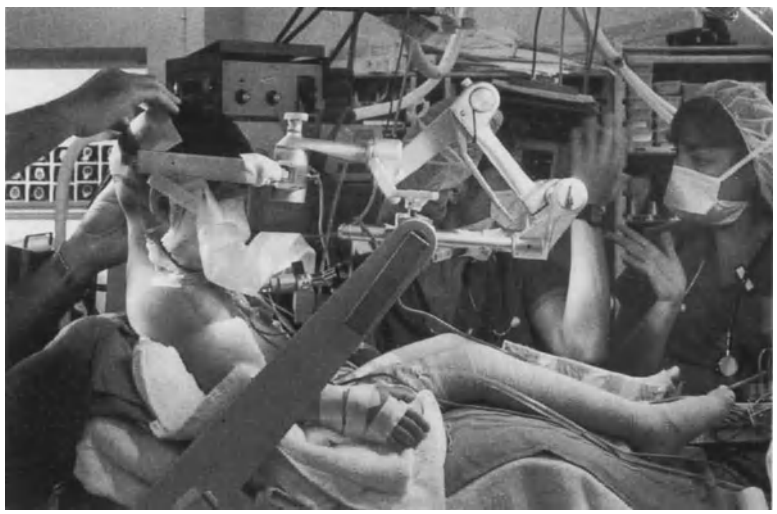
Accommodating Anesthesia

Although the purpose of positioning the child on the operating table is to permit the surgeon maximum exposure of, and access to, the operative site, great attention must be given to providing the anesthesiologist full access to the patient throughout surgery.

Setting heat lamps (Fig. 1.22 A, B) at a safe, yet warming, distance from the child during positioning protects him from potentially dangerous hypothermia, as does placing the plastic drape around the operative field before beginning the prep (Fig. 1.23 A). This prevents heat loss and insulates the child from the cooling effect 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 ten-minute scrub would allow large amounts of surgical soap and water to accumulate along the recumbent surfaces of the child's trunk and neck (Fig. 1.23 B).



A



B

Figure 1.22. 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. One may note the wrapped legs and lounging position.

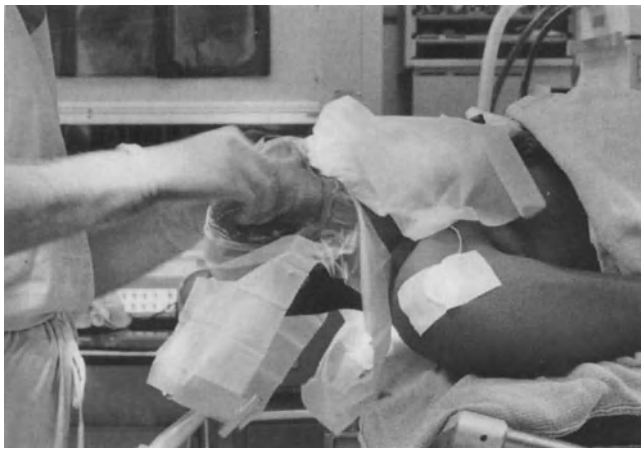
The drapes should be placed, fashioned, around the child, extending from the operative field in such a manner as to allow the anesthesiologist full access to the face, neck, chest, and limbs. This entails the use of an overhead table, either for the instruments or to which the drapes may be fixed. Placement 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 purposes of controlling the position of the endotracheal tube, functioning of central venous pressure lines, and the 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. This eliminates the need for lowering the head of the table when working at the optic chiasm or optic foramina. Conversely, flexing the head slightly provides more direct visualization of the convexity of the posterior frontal and anterior parietal lobes. Avoid excessive lowering of the head in prone or supine positions, and excessive elevation in the lounging position.

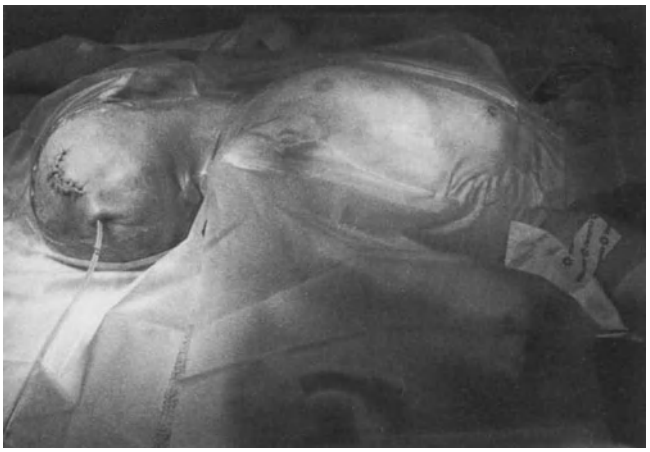
In Figure 1.24A–I one may study the actual positions of the patients from the perspective of the anesthesiologist, taking into consideration the positioning of the anesthesiologist's instruments and equipment, in each of the three basic surgical positions: sitting, prone, supine. The provision of a color television monitor permits the anesthesiologist to observe directly the operative procedure.

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 positoned on the



A



B

Figure 1.23. (A) The juvenile is in the supine position and the Vi-drapes have been placed bordering the operative field. Preparation of the scalp, after Vi-drape has been applied and the eyes protected. The face is covered. (B) The infant is supine, for conversion of an external ventricular drain into a ventriculoperitoneal shunt. Vi-drapes border the operative field, protecting the child from the cooling effects of pooled surgical soap and water.

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

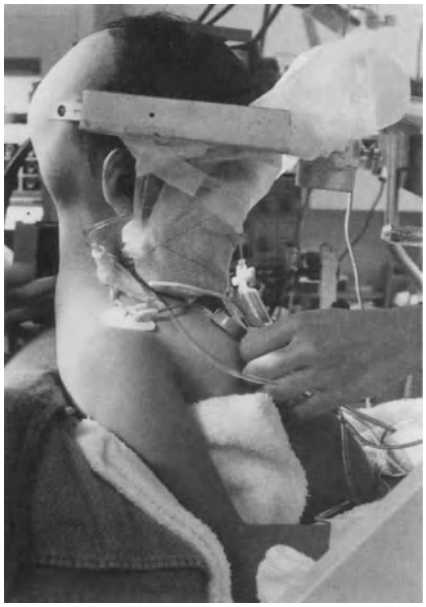
The head may be manipulated on the supine body for performance of frontal, bifrontopterional, craniofacial, parietal, biparietal, and temporal craniotomies as well as procedures demanding access to the craniocervicothoracoabdominal areas (such as for ventriculojugular or ventriculoperitoneal shunts). The 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.

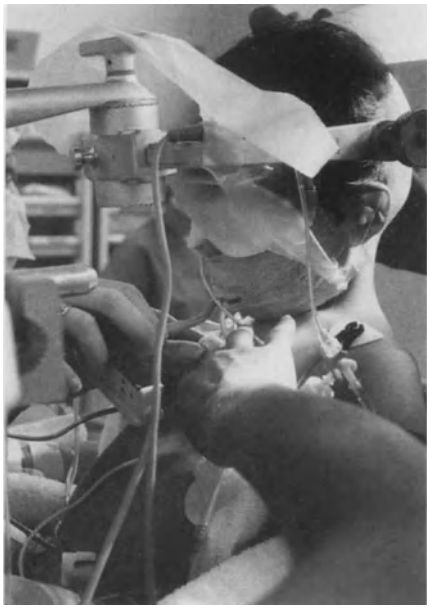
Anterior Fossa and Parasellar Area: Frontal Craniotomies

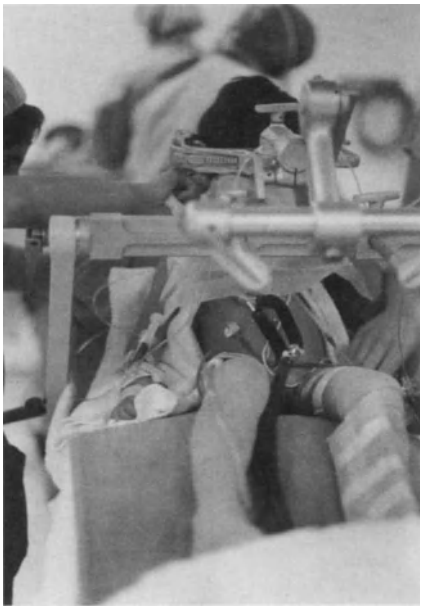
The supine position is best used for access to the frontal lobes, the orbit, the optic foramina, the intraorbital and intracranial optic nerves, and the optic chiasm. Retrochiasmatic extension of such parasellar tumors as craniopharyngioma may be resected more effectively through the interopticocarotid space, with the child supine, by resorting to the use of "pitch" and "role" 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. 1.25A). Consequently, slight extension of the head on the neck, as illustrated in Figure 1.25B, 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. 1.26A–C).

Since slight extension of the head makes for somewhat awkward hand positioning (Fig. 1.27A–C) for the scalp flap, one may simply elevate the head of the table for the skin incision and the posterior burr holes. The


A


B


C


D

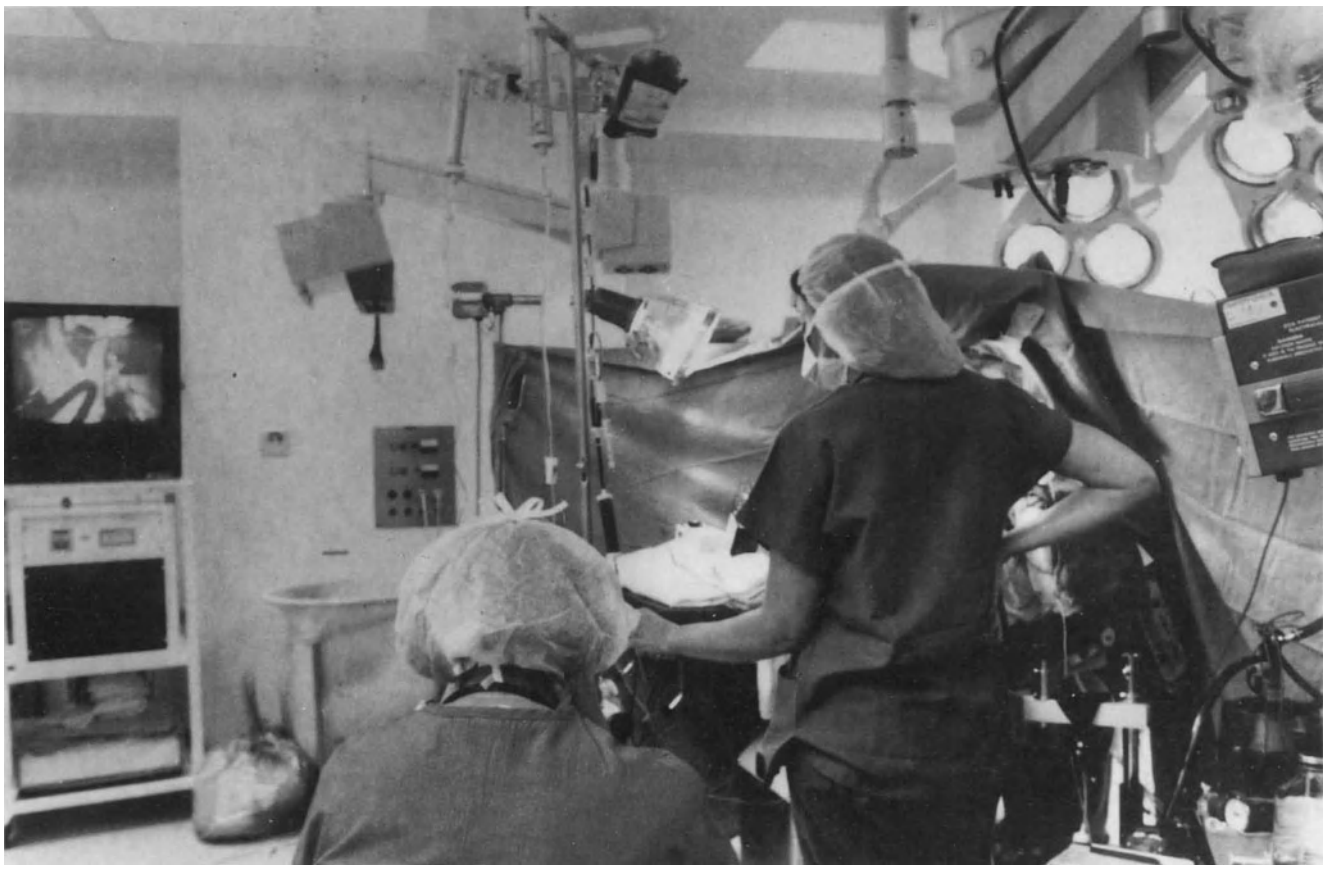

E


F

Figure 1.24. The lounging child, with his head fixed, as his face is protected and endotracheal tube secured, in progressive stages of draping. The anesthesiologist has full access to the face, neck, chest, abdomen, and limbs. This is essential. (A) The head is locked in position, so the anesthesiologist may now place his monitoring receivers and electrodes. Note that the head is distracted and flexed on the neck at the craniocervical junction. The Vi-drape is brought over the headholder to assure free access to the eyes and nose. (B) All lines and tubes have been positioned. The Vi-drape has seen brought over the headholder to assure free access to the eye and nose. (C) This is a view of the child behind the headholder and monitoring devices, illustrating fully exposed abdomen: anesthesiologist's view of trunk and extremities. (D) Anesthesiologist's view of face after child has been positioned lounging

G

H



I

and completely draped. The eyes are covered, the esophageal thermister has been passed through the nose, and the endotracheal tube is secured by elastoplast. (E) Anesthesiologist's view of fully draped child in the lounging position. (F) Anesthesiologist's view of fully draped child in the supine position. The same access exists when prone. (G) Placement of anesthesiology equipment and intravenous poles. (H) Anesthesiologist's available working area after child has been draped. The warming lamps (*1*), jointed mount for headholder (*2*), and microscope (*3*), draped and set in "park" position for immediate use. (I) Placement of TV monitor so anesthesiologist may observe entire (macro and micro stages) operation.

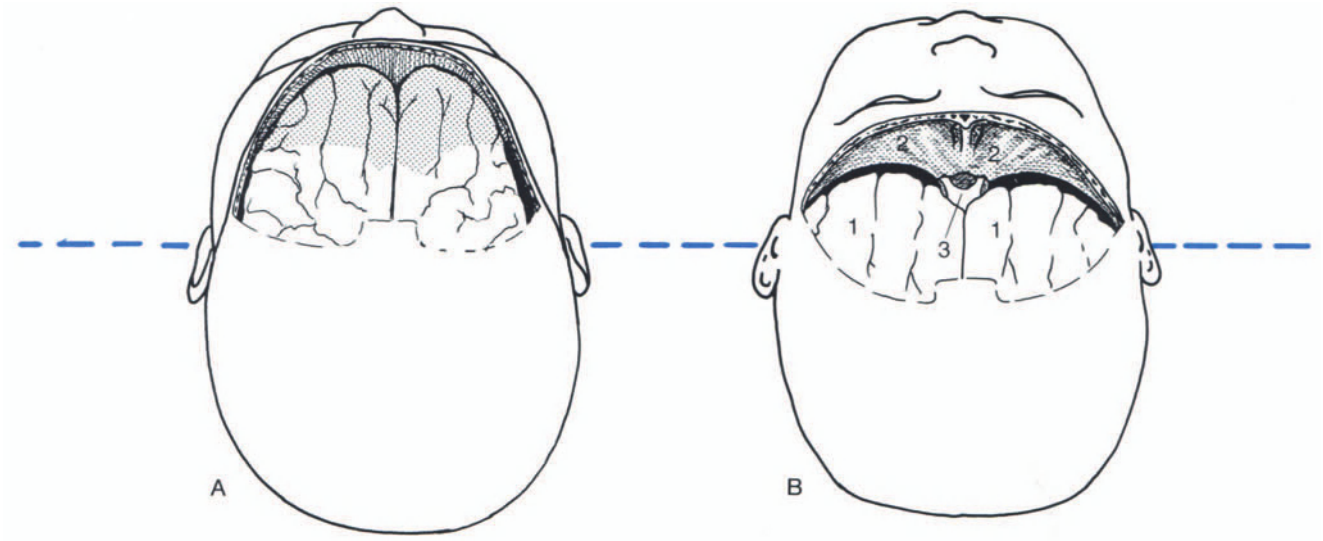


Figure 1.25. The falling away of the frontal lobes results from extending the head around its interauricular axis, not by lowering it beneath the level of the right atrium. On occasion, however, especially if a preoperative shunt has not been inserted, it may become necessary to lower the head by manipulating 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 covered by frontal lobes. (B) After the head has been rotated, extending it around its interauricular axis, the frontal lobes (*1*) fall posterioinferiorly, and the entire floor of the anterior fossa (*2*) with the chiasm (*3*) come into view.

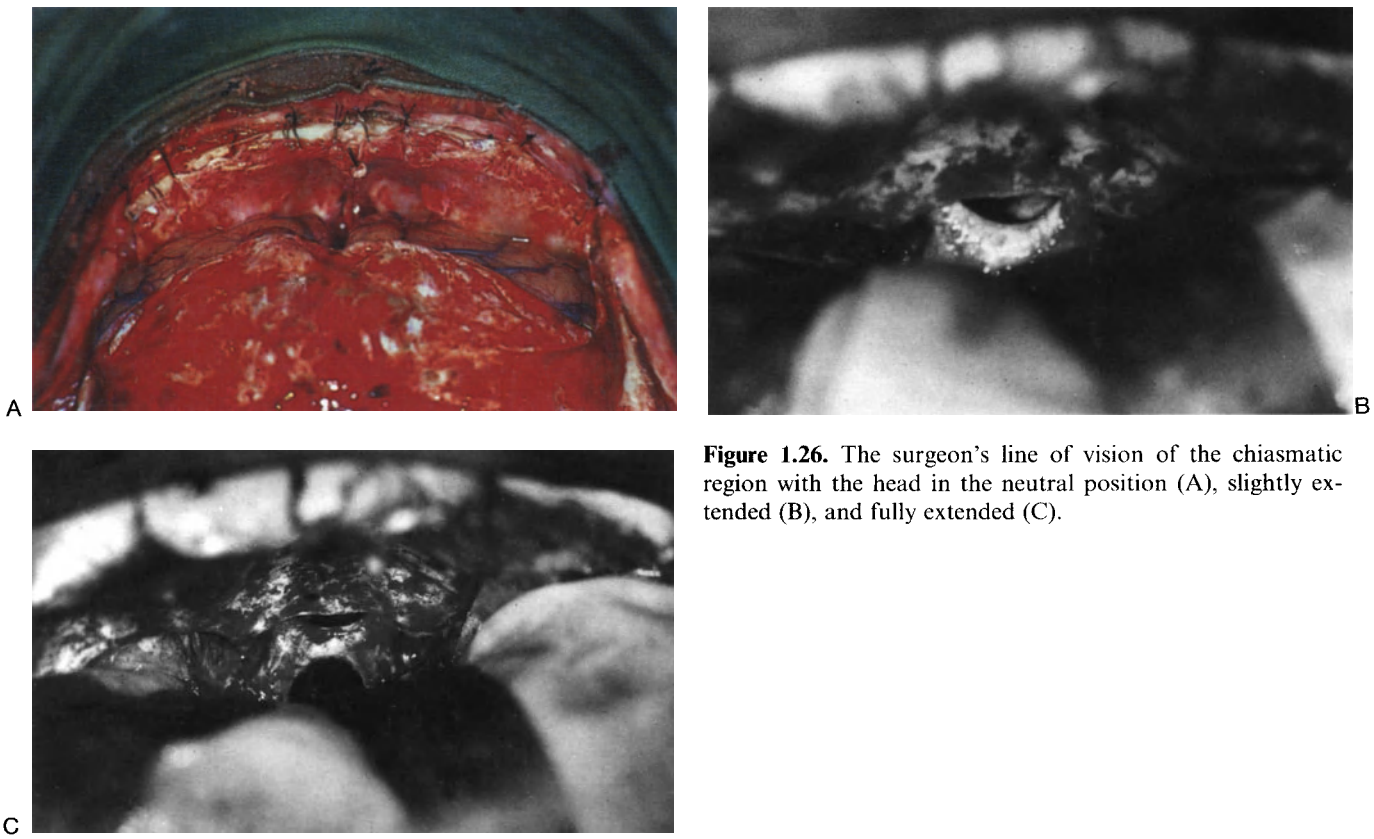


Figure 1.26. The surgeon's line of vision of the chiasmatic region with the head in the neutral position (A), slightly extended (B), and fully extended (C).

head of the table then is lowered to the desired level for placing the "keyhole" and glabellar burr holes, passing the Gigli saw and reflecting the osteoplastic bone flap, whether unilateral or bifrontopterional (Fig. 1.28A–G). 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), and optic nerve(s) and chiasm, one may "roll" the operating table from right (Fig. 1.29A) to neutral (Fig. 1.29B), to left (Fig. 1.29C) so as to facilitate visualization of the lesion and parasellar anatomical structures.

Unilateral Frontopterional Craniotomy
The head is extended on the body and rotated slightly (Fig. 1.30A–D) so as to allow the surgeon to visualize directly the frontal eminence of the side to be operated. 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. 1.31A) so that the surgeon's direct line of vision is at the metopic suture (Fig. 1.31B). 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. 1.31C) 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. 1.31D).

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, and there must 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. 1.32).

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. Parietal lobe and corpus callosum lesions,

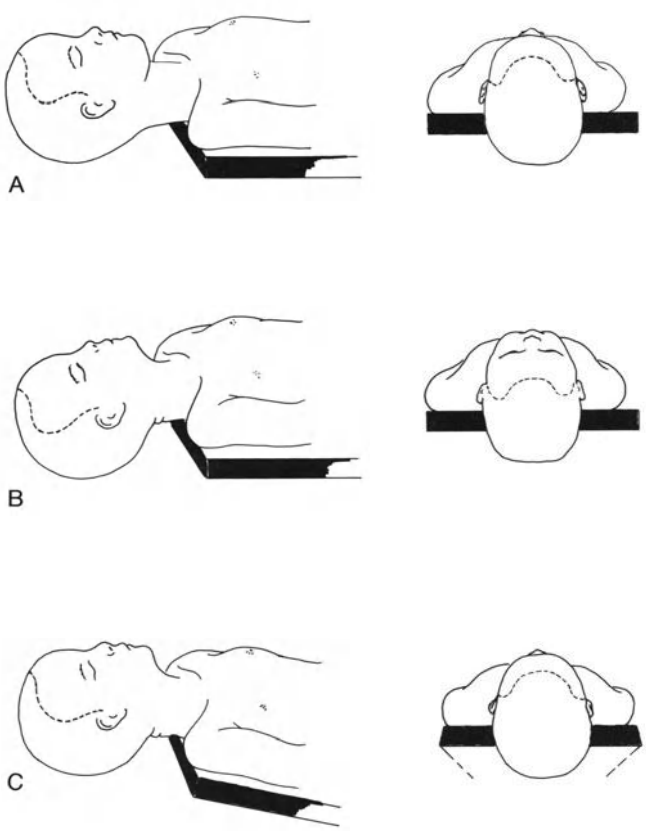


Figure 1.27. (A) Head in neutral position, (B) extended, and (C) extended but elevated using table pitch.

as well as those within the III ventricle or the 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 burr 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.

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 parietofrontal, parietotemporal, and parietooccipital lesions.

Unilateral Parietal Craniotomy
The head is rotated 90° so as to bring the operative side into the midsagittal 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. 1.33).

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. 1.34A, B). 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

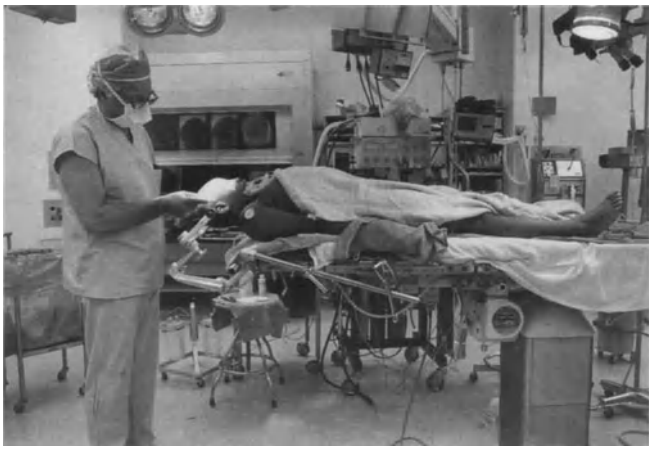
By positioning the child supine and rotating the head a full 90° so as to bring the coronal plane of the head parallel to the sagittal plane of the body, one may expose completely the convexity of the skull and cerebrum. Flexion of the head on the cervical spine to approximately 5°, after it has been turned fully, places the lateral aspect of the calvarium in a plane perpendicular to the surgeon's line of vision. This provides him the possibility of exposing the lateral aspects of the frontal, parietal, 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. 1.35A–D).

Temporal Craniotomy

The positioning of the head for a temporal craniotomy is much the same as that for a unilateral parietal craniotomy (Fig. 1.34), 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. 1.36A). This provides the surgeon a direct line of vision to the superior and inferior temporal lines, the zygomatic arch, and the external auditory canal (Fig. 1.36B). Elevating and lowering the head facilitates access, respectively, to the sylvian fissure and the entirety of the tentorial surface and ring.

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


A

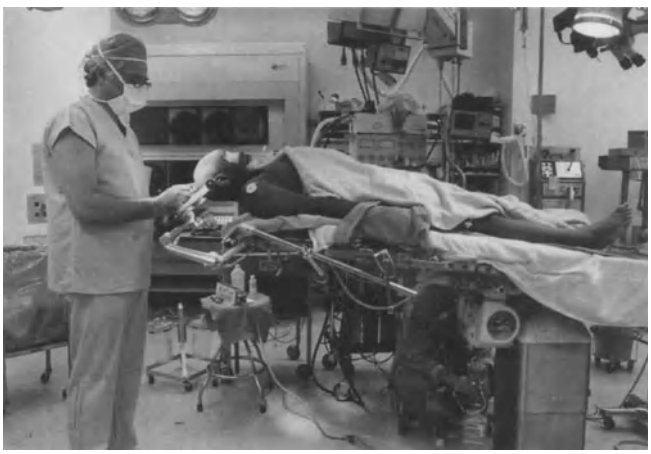
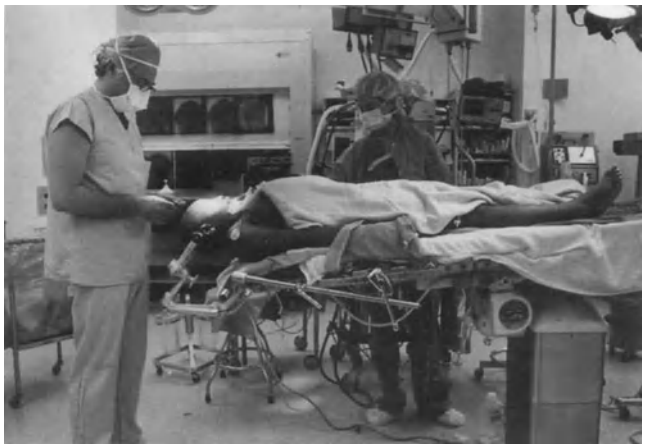
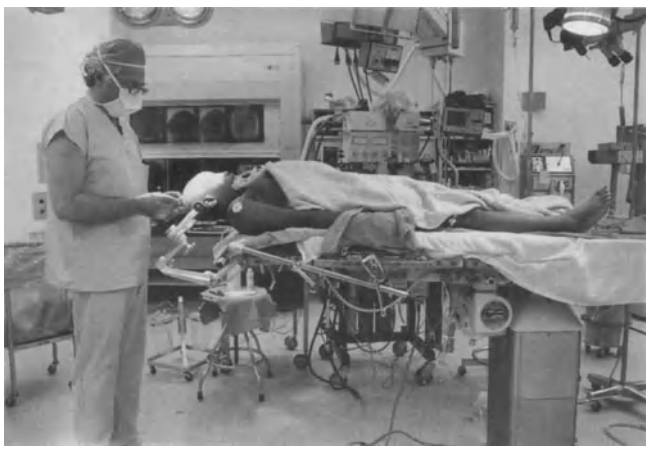

B

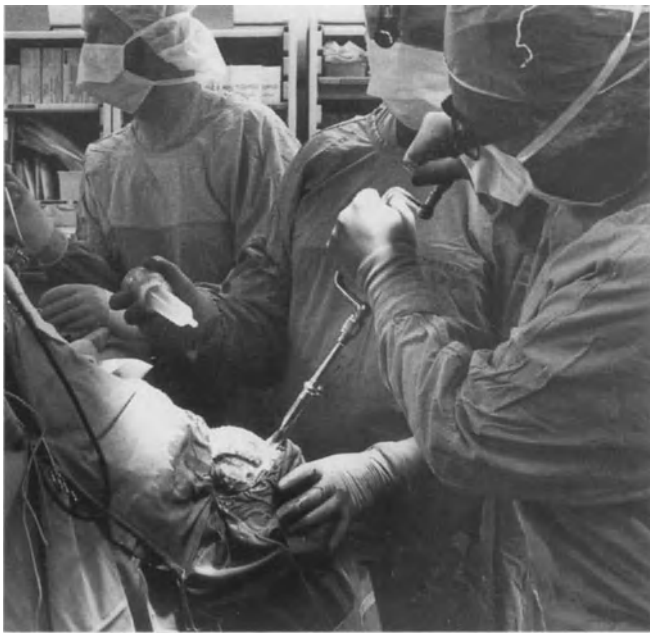
Figure 1.28. (A) Minimal body pitch is used to elevate head of table for skin incision. (B) More accentuated head elevation brings the most posterior portion of the parietal area into efficient working range. (C) Head lowered slightly, providing good hand positioning for anterior parietal and posterior frontal surface areas. (D) Head slightly elevated providing good



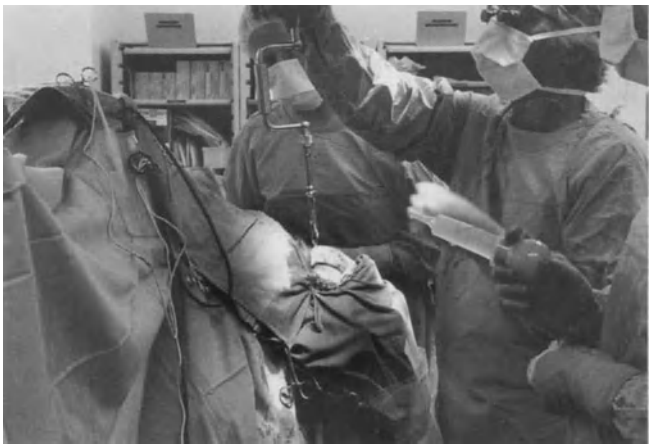
C



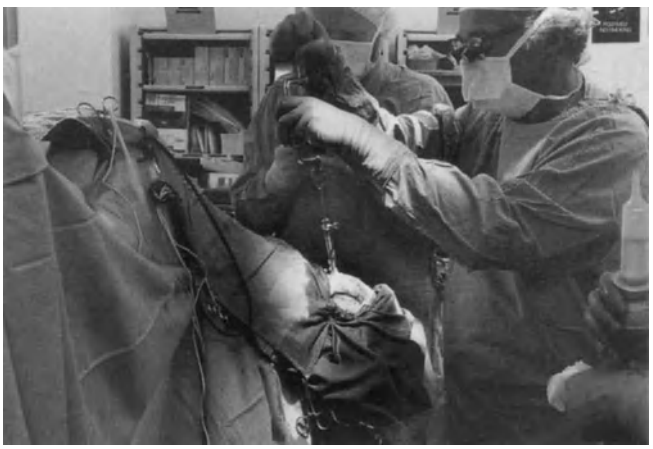
D



E



F



G

hand positioning for midparietal surface areas. (E) Convenient body and arm posturing for posterior frontal burr holes is attained by lowering the head. (F) The head is at the same level as in E, which is inconvenient for glabellar burr opening, so (G) the head has been lowered to facilitate arm positioning for the glabellar opening.

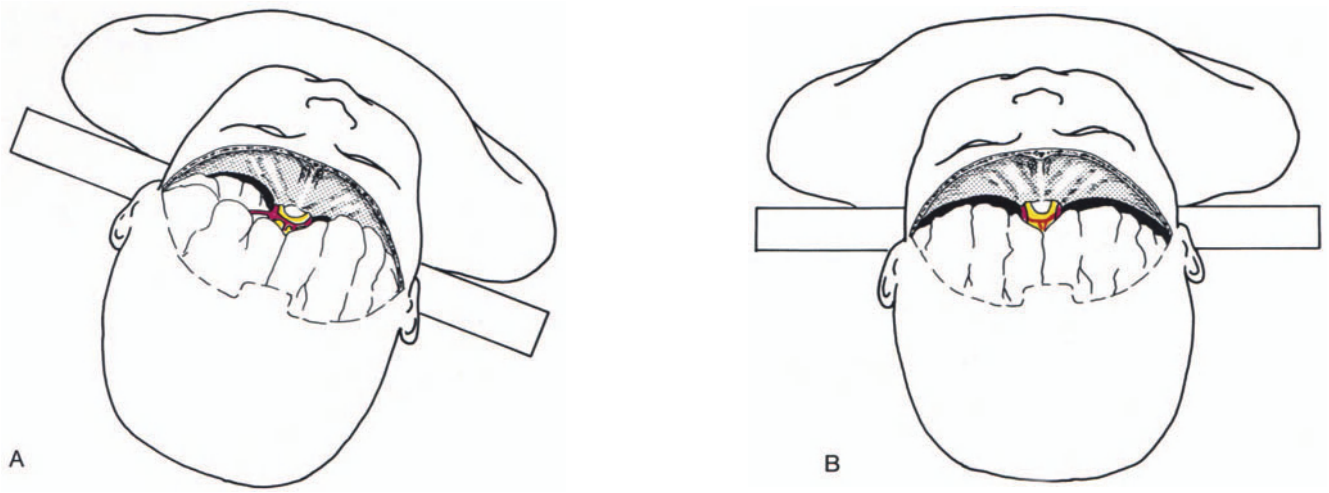


Figure 1.29. (A) Roll to the right permits visualization of the left sphenoid wing from the pterion to the anterior clinoid, internal carotid, and middle cerebral arteries after bifrontopterional craniotomy has been performed. (B) Neutral position is best for planum sphenoidale, optic nerves and chiasm, carotids, and anterior cerebrals. (C) Roll to left permits exposure of right homonyms to those illustrated in A.

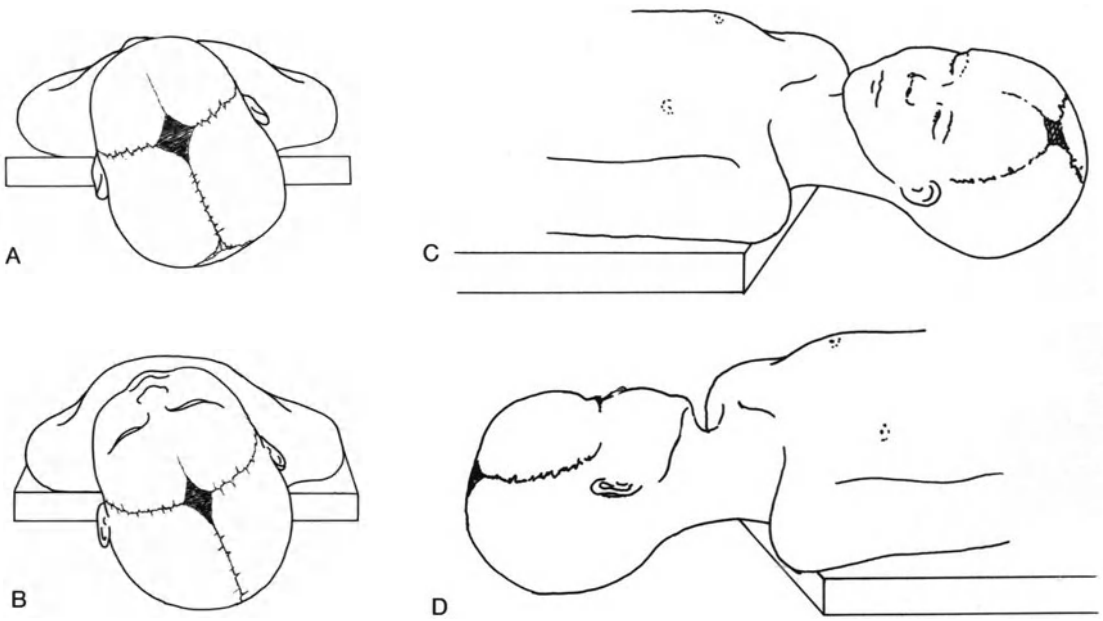
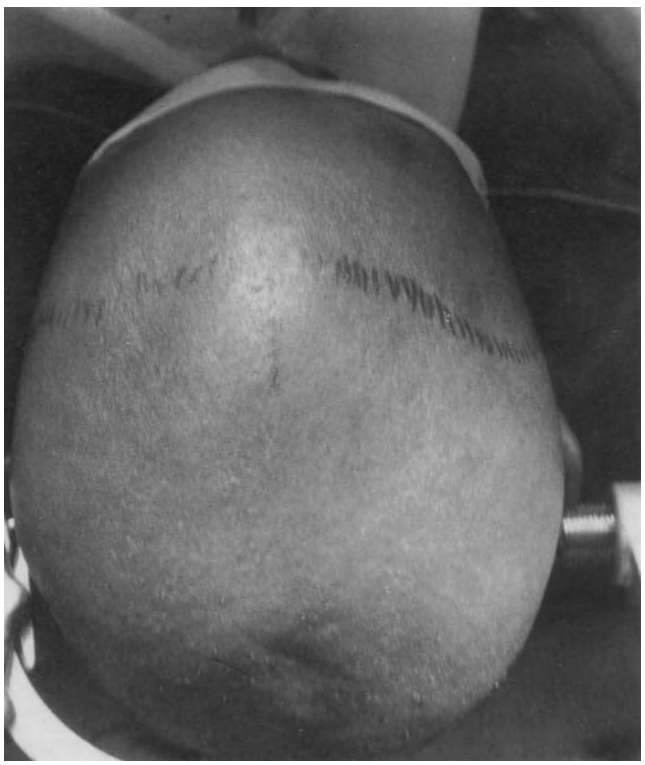
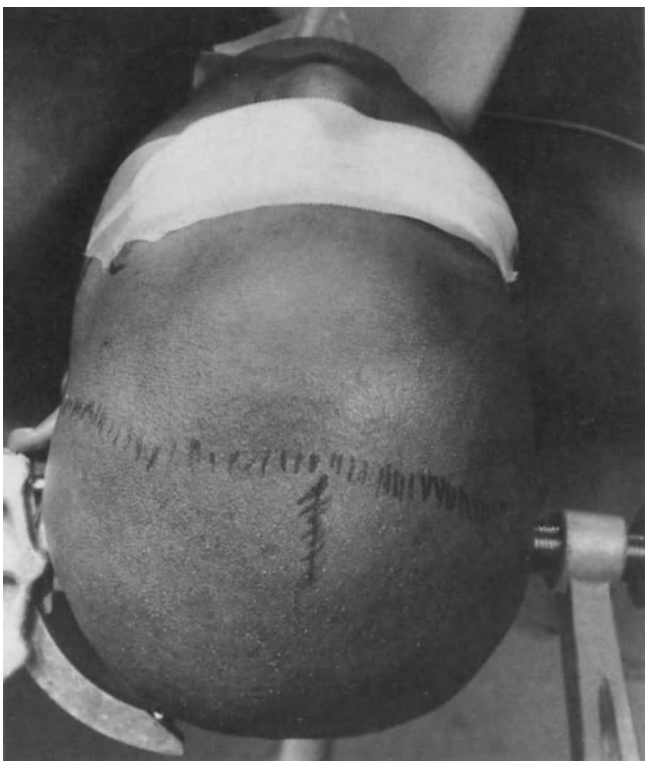


Figure 1.30. Unilateral frontopterional craniotomy. The child's head is positioned for unilateral craniotomy, showing the frontal eminence, coronal, and sagittal sutures. (A) Coronal view of skull. (B) Surgeon's view of A. (C) Lateral view of A from the left. (D) Lateral view of A from the right showing pterional area.

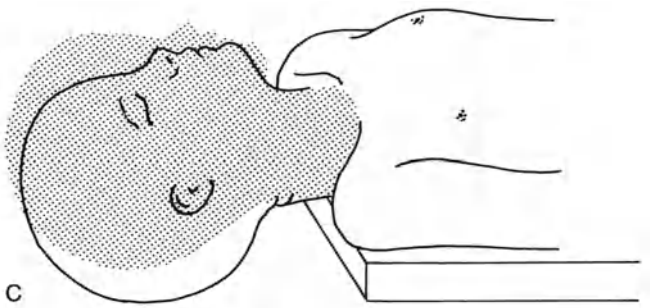


A

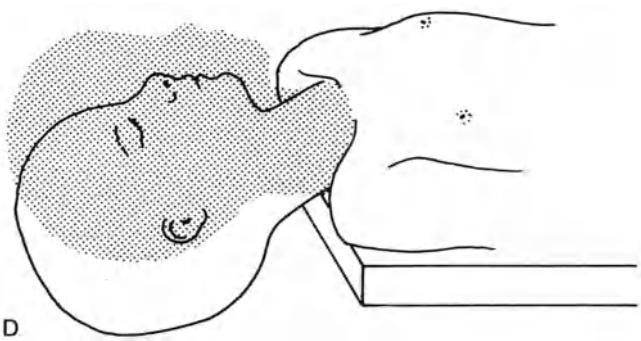


B

Figure 1.31. Bilateral frontopterional craniotomy, showing surgeon's view with (A) head neutral and (B) extended. (C) The 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.



C



D

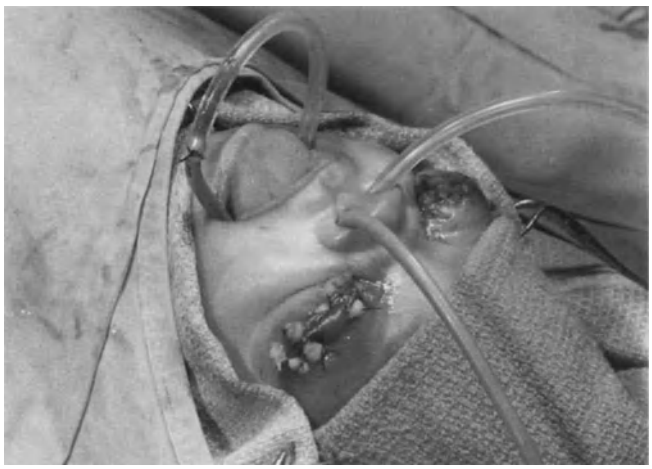


Figure 1.32. Exposure of the orbital and malar areas, which may be attained by slightly extending the head.

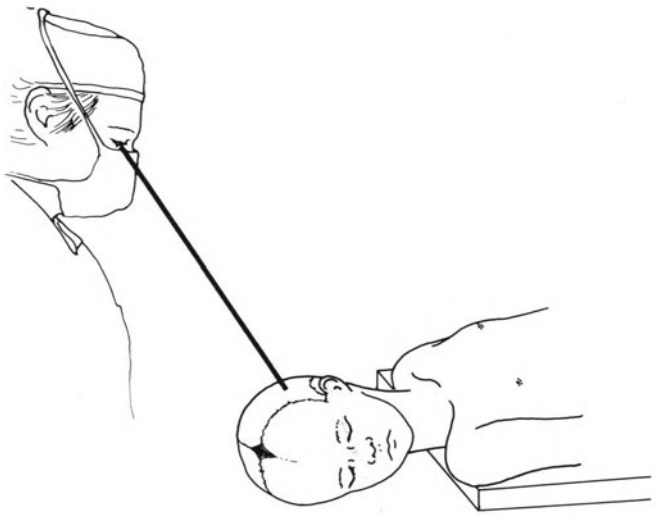
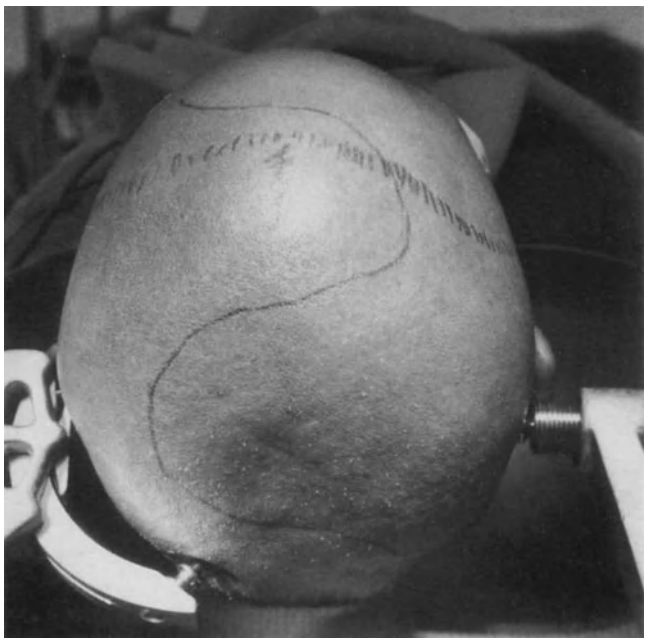
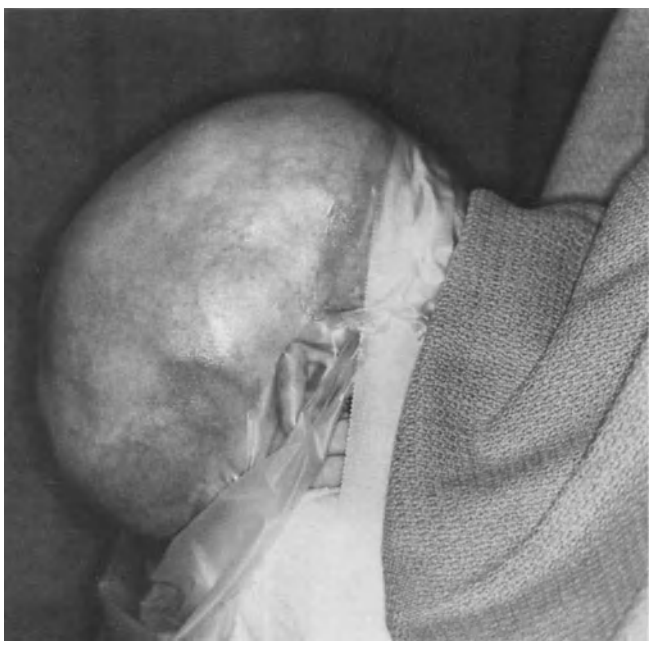


Figure 1.33. The child is supine with the head flexed slightly, after having been rotated so as to bring the sagittal suture parallel with the coronal plane of the body, providing the surgeon a straight line of vision to the temporal lobe but not to the floor of the middle fossa.

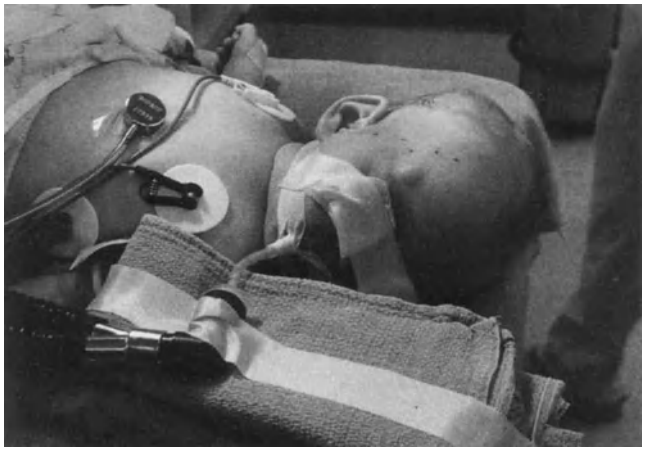


A

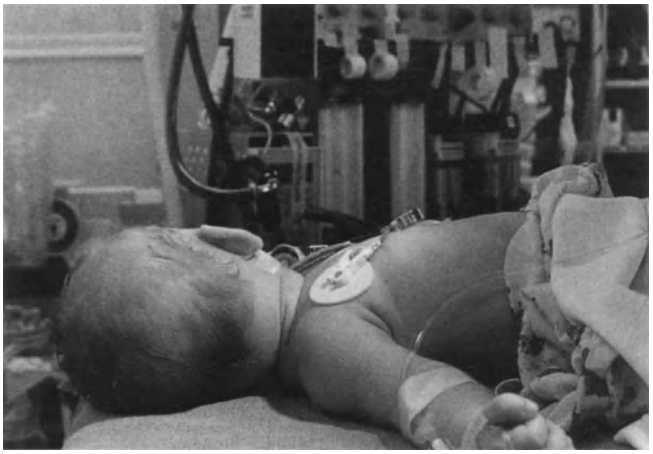


B

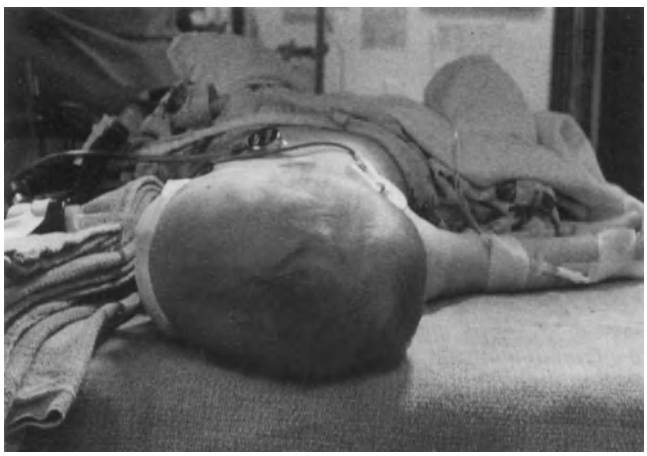
Figure 1.34. (A) Biparietal craniotomy. Slight flexion of the head on the neck rotates the superior surface of the skull into excellent position for the S shaped incision, and access to the frontal and parietal areas bilaterally. (B) A lateral photograph of an infant with his head in this position, illustrating exposure of the inion.



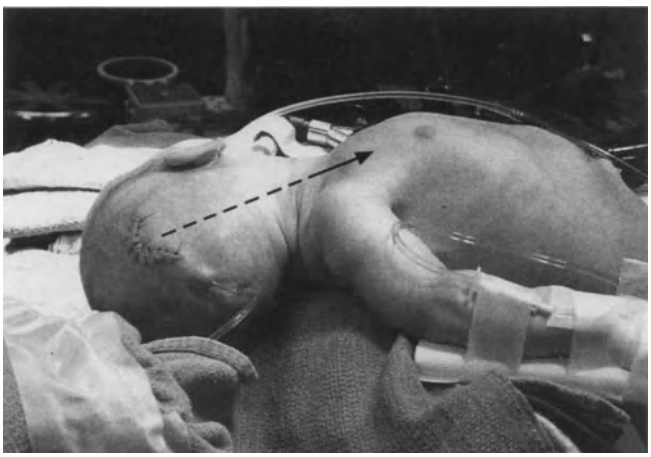
A



B



C



D

Figure 1.35. (A) Anterooblique view. Supine child with head rotated 90° to the left, bringing the coronal plane of 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 in the neck). (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. This is viewed from the vertex of the skull. (D) When a ventriculoperitoneal shunt is to be performed, a role should be placed under the neck so as to "unfold" it, permitting easy passage of the subcutaneous guide from the parietal eminence to the supraclavicular area (arrow). In this child a shunt revision revealed ventriculitis, so it was converted to an external ventricular drain.

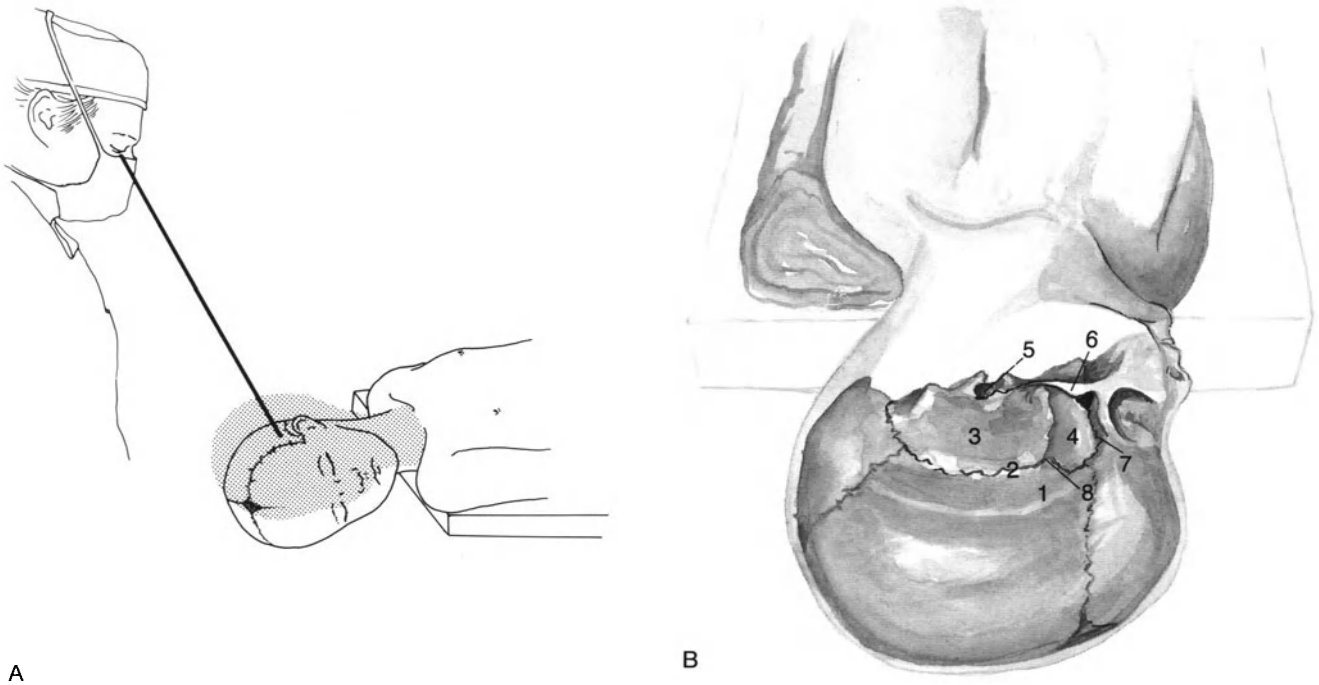


Figure 1.36. (A) The head has been lowered slightly, approximately 15°–20°, giving the surgeon a direct line of vision to the squamous temporal area. (B) Surgeon's view of the operative area. The bony landmarks are the superior (*1*) and inferior (*2*) temporal lines, the squamous temporal (*3*) and greater wing of the sphenoid (*4*) bones, the external auditory canal (*5*) and zygomatic arch (*6*), the "keyhole" area (*7*), and the pterion (*8*).

(Fig. 1.37A). 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. 1.37B). This position permits access to the parietal eminence as well as visualization of the vertex and inion posteriorly (Fig. 1.37C) and of the glabella anteriorly (Fig. 1.37D). 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 of 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 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.

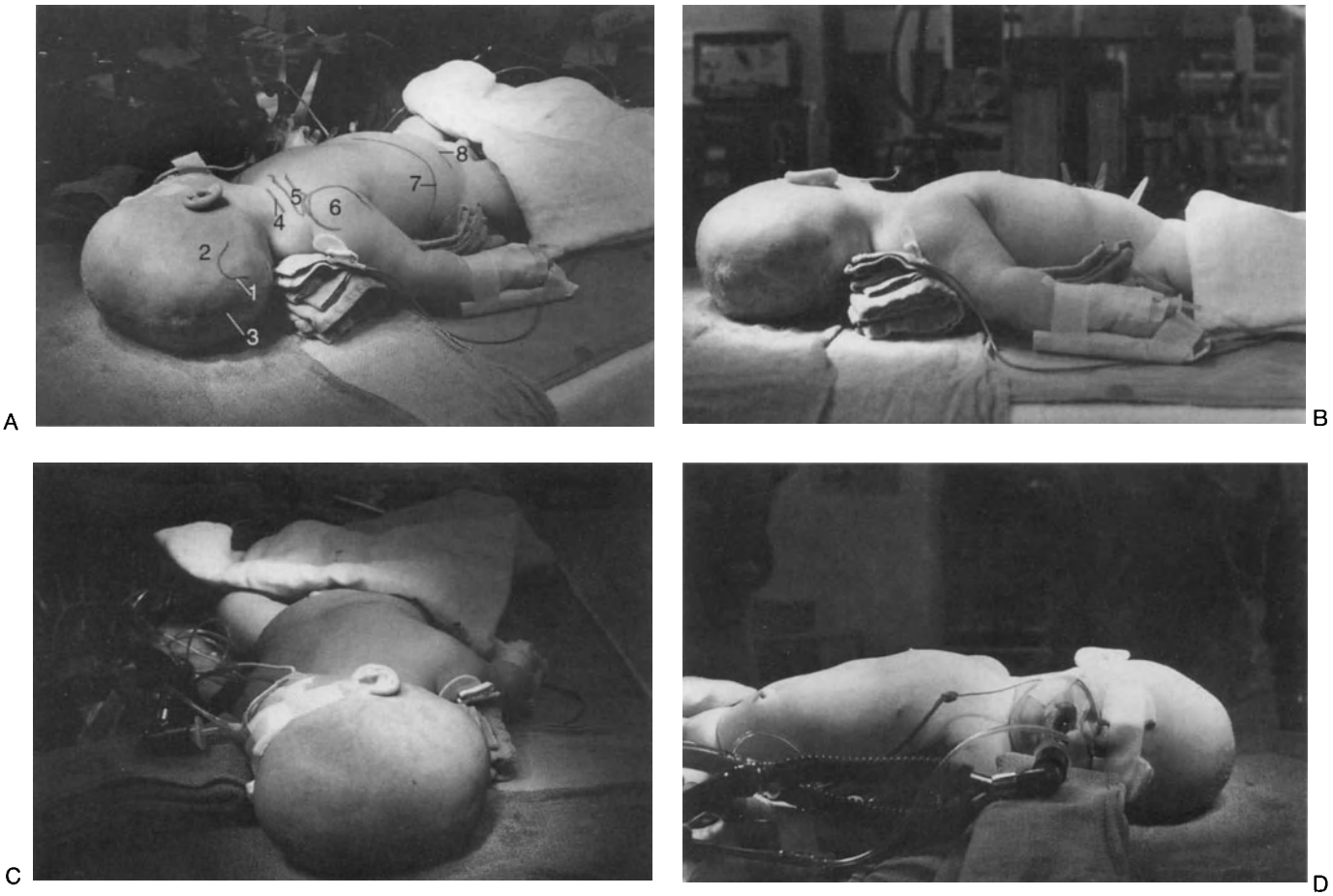


Figure 1.37. Various perspectives of positioning of a child for performance of a ventriculoperitoneal shunt. (A) The horseshoe scalp incision (*1*) is placed midway between the parietal eminence (*2*) and the midsagittal plane (*3*). The supraclavicular incision (*4*) is placed approximately 2.5 cm above 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*.

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. 1.38 A–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 himself so that his line of vision centers along a plane running 45° upward from the horizon (Fig. 1.38 D). Consequently, the decision to operate on the posterior fossa with the child supine 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!

In 1974 Meridy and coworkers[1] reported complications during neurosurgery in the prone position in children. They noted 8% incidence of cardiac arrhythmias, 3% incidence of respiratory complications (one of whom died) 2% incidence of cardiac arrests (two of whom died), and 1.6% incidence of air emboli. They state that "many anesthetists and neurosurgeons advo-

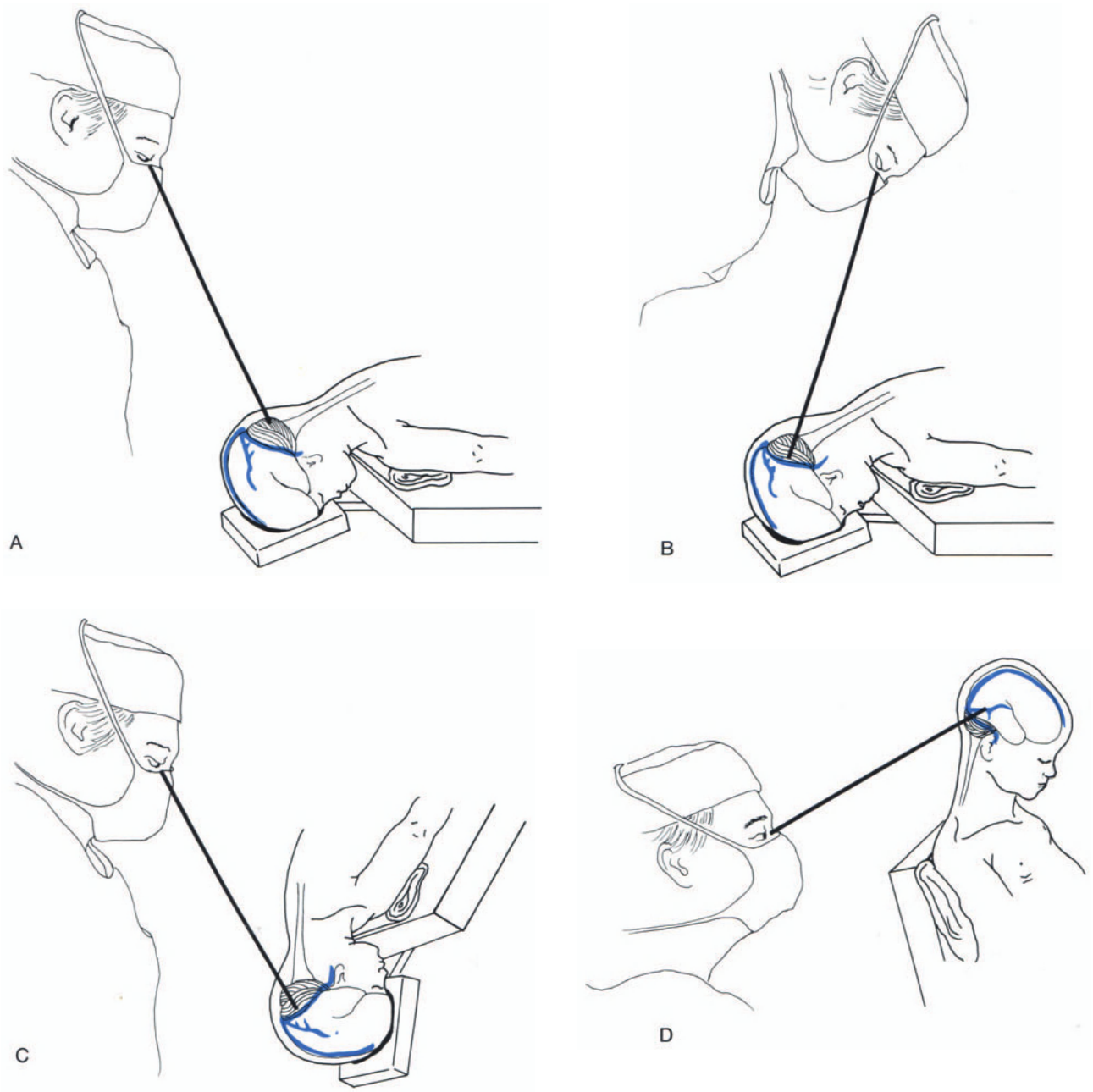


Figure 1.38. With the child prone (A), the surgeon cannot position himself to view the superior cerebellar triangle contents, the superior IV ventricle, or the aqueduct because he is obliged to work from the head of the patient. In order to obtain a line of vision to the tentorial opening, the surgeon would have to elevate his head and move it caudad (B), or bring the child's head so low as to have him 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.

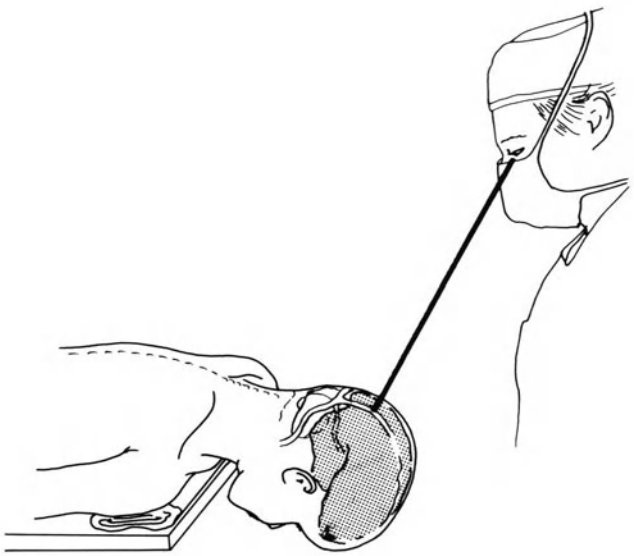


Figure 1.39. Prone position for occipital craniotomy: The head is distracted and then slightly flexed, permitting direct visualization of the inion and both parietal eminences.

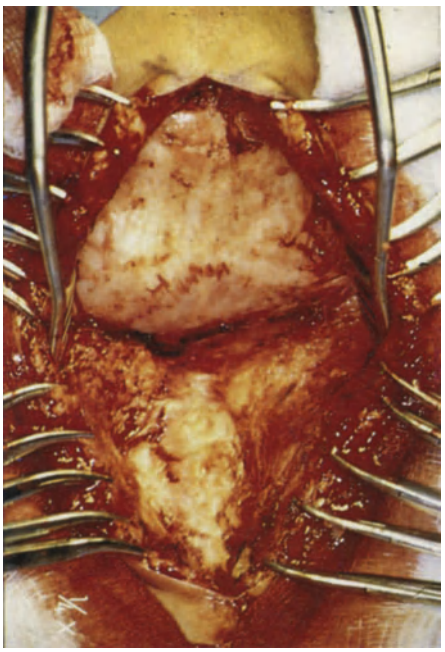


Figure 1.40. The head was not adequately distracted from C-1, so that the latter has come to rest within the foramen magnum.

cate 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 the sitting position. Such pitfalls include the risk 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, *reveal 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] holding over 2000 patients studied.

The most experienced posterior fossa neurosurgeons[3-8] prefer the lounging position. Bucy[9] stated, "For many years I operated in the posterior fossa with the patient lying prone and with the upper part of the body raised so that the long axis formed 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 distracting it. 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. 1.39).

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. 1.40), to permit the surgeon as cephalad a line of vision as possible (Fig. 1.41). 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.

Laminotomy

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

Cervical Laminotomy

Maximum exposure of the cervical spine and spinal

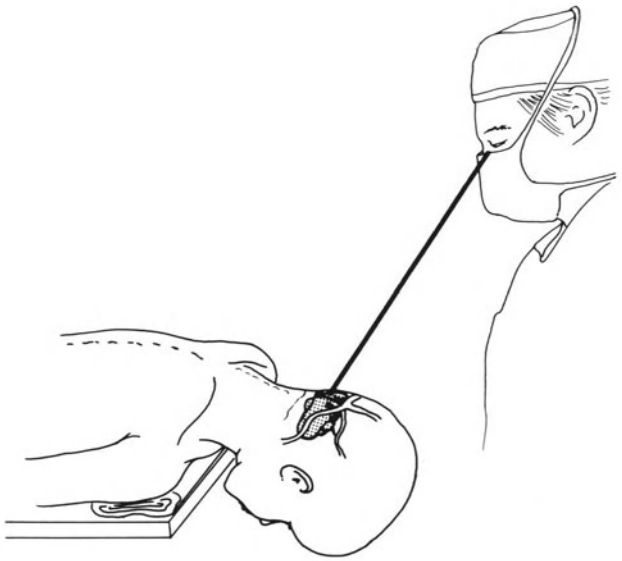


Figure 1.41. The prone position for suboccipital craniotomy, and exposure of inferior cerebellar triangle, with head distraction, separating widely C1 from the foramen mangum, provides maximum view of inferior cerebellar triangle contents.

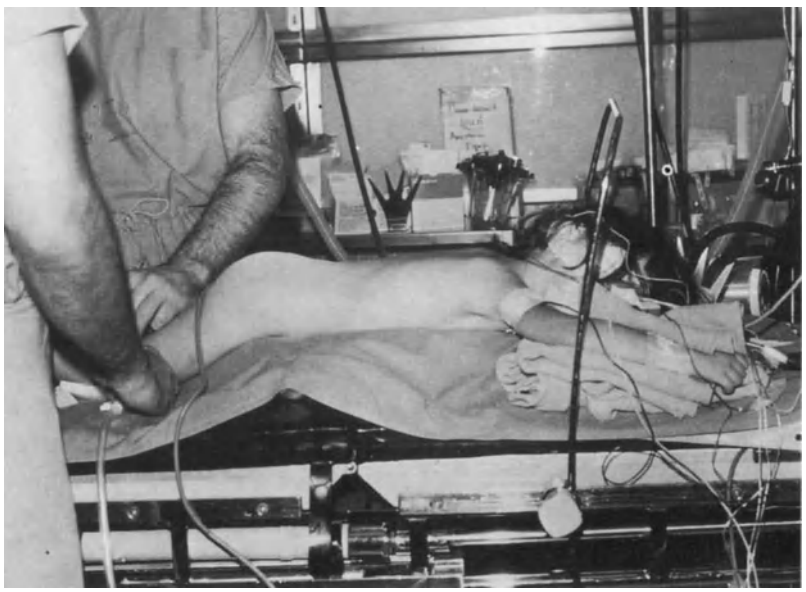


Figure 1.42. Prone position for lumbar laminotomy, thoracic costotransversectomy, or lumbar posterolateral approach for hourglass neuromas extending from the spinal canal into the retropleural or retroperitoneal spaces.

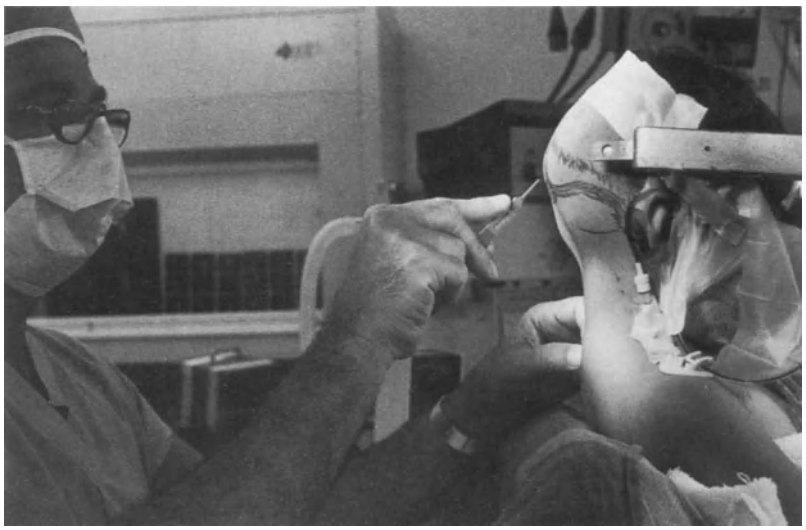
cord is obtained by positioning the child in identically the same manner as for suboccipital craniotomy (Fig. 1.41). This brings the inion into a position such that it does not obstruct the surgeon's line of vision to the atlas, permits him entry into the posterior fossa if needed, and gives him a very complete and direct view of the entire cervical cord.

Thoracic Laminotomy

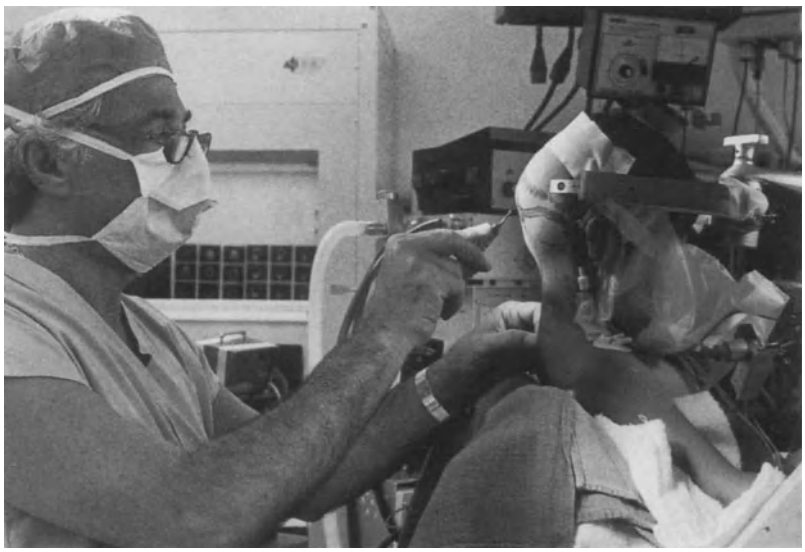
For upper thoracic laminotomies it is best to place the head in a neutral position, distracting it only slightly, but not turning it to either side, whereas for mid- and lower-thoracic laminotomies the head may safely be turned to either side, depending exclusively upon the preference of the surgeon 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. 1.42).



A



B

Figure 1.43. (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 potential range of motion from pronation to supination, from flexion to extension.

Lounging (Sitting) 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. 1.43A, B) and dissecting the muscle from the skull (Fig. 1.44) and atlas, but the setting should allow the table to be elevated when the craniotomy is performed and elevated and tilted forward when opening the dura and entering the superior cerebellar triangle. Also, convenient regulation of the surgeon's height with the operating table, raising or lowering it, adds a significant amount of security to control of the Hudson brace (Fig. 1.45A, B) for either perforator or burr 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 0% when positive/negative ventilation was used. Michenfelder 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

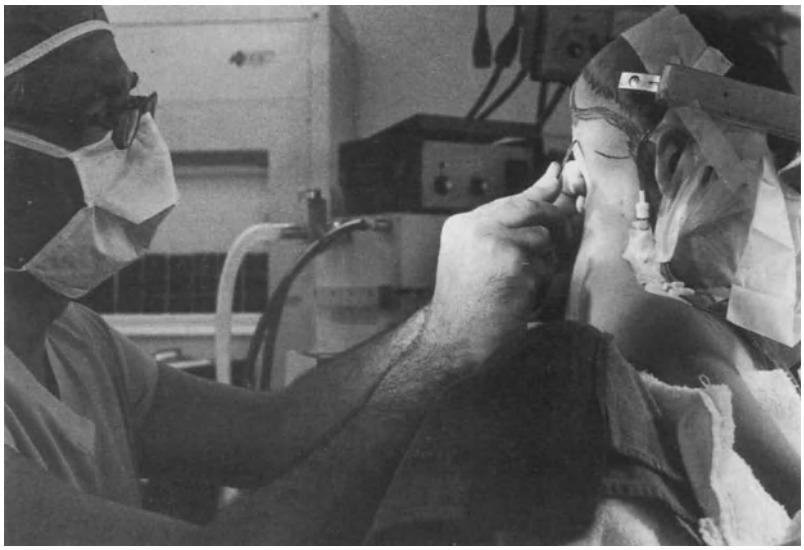
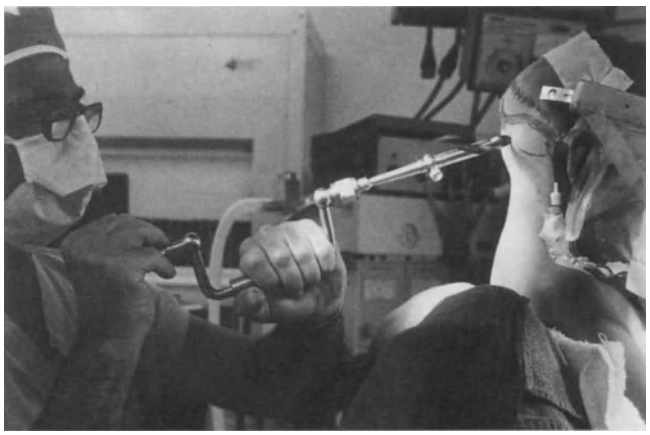


Figure 1.44. This photograph permits the reader to appreciate the strength and sureness of instrument control, which proper elevation of the operating table permits.



A

B

Figure 1.45. (A) The line of force extends from the shoulder through the bracing hand and perforator, with the turning hand moving circumferentially through this axis: maximum strength and control. (B) The same as A but left-handed brace control and cone-down view.

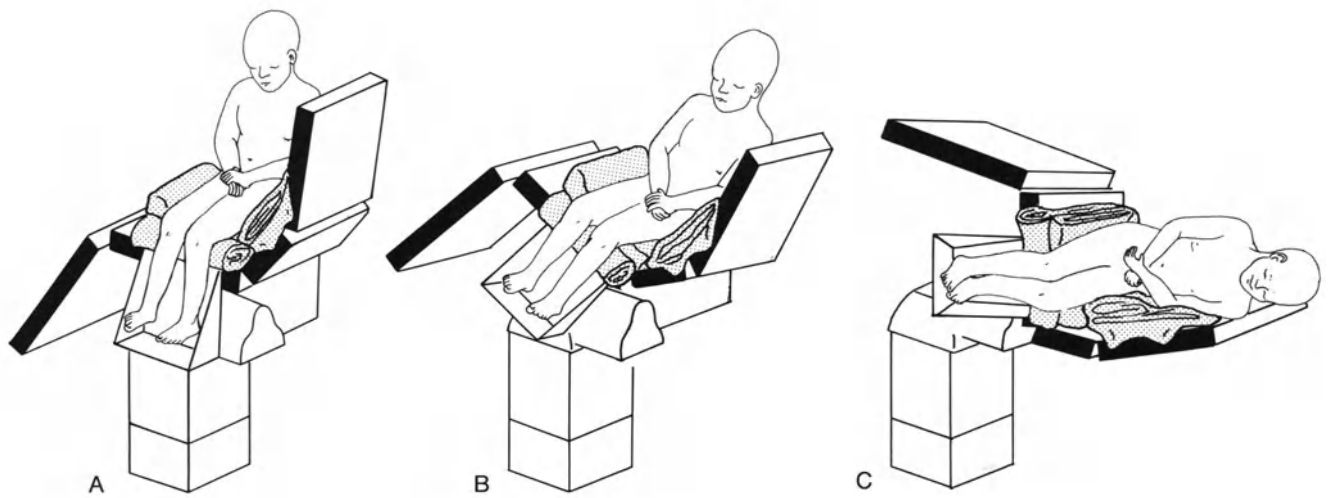
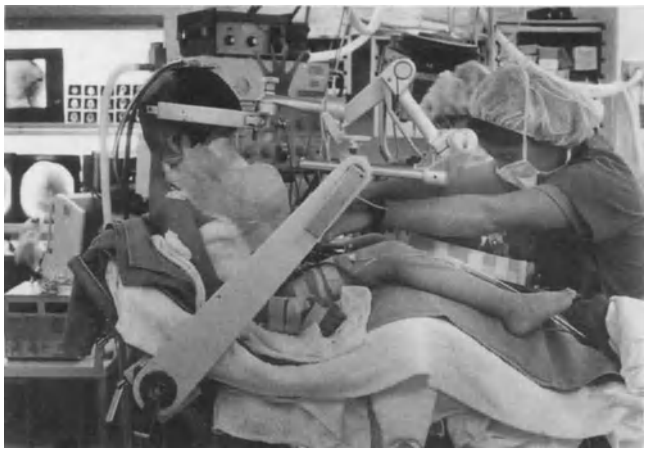
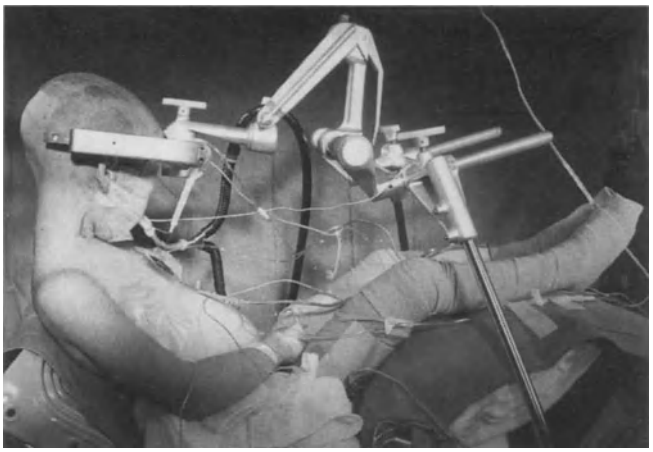


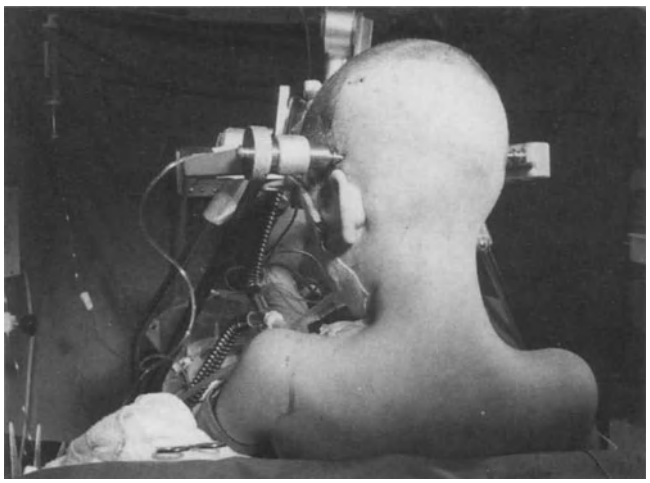
Figure 1.46. (A) The child is positioned sideways, with the vertical axis of his body perpendicular to the long axis of the table, so that his arms are resting on the backrest and his legs are extended perpendicular to the table. Then, pushing the table downward (B) brings the child into the horizontal position (C), without obliging the surgeon to stop his operative procedure.



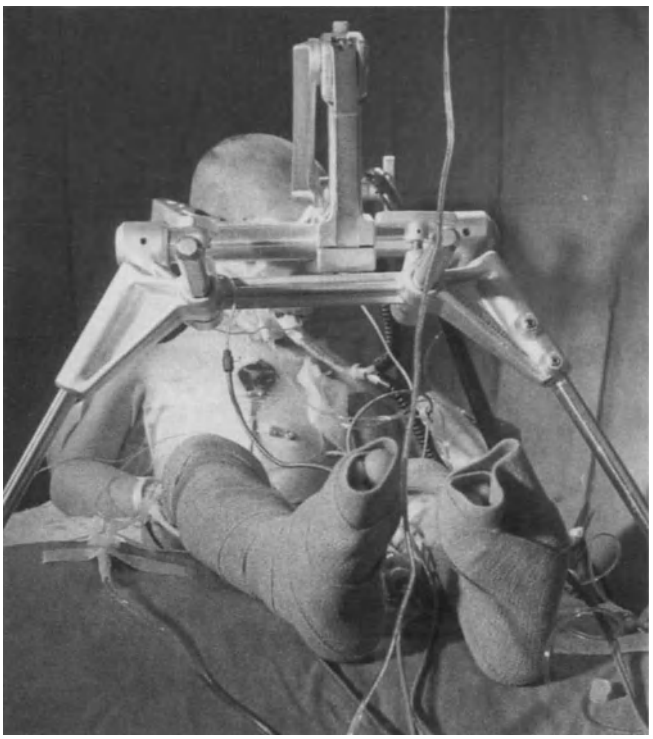
A



B



C



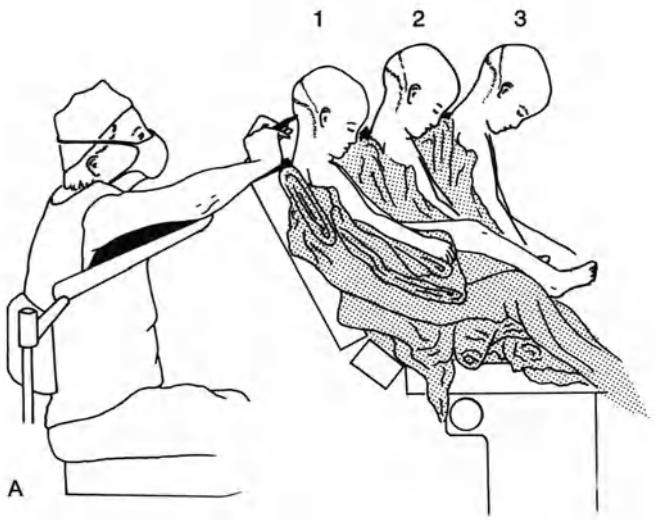
D

Figure 1.47. (A) The lounging position. This is a variation of the sitting position, one which minimizes potential disadvantages of the sitting position but does not eliminate them. Here, the table has been pitched slightly forward. (B) The table is pitched backward, bringing the knees in the same plane as the shoulders. (C) Turning the head slightly on the cervical spine exposes the lateral occipital bone for cerebellar hemisphere or pontocerebellar angle lesions. (D) The anesthesiologist's view.

clinically 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 the patient in the sitting position." In fact, 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 extremely interesting variant of the "sitting" position is one reported by Garcia-Bengochea and coworkers.[12] In brief, it consists of positioning the patient sitting, or, preferably, lounging, but seated sideways on the operating table (Fig. 1.46). Lowering of the table in the event of an air embolus, or other intraoperative complication necessitating positioning the patient horizontal, may be easily, safely, 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 Figure 1.47. This position is used whenever one wants to sit the child up, whether for a midline or lateral posterior fossa craniotomy. It is easier to place a child, especially an infant, in the lounging position than it is to sit him up, since one need only distract and flex the head on the neck at C-1, place a pillow or sandbag across the thoracolumbar area, and center a pillow at the popliteal fossae. Though an upper cervi-



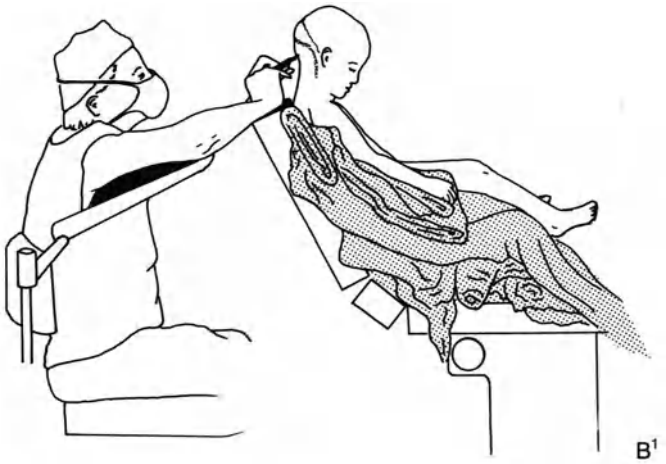
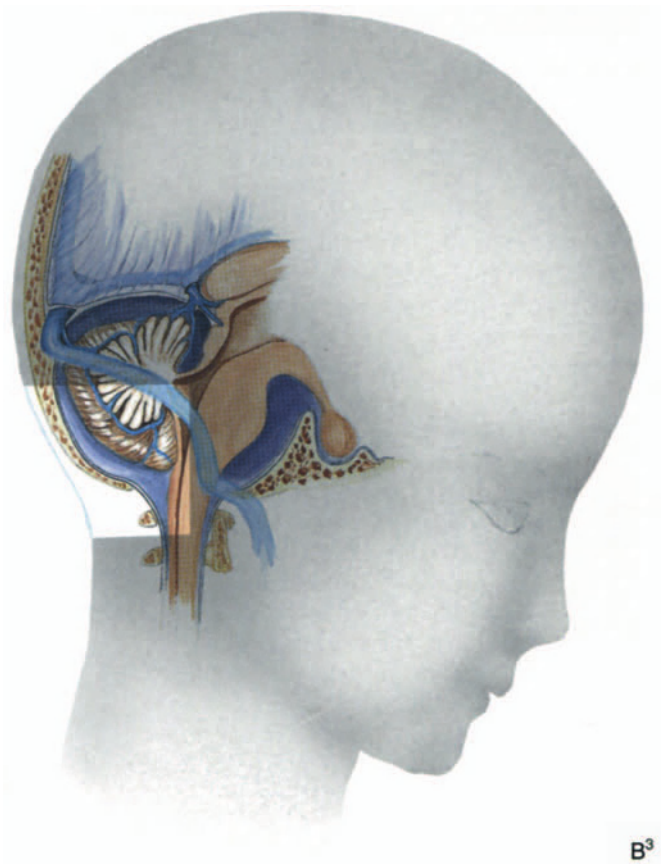
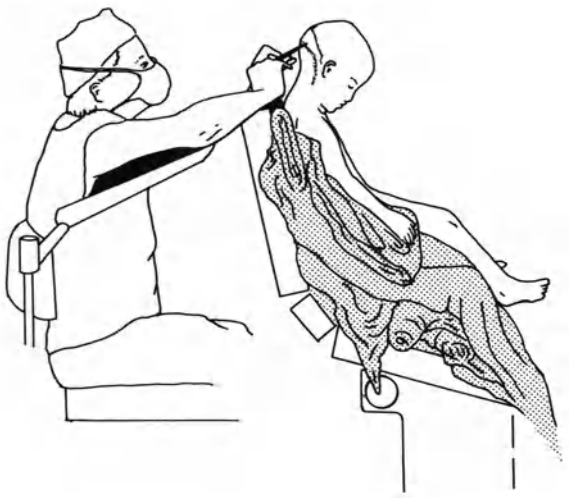


Figure 1.48. (A) The surgeon is seated, his arms on a rest, for skin incision, muscle dissection, craniotomy, and inferior cerebellar triangle work (*1*). Pitching the operating table forward permits access to the IV ventricle and aqueduct (*2*) and superior cerebellar triangle (*3*), Notice that, as this is done, the child's head is progressively elevated. (B) Photographic and diagrammatic demonstration of relative positions of surgeon, child, and posterior fossa contents for opening and exposure of inferior cerebellar contents. (C) As the operating table is rotated forward, the surgeon's arms are somewhat extended to permit work within the IV ventricle. (D) Exposure of the superior cerebellar triangle structures and the region of the quadrigeminal cistern necessitate further forward rotation of the child and extension of the surgeon's arms.

cal laminotomy may be performed 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. 1.48 A–D). As the child is rotated forward, the surgeon must both elevate his chair and extend his arms.

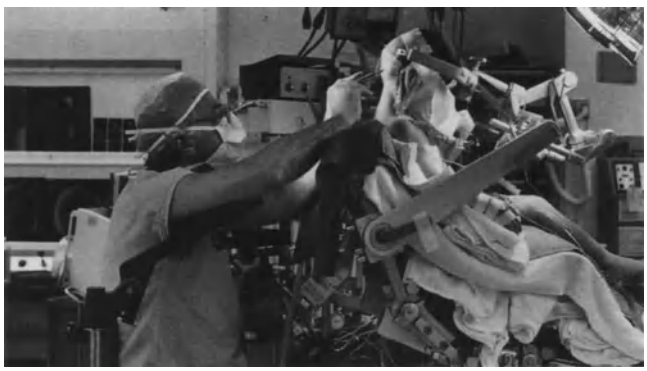
This position is ideal for occipital and suboccipital craniotomies (whether midline or lateral), burr holes (unilateral or bilateral, diagnostic or therapeutic), and mid- or lower-cervical laminotomy, and for bilateral ventriculoperitoneal shunts (whether shunting from occipital or frontal horns). It 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.





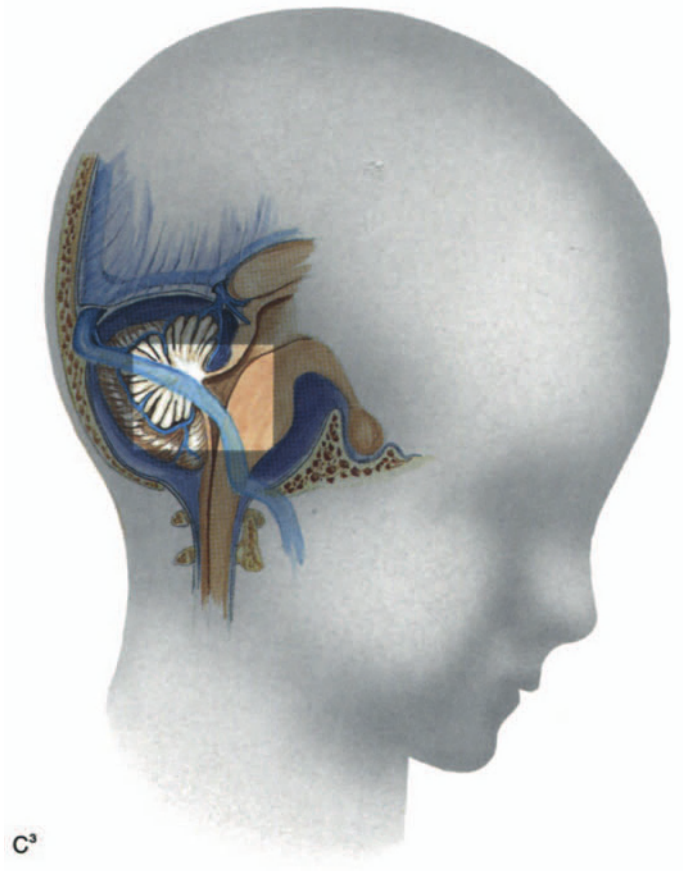
C¹

D¹

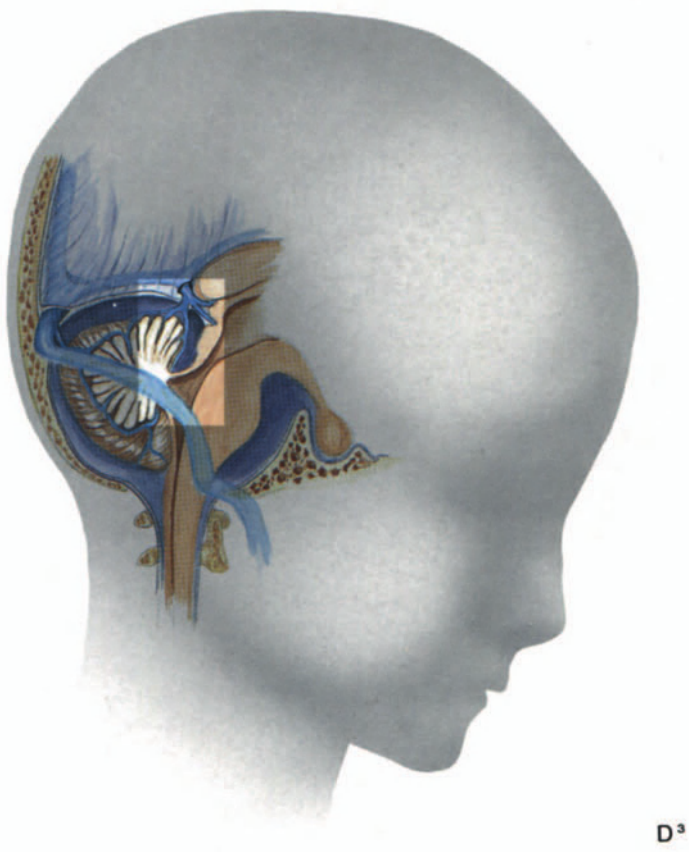


C²

D²



C³



D³

References

1. Meridy HW, Creighton RE, Humphreys RP: Complications during neurosurgery in the prone position in children. Can Anaesth Soc J 21:445–453, 1974.
2. Michenfelder JD, Martin JT, Altenburg BM, Rehder K: Air embolism during neurosurgery. An evaluation of right-atrial catheters for diagnosis and treatment. JAMA 208:1353–8, 1969.
3. Drake CG: Total removal of large acoustic neuromas. A modification of the McKenzie operation with special emphasis on saving the facial nerve. J Neurosurg 26:554–61, 1967.
4. Koos WT, Miller MH: Intracranial Tumors in Infants and Children. St. Louis, Mosby, 1971, p 415.
5. Yasargil CG: Microsurgery Applied to Neurosurgery. Stuttgart, Thieme, 1969, p 230.
6. Kempe LG: Operative Neurosurgery. New York, Springer-Verlag, 1970, vol 2, p 281.
7. Symon L: Control of intracranial tension. In operative neurosurgery. In Logue (ed): Neurosurgery. London, Butterworths, 1971, p 1.
8. Decker RE, Malis LI: Surgical approaches to midline lesions at the base of the skull; A review. J Mt Sinai Hosp 37:84–102, 1970.
9. Bucy PC: Exposure of the posterior or cerebellar fossa. J Neurosurg 24:820–32, 1960.
10. Marshall BM: Air embolus in the neurosurgical anesthesia: Its diagnosis and treatment. Can Anaesth Soc J 12:255–61, 1965.
11. Michenfelder JD, Miller RH, Gronert DA: Evaluation of an ultrasonic device (doppler) for the diagnosis of venous air embolism. Anesthesiology 32:164–7, 1972.
12. Garcia-Bengoechea F, Munson ES, Freeman JV: The lateral sitting position for neurosurgery. Anesth-Analg (Cleve) 55:326–30, 1955.

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

SOULEYMANE CISSÈ
Thoughts

Chapter 2

Incisions

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

Skin (Figures 2.1 and 2.2)
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 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 fontanels in the newborn, infant, and toddler determine the differences in technique for skin incisions 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 fontanels 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 Figure 2.1, showing scalp characteristics, the presence or absence of a fontanel, and the state of the sutures are illustrated in a comparative manner so as to put into relief the existence of thin scalp, the presence or absence of fontanel, and opened or closed sutures. The very thin scalp, open fontanels, and sutures are illustrated in Figure 2.1 A, thickening of the scalp and disappearance of the fontanels are illustrated in Figure 2.1 B, and further thickening of the scalp with closure of the sutures are illustrated in Figure 2.1 C.

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 # 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 two former 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 newborns 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 that the sutures may be open from a few millimeters to several centimeters, so that the suture

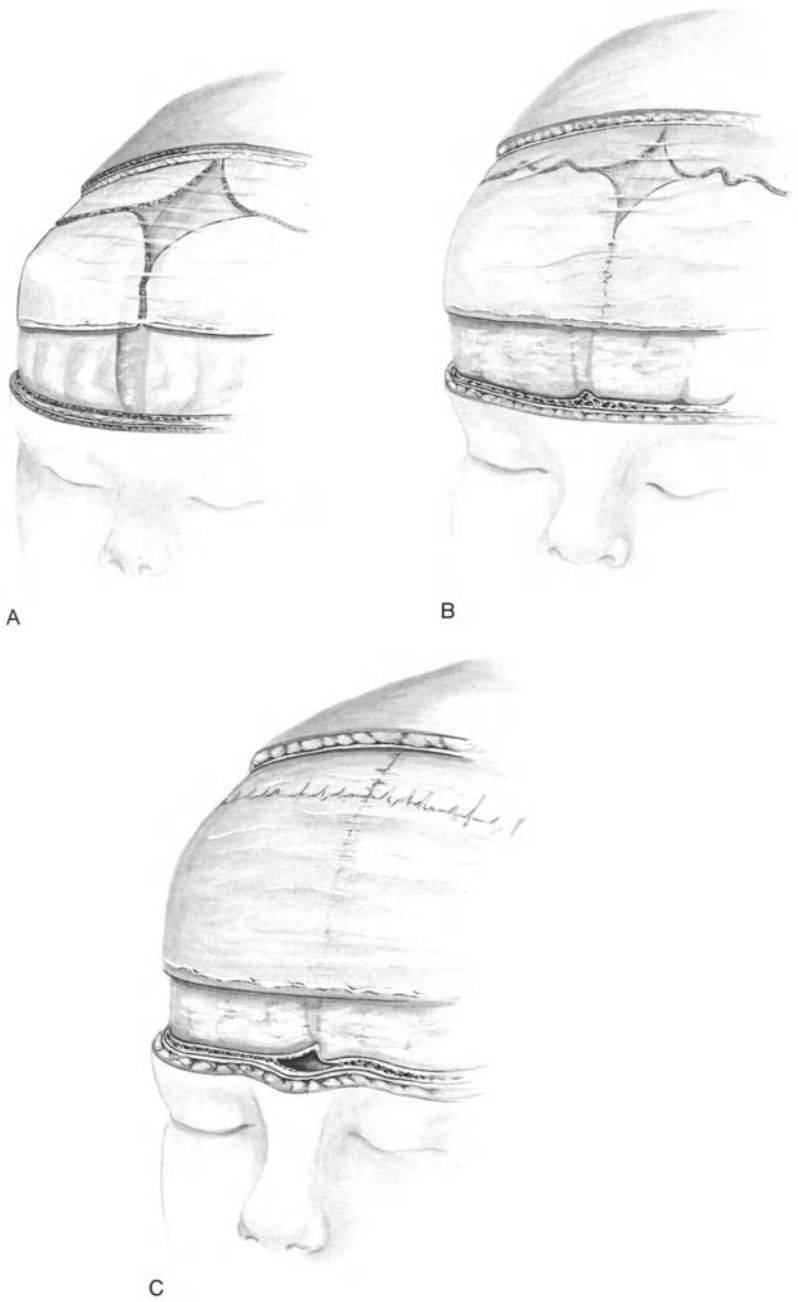


Figure 2.1. The relative thickness of scalp, presence of fontanel, and state of the sutures in the newborn, infant, and toddler. Note that in the newborn the skin, connective tissue, and aponeurosis (Galea) are quite thin, whereas the fontanels and sutures are open. With progressive age, the skull thickens, the sutures close, 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, explaining the mobility of the outer layers of the scalp, the periosteum, and bone. (B) Infant: The connective tissue thickens and becomes more vascular, the fontanels disappear, 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 indentifiable tables (outer and inner) and diploë.

line—namely, periosteum and the outer layer of the dura—a purely membranous structure, is the only anatomical barrier between Galea and the surface of the brain.

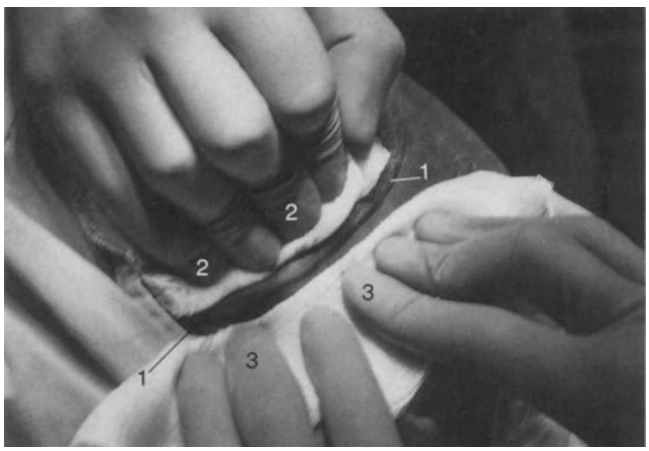
Also, when attaining scalp hemostasis (during the time of skin incision) by compression of the scalp, one must take care to compress *very gently,* since the underlying skull is thin and fragile, susceptible to compression fracture. Consequently, the pressure put upon the scalp should be just enough 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 down upon 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.

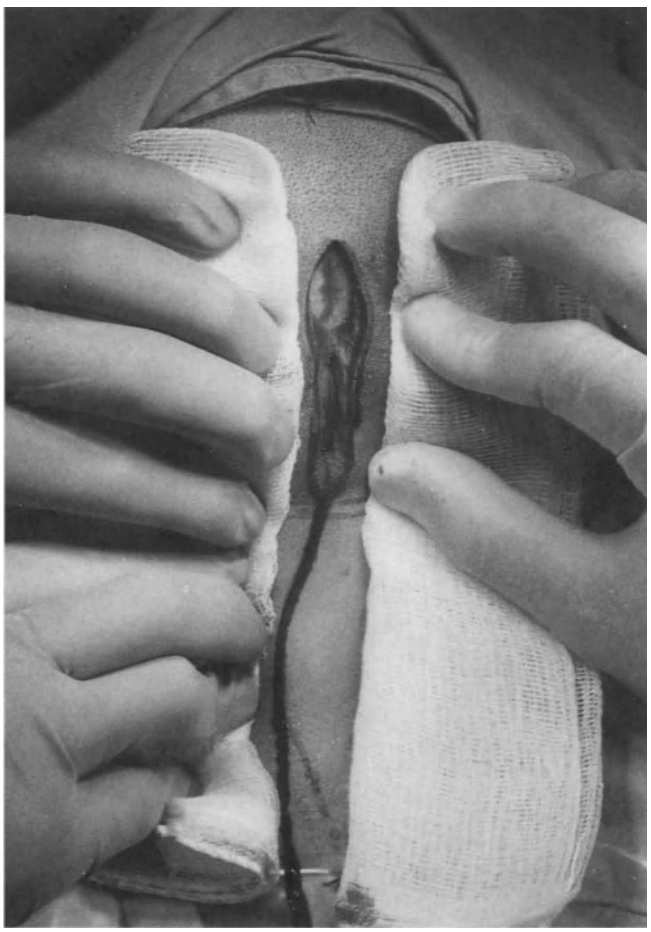
In the toddler, juvenile, and adolescent, scalp compression must be more forceful, so as to wedge smartly the vessels within the loose connective tissue of the scalp between the surgeon's fingers and the skull.

Galea (Figures 2.3 to 2.7)

The application of Kolodny and Dandy clips (to the Galea) for scalp hemostasis in the newborn and infant

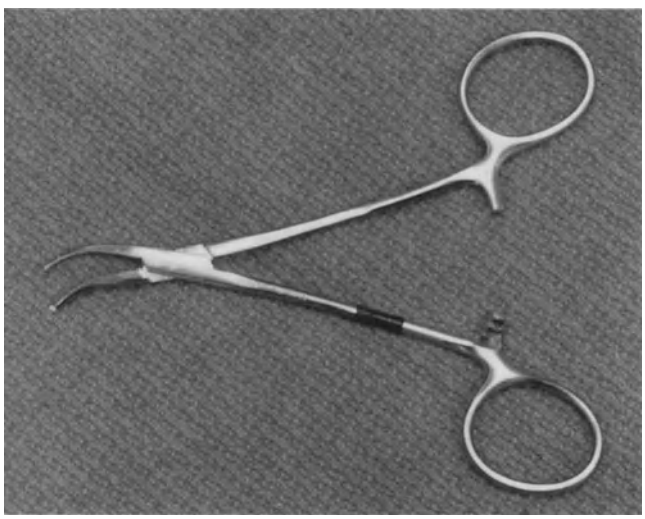


A

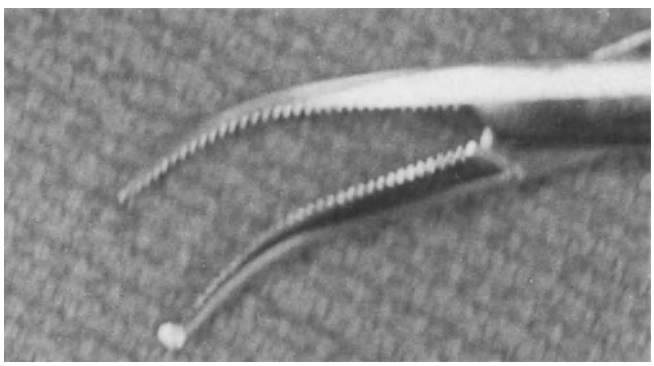


B

Figure 2.2. (A) The incision line (*1*) has been drawn with a marking pen. The full 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 fingertips. Note that the fingers compressing the scalp at the reader's left are "cutting" into the skin, with the compression being exerted entirely 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 (at the right) is undesirable.



A



B

Figure 2.3. (A) The lightweight (aluminum)-toothed Raimondi clip. Note that the curvature of the tip is in the same plane as the rings of the shank. (B) A cone-down view illustrating the serrated jaws with a toothed tip, designed to grip the Galea.

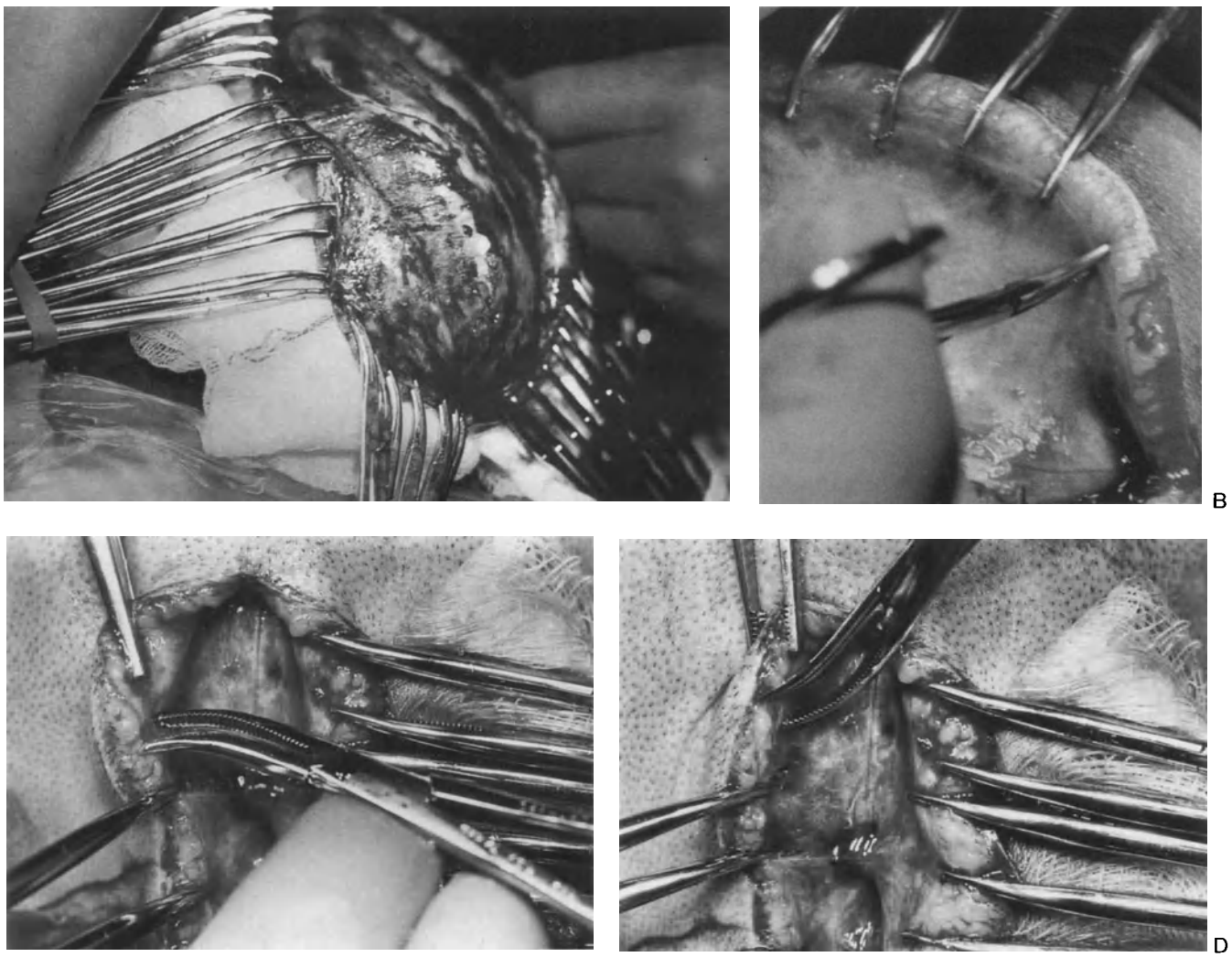


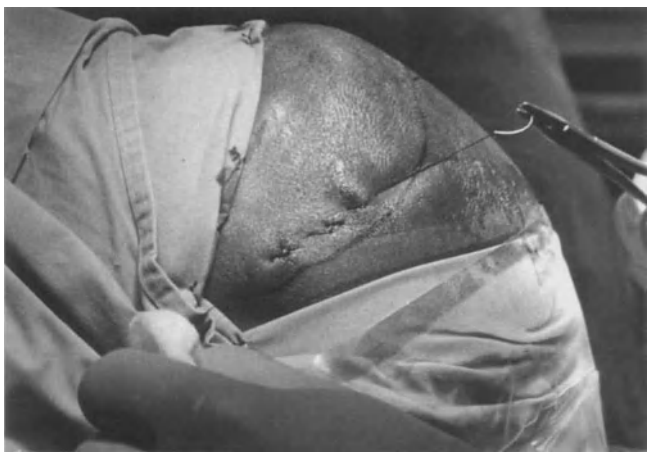
Figure 2.4. (A) The clips are applied to the Galea as the tooth forceps is used to reflect the skin, dense connective tissue, and Galea in such a manner as to expose the cut Galea edge. The interval between clips is approximately 4 mm. Note that the applied clips are gathered into clusters (which are held together with a rubber band). All of the clips have been applied to the edges of an S-shaped incision and collected into clusters. These are then splayed over a sponge which serves to eliminate the risk of kinking the scalp. (B) The delicate, toothed clips permit locking onto the Galea without damaging the dense connective tissue on skin. (C) 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. (D) Same as C, but a cone-down view.

is of no value whatsoever 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, specially designed for use in the newborn and infant*, 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 they are made of light aluminum alloy. They neither take up a great amount of space over the small cranium nor weigh heavily on it.

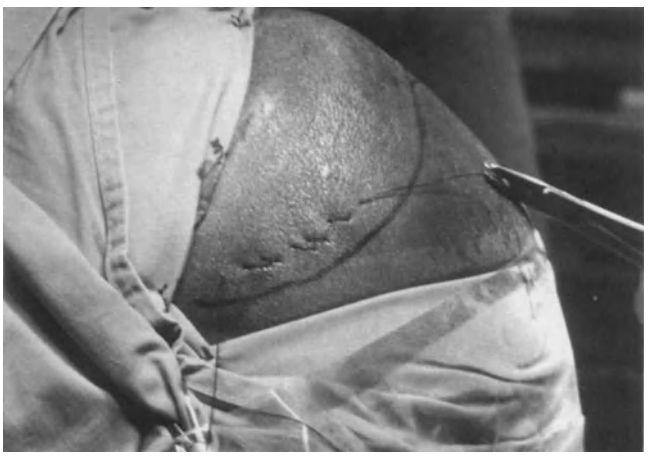
After the clips have been applied to the Galea, at approximately 4-mm intervals, and fastened to one another by a rubber band, they may be used to retract the scalp flap. Caution must be taken not to pull on them, since the Galea in the newborn and infant is almost membranous and, consequently, may easily be torn, allowing bleeding to continue. The clips are applied serially, at 4-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 prevents kinking of the scalp flap, something that could result in devascularization of the flap.

* Codman (catalogue # 30-1360), Codman & Shurtleff, Inc., Johnson & Johnson Company, Randolph, Massachusetts 02368.

A



B



C

Figure 2.5. The continuous overlapping suture is placed about 1 cm from the planned line of the skin incision. Placing it obliquely provides greater vascular occlusion. (A–C) The progressive suture placement.

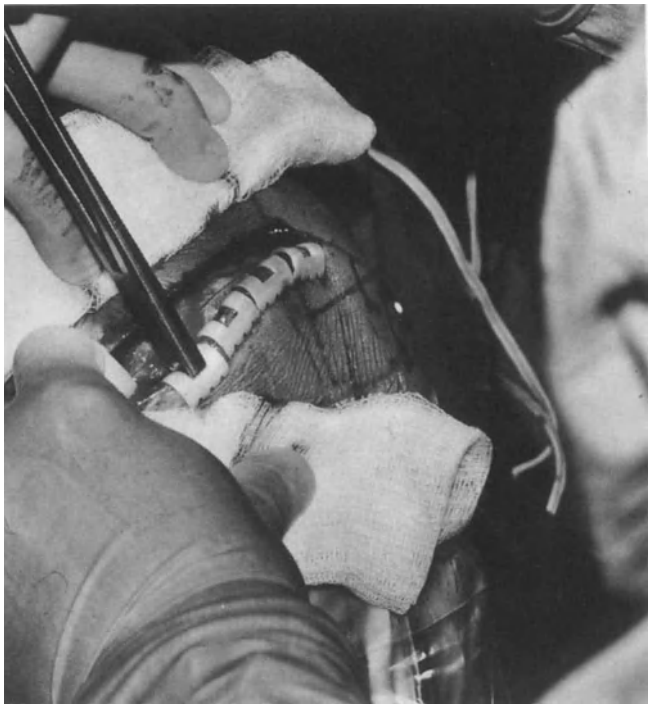


Figure 2.6. The Rainey clips are placed so that they embrace the skin and Galea, constricting the vessels within dense connective tissue between their jaws. They have the advantage of being lighter in weight and less bulky than the Galea clips, permitting the surgeon to move more rapidly. Unfortunately, they are so designed as to be of value only in adolescents, not in younger children.

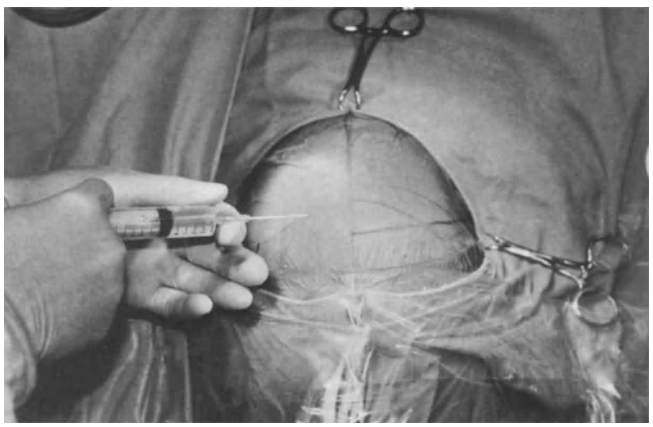


Figure 2.7. Saline is being injected into the dense connective tissue between the skin and the Galea. If one chooses to use this technique to minimize bleeding during the skin incision, it is essential that the saline be injected *into* the dense connective tissue, not into the subGaleal space. The surgeon may know that he is injecting into the dense connective tissue when he finds that it is extremely difficult to inject the saline and that the skin blanches. If he sees that the skin swells up from the underlying skull, the fluid is being injected into the subGaleal space, where it will be of no value with regard to hemostasis.

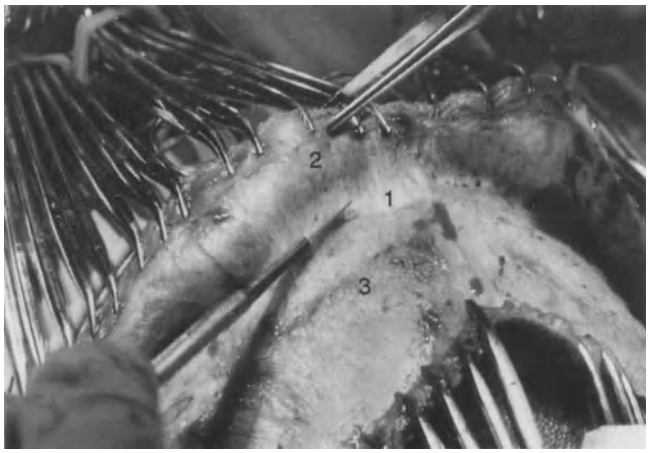
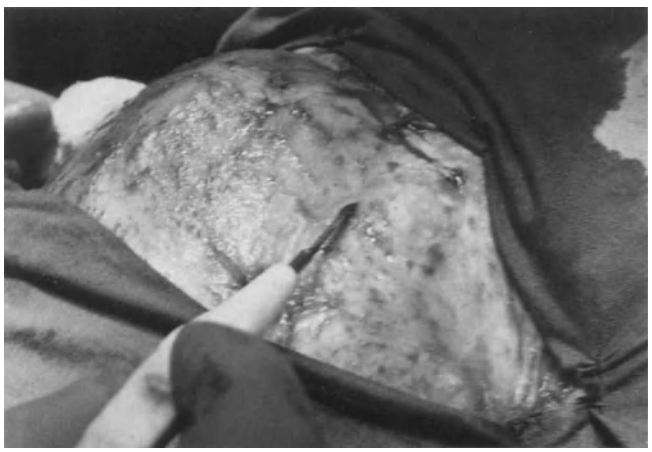
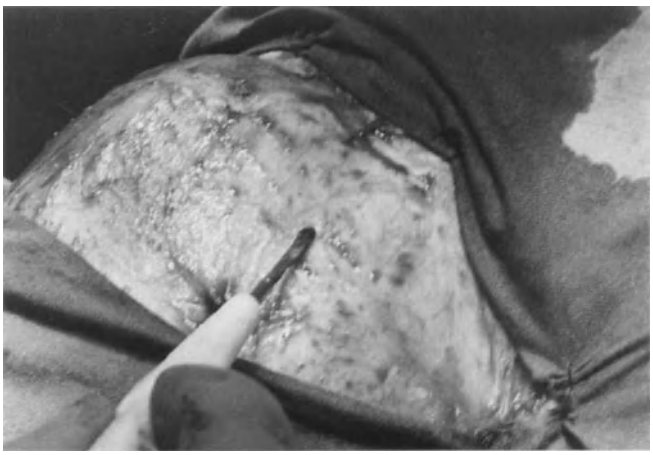


Figure 2.8. The loose connective tissue (*1*) is cut with a # 15 blade, preserving Galea (*2*) and periosteum (*3*). The frontal portion of a bifrontal flap is being separated.



A



B

Figure 2.9. The monopolar thermocautery unit cuts the periosteum very effectively, minimizing periosteal bleeding. One should use the sharp (A), not the flat (B), surface of the blade.

Besides the use of Galea clips, scalp bleeding may also be controlled by inserting a continuous, overlapping suture approximately 5 mm from, and parallel to, the planned line of the skin incision before cutting the skin. This is, in my opinion, a less desirable procedure than application of Galea clamps, since it often results in occluding proximal segments of the subGalea vessels (branches of the frontal, superficial temporal, and occipital arteries). These vessels, if ligated, will not bleed when they are cut. Therefore, they may not be coagulated during the reflection of the scalp flap, presenting the risk of subGalea bleeding after the scalp flap has been closed. Rainey clips may be used in adolescents, but are of no value in younger children because the jaw closure is too wide. Some surgeons infiltrate the dense connective tissue between the skin and the Galea with saline, so as to compress the vessels and diminish bleeding. This is of no value. It is also extremely difficult in newborns and infants.

Loose Connective Tissue and Periosteum

(Figures 2.8 to 2.10)

After the skin incision has been made, the separation of the Galea from the periosteum is quite easy and is very often completed by blunt dissection with a gauze sponge to fray and tear loose connective tissue bridging the two. This is rough and damages the tissue. Moreover, it causes bleeding from the Galea and the periosteum, tearing vessels within and between the two, tiny vessels over a large surface area, which would escape damage if a cutting blade were used. Blunt dissection also disrupts adjacent tissue. 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, most important of all, 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. Temporal and posterior frontal branches of the trifacial nerve may be damaged within the loose fatty tissue just posterior to the zygomatic process of the frontal bone. This causes flattening of the scalp over the forehead and results in an inability to raise the eyebrows.

One may save the frontal artery, nerve, and vein by using sharp dissection of the loose connective tissue that

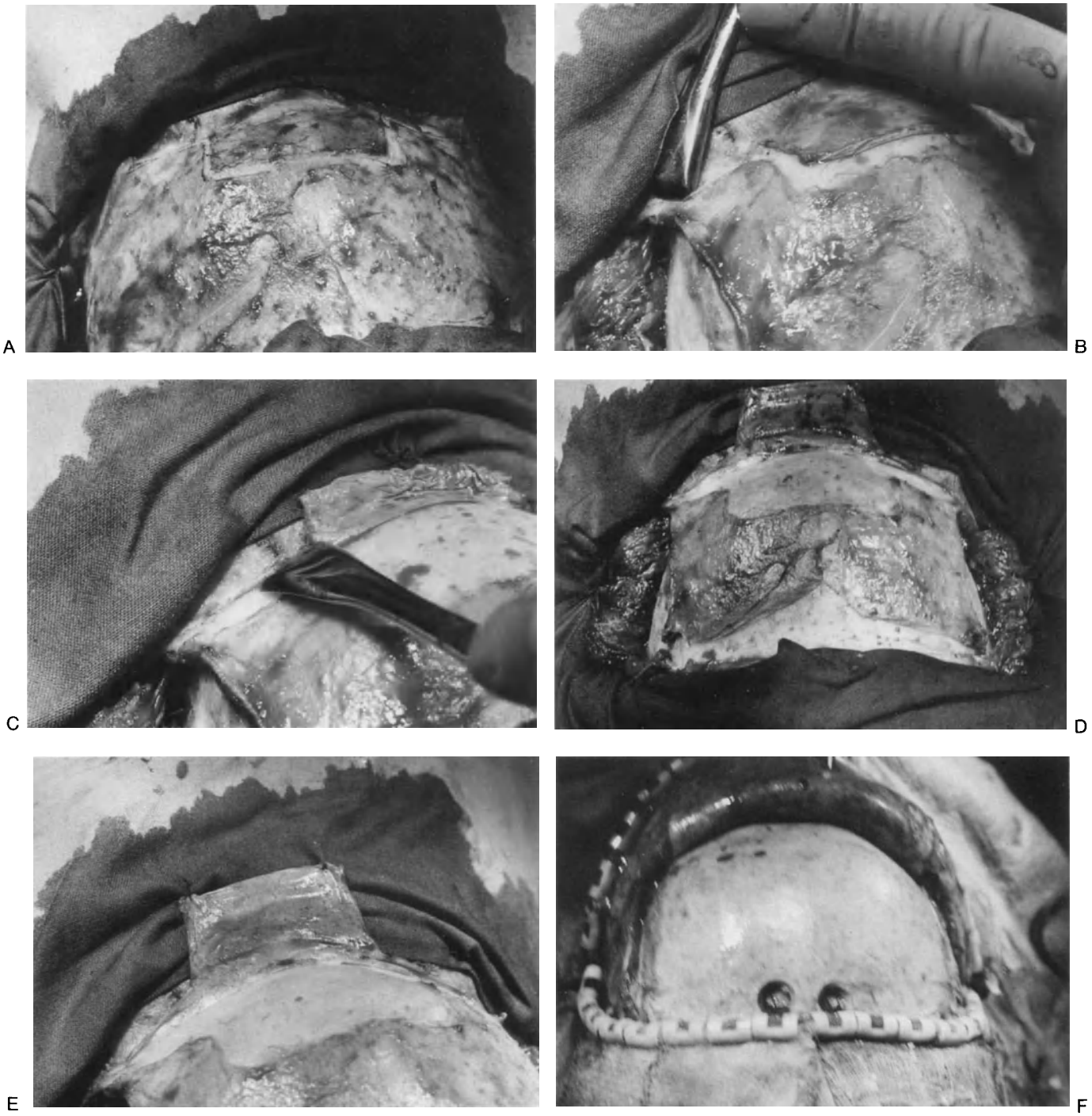


Figure 2.10. A periosteal flap, large enough to be sewn over the frontal air sinus and onto the dura, so as to protect the epidural space form empyema, is being fashioned. (A) The periosteum has been incised. (B) A periosteal elevator is used to separate periosteum from skull without fraying the former. (C) The periosteal elevator is separating periosteum from the superior orbital ridge without damaging the superior orbital artery, nerve, or vein. (D) All of the periosteum has been incised and stripped from the skull along the planned craniotomy line. (E) This is a view of the periosteal flap, illustrating the technique for sewing it up to the drapes so that it may be kept stretched—to avoid shriveling—throughout the operation. (F) This illustrates the *incorrect* technique for reflecting the scalp flap: the periosteum should not be taken with the Galea, since the free bone flap then is completely denuded and healing impaired.

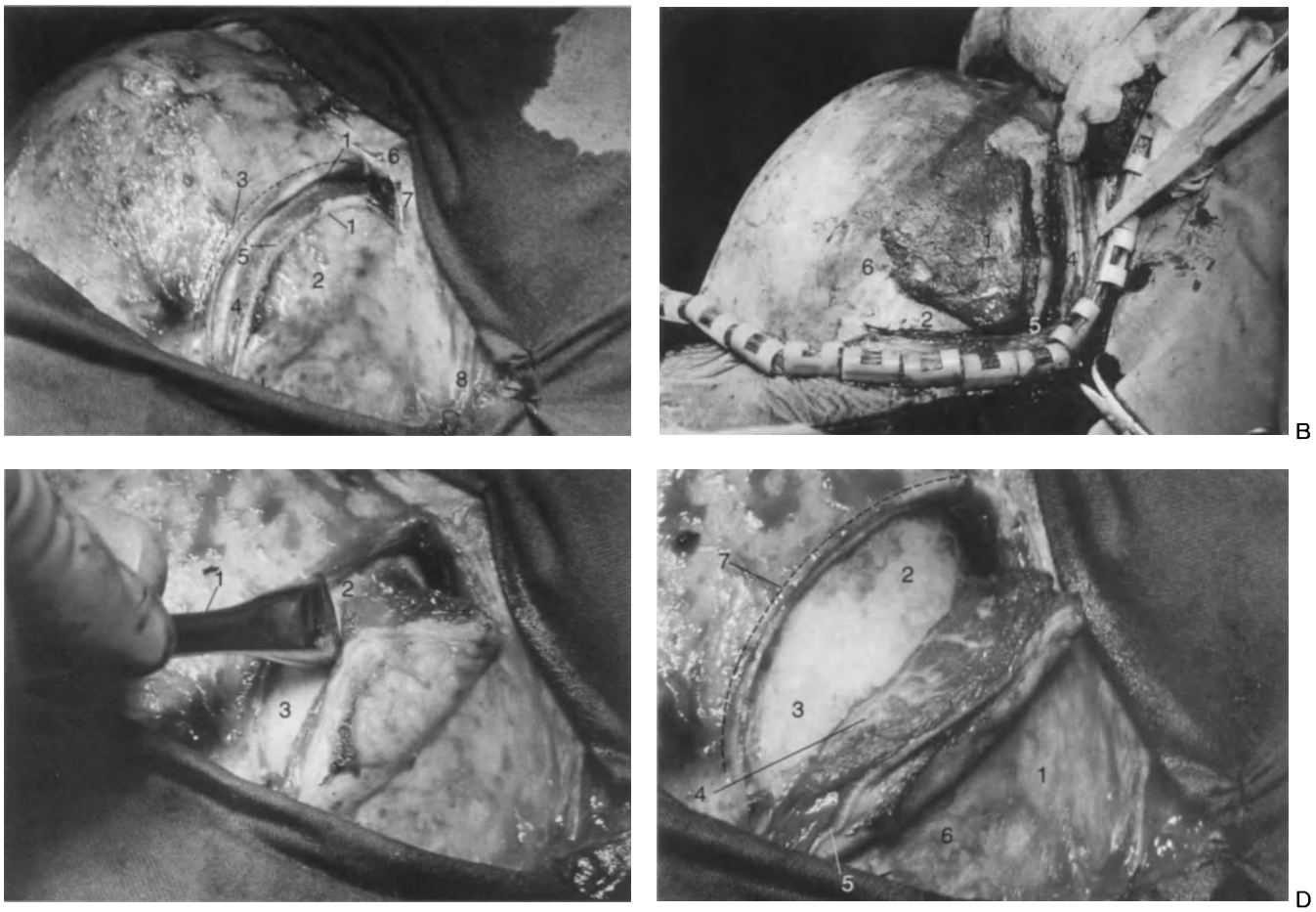


Figure 2.11. (A) Line of incision (*1*) in temporalis muscle (*2*), so as to minimize bleeding and facilitate anchoring bone flap into position at the time of closure. The fascia of the temporalis muscle has been cut about 3 mm from the superior temporal line (*3*) and then reflected to show full thickness 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 superfical (*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 of closure.

bridges the potential space between the periosteum and Galea. Ifl 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 burr holes. This assures complete union of the free bone flap to the surrounding skull.

As will be described subsequently in the section on craniotomy, the craniotome is not to be used to cut

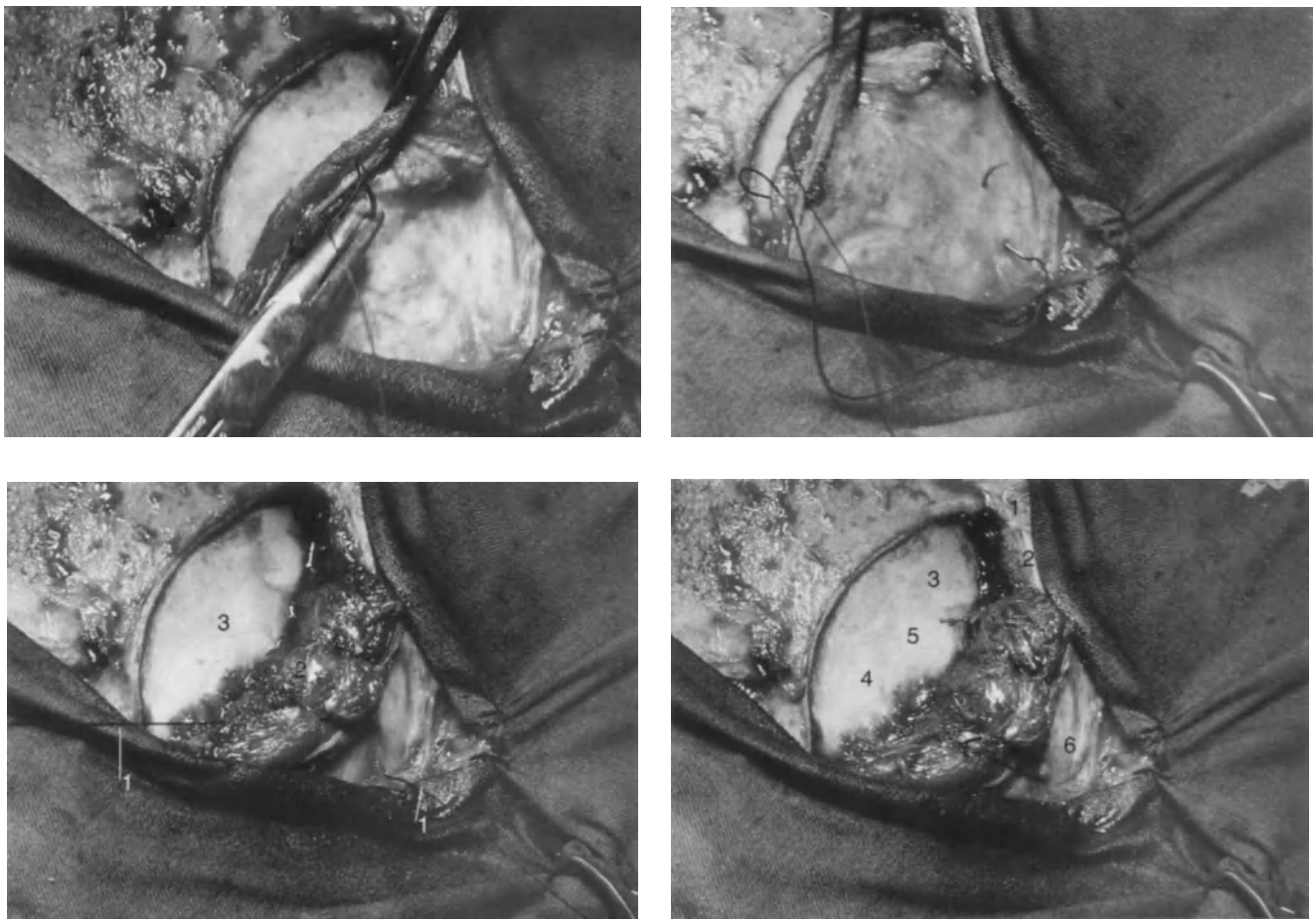


Figure 2.12. Technique for retraction of the temporalis muscle from the pterional region. (A) A suture is sewn through the full thickness of the temporalis muscle, which is held with a tooth forceps. (B) The suture is then brought over the surface of the temporalis and sewn onto the same muscle immediately 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.

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.

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 (Figures 2.11 and 2.12)
When soft tissue preparation for reflection of the bone flap entails incision of the temporalis muscle, this 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 zygomaticofrontal suture. This cut should run superiorly for about 5 or 6 mm, and then extend posteriorly. It is run 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

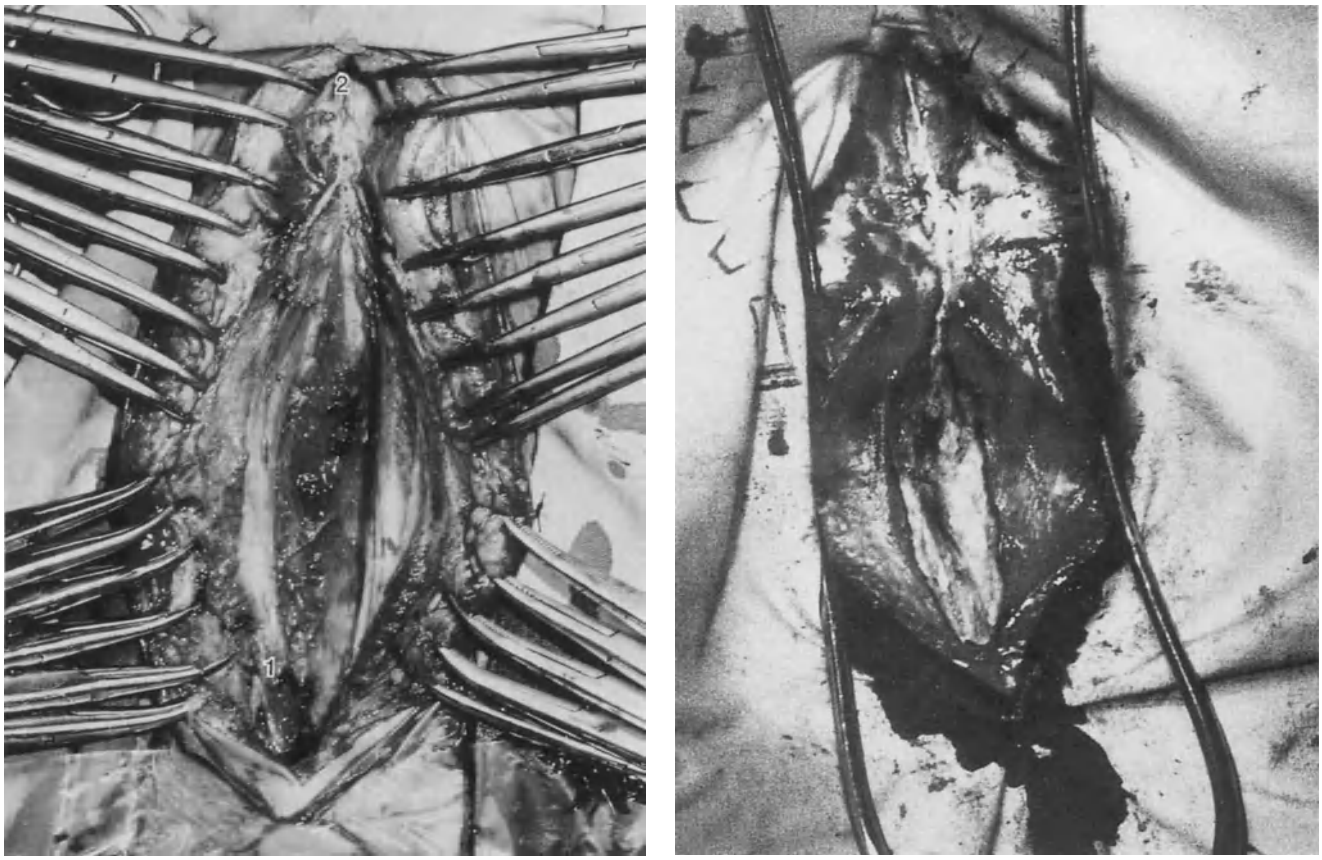
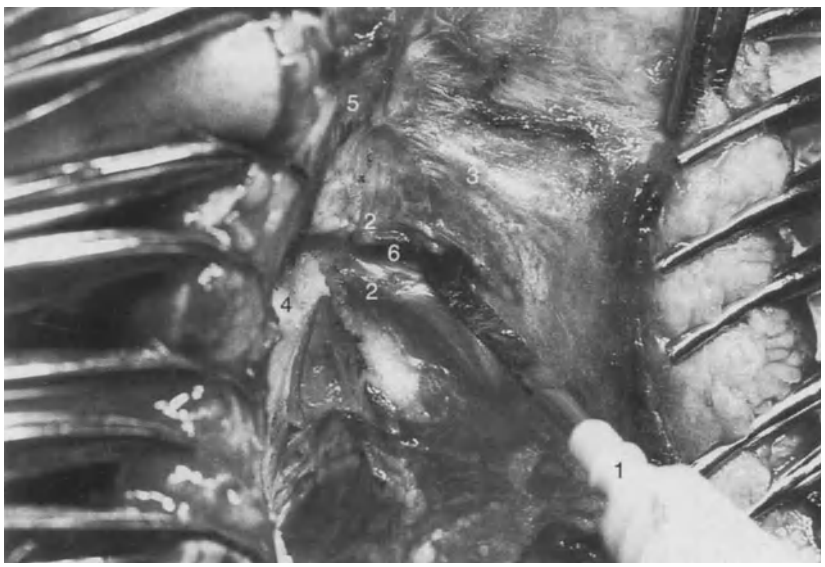


Figure 2.13. (A) After the skin incision is made and the Galea clamps applied, the Bovie knife is used to cut through the ligamentum nuchae from C-2 (*1*) to the inion (*2*). This permits one to separate bloodlessly the erector capitis muscles from one another. It is recommended that self-retaining retractors not be used throughout this stage of the procedure because they deform the normal anatomy and stretch cut blood vessels. This latter often results in providing *temporary* hemostasis, a distinct disadvantage, since the surgeon does not stop these potential bleeders until the end of the operative procedure. Those vessels that may have retracted into muscle may not be completely clotted. This is a source of postoperative hematoma. (B) After the dissection has been brought to the periosteum and both the erector capitis and erector cervices muscles (to the level of C-2) separated from one another, drapes are sewn to the skin edges and the self-retaining retractors are applied. (C) 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*). (D) The periosteal elevator is being used to free the left superolateral musculotendinous

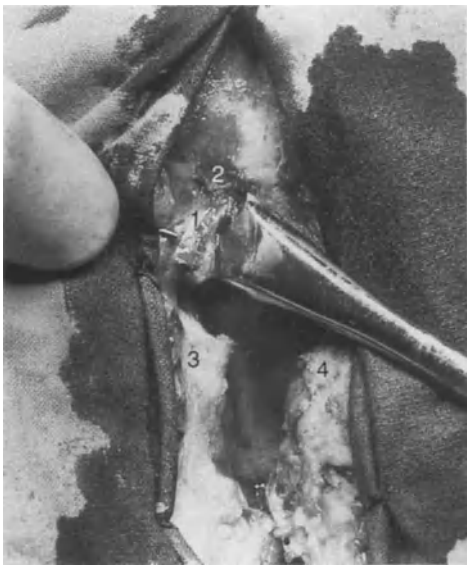
for the sutures, which will bring the temporalis muscle back into anatomical position at the time of closure. The temporalis muscle should not be cut through its belly nor along its origin from the zygomatic arch. This devascularizes it.

The periosteal elevator may then be used to sweep the temporalis from its insertion onto the bones around the pterion, exposing the frontal, parietal, squamous temporal, and greater wing of the sphenoid. Retention sutures may now be used to hold the temporalis muscle flap away from the pterional area.

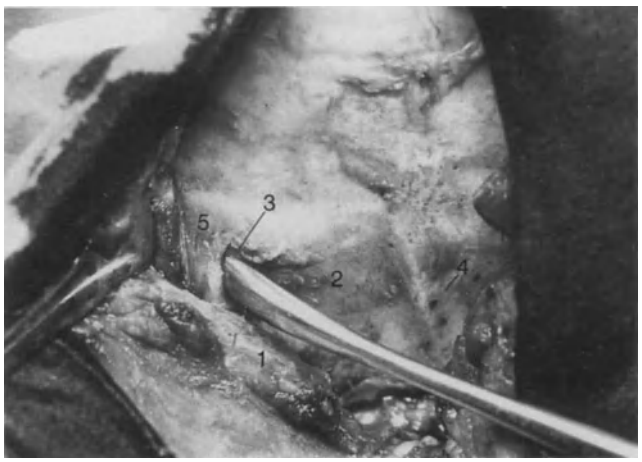
Erector Capiti Muscle (Figures 2.13 and 2.14)
Dissection of the erector capiti muscles and the trapezius from the lowest and highest nuchal lines results in stripping the periosteum from the squamous portion of the occipital bone. If these are stripped with chopping or sawing movements, it becomes impossible to reapproximate them at the time of closure, adding to the deadspace 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



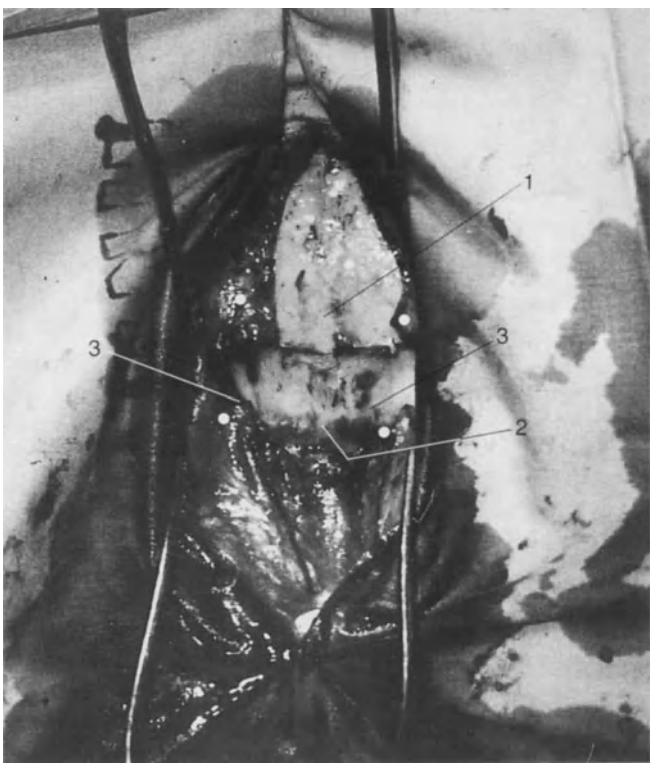
C



D



E



F

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. (E) 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*). (F) Four musculoperiosteal flaps have now been separated from the squamous occipital bone, exposing the external occipital protuberance (*1*), rim of the foramen magnum (*2*), and digastric grooves bilaterally (*3*).

grooves on either side, preserving musculotendinous integrity for complete anatomical osteomuscular reconstruction at the time of 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 carniotomy 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, parietal-temporal, 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 performed with a sharp-edged periosteal elevator (such as

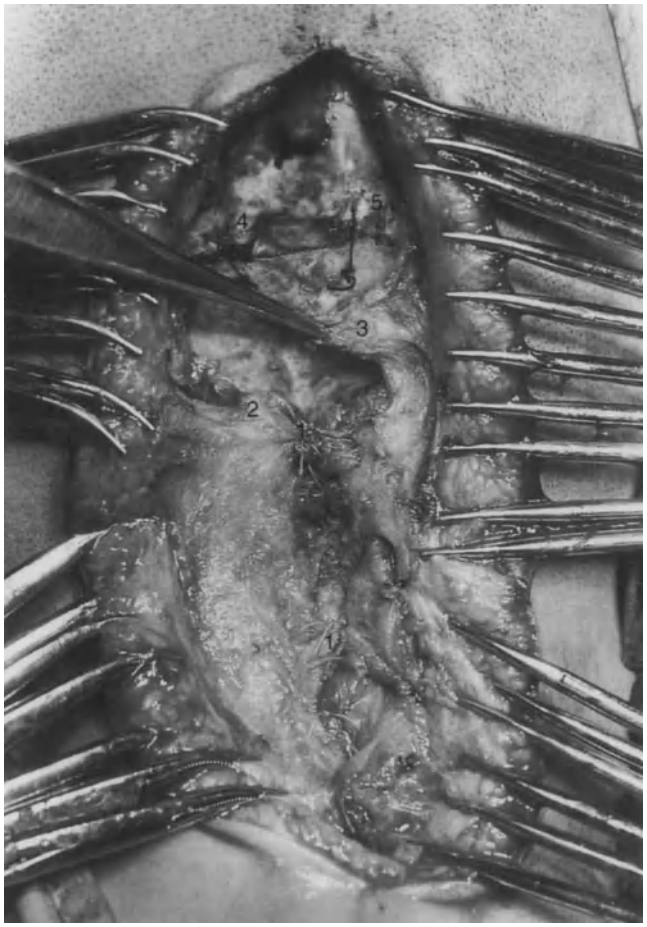


Figure 2.14. The closure that may be attained by preparing four musculotendinous flaps. The line of closure is brought superiorly along the ligamentum nuchae (*1*) to the left (*2*) and right (*3*) inferolateral flaps, which are, in turn, closed. These latter are then anchored to the left (*4*) and right (*5*) superolateral flaps, thereby bringing the erector capiti muscles back into anatomical position.

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 Chapter 4, it is much simpler to use a heavy scissors, such as a Mayo, to cut suture or bone.

Specific Incisions for Surgical Approaches

The individual skin incision (bifrontal, suboccipital, temporal, etc.) is planned so as to expose completely 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 latral occipital craniotomy, the hemispheral skin incision for frontotemporoparietooccipital craniotomy. The frontal and bifrontal skin incisions, and flaps, are used for access to the orbit(s).

Bifrontal Incision (Figures 2.15 and 2.16)
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 bones, a bifrontopterional bone flap.

Draping. Draping the child for this 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 the 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. This 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 completely the glabella, and as far inferiorly, on either side, as the zygomatic arches, so 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 lifted off, *en bloc,* with both pterional areas coming away with the single bone flap.

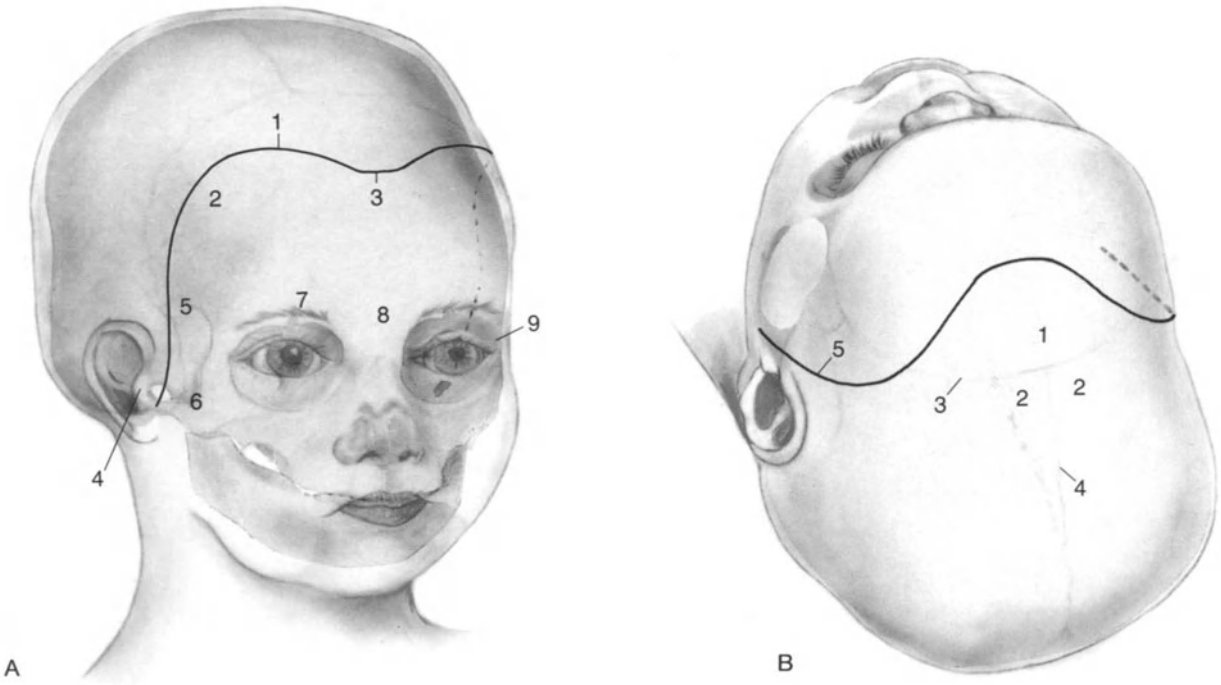


Figure 2.15. (A) If the skin incision (*1*) is made a few millimeters behind the hairline (*2*), it both hides the scar and, because of the horseshoe form it takes in approaching the sagittal plane (*3*), assures complete exposure of the frontal bone and the pterional regions bilaterally. Extending the 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. (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.

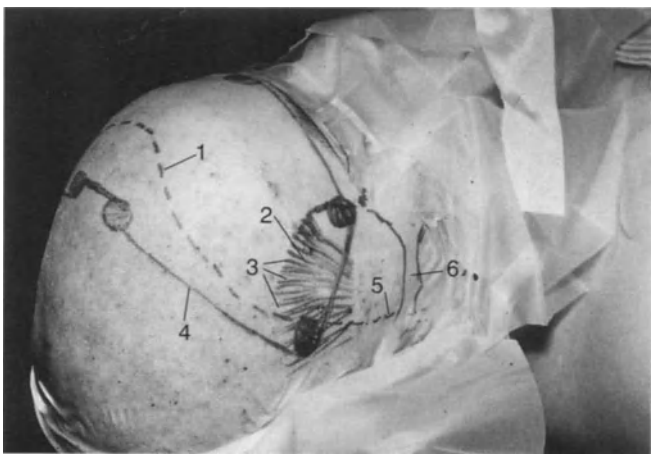


Figure 2.16. The surgeon's view of the bifrontal skin flap shown from the right. The skin incision (*1*) is represented by a broken line. Note the fanning of the temporalis muscle (*2*) and its insertion along the superior temporal line (*3*). The burr holes and craniotomy line (*4*) are drawn onto the 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.

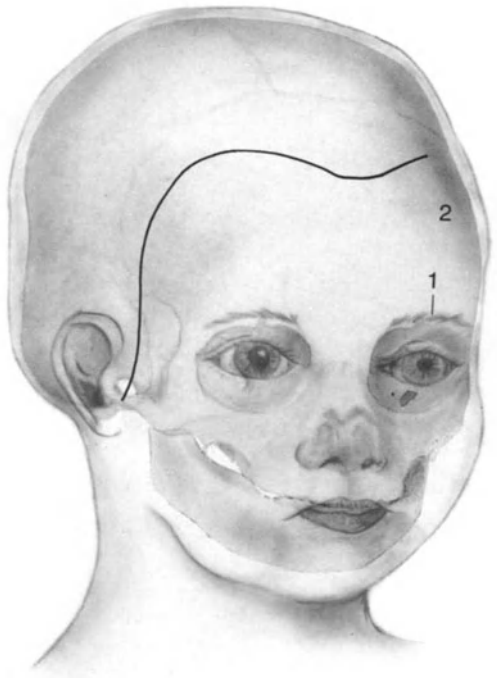


Figure 2.17. The skin incision for a unilateral frontal craniotomy is extended across the midline to the parasagittal plane running through the center of the opposite orbit (*1*), just medial to the frontal eminence (*2*).

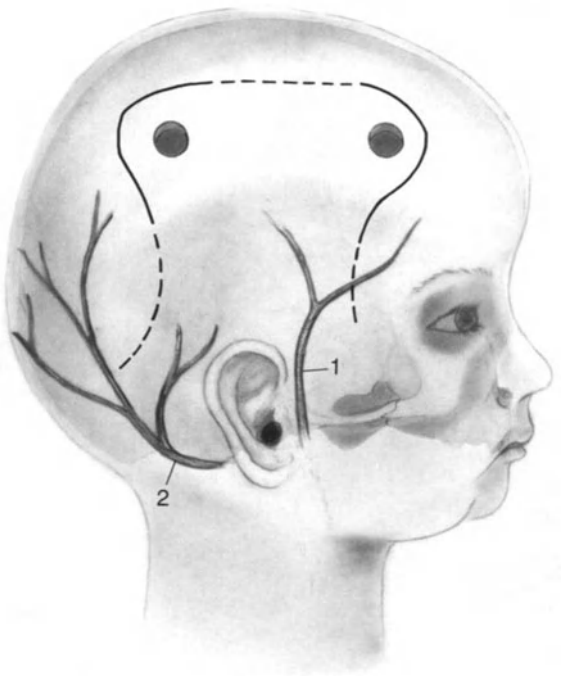


Figure 2.18. The incisions for the frontal and parietal burr holes are marked with the solid lines. The interrupted lines indicate extension of the skin incision to permit reflection of a frontotemporoparietal flap if the operative findings suggest this to the surgeon. The superficial temporal (*1*) and small branches of the occipital (*2*) arteries are included in the flap to assure healing without necrosis along the flap's edges.

Frontal Incision (Figure 2.17)
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 identical to the bifrontal incision, with the exception that it is extended only to the parasagittal plane running through the center of the opposite orbit.

Frontoparietal Skin Incision for Frontoparietal Burr Holes (Figure 2.18)
If one considers that it is often necessary to reflect a frontotemporoparietal skin flap so as to perform a frontoparietal craniotomy after frontoparietal burr holes have been placed, *the desirability of planning skin incisions for frontoparietal burr holes which may be extended into a skin flap becomes clear.* Therefore, curvilinear skin incisions for frontoparietal burr holes are preferable because they may serve as either limb of the frontotemporoparietal flap if this becomes necessary.

Draping. The draping for frontoparietal burr 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 burr 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 he chooses to go as far inferiorly as the zygomatic arch

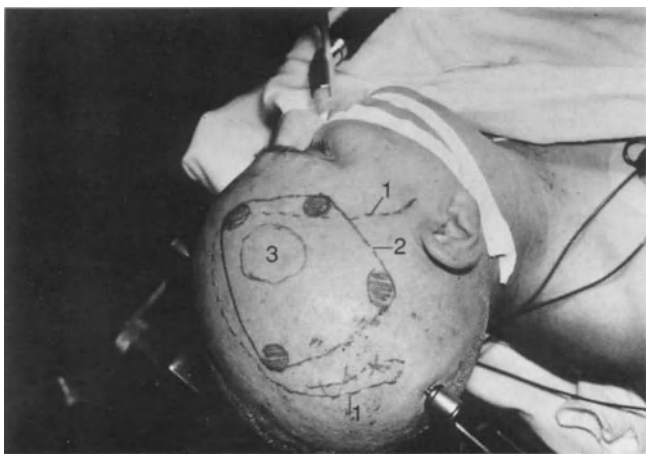


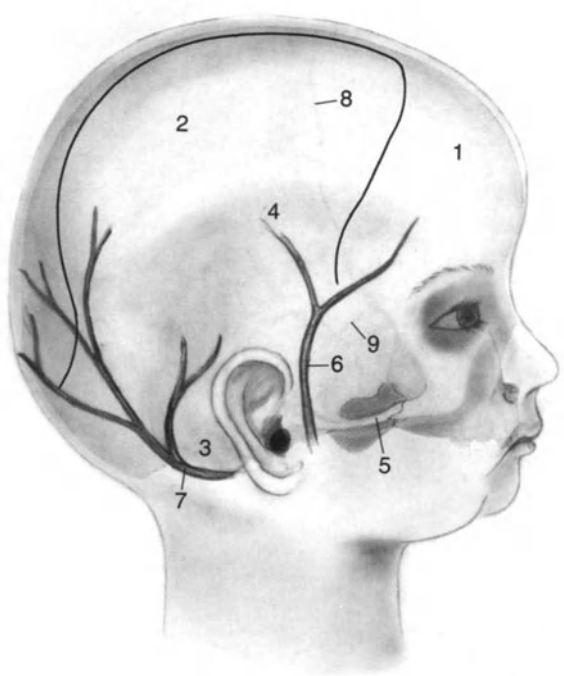
Figure 2.19. For frontoparietal flaps the anterior limb of the skin incision is identical to that for frontal flaps, but the medial limb curves posteriorly (contralateral to the midsagittal plane) to the midcoronal plane of the head, where it is redirected inferiorly and then back over the original side (*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.

in order to perform a temporal craniotomy/craniectomy.

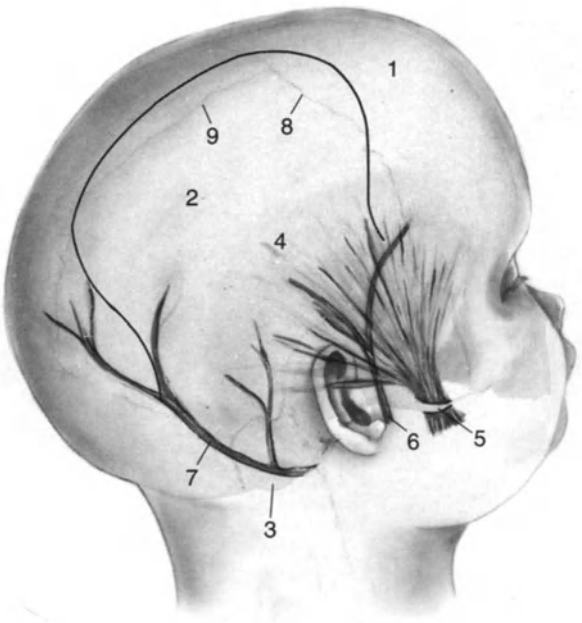
Frontoparietal Incision for Posterofrontal or Anteroparietal Lesions (Figure 2.19)
Lesions within the posterior frontal or anterior parietal areas are approached through a frontoparietal flap.
Incision. The skin incision extends from approximately 1 cm anterior to the antitragus superiorly and medially 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 in front and when he feels it in back.) It is then curved broadly back to the original side and extended as far inferiorly as the superior temporal line.

Parietal Incision Figure 2.20)
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. This incision provides a wide pedicle, access to the entire parietal bone, and exposure of the sagittal suture.

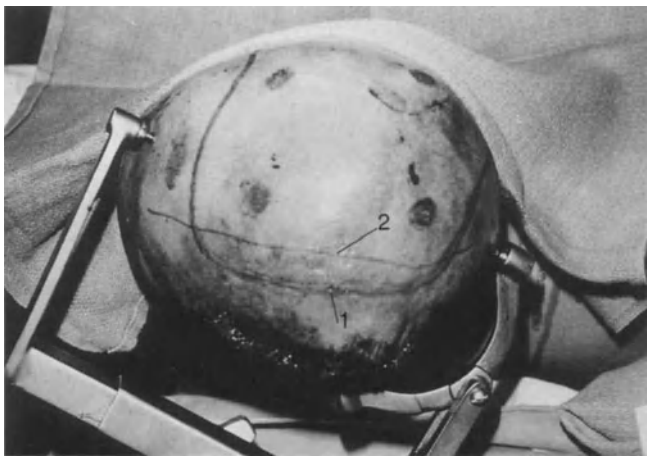
Parasagittal Incision (Figure 2.21)
The parasagittal incision offers excellent exposure of the superior surface of the posterior portion of the frontal bone, and the entirety of the medial aspects of the two parietal bones. Thus, the sagittal suture and the medial third of the coronal and lambdoidal sutures may be exposed.
Draping. Draping for the 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, as indicated in Figure 2.23 B by the broken line.) The surgeon should take care to assure the gentle curvilinear course of the parasagittal incision, rather than cutting sharp angular routes: the former guarantees 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.



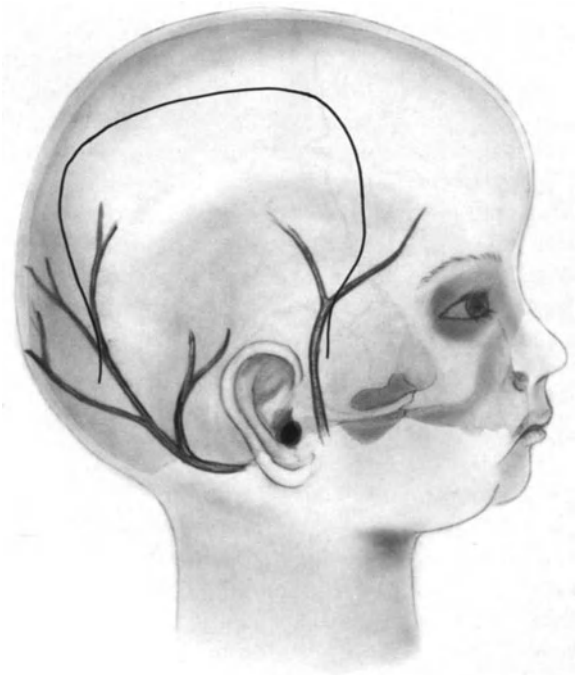
A



B

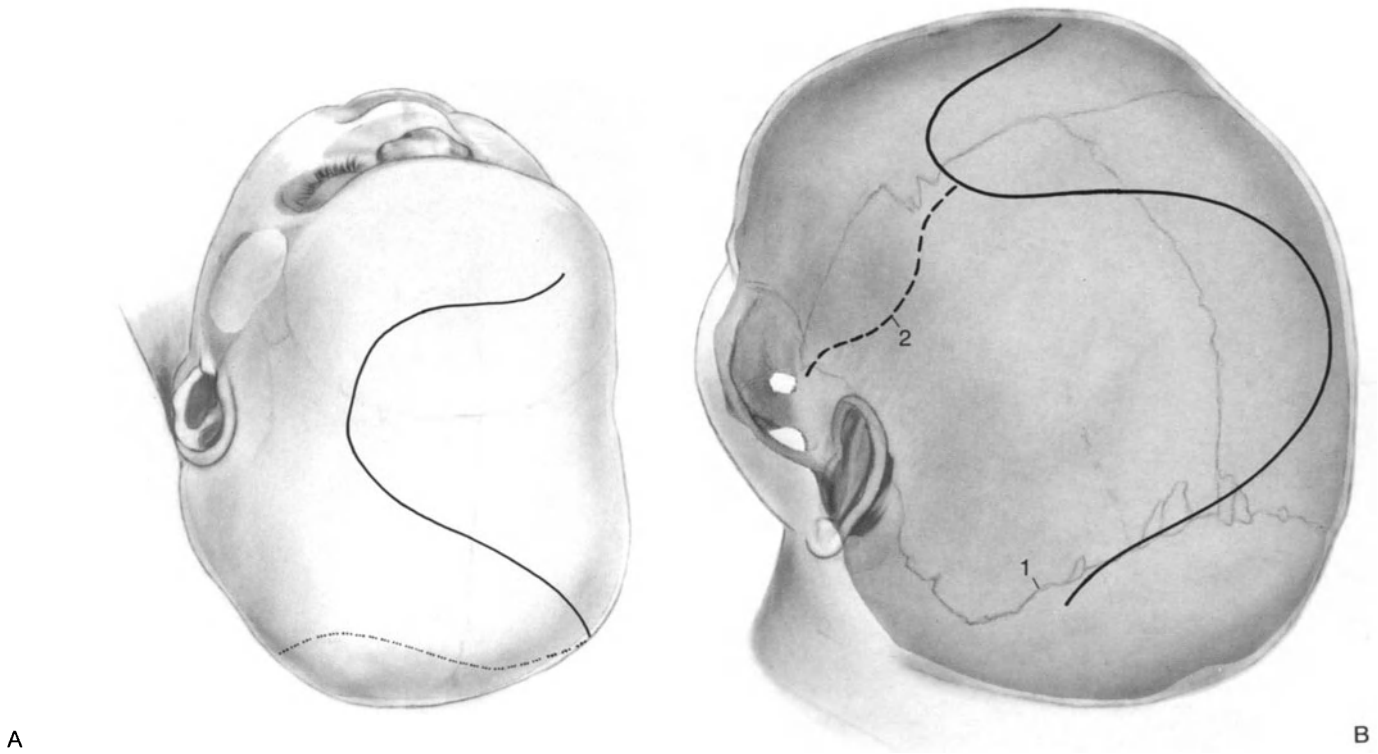


C



D

Figure 2.20. 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.



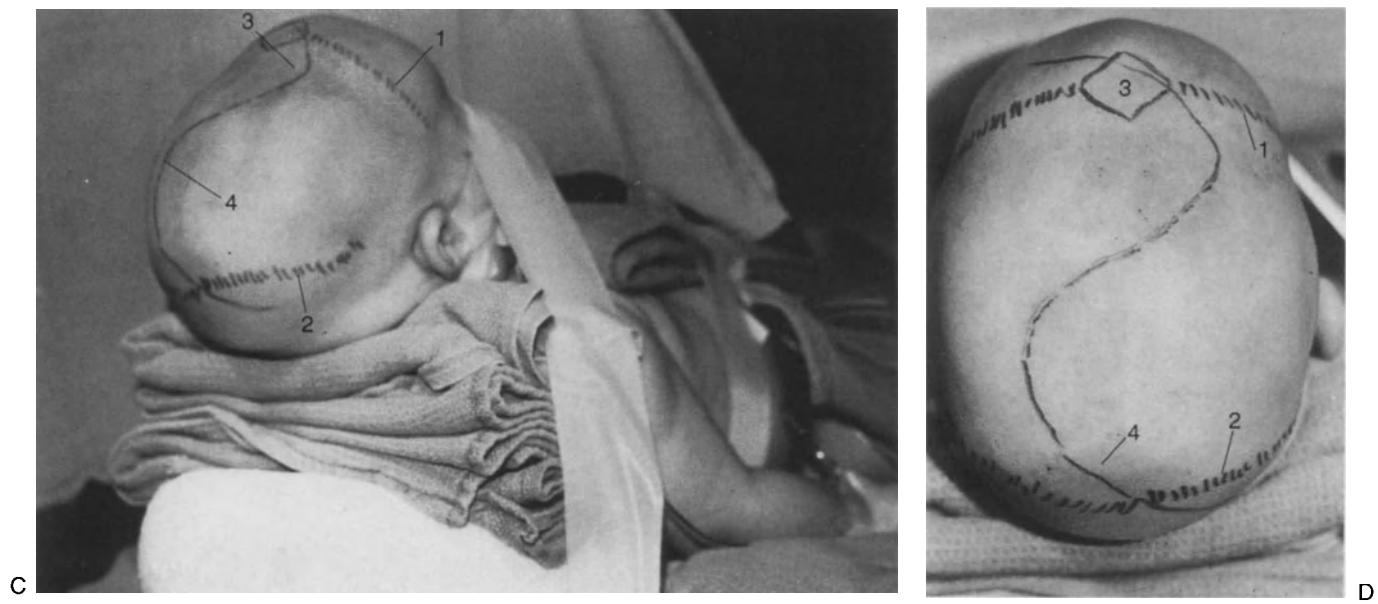


Figure 2.21. (A) This shows the line of the sinusoidal parasagittal skin incision with the child in the supine position, as he would be during the operative procedure. The broken line indicates extension of the incision along the squamous occipital area, from right to left. (B) This drawing illustrates 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. (C) The placement of towels beneath the child's shoulders and neck (so as to bring the inion into easy working distance), the coronal (*1*) and lambdoidal (*2*) sutures, and the open anterior fontanel (*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 parietal 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. (D) This perspective permits one to identify the coronal suture and the anterior fontanel, 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.

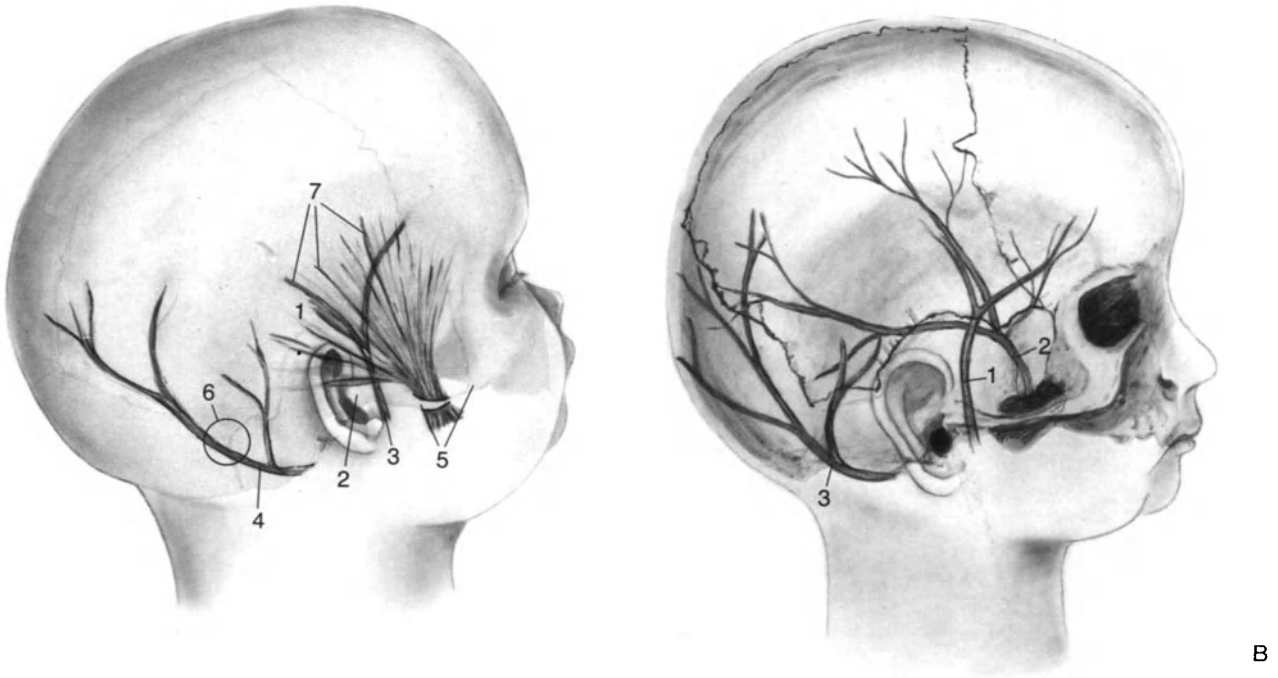
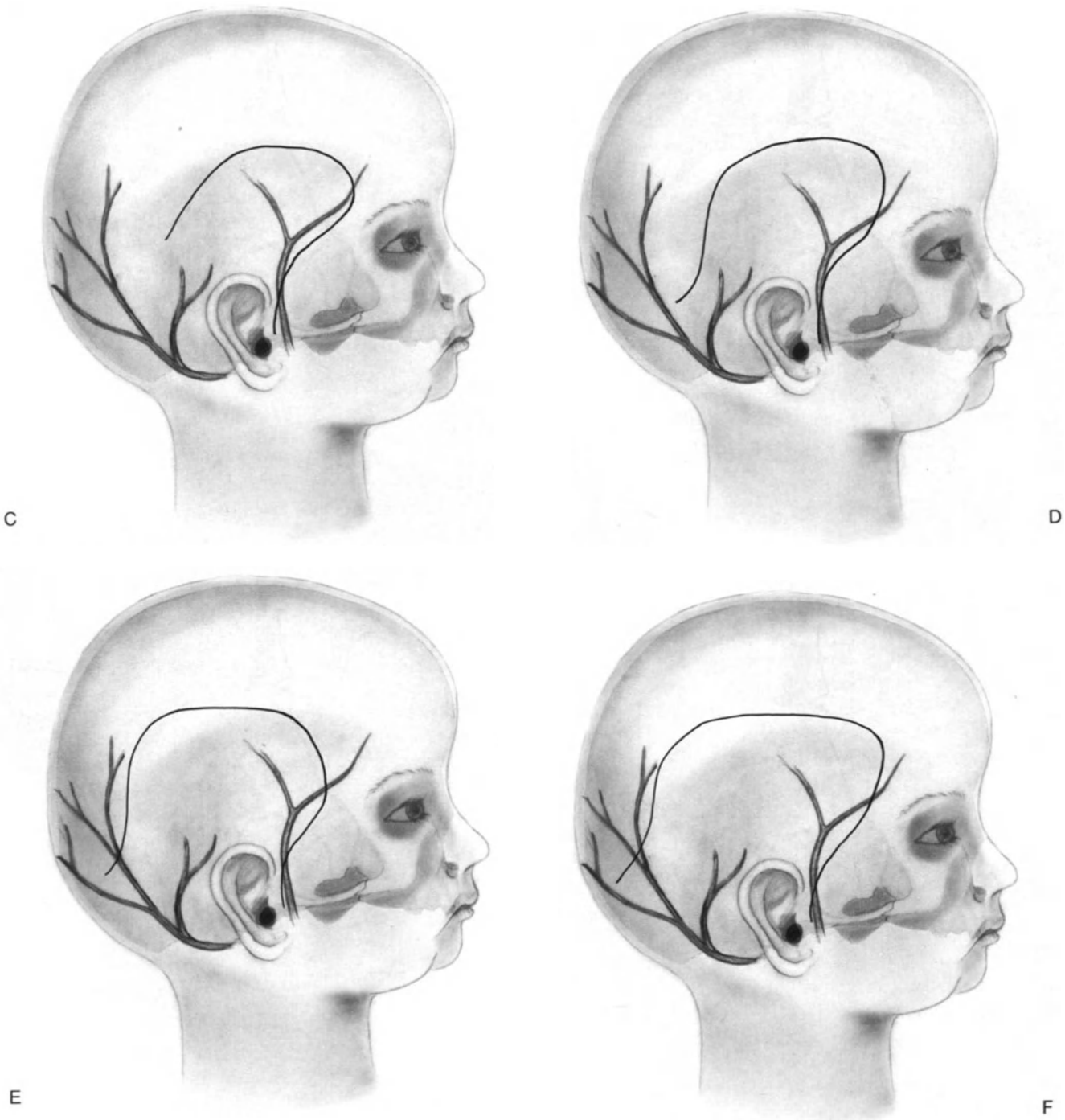


Figure 2.22. Relative location of extra- and intracranial branches of the external carotid vessels. (A) The posterior oblique view illustrates relative locations of the temporalis muscle (*1*), antitragus (*2*), superficial temporal (*3*) and occipital (*4*) arteries, zygomatic arch (*5*), and occipitotemporoparietal 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) 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.

Temporal Incision (Figure 2.22)

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 on 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, 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.

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, 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.

Incision. 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 tempo-

ral 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.

Occipital Incision (Figure 2.23)
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 posterioinferior portion of the sagittal suture, and the entirety of the homolateral lambdoidal suture. There are medial and lateral 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. One *lateral* incision extends from just lateral to the torcular Herophili superiorly, and parallel to the superior sagittal sinus to over the parietal eminence. It is then extended inferiorly and anteriorly to just above the helix.

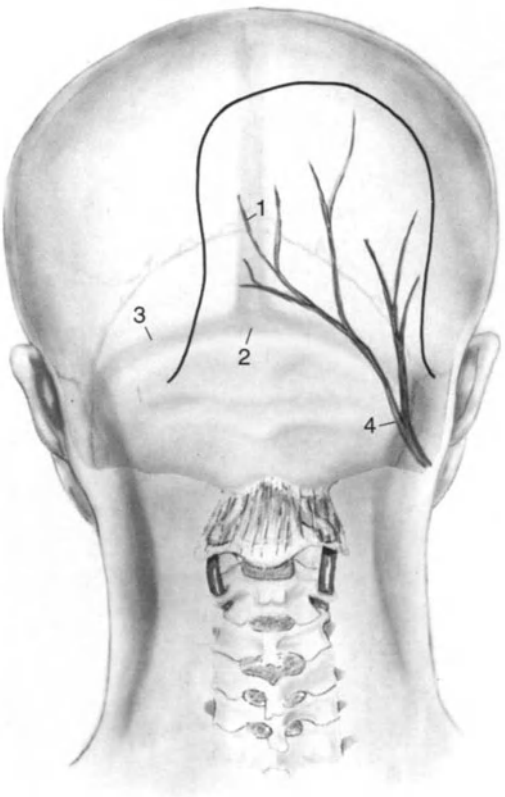
Suboccipital Incision (Figures 2.24 and 2.25)
Suboccipital skin flaps may be either medial (midline) or lateral, depending upon whether one must reflect the squamous portion of the occipital bone for a vermis tumor (medial), a cerebellar hemisphere or pontocerebellar angle tumor (lateral). The medial 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.

Combined Supra- and Infratentorial Incision
(Figures 2.26 and 2.27)
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.

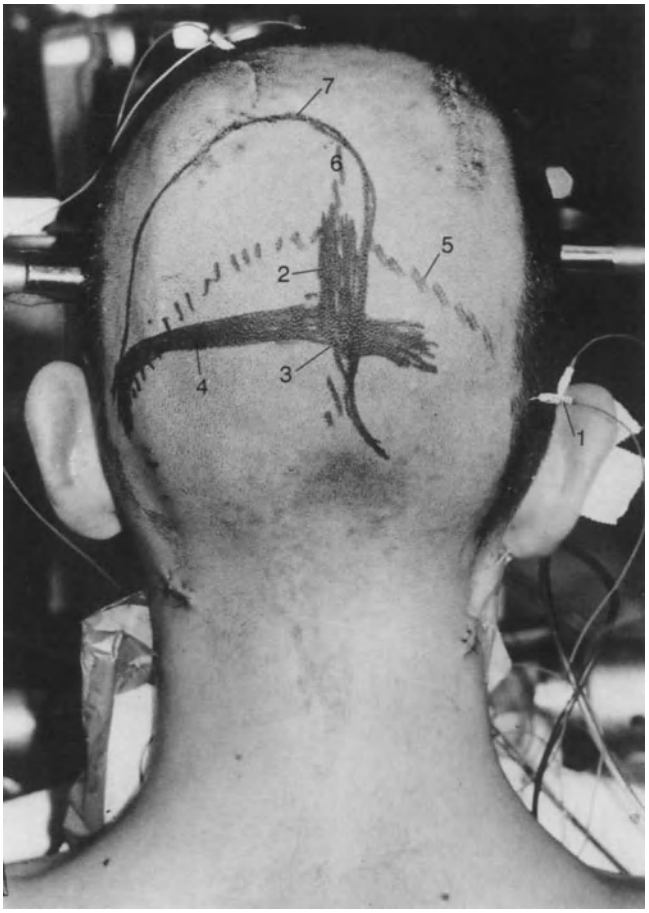
For such tumors as meningioma, acoustic neuroma, and glomus jugulare, which may grow within the supra- and infratentorial spaces as independent tumors, dumbbell tumors growing on either side of the tentorium, or particularly large extraparenchymal tumors extending into the supratentorial compartment 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 question-mark incision can 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 that 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.

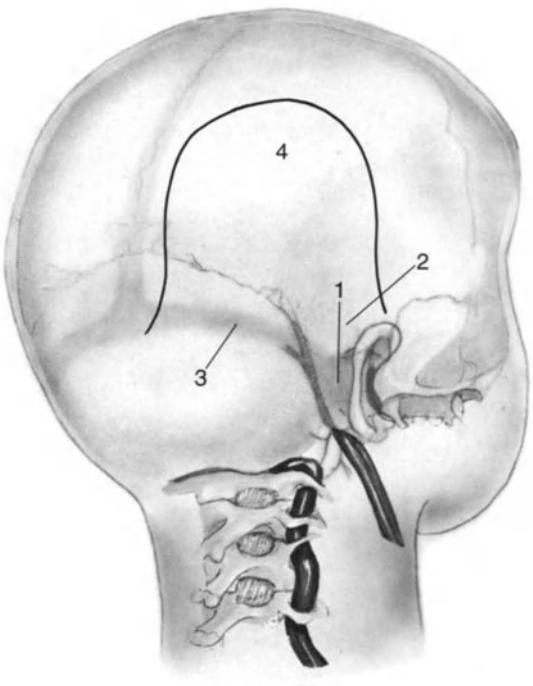
Hemispheral Incision (Figure 2.28)
The hemispheral skin incision permits the surgeon to expose half of the frontal and occipital bones and the entirety of the parietal bone, as well as portions of the greater wing of the sphenoid and the squamous temporal bones. This incision is used for hemispherectomies 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 mid-



A

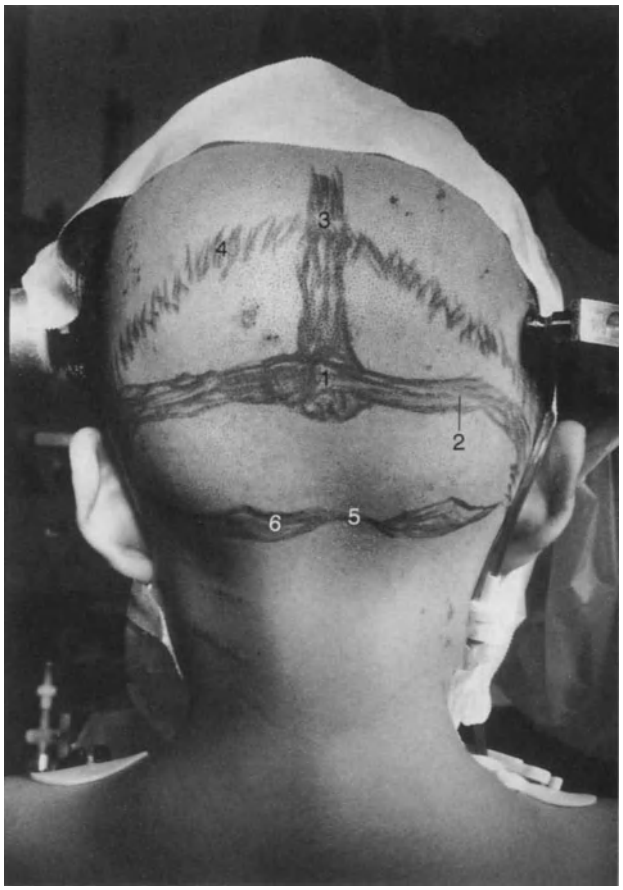


B

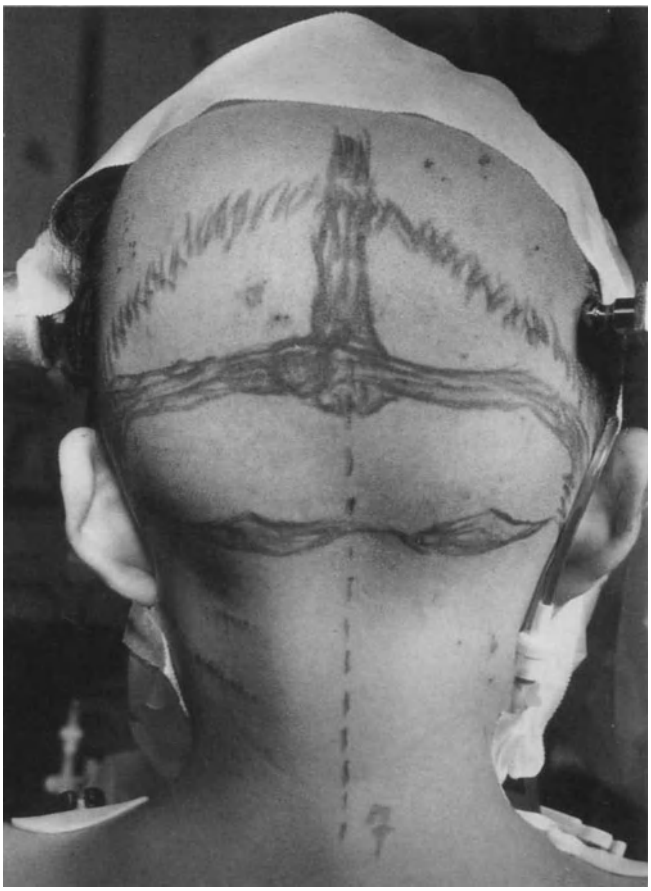


C

Figure 2.23. (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 MEP's 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*).

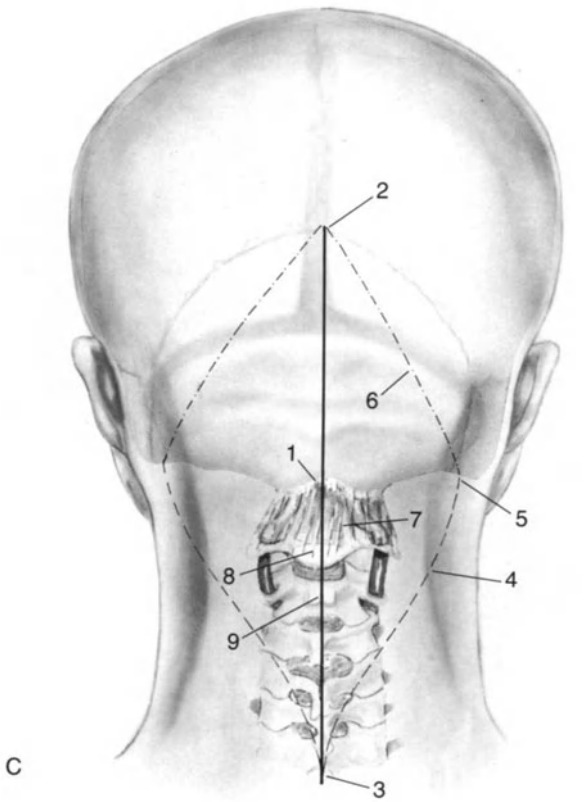


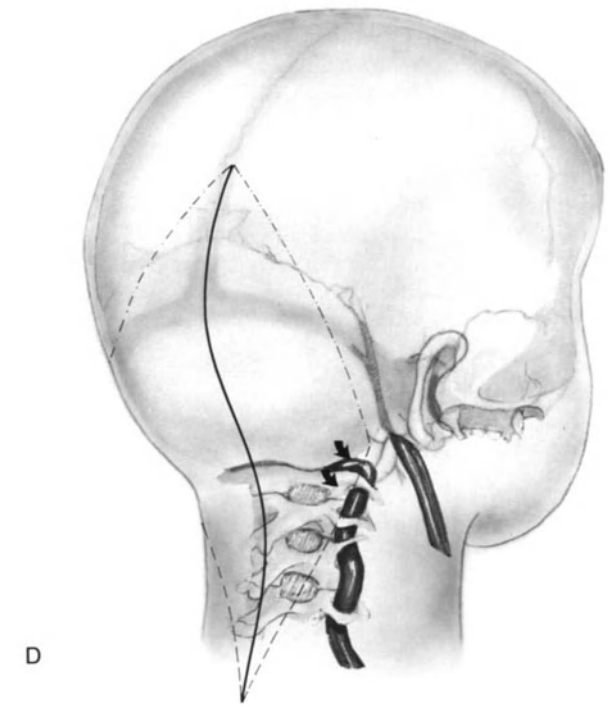
A



B

Figure 2.24. (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 appropriately 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 atlantooccipital 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 (–●–), 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 Figure 2.27C. The vertebral artery, and the entrance of Batson's plexus into the dural sinuses is at the most lateral exposure of the field (arrows). ►





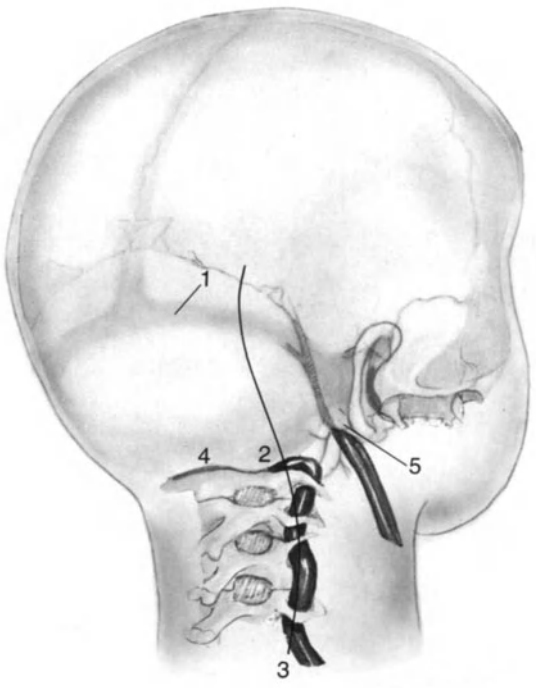
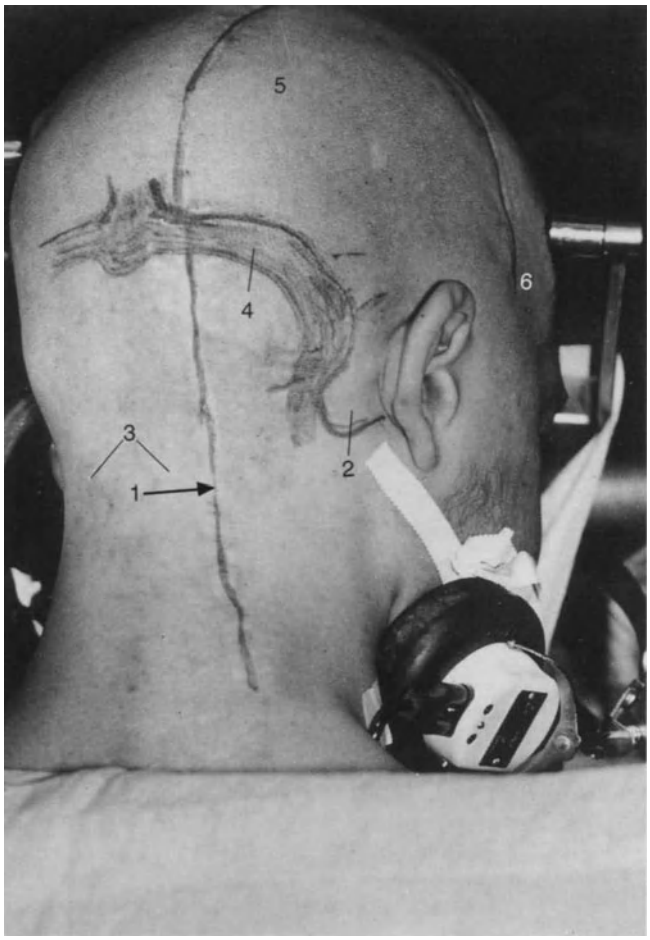
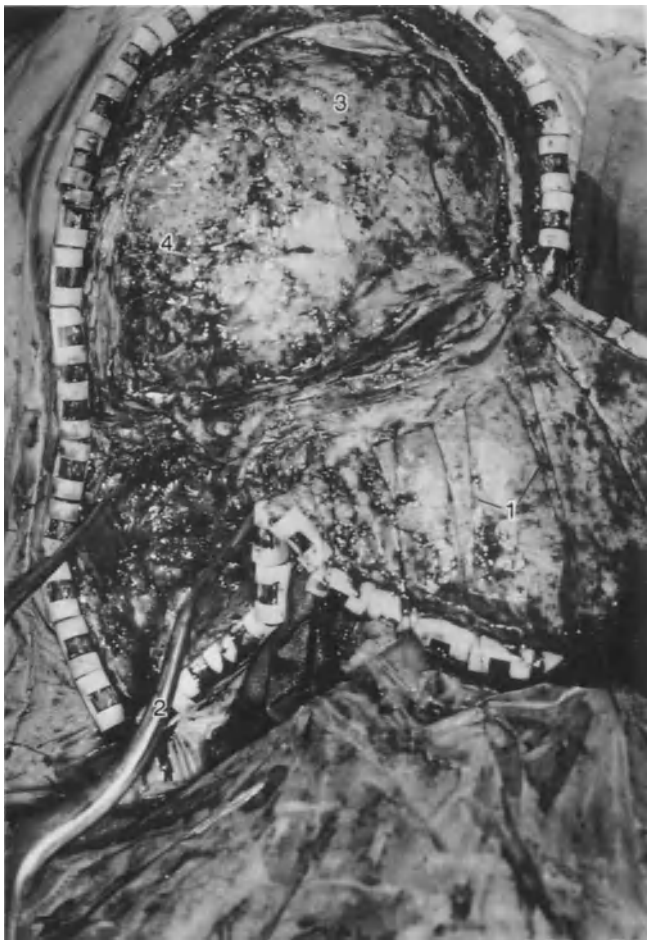


Figure 2.25. The lateral suboccipital skin incision is illustrated diagrammatically, here from an oblique view, in a transparency drawing. The incision extends from above the transverse sinus (*1*), across the junction of horizontal and vertical segments of the squamous occipital bone (*2*), and down 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*).



A



B

Figure 2.26. (A) The question-mark skin incision for combined supra- and infratentorial approach to the tentorial ring at the anterior clinoid and the pontocerebellar angle is herein illustrated, 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*), to end at the pterion (*6*). (B) The temporoparietooccipital portions of the scalp flap have been reflected inferiorly, and retracted from the field with the use of retention sutures (*1*). A self-retaining retractor (*2*) has been placed between the cut segments of the erector cervicis and erector capitis muscles. The coronal suture (*3*) and parietal eminence (*4*) are already exposed.

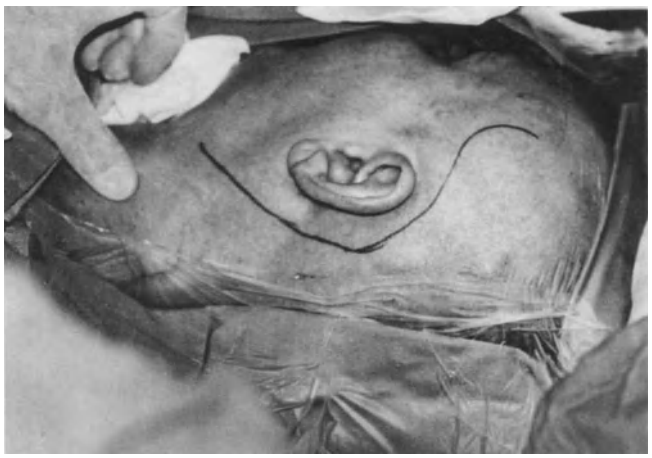


Figure 2.27. The "sine-wave" incision has been marked off. Note that it extends from the angle of the mandible, posteriorly to the mastoid apex, and then superiorly to the base of the mastoid, before turning anteriorly across the occipital and parietal bones to curve over the superior temporal line. 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. Once these bones are removed, one has visualization of the sigmoid sinus, transverse sinus, the supratentorial and infratentorial compartments.

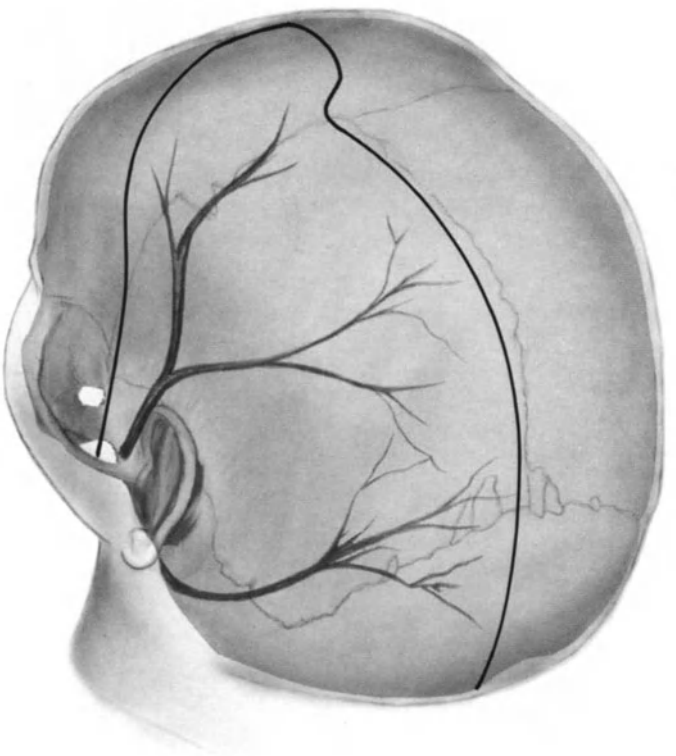


Figure 2.28. Placement of the skin incision for an hemispheral craniotomy.

line 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 (Figure 2.29)

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.

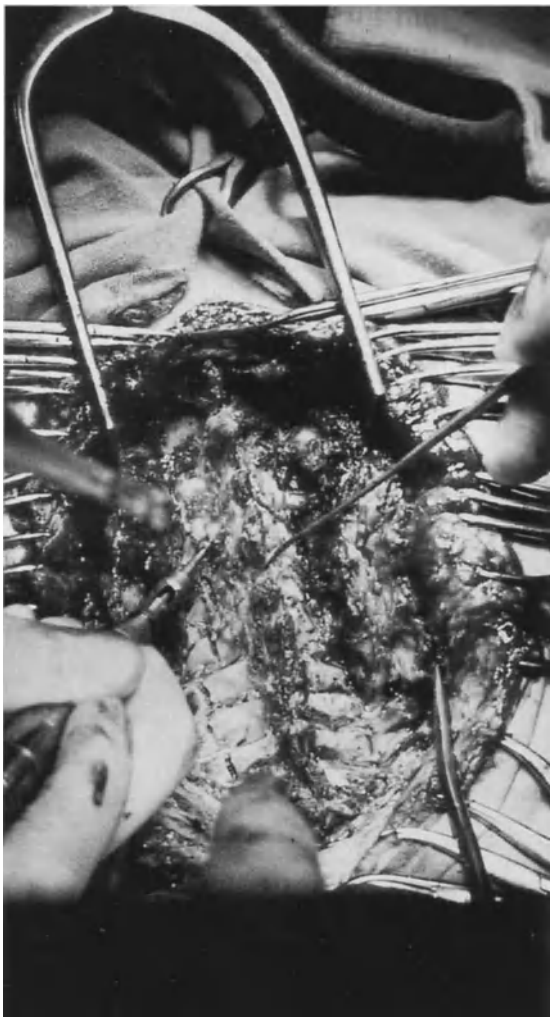
Incision. The skin incision is midline, extending the full length of the planned laminotomy plus 4 cm cephalad and 4 cm caudad. If one is operating on a neuroma that extends into both the spinal canal and either the retropleural or retroperitoneal spaces, a "hockey stick" incision is ideal. Its short 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, ligamentum flavae, spinous processes, interspinous and suprasinous ligaments, and paraspinal muscles. In the adult, stability depends mostly on 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 imbalances 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 *substitute scar tissue for insertion of paraspinal muscle masses onto the laminae and spinous processes.*

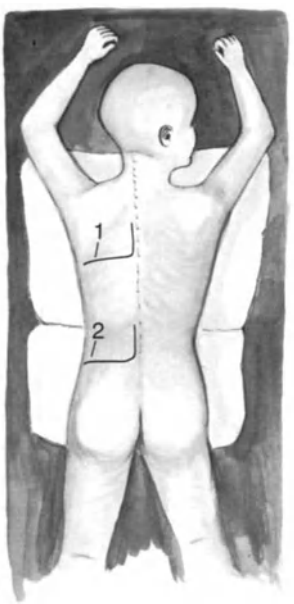
After the skin incision has been made and clips applied to the subcutaneous connective tissue, the very thin paraspinous muscles are cut from their insertion along the midline of the vertebral arch and the stripped free.

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 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 spinal apophyses.

A

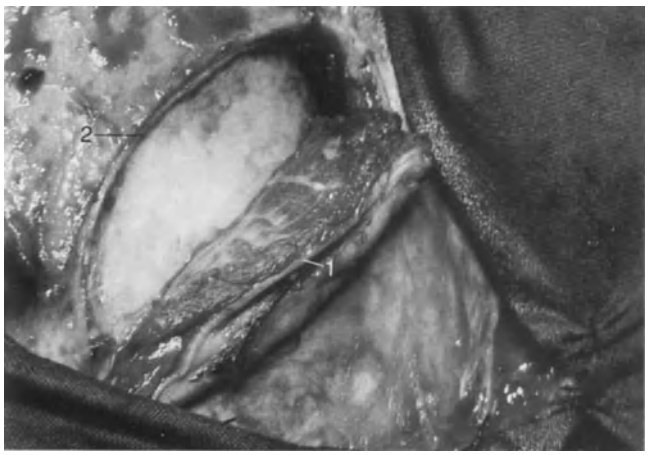


B

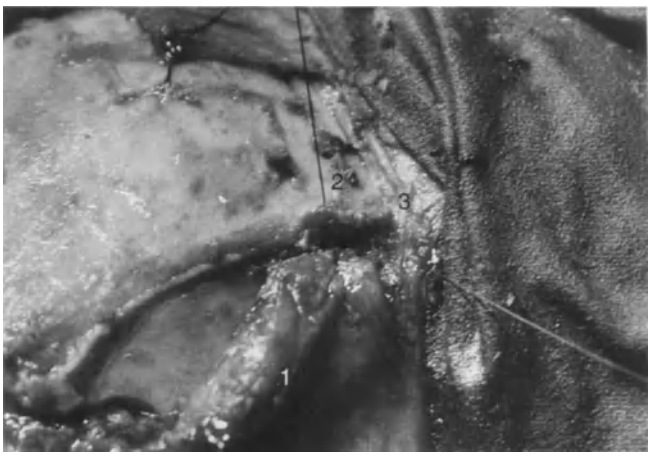


C

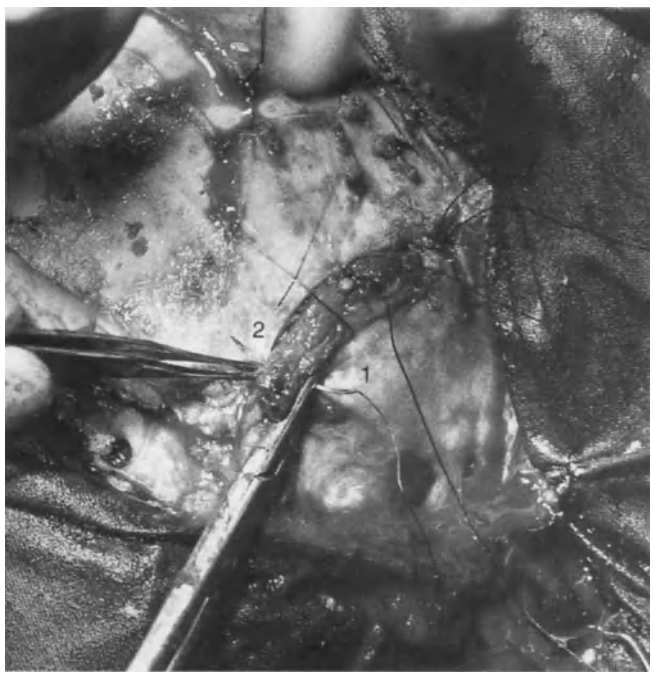
Figure 2.29. (A) The skin incision has been made and clips applied to the subcutaneous tissue. The very thin paraspinous muscle mass (*1*) is incised at its point of attachment to the midline of the posterior vertebral arch (from which the spinous processes will develop), taking care to incise along the full width of the spinous processes (*2*). (B) 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. (C) 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 of dumbbell neurofibromas is possible with these hockey-stick incisions.



A



B



C

Figure 2.30. (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" burr 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*).

In newborns 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.

Closure

Cranial Closure

Fascia and Muscle Closure (Figures 2.30 and 2.31)

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.

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 fascial and muscular tissues that had been preserved at the time of opening (Fig. 2.13, 2.14). Since it is not necessary to remove the erector cervicis muscles 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 of

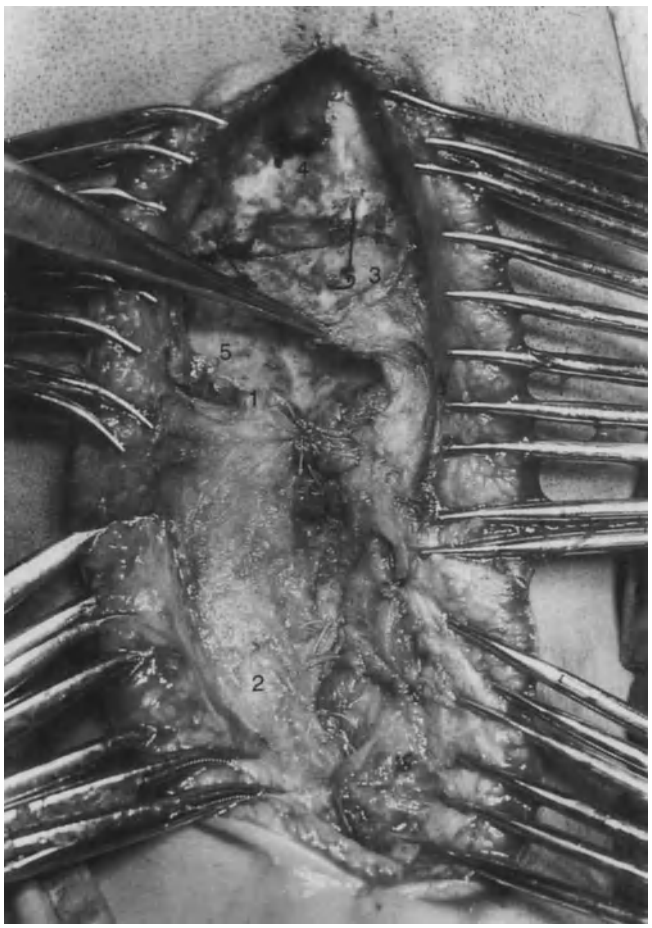


Figure 2.31. The erector capitis muscle masses have been reapproximated and sewn together in the area extending from over the squamous occipital bone (*1*) and C-2 (*2*). Also, the fascia from these masses (*3*) has been sewn to the periosteum, which was left intact, and attached to the squamous occipital 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.

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 (Figure 2.32)
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 noncoagulated 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, the author recommends 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.

Laminotomy (Figures 2.33 and 2.34)
Muscles and Fascia Closure
The paraspinous muscles are allowed to fall into place by removing the self-retaining retractor. Then the mus-

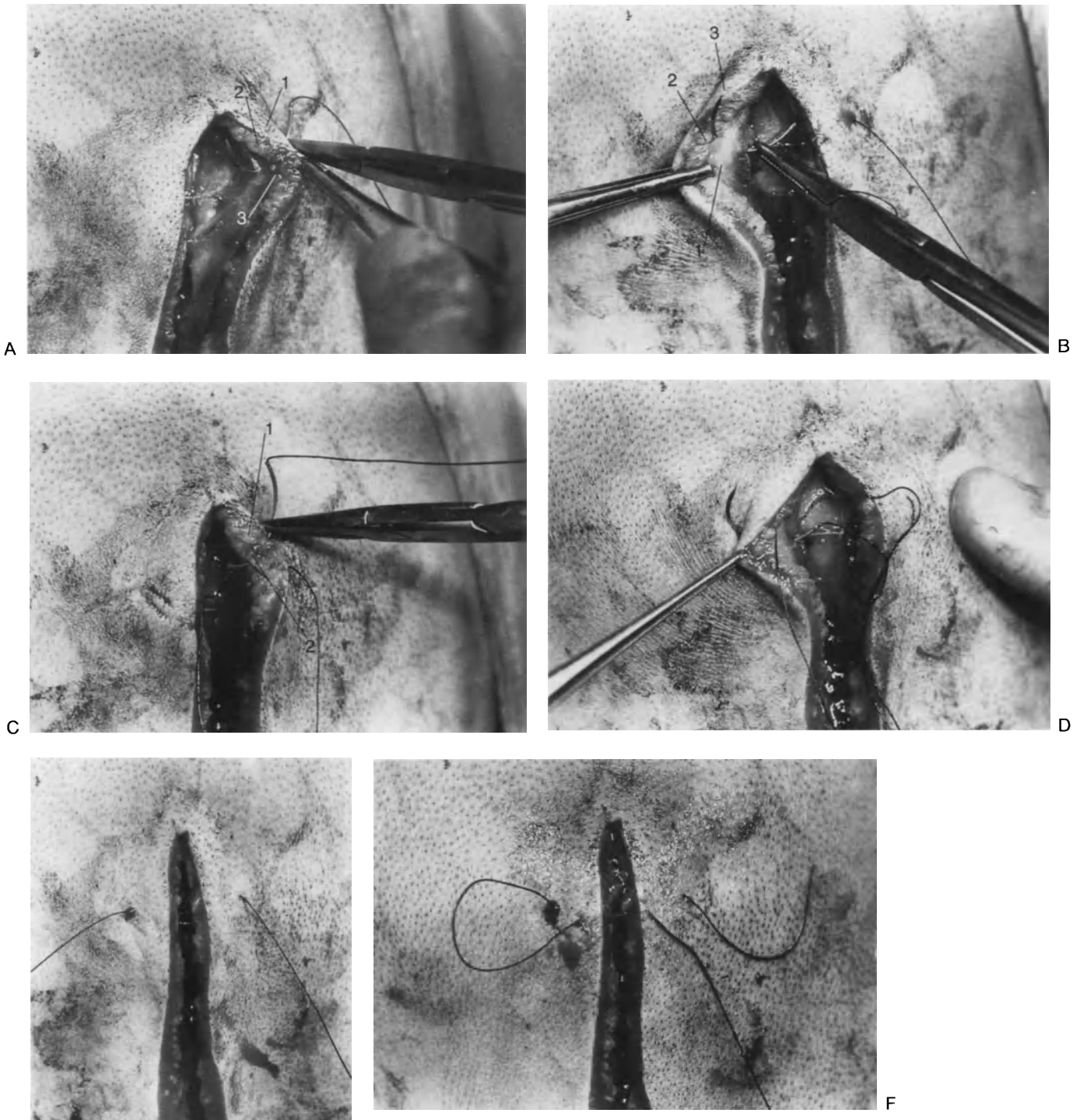


Figure 2.32. Technique for the Cloward suture, which closes the Galea and the skin without necessitating burial of suture material. (A) The needle is brought through the full thickness of skin (*1*), dense connective tissue (*2*), and Galea (*3*). (B) The needle is brought only through the Galea (*1*) 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 I recommend for children.

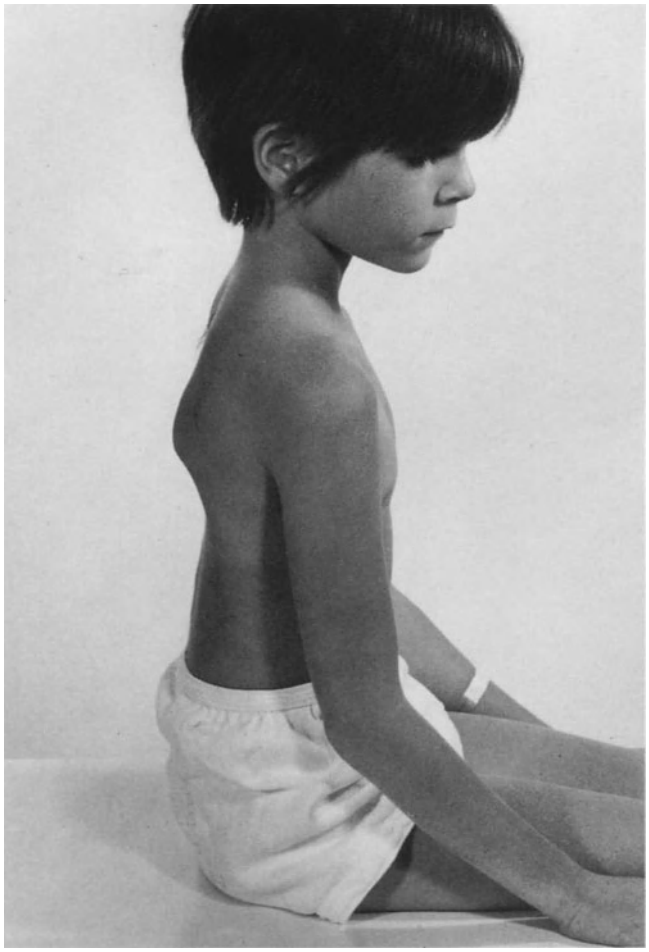


Figure 2.33. Postoperative kyphoscoliosis in a child who has had a laminectomy.

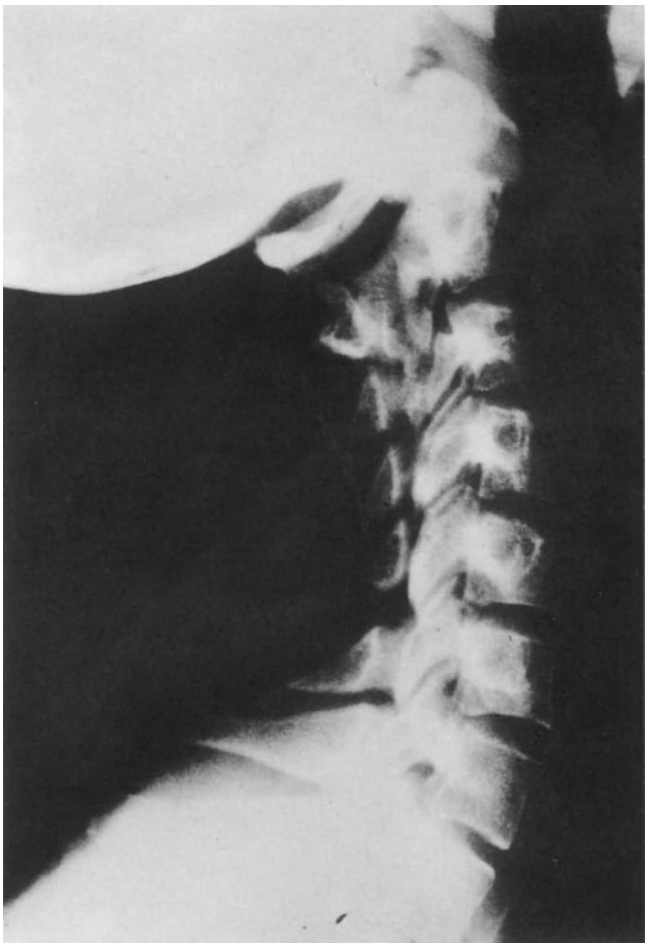


Figure 2.34. Lateral cervical spine film of a child on whom an extensive (see Fig. 2.7) cervical laminotomy had been performed. No malalignment is present.

cles 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.

Skin Closure

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

"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

Chapter 3

Burr Holes and Flaps

Unfortunately, the terminology of anterior fossa, middle fossa, 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, frontotemporal craniotomies; the middle fossa includes anterior and posterior temporal craniotomies, 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.

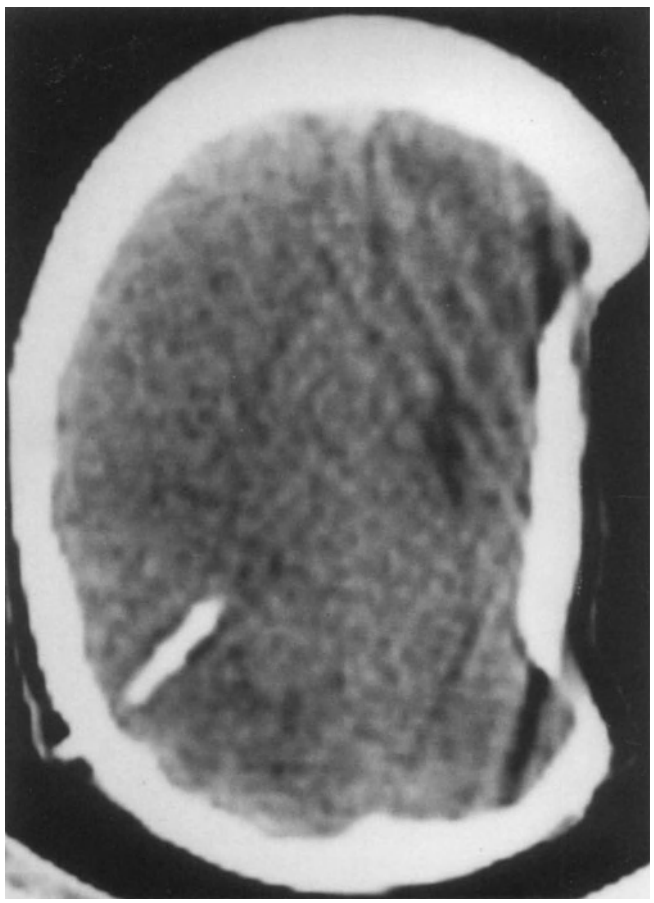
In planning the bone flap, one must correlate the "target area" with the skin incision, so as to attain ideal placement. For example, a bifrontopterional craniotomy for access to a *retrochiasmatic craniopharyngioma* entails exposure of the clinoid processes, optic nerves, and internal carotid arteries, 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 middle 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 in this text to illustrate this manner of conceptualizing the lesion and approaching it.

Burr holes and either craniotome or Gigli saw are not necessary in the newborn or very young infant, since the fontanels 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 a 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 Chapter 4). This, of course, does not apply when an infant has craniosynostosis. The techniques for these specific procedures are described subsequently.

The use of power instruments for making burr 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 sensi-



A

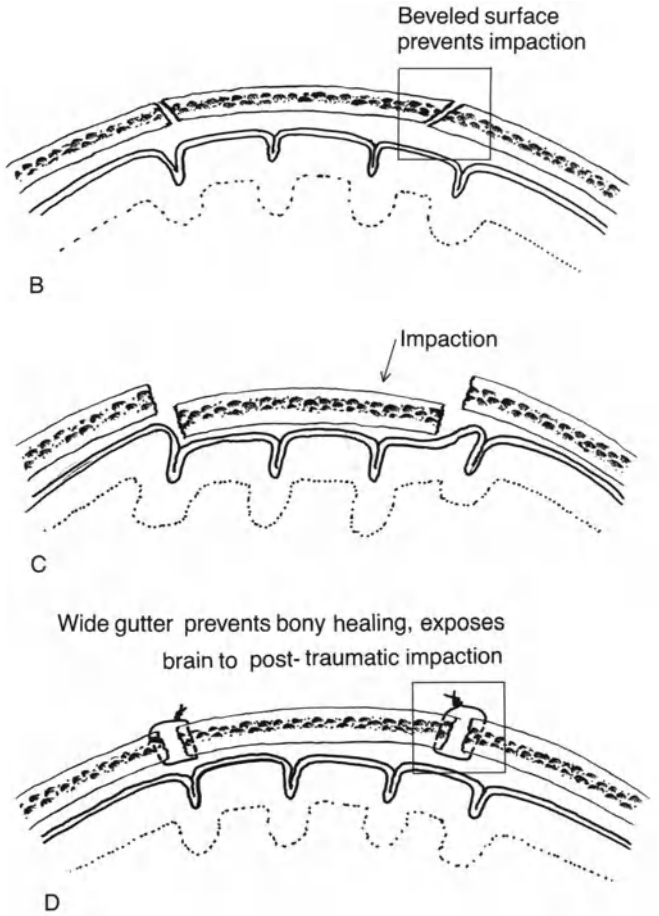


Figure 3.1. (A) This computerized transmission tomography (CTT) scan shows the unacceptable degree of flap depression that occurs when the power craniotome is used. The Gigli saw permits adequate beveling, so that the flap nestles into place irrespective of placement of sutures through the skull or a 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 sutures placed through drill holes may suspend the free flap, but still bony growth across the gutter only rarely occurs. The defect is permanent, and thereby susceptible to dislocation, impacting the underlying cerebrum from injury.

tive enough to guarantee release of the clutch in children with thin skulls. Because it is possible to refill the burr 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 on Bone Closure (Figs. 3.49 and 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 pressure 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 particularly 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 is too wide to permit bony bridging and the movement is 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.

Burr Holes: Frontoparietal (So-Called "Diagnostic") (Figure 3.2)

The theoretical considerations for placing the skin incision for frontoparietal burr 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 burr hole openings reveal no visible epidural or subdural hematoma because the clot may be located between the burr holes. Also, inferior extension of both the anterior and 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.

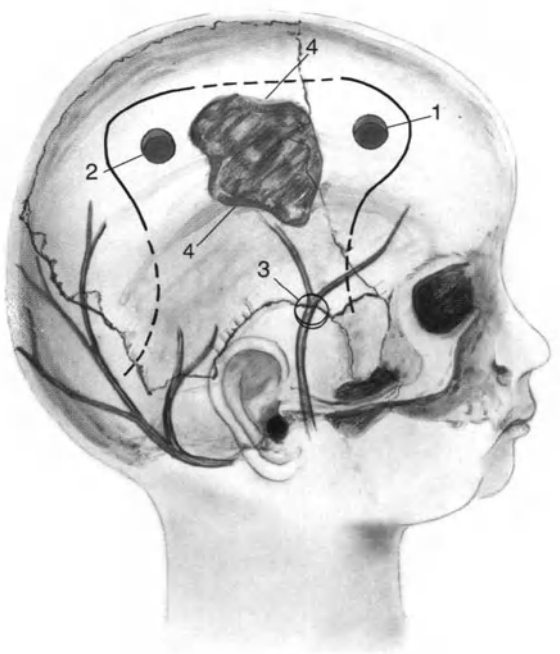


Figure 3.2. This drawing illustrates how frontotemporoparietal burr holes (*1*, *2*, *3*) may be placed around an epidural clot (*4*), so that the surgeon may not see the clot at the time of surgery if he limits his procedure to burr holes. If the clot is not seen when the burr holes are placed, one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

Flaps

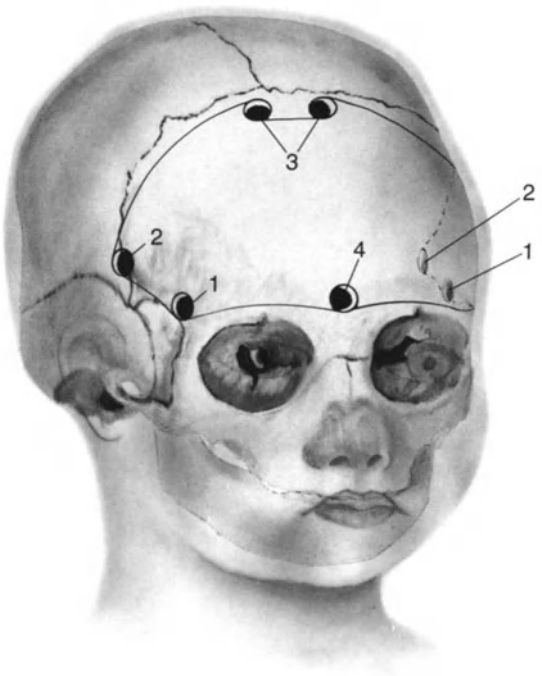
The placement of burr holes and outlining of the Gigli saw osteotomy lines are discussed in the following sections.

Bifrontal Flap (Figures 3.3 and 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 their 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 7 burr holes of the bifrontal flap are located at the glabella, immediately lateral to the superior sagittal sinus (SSS) on either side, just posterior to the pterion on either side, and immediately posterior to the zygomatic processes of the frontal bone (the "keyhole") bilaterally.

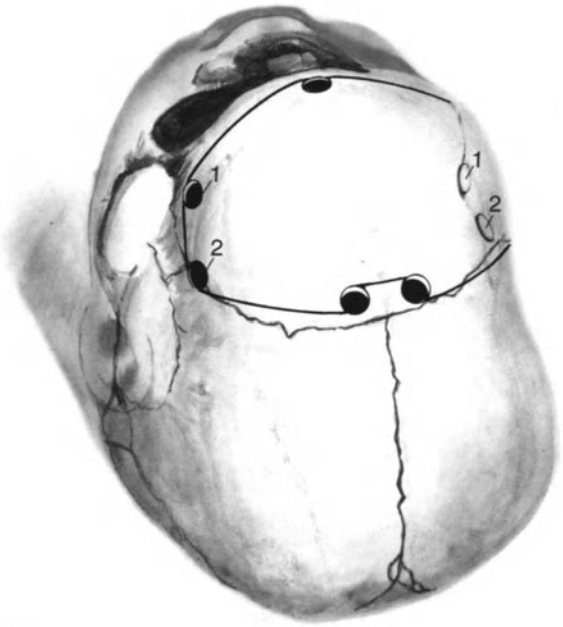
For early childhood, the frontal air sinuses are not developed. The burr hole at the glabella runs no risk of penetrating one of them. The two parasagittal burr holes should be placed so that their medial aspects border upon the lateral aspects of the SSS. Orientation concerning the width of the SSS may be obtained by studying the cerebral angiogram, which also permits one to study the number, caliber, and points of entry of the bridging cortical veins into the SSS. This is of great assistance.

The two pterional burr 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 burr 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 lacrimal 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 burr 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.

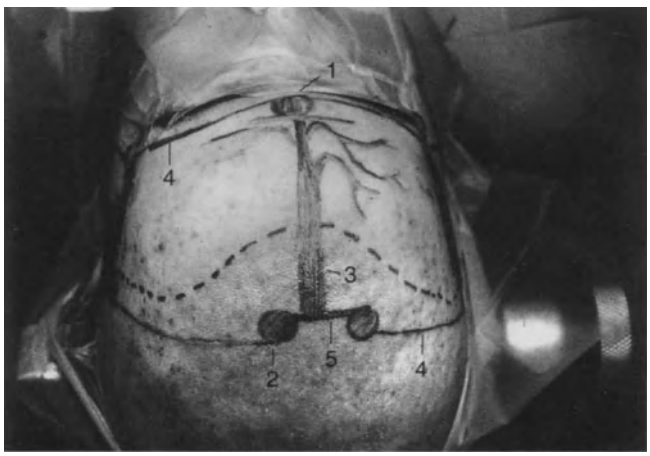
After the Gigli saw guide has been passed from burr hole to burr hole and the Gigli saw is brought into position between the inner table of the skull and the guide, the saw itself should be set snugly at the extreme periphery of each hole so as to connect their *outer* arcs and thus assure maximum size of the bone flap. The exception to this technique is the osteotomy line between the two parasagittal burr 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



A



B



C

Figure 3.3. (A) Burr holes and osteotomy lines for bifrontal bone flap with child in the anatomical position. The seven burr 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 burr 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*) burr 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 burr holes along the most peripheral arc of the burr holes, with the exception of the two parasagittal ones (*5*), where they connect the most medial arcs.

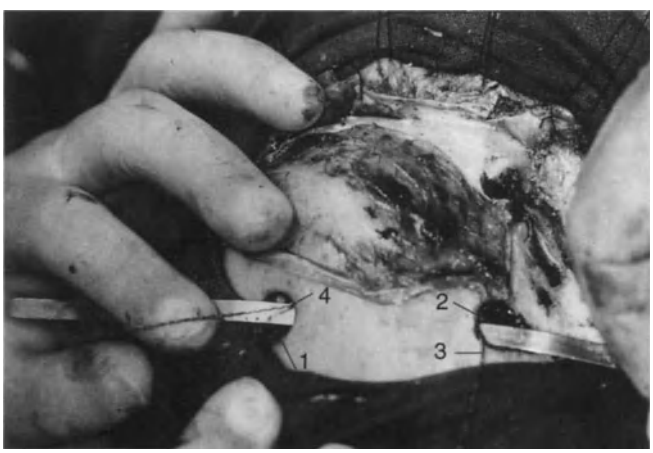
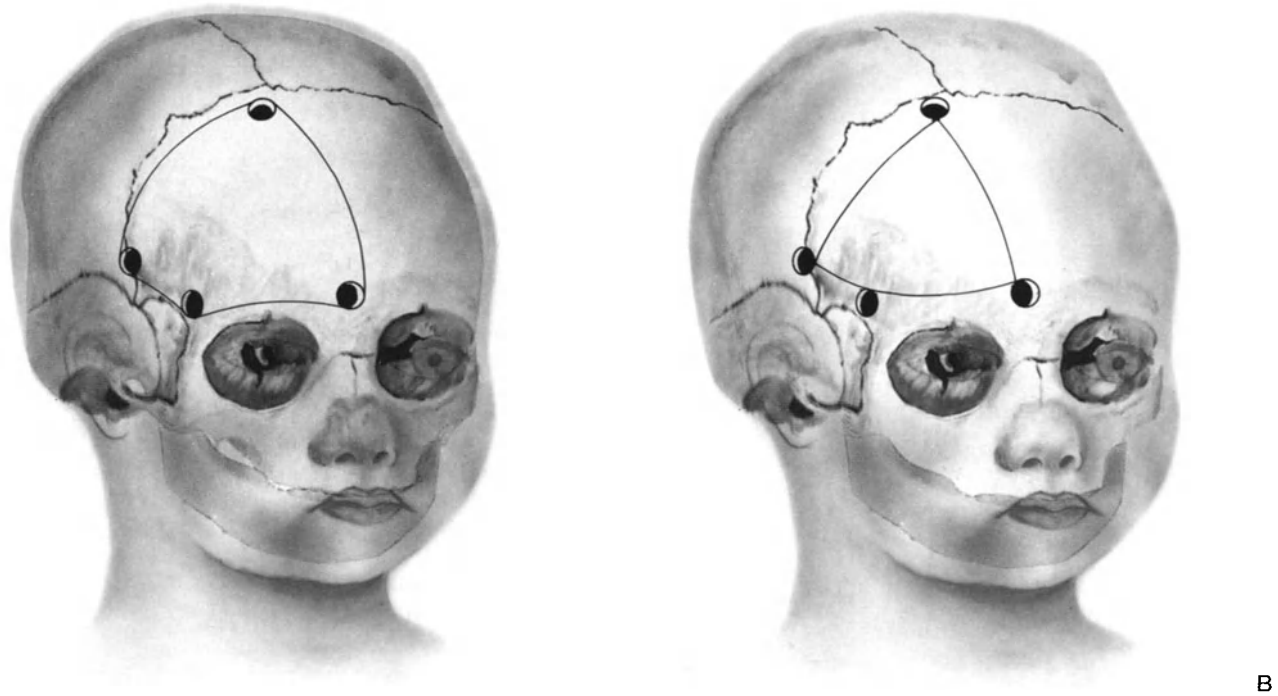


Figure 3.4. The Gigli saw guide has been passed from the parasagittal (*1*) to the pterional (*2*) burr holes and the saw subsequently brought into place. At the right the saw is positioned properly, at the peripheral arc of the burr hole (*3*), whereas at the left it is positioned improperly, at the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



A B

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

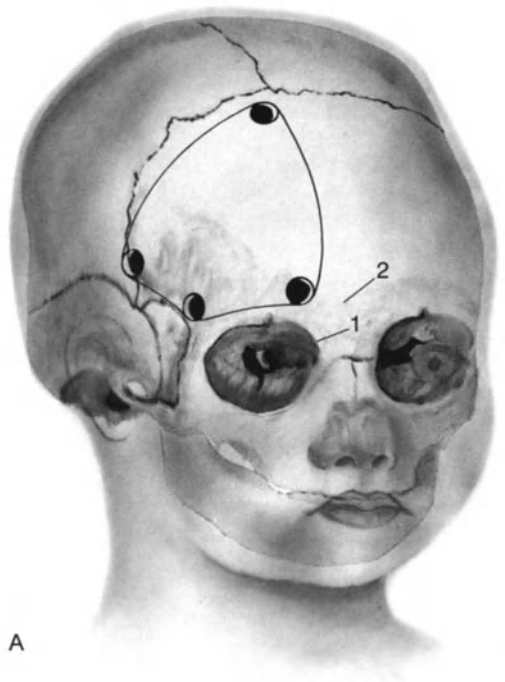
that may be removed in order to gain access to the proximal portion of the sinus in the event its distal portion is damaged during reflection of the free bone flap. If this does happen, the surgeon need only use a rongeur (to bite away the bony spur) to gain immediate access to the SSS. This precaution is best taken because of the proximity of the coronal suture to these parasagittal burr 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.

Frontal Flap (Figures 3.5 and 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 burr 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 burr 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.

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, I will 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 at 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 lacrimal 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.



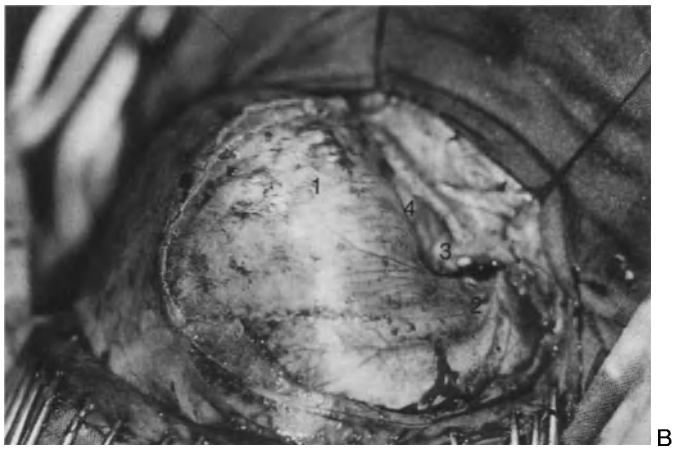


Figure 3.6. (A) Lateral frontal flap, illustrating placement of inferomedial burr hole over medial aspect of supraorbital ridge (*1*), lateral to the glabella (*2*). (B) The exposure of the frontal (*1*) and temporal (*2*) lobes as well as the pterion (*3*) and lesser wing of the sphenoid (*4*), which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

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 lacrimal 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 burr 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 under the transfrontal approach.

Lateral Orbitotomy (Krönlein Approach)

For the Krönlein approach,[1] 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 lacrimal 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 burr (though more time-consuming) permits a relatively precise opening with minimal risk of damaging underlying soft tissue.

Extended Lateral Orbitotomy of Jones

For Jones's extended lateral orbitotomy,[2] 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 arch. 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 bisects 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. Burr holes are placed at the glabella and the "keyhole", and then a craniotome 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 (Figures 3.7 to 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.

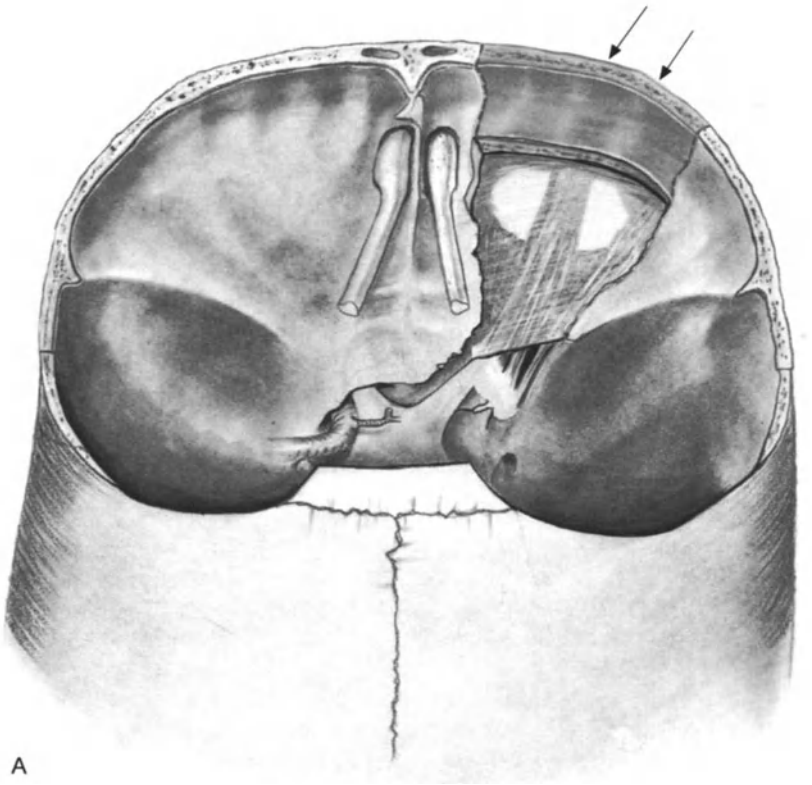
After the orbital roof has been exposed, one may choose only to unroof the orbit if he is certain that the intraorbital tumor is located posteriorly, at the apex of the *cone*, or to remove the supraorbital rim if more complete access to the globe and medial portion of the orbital cone is desired. Leaving the supraorbital rim in place limits considerably the exposure of the intra- and extraconal contents, does not permit exposure of either the lacrimal gland or the trochlear region, and inhibits "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 matter 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.

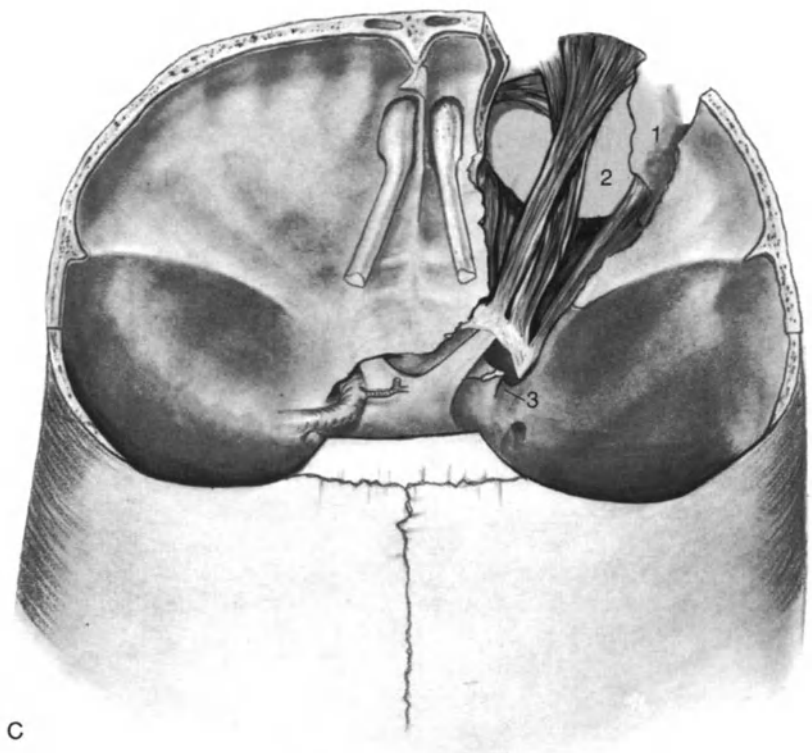
One now has a complete view of all of the orbit as well as the globe and its adnexa. If it is desirable, for example, when beginning resection of a nerve sheath tumor, an angioma, and so on, one may expose the apex of the cone by transecting the levator palpebrae superioris and the superior rectus muscles, sewing them out of the way, and proceeding to work within the base and 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.

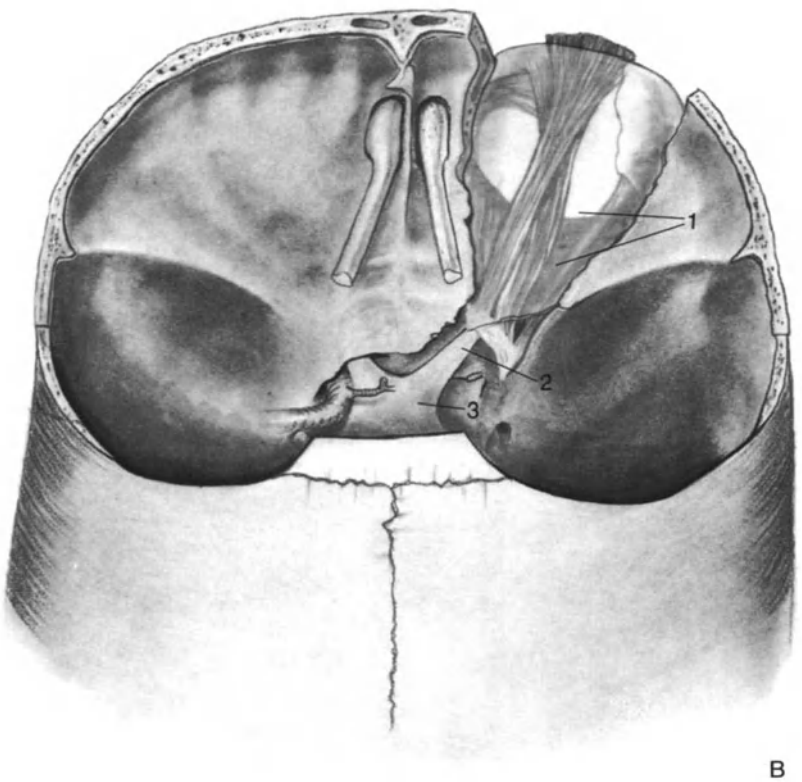
On occasion, when dealing, for example, with optic nerve tumors that involve the intraorbital and intracranial portions of the optic nerve but spare the optic chiasm, one may choose to perform a combined extradural approach to the orbit (in order to resect the intraorbital portion of the tumor) and an 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 the 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 Figure 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. This unroofing completely opens the superior orbital fissure and exposes 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 optic strut so as to expose completely the optic nerve.

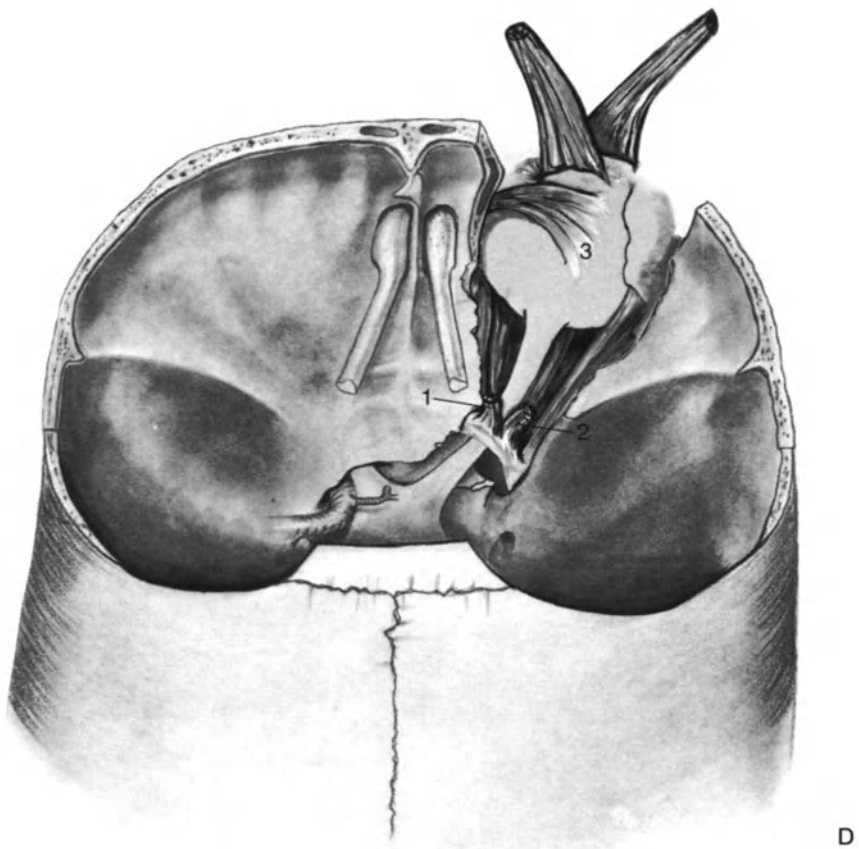
Figure 3.7. (A) The surgeon's view of the anterior fossa if a bifrontal craniotomy is followed by an extradural dissection of the orbital roof, with opening of the latter but leaving supraorbital rim intact (arrows). (B) The supraorbital rim has been removed *en bloc*, after the dura was separated from 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 he wishes to follow the optic nerve into the optic chiasm. (C) The periorbita has been removed, bringing into relief the extraocular muscles, lacrimal gland (*1*), 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, exit of optic nerve from the globe, and the insertion of the superior oblique muscle (*3*) onto the globe.



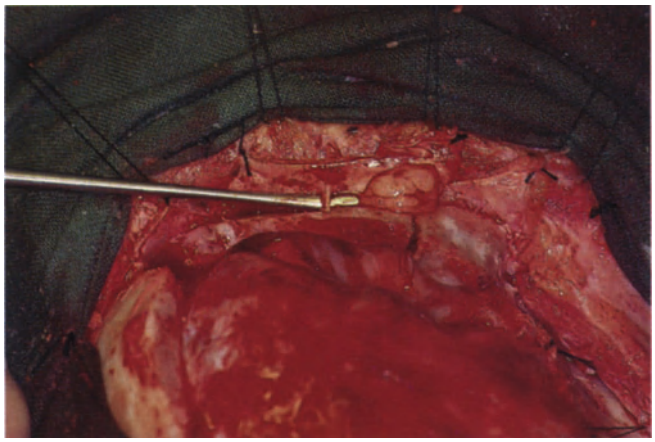




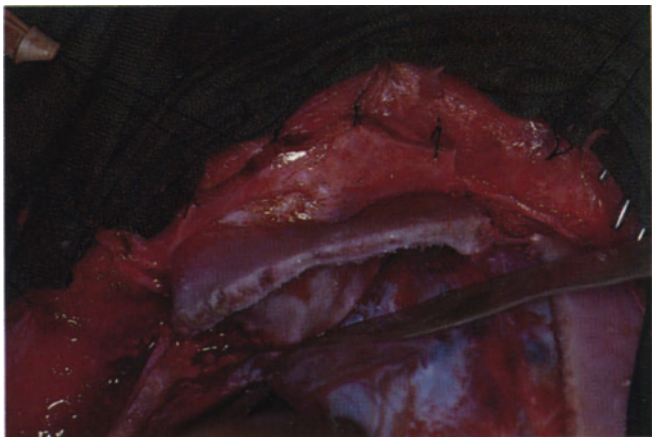
B



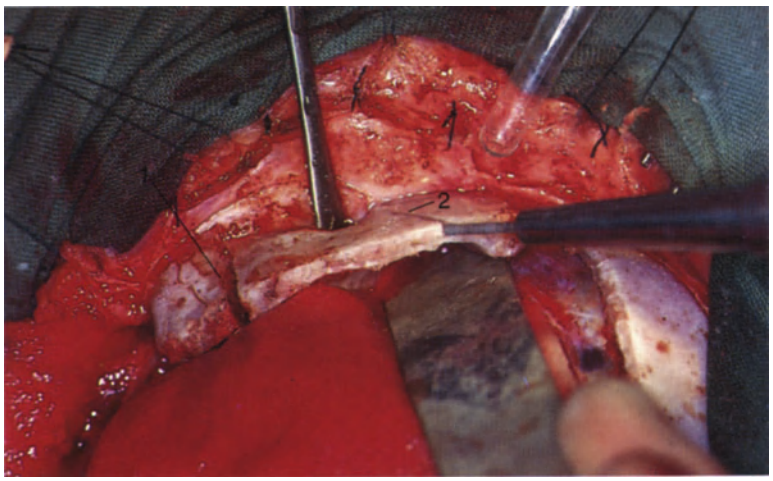
D



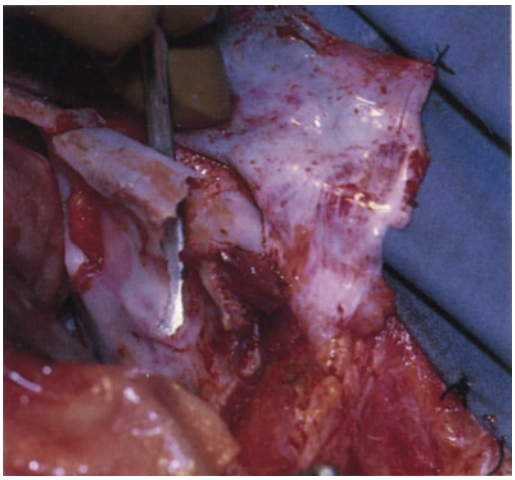
A



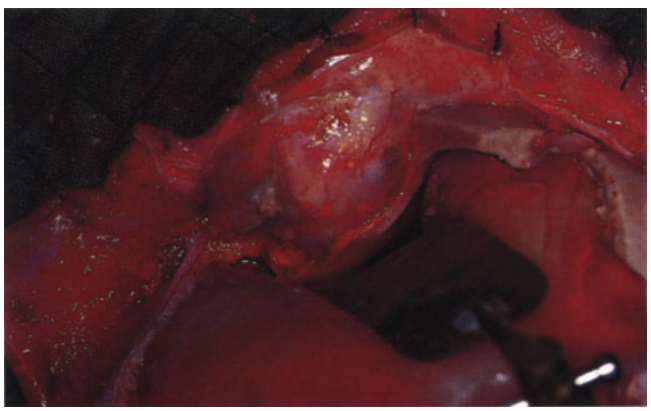
B



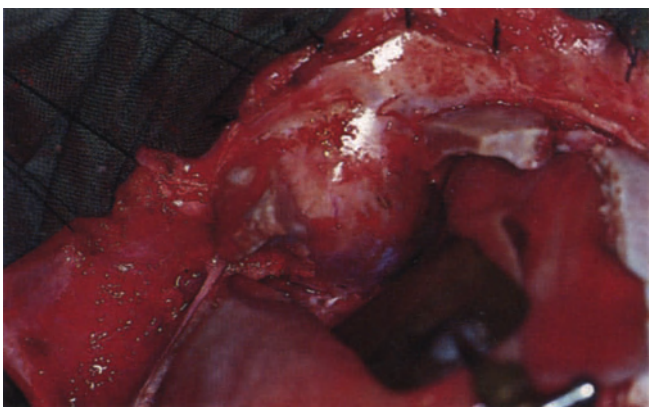
C



D



E



F

◀ **Figure 3.8.** 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 is 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, 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. At the left (*1*), the osteotomy has already been performed with a high-speed drill. At the right (*2*), the high-speed drill is being used to cut the frontal bone. Subsequent to this, a Penfield #1 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. (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.

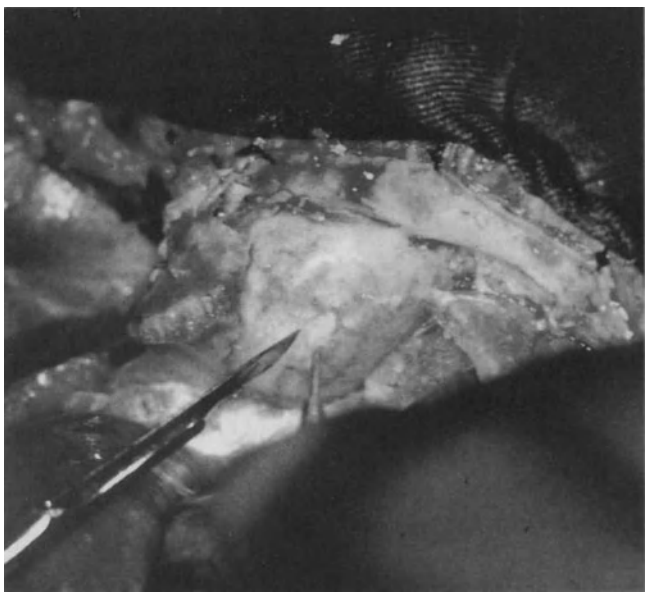
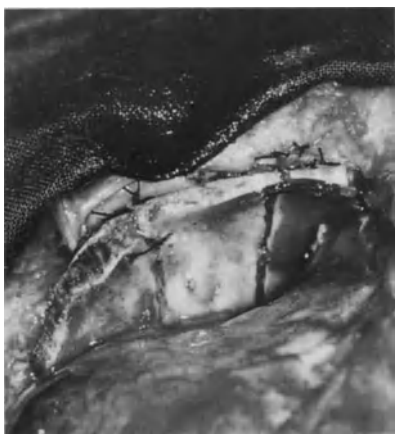
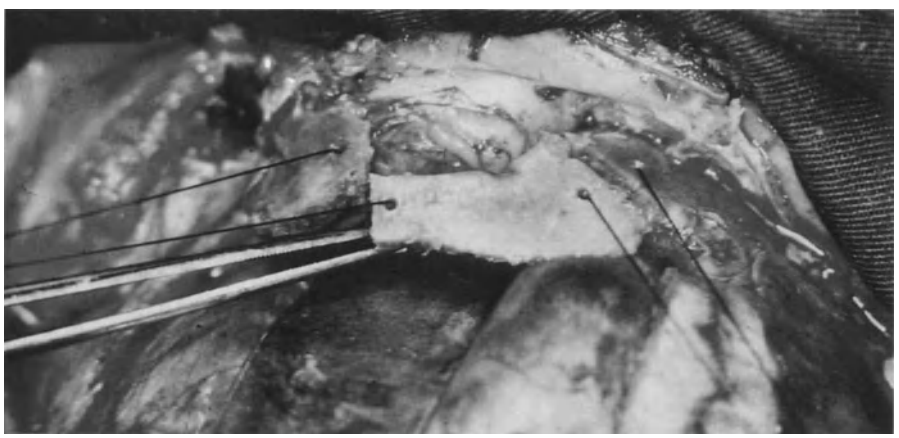


Figure 3.9. The periorbita is opened exactly as one would the dura mater, using a #15 blade, with or without a dural hook. The periorbita is being incised with the assistance of a dural (scleral) hook.

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).



A



B

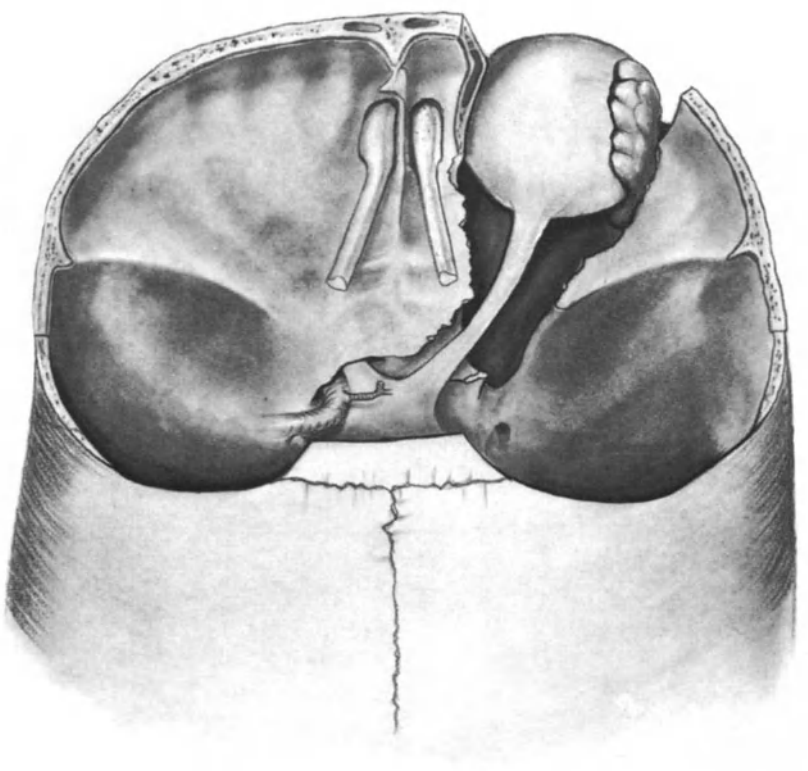


Figure 3.11. Anatomical representation of the relative size, location, and course of the optic nerve, diagrammatically exposed by removing the extraocular muscles and adnexa.

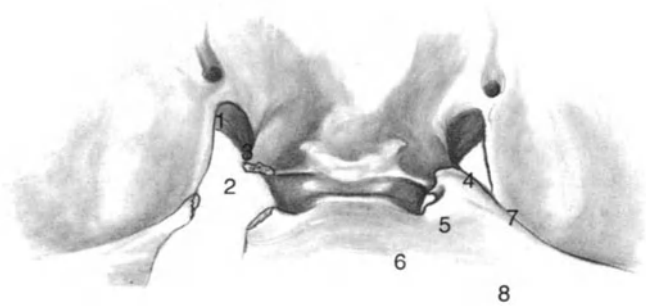


Figure 3.12. Compare resected side (left) to intact side (right). Anatomical representation of unroofing of the most posterior portion of the orbit (*1*), removing the anterior clinoid process (*2*) with the lateral strut of the optic canal (*3*). This opens completely the superior orbital fissure (*4*). 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.

Parietal Flap (Figures 3.13 to 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 burr hole does not exist. Parietal flaps allow access to the convexity of the parietal lobe or to the superior sagittal sinus (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

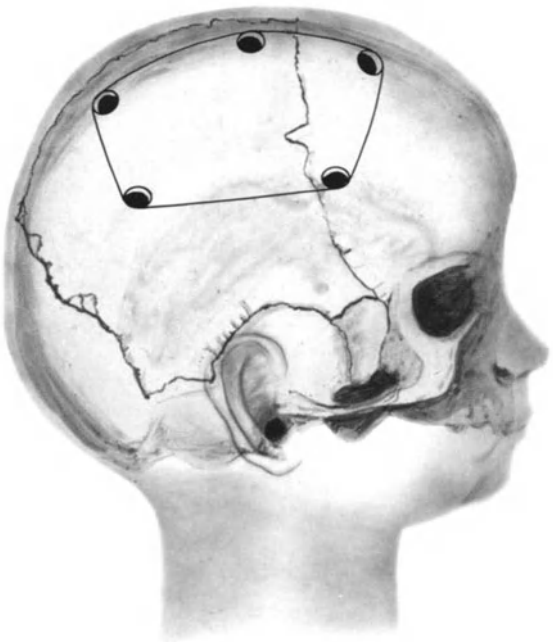


Figure 3.13. This illustrates a pentalateral parietal flap, with the most anterior limb extending across the coronal suture, into the frontal bone.

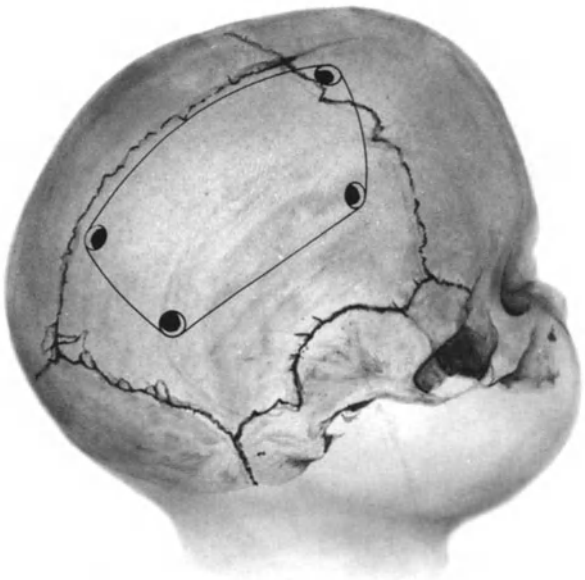


Figure 3.14. A quadrilateral parietal flap offers exposure of the parietal convexity, parasagittal area, corpus callosum, and III ventricle.

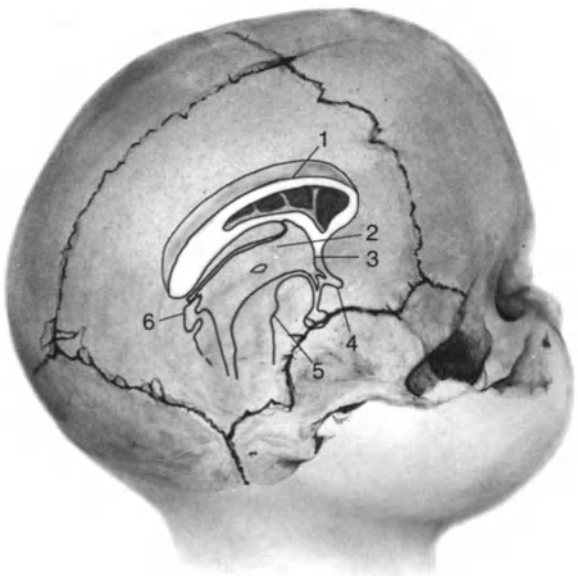


Figure 3.15. This is an anatomical representation, depicting diagrammatically, in three dimensions, the access a quadrilateral bone flap affords to the corpus callosum (*1*), III ventricle (*2*), septum pellucidum, (*3*) optic chiasm (*4*), midbrain (*5*), pineal gland (*6*).

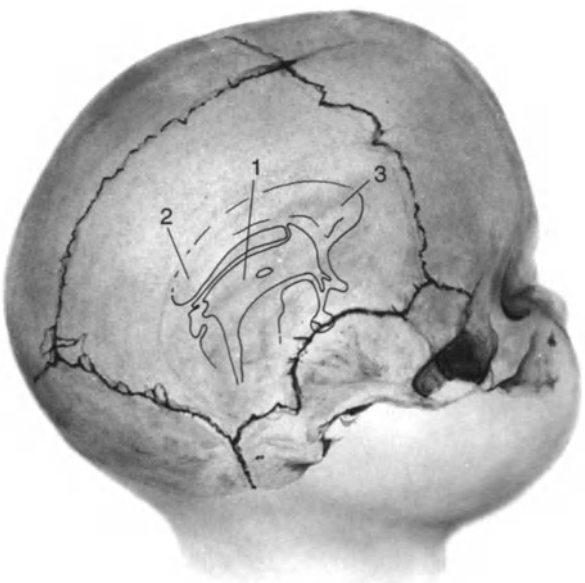


Figure 3.16. The III ventricle (*1*) is projected into its anatomical position in this three-dimensional drawing to illustrate access to it through a parietal flap. Either transventricular or parasagittal approaches may be used. The splenium (*2*) and genu (*3*) are shown at the posterior and anterior surfaces of the III ventricle.

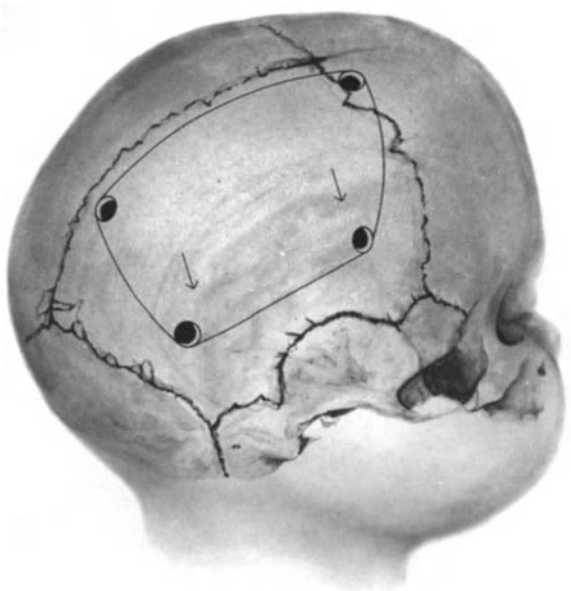


Figure 3.17. One may gain access to the supramarginal and angualar gyri as well as the superior temporal convolution through a superior parietotemporal flap, by placing the inferior burr holes below the superior temporal line (arrows).

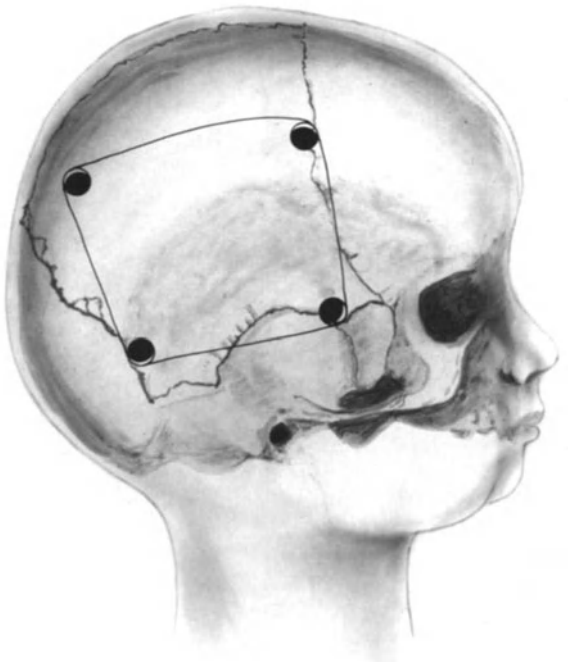


Figure 3.18. Inferior parietotemporal bone flap.

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.

Parietotemporal Flap (Figures 3.17 and 3.18)
The parietal flap may easily be extended into a parietotemporal flap, if the parietal lesion also invades the superior or middle temporal convolutions posteriorly (in correspondence to the supramarginal and angular gyri). Similarly, one may place the burr 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 burr holes at (1) the parietal eminence, (2) the coronal suture, (3) the pterion, and (4) just above the base of the mastoid bone.

Biparietal Craniotomy (Figures 3.19 and 3.20)
The biparietal craniotomy is used for resection of the sagittal suture (really a craniectomy), to gain access to the superior sagittal sinus (SSS) or both sides of the falx cerebri, to attain complete exposure of the great vein of Galen for arteriovenous fistulae, and to lower 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.

Temporal Flaps
Temporal bone flaps may be placed in three basically different positions, depending upon whether the sur-

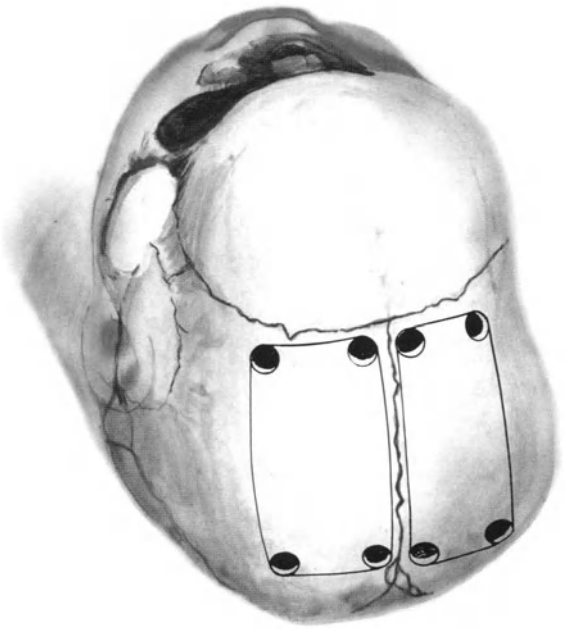


Figure 3.19. Biparietal quadrilateral bone flaps, placed one on either side of the sagittal plane, leaving the bone over the superior sagittal sinus (SSS) intact. In the event access to the SSS becomes desirable, this strip of bone may be quickly removed.

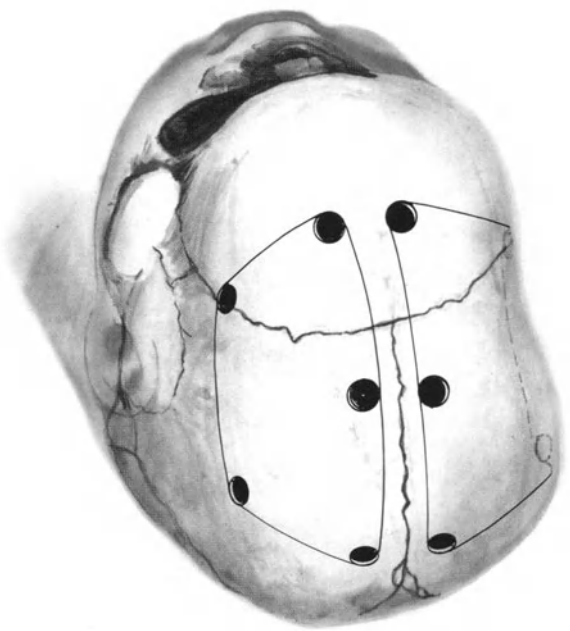


Figure 3.20. Bilateral frontoparietal (pentalateral) bone flaps may be extended across the coronal suture, leaving bone over the superior sagittal sinus.

geon wishes access to the temporal tip, the posterior portion of the temporal lobe, or the entirety of the temporal lobe and the Sylvian fissure.

Anterior Temporal Flaps (Figures 3.21 and 3.22)
The anteroinferior burr 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 preparation of the craniotomy for an anterior temporal approach.

The superior burr holes are placed immediately beneath the superior temporal line, the posteroinferior burr 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, 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.

Posterior Temporal Flaps (Figure 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 burr hole is placed at the superior temporal line, directly above the external auditory canal, and the inferior burr hole is placed at the base of the mastoid process. The anterior and posterior burr 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.

Midtemporal Flap (Figure 3.24)
The midtemporal flap incorporates the anterior and posterior temporal burr holes so as to permit access

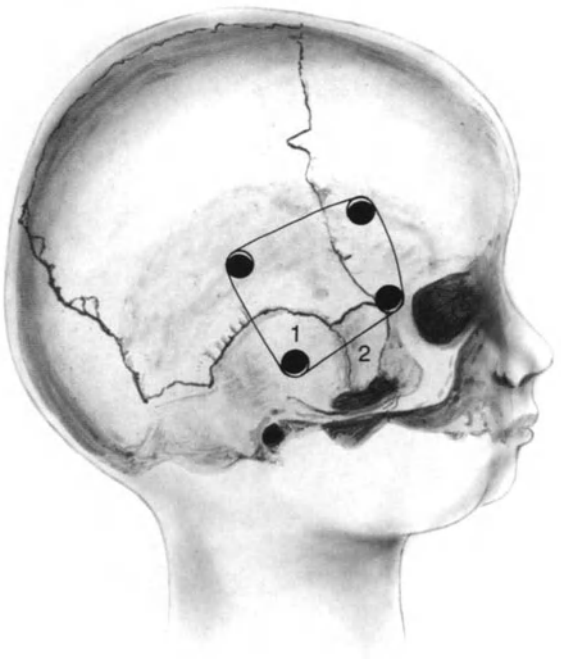
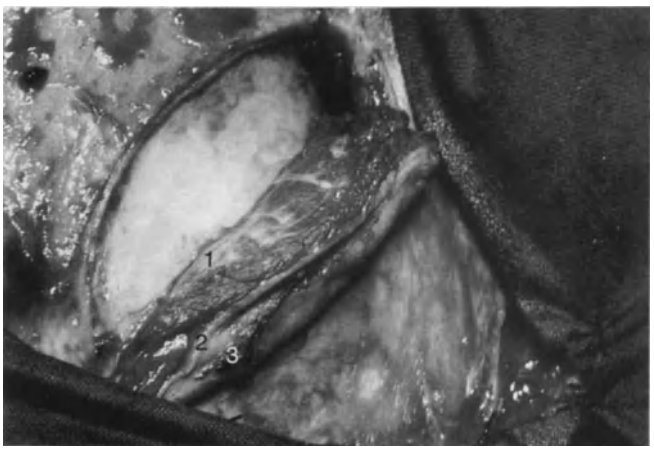


Figure 3.21. Anterior temporal craniotomy. The anteroinferior burr hole is placed at the key hole so as to assure exposure of the temporal pole. Note that the inferior craniotomy line extends through the squamous temporal (*1*) and greater wing of the sphenoid (*2*). Rongeuring these bones gives access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

A



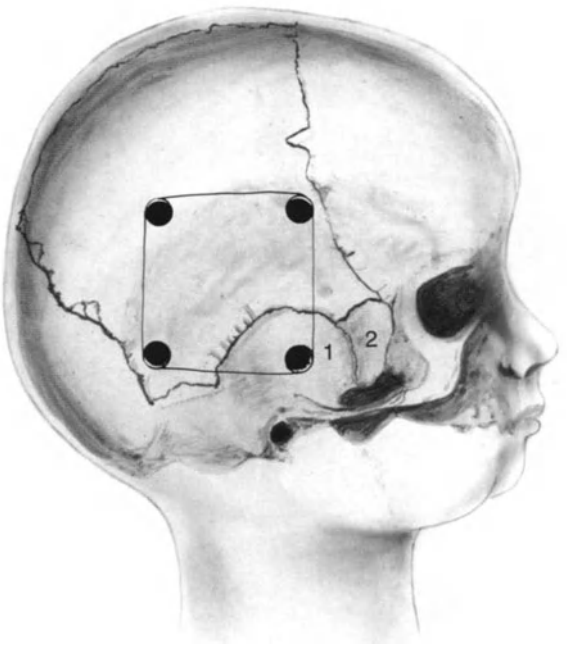
B

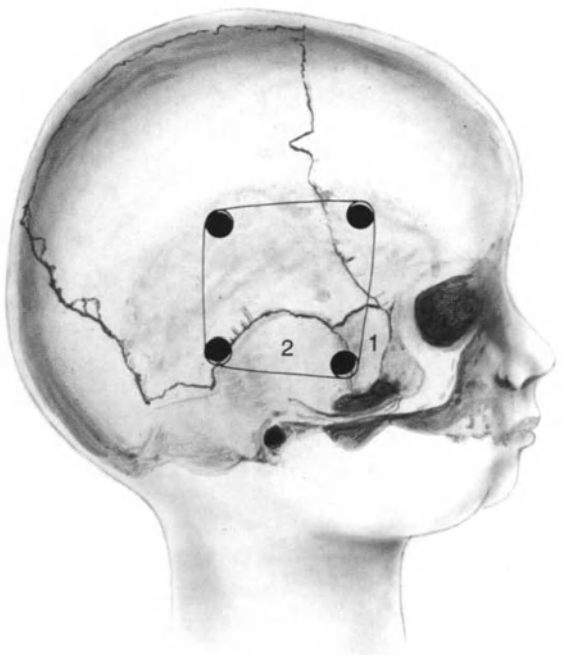
Figure 3.22. (A) Suggested technique for use of periosteal elevator to strip periosteum from the greater wing of the sphenoid and squamous temporal bones, so as to avoid fraying the periosteum: cutting edge to be held firmly and run parallel to muscle insertion. (B) The bipenniform temporalis muscle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

to the entire temporal lobe, from the temporal pole as far posteriorly as the temporooccipital 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.

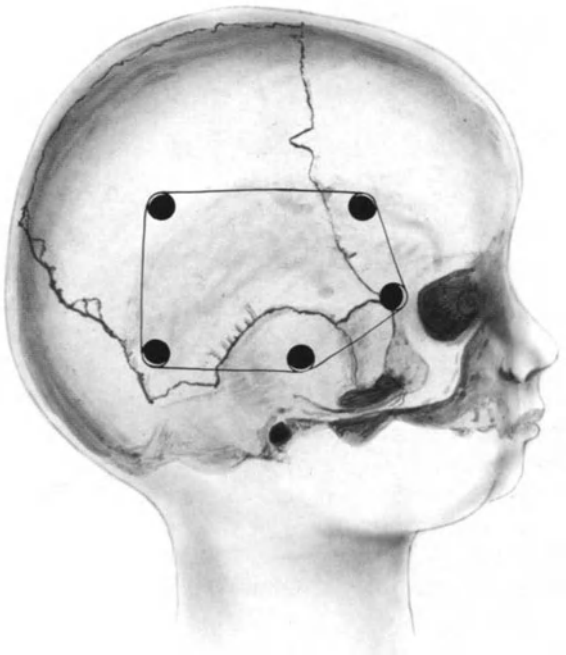
The midtemporal flap is generally pentalateral. The anteroinferior burr hole is placed at the pterion, the anterosuperior burr hole directly above it at the superior temporal line. The posterosuperior burr hole is placed immediately beneath the parietal eminence, and the posteroinferior burr hole just anterior to the lambdoidal suture in a line above the digastric groove. The inferointermediate burr 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

Figure 3.23. Posterior temporal flap, illustrating placement of burr holes. Note that no burr 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.





A



B

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 burr holes are all just beneath the superior temporal line, the anteroinferior burr 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.

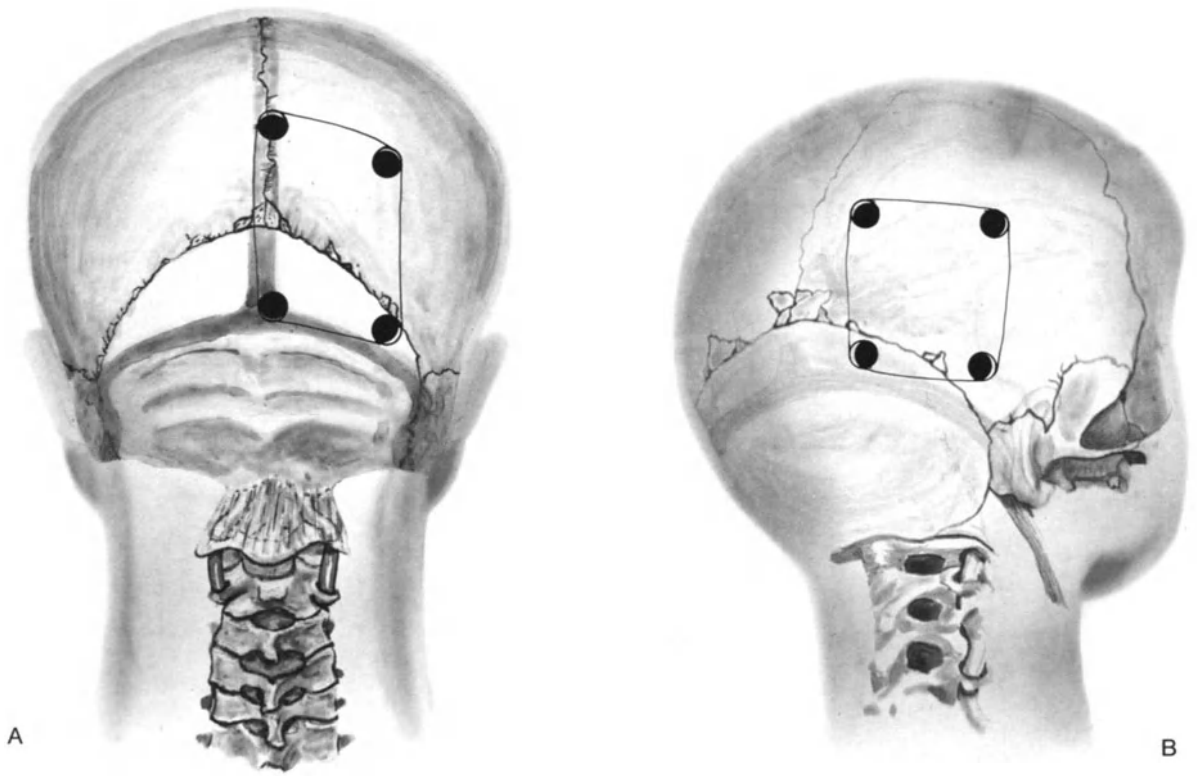


Figure 3.25. (A) Medial occipital bone flap, illustrating location of burr holes and osteotomy lines, as viewed by the surgeon, in a straight posteroanterior view. Note that the lambdoidal suture is within the flap, and that the medial osteotomy follows the sagittal suture. The inferior osteotomy line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

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 Flap (Figure 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 of 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 burr 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 burr 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 burr hole must be placed over the superior sagittal sinus, with its medial surface just touching the sagittal suture. The superolateral and inferolateral burr 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).

Lateral Occipital Flap

The lateral occipital bone flap has the inferomedial burr hole just superolateral to the torcular Herophili, the inferolateral hole above the sigmoid sinus. The superior

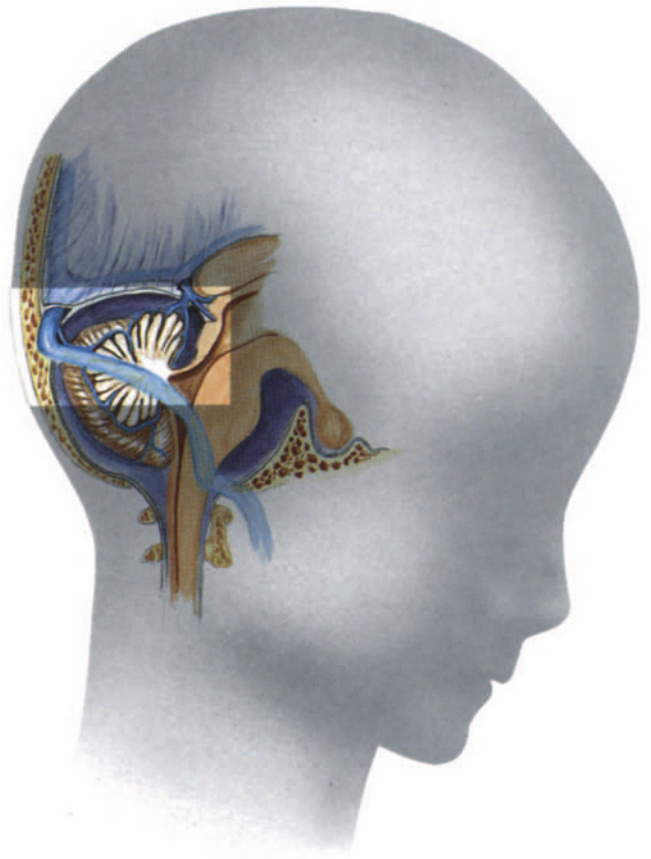


Figure 3.26. Superior cerebellar triangle.

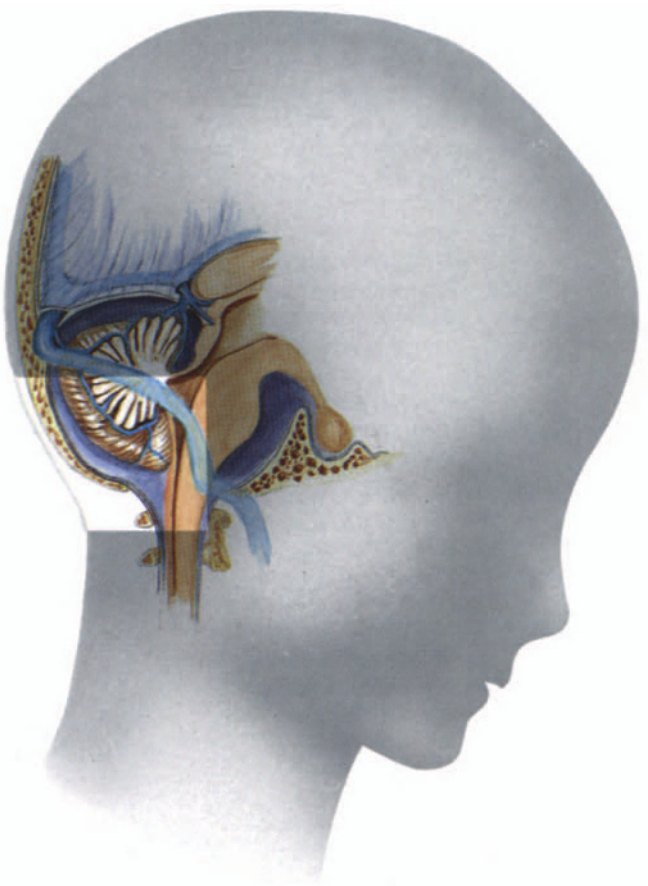


Figure 3.27. Inferior cerebellar triangle.

holes are placed medial and lateral to the parietal eminence.

Suboccipital Flap

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 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 as its apex, and the (inferolateral coursing) superior surfaces of the cerebellar hemispheres as 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, 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! 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

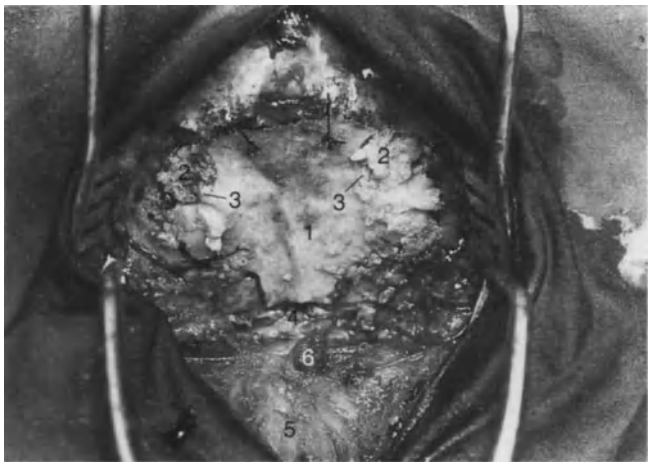


Figure 3.28. Suboccipital craniotomy at time of closure, illustrating replacement of the bone flap (*1*) and filling of the burr holes with bone plugs (*2*) and bone chips (*3*). One notes an intact arch of C-1 (*4*) and erector cervicis muscles (*5*) still attached to C-2 (*6*).

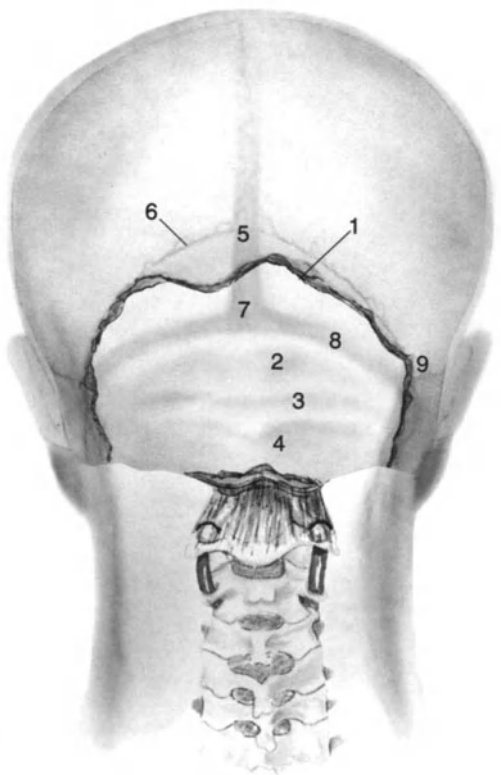


Figure 3.29. Periosteal dissection for suboccipital craniotomy. The periosteal dissection (*1*) has been brought above the highest nuchal line (*2*). This drawing shows both the intermediate (*3*) and lowest (*4*) nuchal lines. Lambda (*5*) and the squamosal suture (*6*) are superior to the torcular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

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 dense scar tissue between muscle and dura, and in an extremely high incidence of suboccipital bulging. This leaves the child with a weakened area over one of the most vital portions of the human 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, the jugular foramen, the 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 from it (Fig. 3.29). However, this 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 parietooccipital (lambdoidal), occipitomastoid, and parietomastoid sutures meet.

Midline Suboccipital Craniotomies

Inferior Suboccipital Craniotomy (Figures 3.30 and 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. 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 sinus, causing profuse venous bleeding or air emboli.*

The Gigli saw guide should be passed horizontally from one burr hole to the other, and then the Gigli saw should be used to connect the burr holes with a linear osteotomy. Passage of the saw guide horizontally presents no difficulties, though great 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

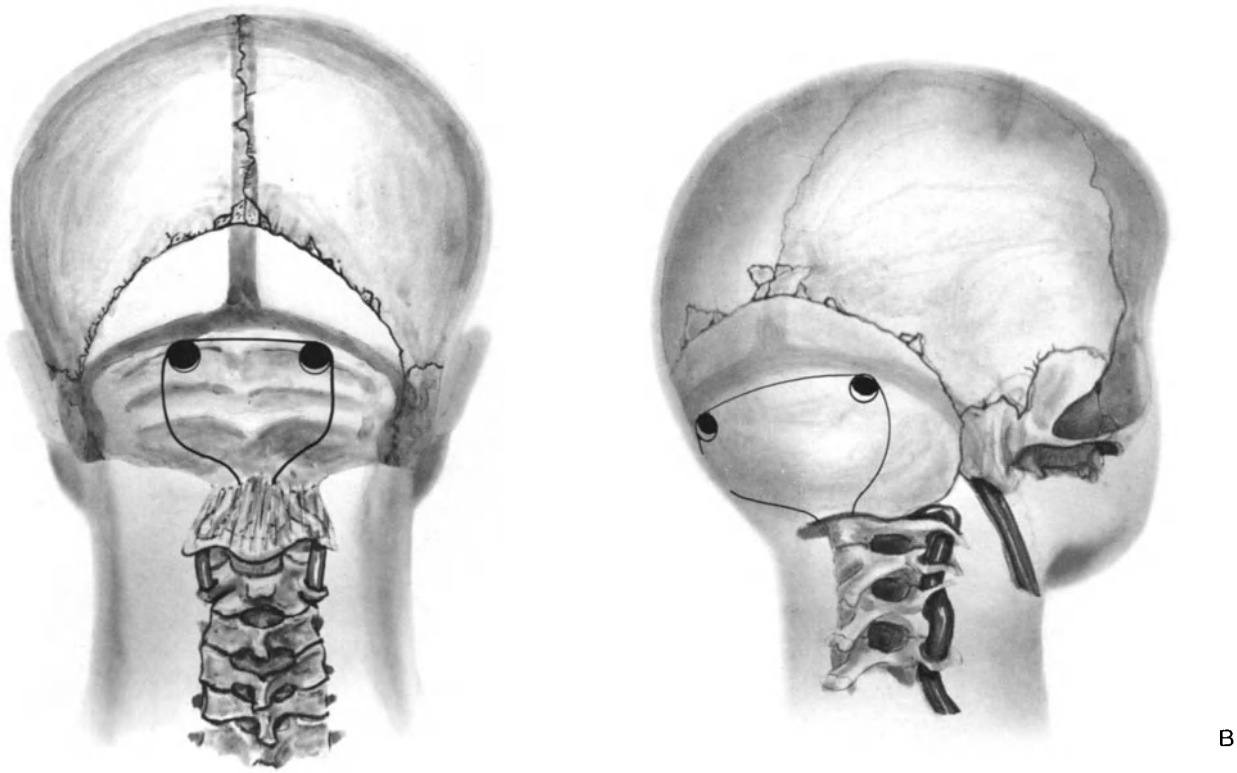


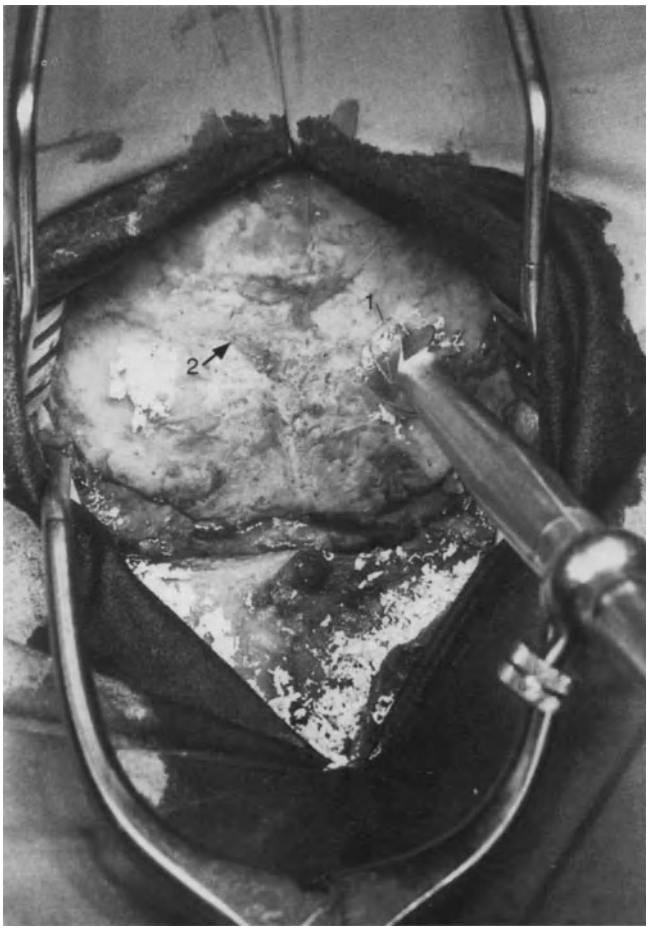
Figure 3.30. (A) Inferior suboccipital craniotomy as viewed in a straight posteroanterior line, illustrating the burr holes and osteotomy lines outlining the free (squamous occipital) bone flap. (B) Inferior suboccipital craniotomy as viewed in a posteroblique line.

then from the left, always feeling the tip of the guide against bone and dura, eases the guide across the midline into the contralateral burr hole. If it proves to be difficult to pass the guide directly horizontally from one burr 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 should then be 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 extraordinarily small (measuring 2.0 cm) and that the lateral surfaces of the foramen magnum consist of thick bony struts which lead to the occipital condyles. Therefore, an attempt to pass the Gigli saw guide directly inferiorly from the burr 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 advisible to attempt to pass the saw guide from a burr hole downward across the rim of the foramen magnum, in the epidural space, since this, too, puts the annular 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 forms the annular sinus.

A craniotome is used to connect the lateral surface of each burr 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, 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, atlantooccipital membrane, and periosteum/pericranium join. This now redundant



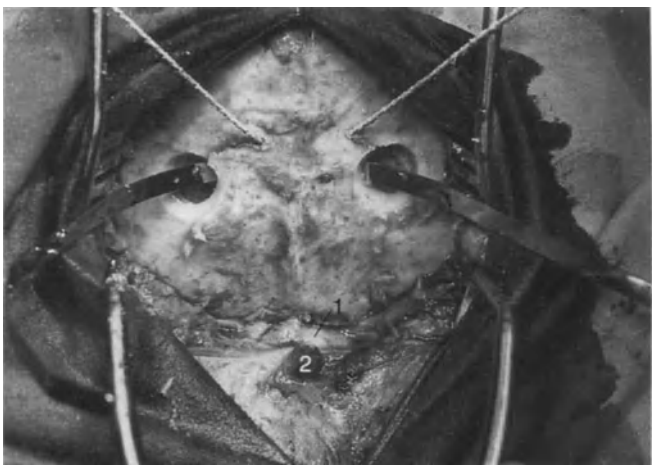
A

B

Figure 3.31. (A) Placement of burr hole (*1*) just at the highest nuchal line (*2*). (B) In passing the Gigli saw guide (*1*) from one burr 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 burr hole (*3*). This facilitates separating dura from the internal occipital crest and frees dura from the inner surface of the flap area. (C) Completion of passage of Gigli saw guide across midline. (D) 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*). (E) 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 at the right has been completed, the one at 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 accomplishes (*2*). (F) The lateral osteotomies have both been brought to the rim of the foramen magnum, so that one now separates squamous occipital bone flap from the underlying dura, cracking the thin slip of bone left at each border of the foramen magnum by gently depressing the base of the bone flap. (G) 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 sinus so as to avoid jamming the bony posterior rim of the foramen magnum into the sinus. (H) The final dissection stage consists of freeing the dura/periosteum from the rim of the foramen magnum, a point of great 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.

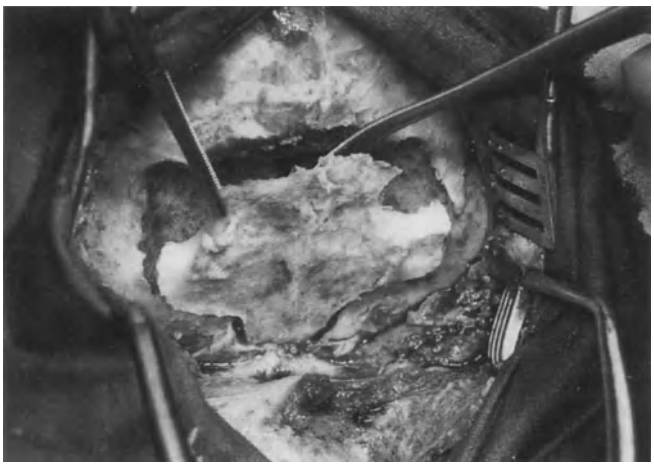
C

D



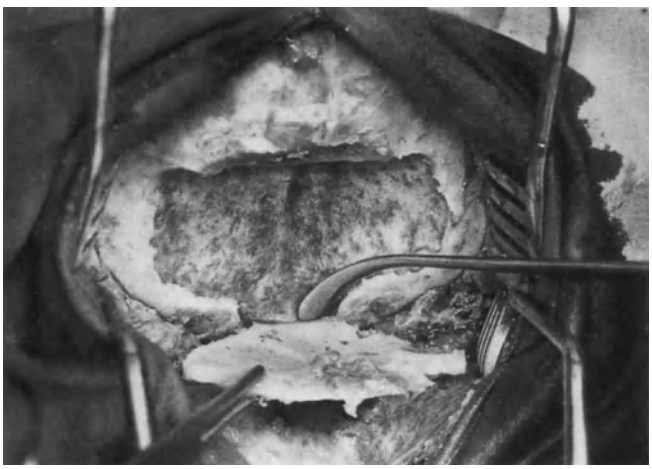
E

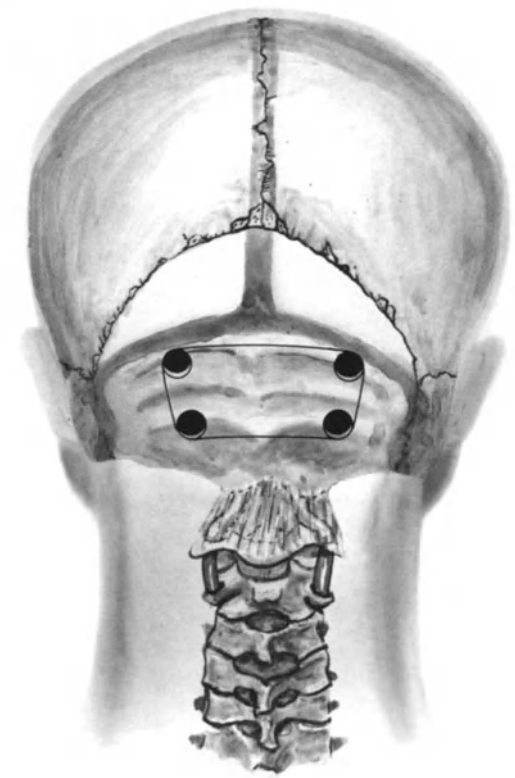
F



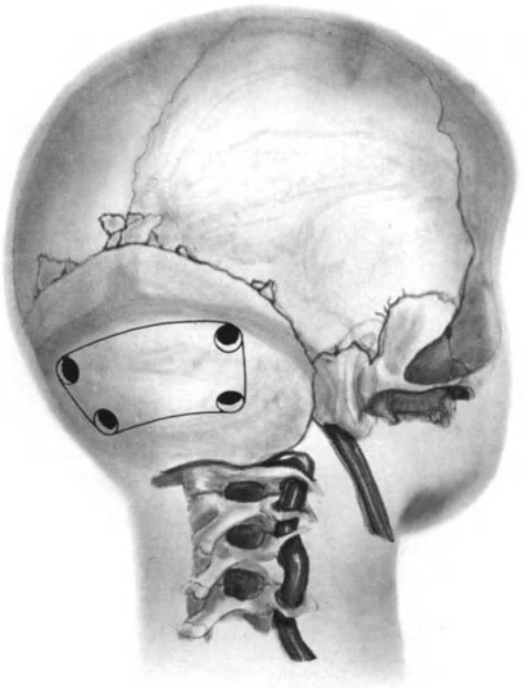
G

H





A



B

C

D

Figure 3.32. (A) Superior suboccipital craniotomy as viewed from the straight posteroanterior line. The upper burr 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) Superior suboccipital craniotomy as viewed from a posterooblique 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.

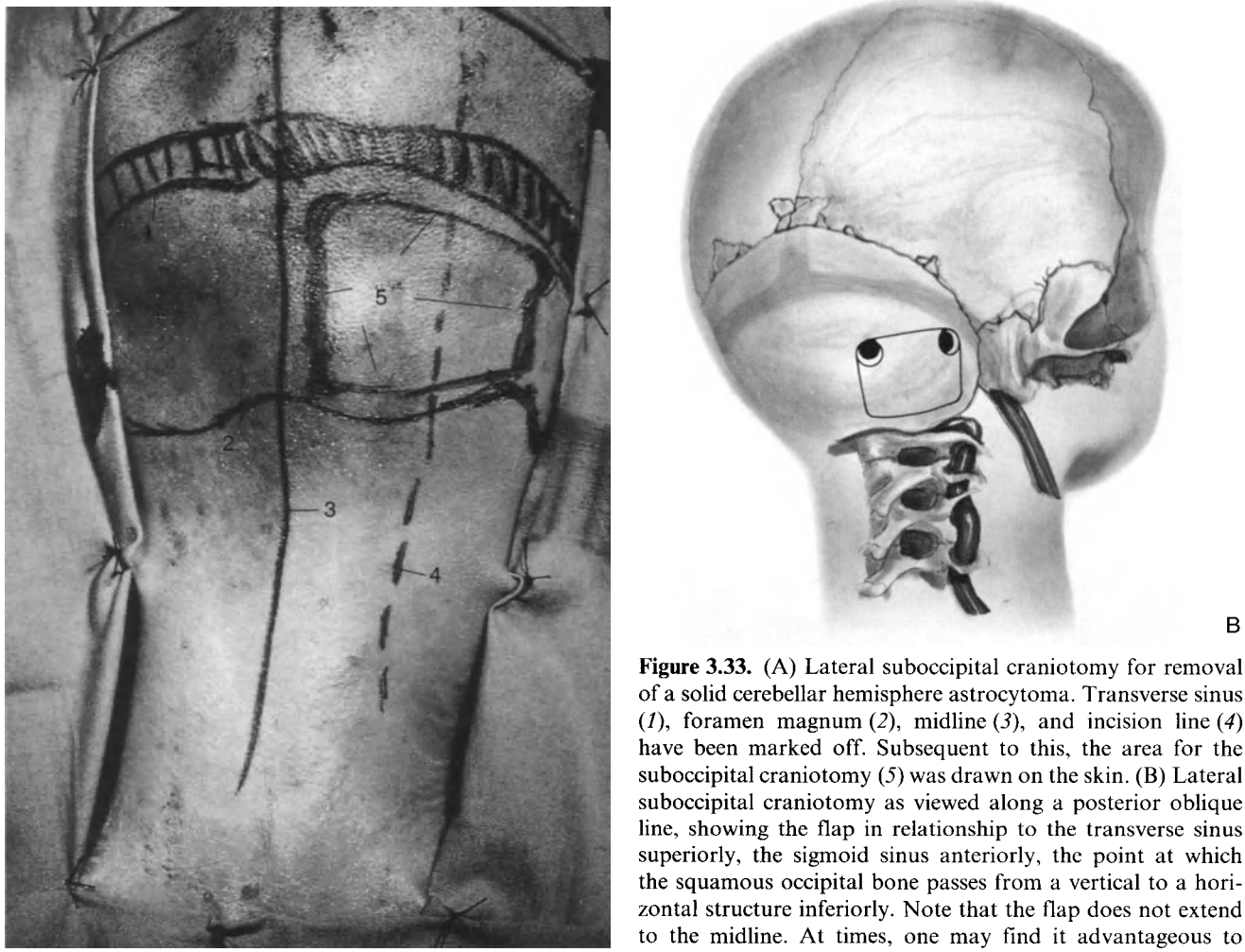


Figure 3.33. (A) Lateral suboccipital craniotomy for removal of a solid cerebellar hemisphere astrocytoma. Transverse sinus (*1*), foramen magnum (*2*), midline (*3*), and incision line (*4*) have been marked off. Subsequent to this, the area for the suboccipital craniotomy (*5*) was drawn on the skin. (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 burr holes, as illustrated here.

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 (Figure 3.32)
The superior suboccipital craniotomy is performed by placing four burr 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 inferior suboccipital craniotomy are taken in passage of the Gigli saw guide across the midline. The Gigli saw is recommended for connecting the burr 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 burr hole in one direction first, then in the opposite direction. This frees all of the dura, and permits safe osteotomies.

Lateral Suboccipital Craniotomy (Figure 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 burr 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

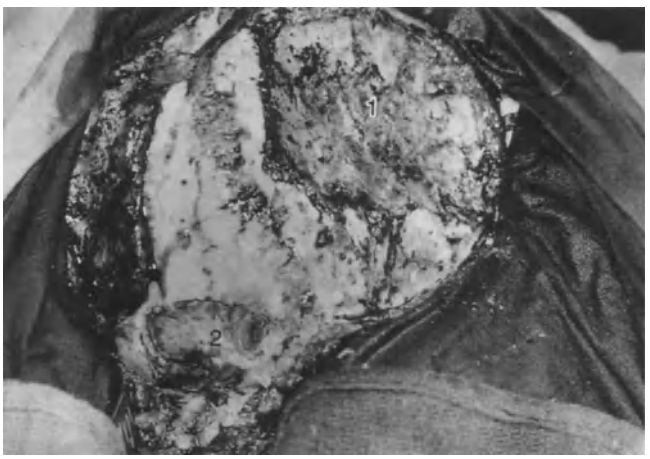


Figure 3.34. Supra- and infratentorial Craniotomies. The parietotemporooccipital (*1*) and lateral suboccipital (*2*) bone flaps have been reflected.

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.

Supra- and Infratentorial Craniotomy (Figure 3.34)
A lateral suboccipital[1] and parietotemporal[2] 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.

Hemispheral Craniotomy (Figure 3.35)
The hemispheral craniotomy is used primarily for hemispherectomy and, when bilateral, for lowering of the superior sagittal sinus (SSS). In this 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 the SSS, 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 hemispheral craniotomy is being used for lowering the SSS, the medial burr holes are brought to the parasagittal plane with their medial surfaces bordering upon the lateral extremity of the SSS. This allows for a median strip of bone and the anterior fontanel to remain in the sagittal plane. This 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

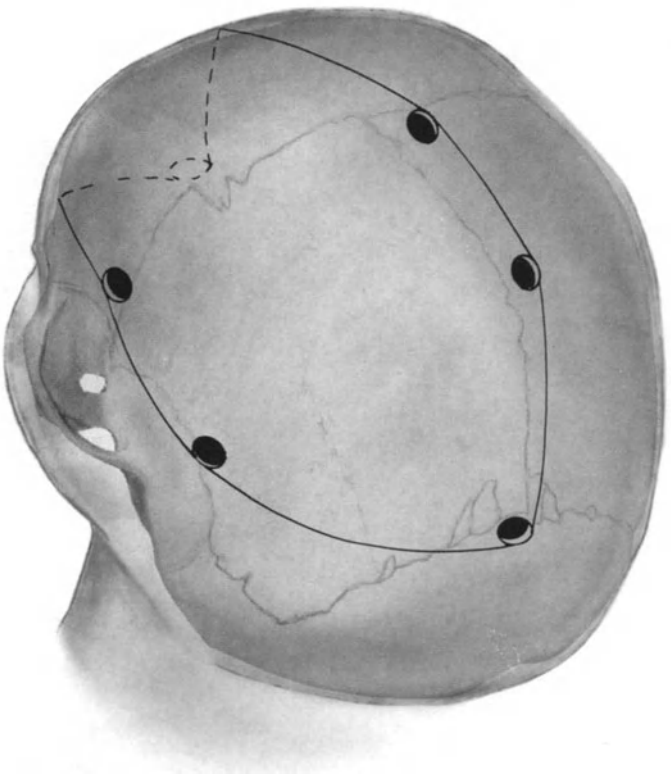


Figure 3.35. Placement of burr holes and osteotomy lines for hemispheral craniotomy are illustrated in this anatomic illustration. The placement of the burr holes permits access to all hemispheral vascular structures and to the entirety of the hemisphere. The superior holes are placed to the contralateral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

when the vault of the skull is reconstructed and molded over the atrophic hemispheres.

The placement of the burr holes for the hemispheral 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."

Laminotomy (Figures 3.36 to 3.45)
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.

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 particularly interested in the fact that the cervical spine, the most mobile segment of the

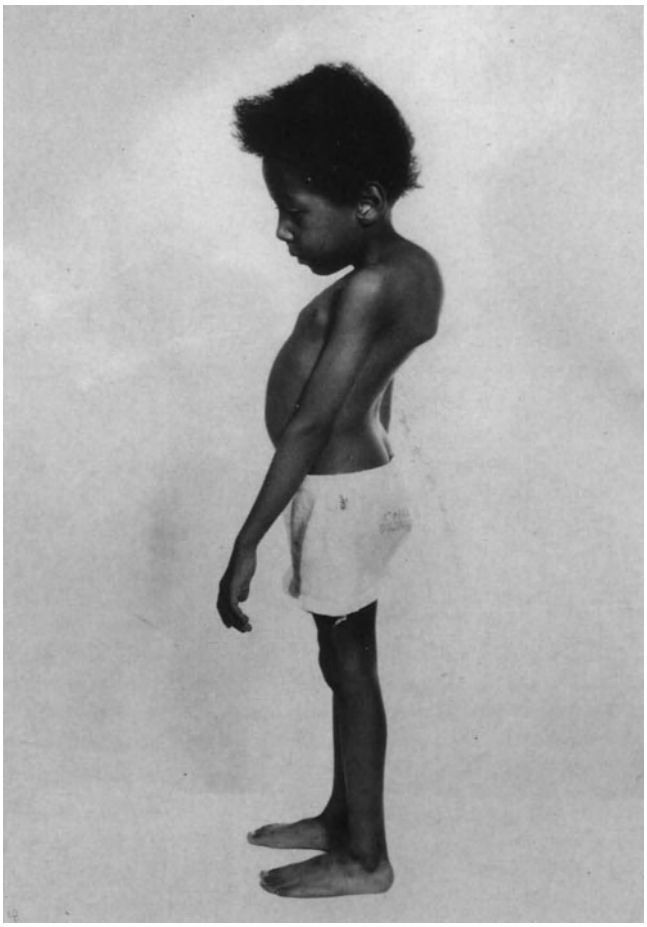
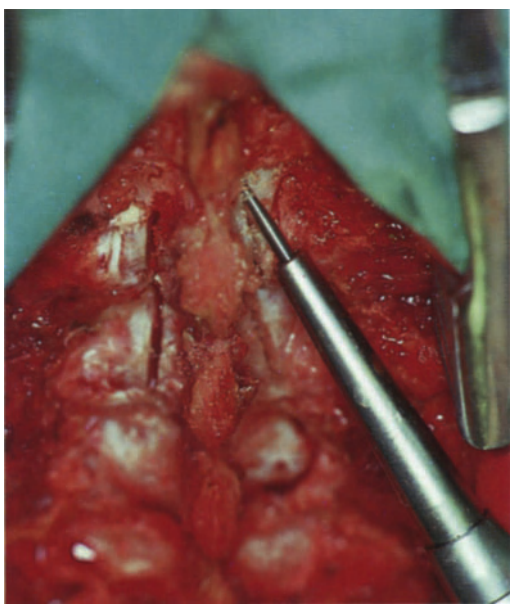
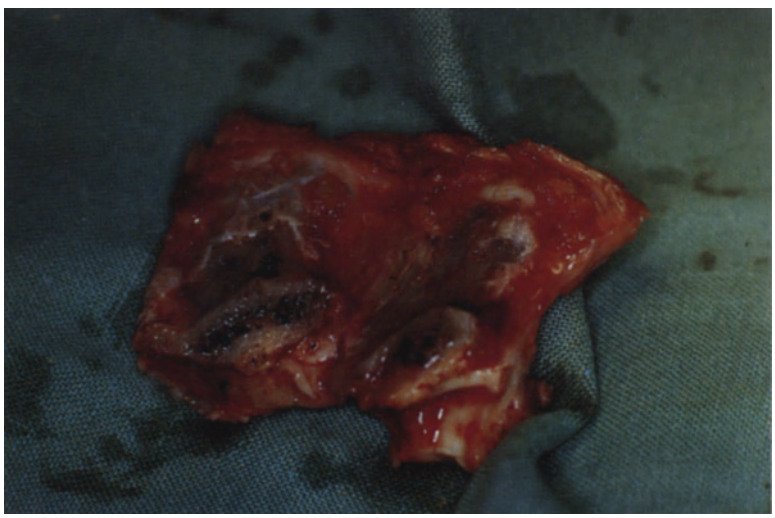


Figure 3.36. This child's kyphoscoliosis developed progressively over an 18-month period of time following multiple level thoracic laminectomy. None was present preoperatively.



A



B

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.

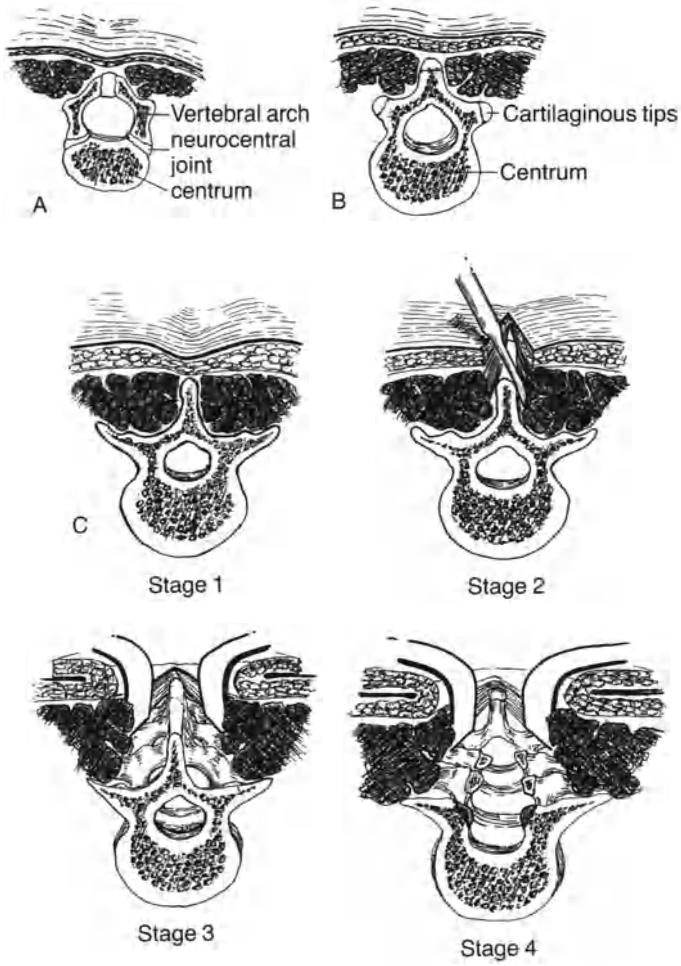


Figure 3.38. (A) Infant: Note that there is almost no spinous process. The laminotomy is made at the same, paraspinous, position. (B) Toddler: The spinous process of the toddler is forming. (C) Juvenile: The four stages of exposure and osteotomy are illustrated in a juvenile to represent reflection of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

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 centra) 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, most important of all, offer purchase to developing muscle masses. It is their conclusion that abnormal growth patterns and greater elasticity of musculoligamentous structures in children are responsible for the rapid and severe deformities of the vertebral column, especially the cervical column, which result following laminectomy.

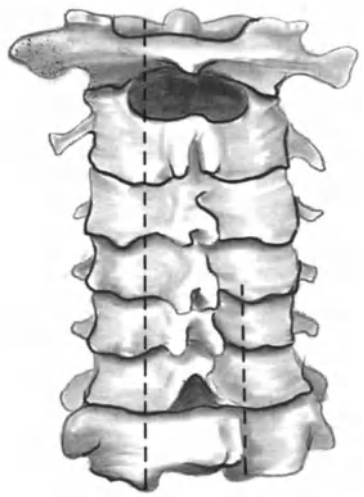
Following the work by Raimondi and coworkers in 1976,[8] concluding that deformities (kyphosis, scoliosis, accentuated lordosis) of the vertebral column in childhood result from laminectomy, and recommending that laminotomy be substituted for laminectomy, Yasuoka and colleagues[9] in 1981, reported that "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.

Figure 3.39. The erector spinae muscle masses of a 10-year-old child have been stripped from the laminae and spinous processes.

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 LaRocca 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 nothing more than 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



A

B

Figure 3.40. (A) The general anatomical characteristics of the posterior spinous portion of the cervical canal are herein represented as is the caudocephalad osteotomy. (B) Laminar osteotomy being performed. One notes that the osteotomy is performed in lateromedial and dorsoventral directions along the line of the pars interarticularis.

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 one year of age the surgeon should perform a laminotomy even if only one level is to be exposed; in children between one and fifteen years for two or more levels, and in patients older than fifteen 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.

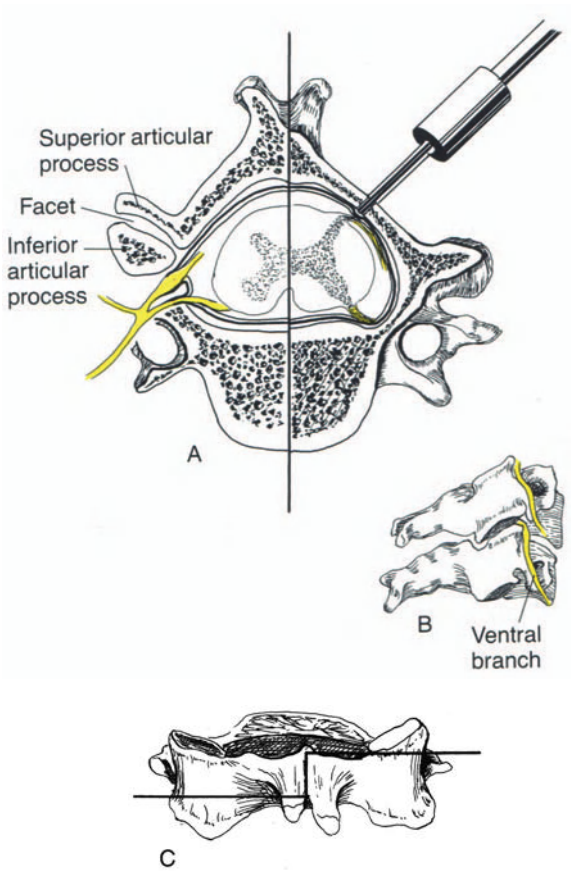


Figure 3.41. (A and B): The fourth cervical vertebra is (diagrammatically) shown cut at two levels, in horizontal section. (C) The line of section, not of recommended osteotomy. This line of section is used to illustrate the inferior level of the vertebra at the left, the superior level at the right.

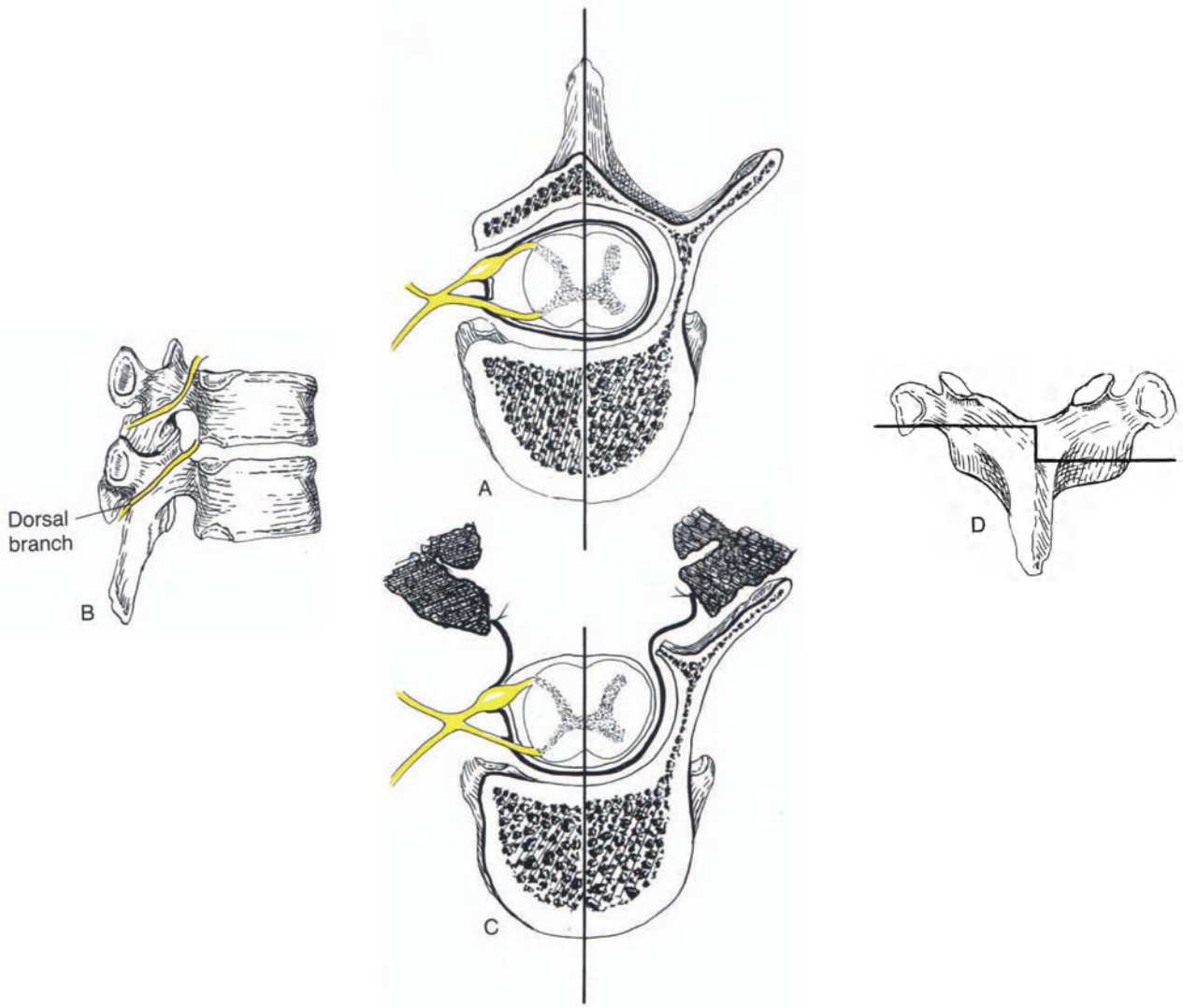


Figure 3.42. (A) The third thoracic vertebra is shown, diagrammatically cut at lower and higher levels to illustrate the different anatomical characteristics of the laminae and to illustrate shingling. (B) The course of the nerves in the lateral view is shown. (C) This illustrates the exposed spinal cord and arachnoid, the opened and reflected dura mater. (D) This depicts the lines of section, superior and inferior.

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 chil-

dren!), 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 to 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 transmission tomography (CTT) 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.

Figure 3.43. The laminar flap is being lifted from its anatomical position, retracting it superiorly and taking care not to allow it to buckle (so as to avoid compressing the spinal cord), as the #15 blade progressively cuts the yellow ligaments at each level. This child is approximately 8 years old.

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, 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 maximum possibility of beveling, not with the expectation to obtain an osteotomy which is wedge-shaped, but with the hope of minimizing the size of the gutter and, thus, facilitate 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 cra-

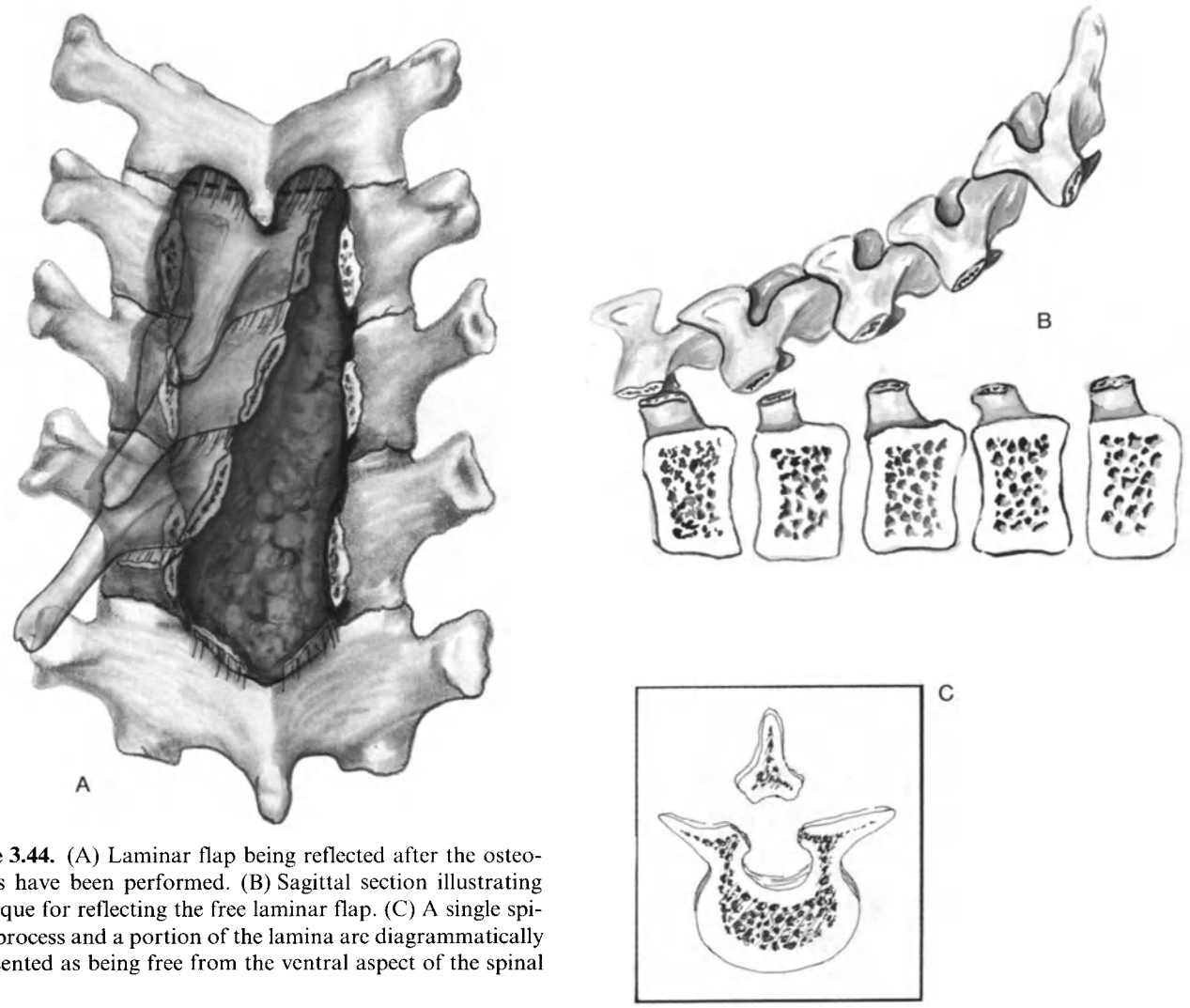


Figure 3.44. (A) Laminar flap being reflected after the osteotomies have been performed. (B) Sagittal section illustrating technique for reflecting the free laminar flap. (C) A single spinous process and a portion of the lamina are diagrammatically represented as being free from the ventral aspect of the spinal canal.

niotome 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 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 caphalad 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 one 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 laminotomy 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 re-

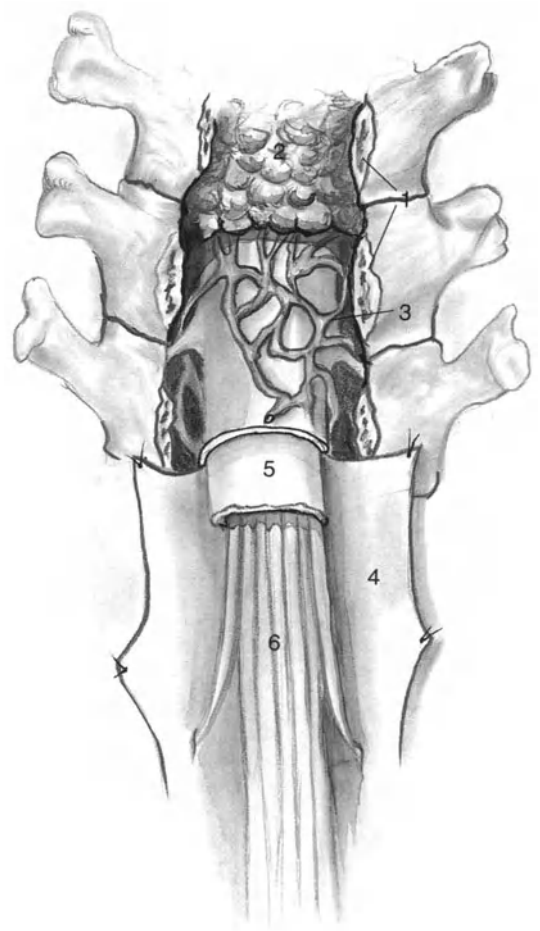


Figure 3.45. An osteotomizied thoracic spine is diagrammatically represented, illustrating the desired inclination of laminar cut (*1*); the epidural fat (*2*) and venous plexus (*3*), reflected dura mater (*4*), and, deeper, the enclosed arachnoid (*5*) and neural elements (*6*).

flected 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 feshly soaked fluffy cotton or precut 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 3.45 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 (Figures 3.46 to 3.49)
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 of any make to cut the bone between burr 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. 3.1 A) 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. 3.1 B). Bony regrowth occurs across the osteotomy lines made by the Gigli saw. Although the vendors of power craniotomies 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 periodically 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

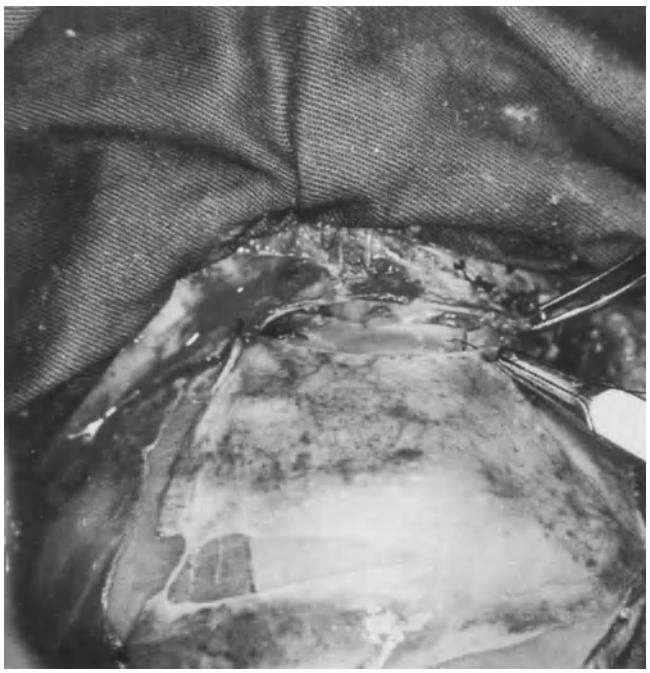


Figure 3.46. This medial frontal craniotomy is being closed with 4-0 sutures, through the periosteum which had been left on the free bone flap and periosteum which had been reflected over the supraorbital rim.

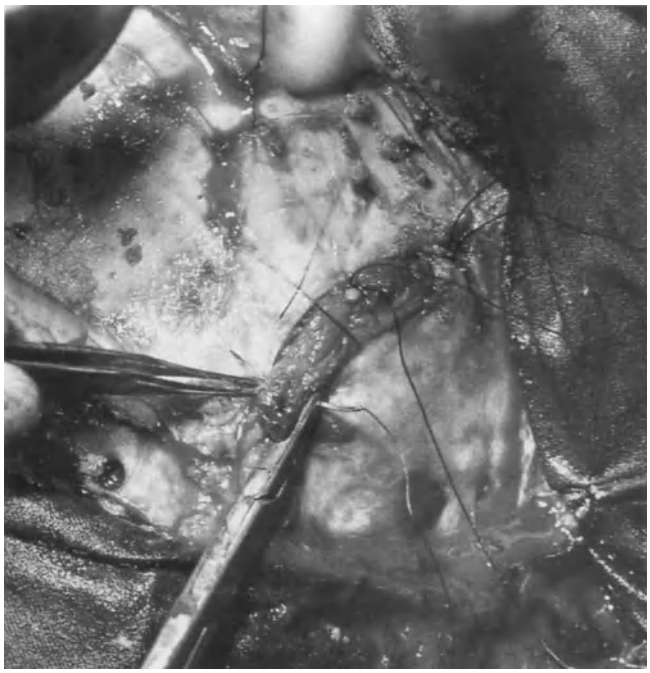


Figure 3.47. 3-0 sutures are being used to anchor the temporalis muscle flap to the periosteum of the frontal bone along the superior temporal line.

indicated! Indeed, now that we are in the computerized transmission tomography (CTT) era and about to enter the era of nuclear magnetic resonance, 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 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 burr holes during the opening should be placed into the burr 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 burr 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, is laying 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 burr 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 (Figures 3.50 and 3.51)
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 completely 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 and at

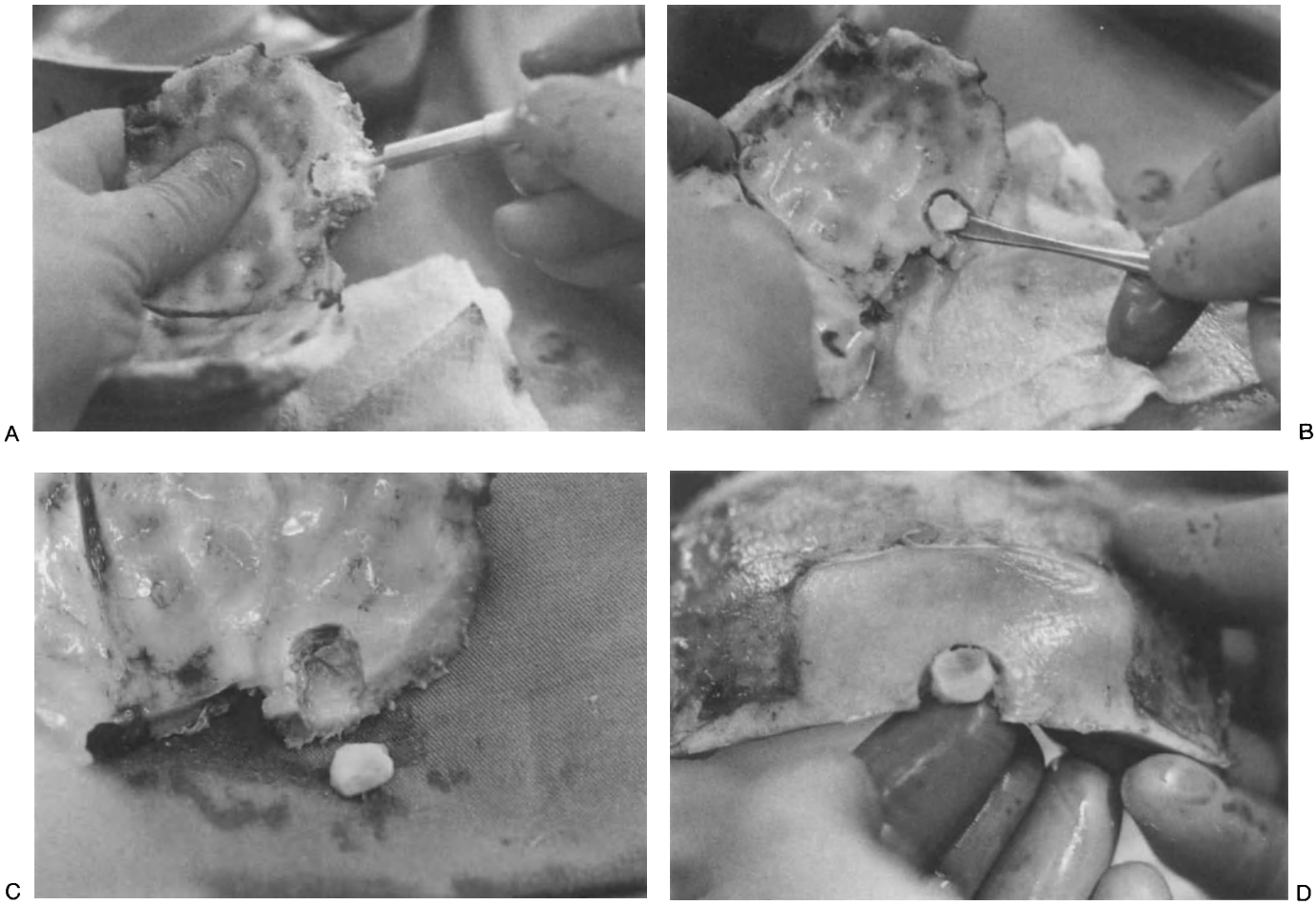


Figure 3.48. Preparation of bone plug. (A) The high-speed drill is being used to fashion a plug from the inner table of the bone flap. (B) After the cut has been made through the inner table and into the diploic layer, a periosteal elevator is used to pry it free, taking care not to crack it. (C) This is a photograph of the freed plug and bone flap from which it was taken. (D) The bone plug is being held in the burr hole defect it will be used to close—in this case the glabellar hole for a bifrontal flap.

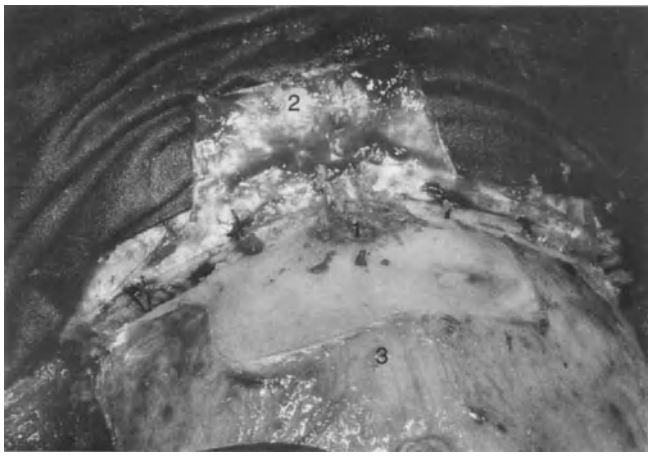
each level, first on one side then the other. The tying down of the laminar sutures one at a time from caudal to rostral unfolds the flap as an open accordion, and brings each of the laminae to rest at its appropiate anatomical level.

Since it is not always possible to perform an osteotomy in a medial lateral, dorsalventral, 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.

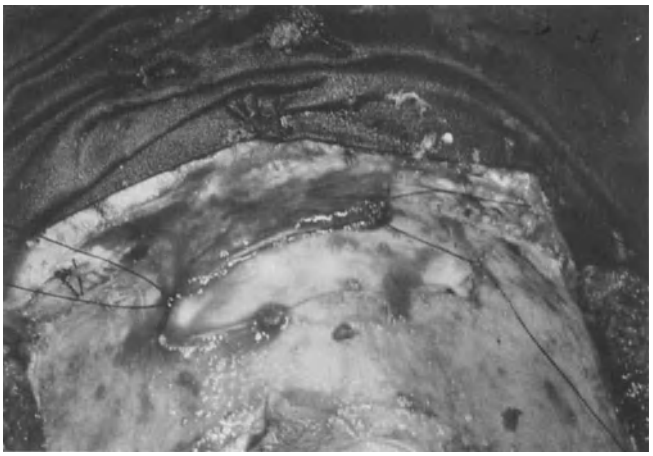
The paraspinous muscle masses are then allowed to fall into their normal position by removing the self-retaining retractors. They are sewn to the respective interspinous ligaments in two layers, deep and superficial, and the fascia over the paraspinous muscles is sewn to the supraspinous ligaments. This 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 "fourposter" 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.

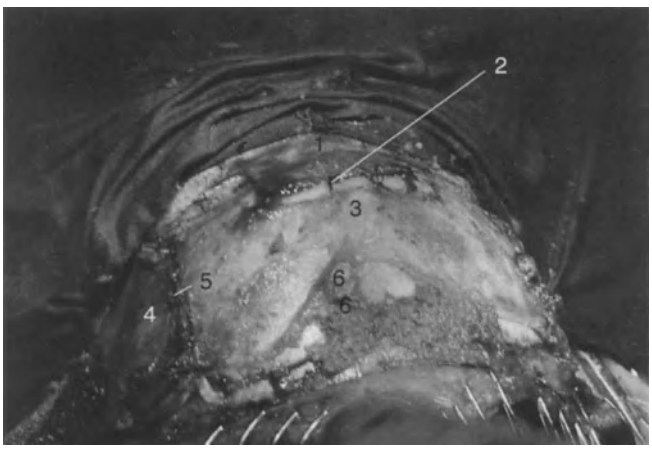


A

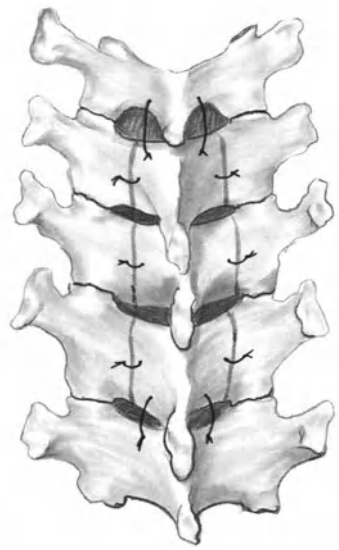


B

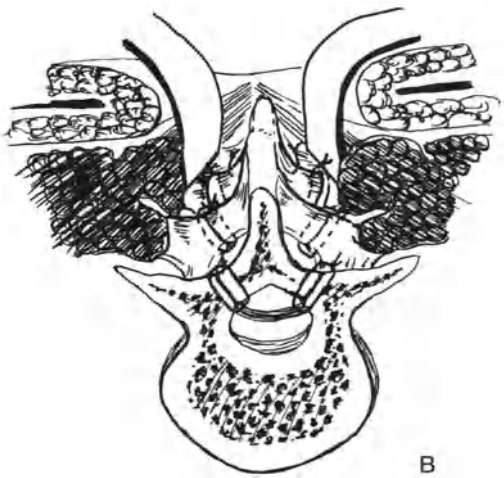
Figure 3.49. Bone chips taken from the burr holes during the opening should be placed into the burr hole sites after the bone flap has been securely anchored in position. Packing of the chips into the hole offers a base upon which the bone plug may rest. (A) A bifrontal free bone flap has been repositioned, and the bone plug (*1*) placed into the burr 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 plugged burr 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 burr hole openings.



C



A



B

Figure 3.50. (A) This is an 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.

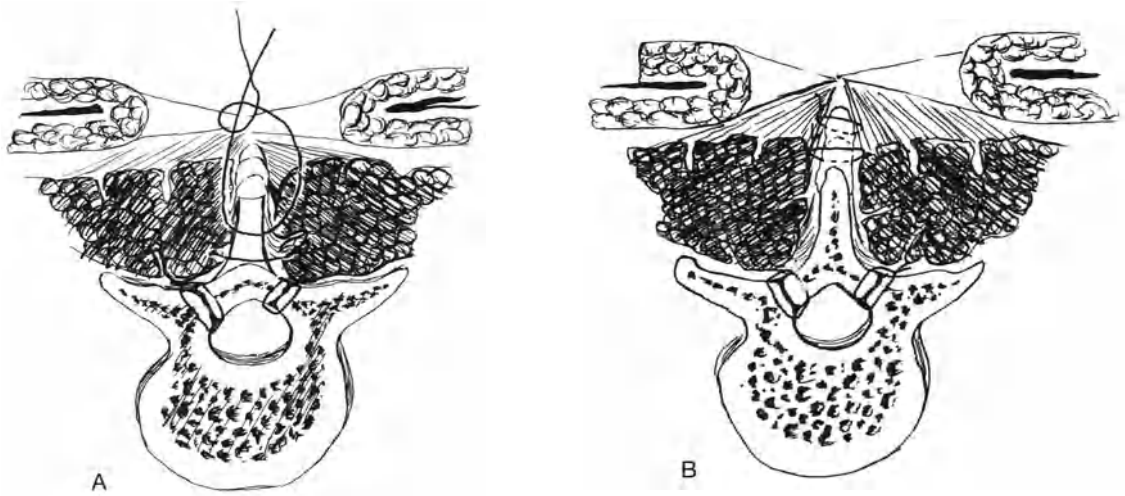


Figure 3.51. (A) The paraspinous muscle masses have been allowed to fall into their normal anatomical position, over the laminae and the spinous processes, and sutures placed so as to anchor these muscle masses, in two layers, to the interspinous ligament. (B) Lastly, just before closing the skin, the fascia of the paraspinal muscle masses is sewn to the ruffle of ligament left on the supraspinous process.

References

1. Krönlein RV: Zur Pathologie und operativen Behandlung der Dermoidcysten der Orbita. Beitr Z Klin Chir 4:149–63, 1888.
2. Jones BR: Surgical approaches to the orbit. Trans Ophthalmol Soc UK 90:269–81, 1970.
3. Jane JA: Supraorbital approach: Technical note. Neurosurgery 11:537–42, 1982.
4. Raimondi AJ, Gutierrez FA: A new surgical approach to the treatment of coronal synostosis. J Neurosurg 46:210–14, 1977.
5. Tachdjian MO, Matson, DD: Orthopaedic aspects of intraspinal tumors in infants and children. J Bone Joint Surg (Am) 47:223–48, 1965.
6. Cattell HS, Clark GL, Jr: Cervical kyphosis and instability following multiple laminectomies in children. J Bone Joint Surg (Am) 49:713–20, 1967.
7. Bette H, Englehardt H: Folgezustände von Laminektomien an der Halswirbelsäule. Z Orthop 85:564–73, 1955
8. Raimondi AJ, Gutierrez FA, DiRocco C: Laminotomy and total reconstruction of the posterior spinal arch for spinal canal surgery in childhood. J Neurosurg 45:555–60, 1976.
9. Yasuoka S, Peterson HA, Laws ER, MacCarly C: Pathogenesis and prophylaxis of postlaminectomy deformity of the spine after multiple level laminectomy: Difference between children and adults. Neurosurgery 9:145–51, 1981.
10. Barbera J, Gonzalez J, Esquerdo J, Brosela J, Barcia-Solario JL: Prophylaxis of the laminectomy membrane: An experimental study in dogs. J Neurosurg 49:419–24, 1978.
11. LaRocca H, Macnab I: The laminectomy membrane. Studies on its evolution, characteristics, effects and prophylaxis in dogs. J Bone Joint Surg 56B:545–50, 1974.
12. Raimondi AJ: Reflection of a laminar flap for exposure of spinal canal in children. Clin Neurosurg 25:504–11, 1978.
13. Raimondi AJ, Gutierrez FA: Reconstruction of the posterior vertebral arch laminotomy for intraspinal surgery: In Ransohoff J (ed): Modern Techniques in Surgery Neurosurgery. Mt. Kisco, NY, Futura, 1979, pp 10–11.

"Nothing may fail like success."

VARÉ
Italian Diplomat

Chapter 4

Suturotomy for Various Flaps in the Newborn and Infant

In the newborn and infant reflection of frontal, bifrontal, parietal, and other flaps with the use of the perforator and either Gigli saw or craniotome is unnecessary and dangerous, since the skull is composed entirely of membranous bone. The skull thickness measures less than 3 mm, and each bone rides freely over the 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 burr. (See Figures 4.1 to 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 fontanel and the 3 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 fontanel. 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 fontanel 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 fontanel to the posterior fontanel, taking care to cut along the junction of bone and suture, so as to avoid damage to the superior sagittal sinus.

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.

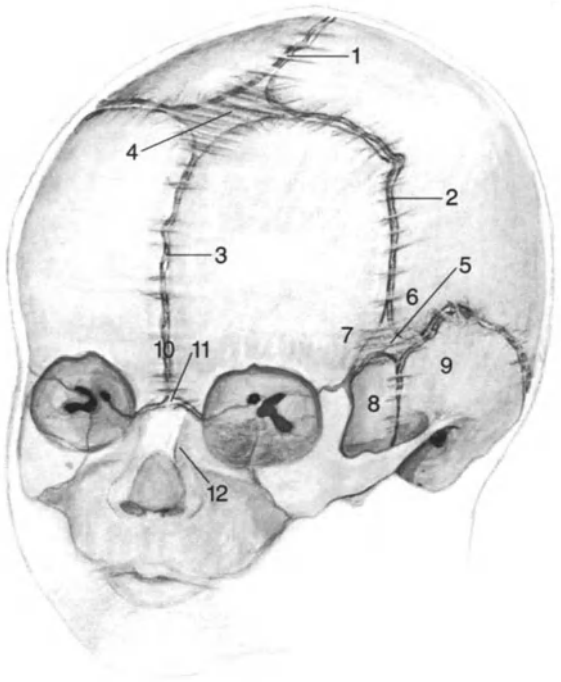


Figure 4.1. Anatomical drawing of the infant's neurocranium, illustrating the bones of the calvarium and the sutures continuous with it. The sagittal (*1*), coronal (*2*), and metopic (*3*) sutures are continuous with the anterior fontanel (*4*). The coronal suture extends anteroinferiorly in the coronal 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*).

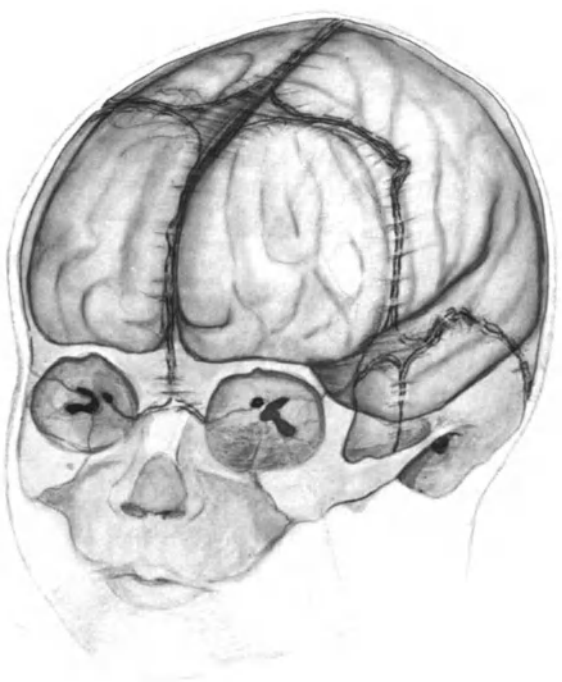
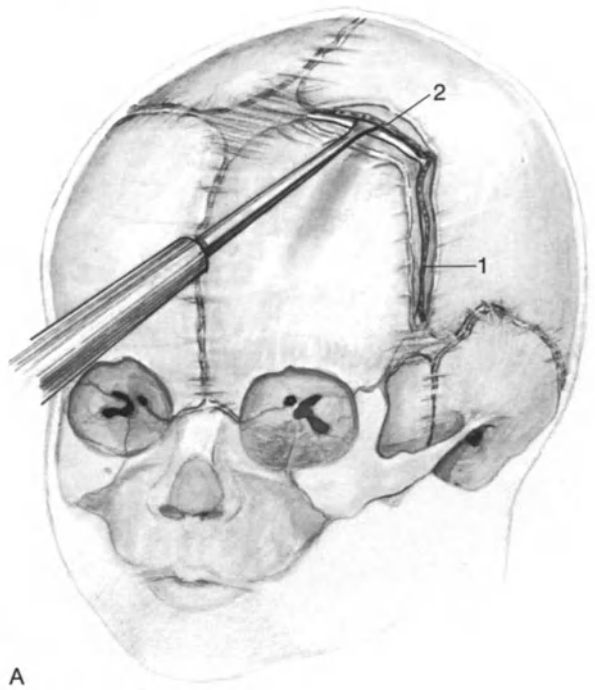
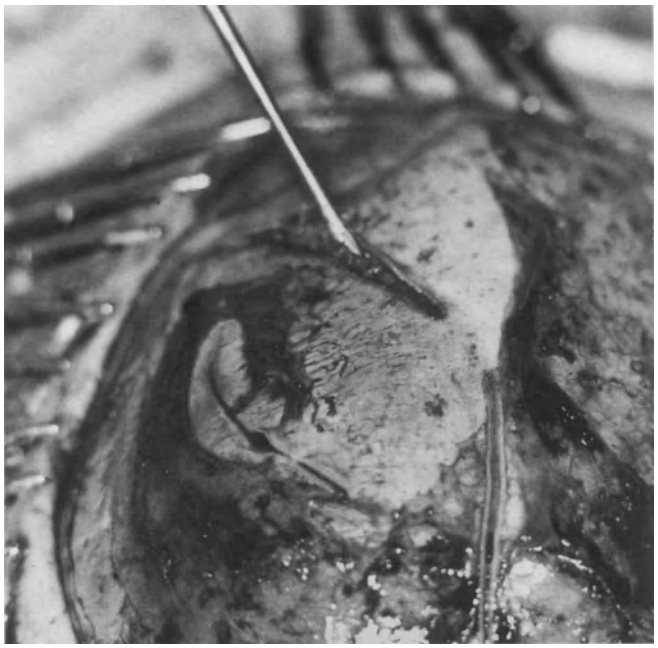


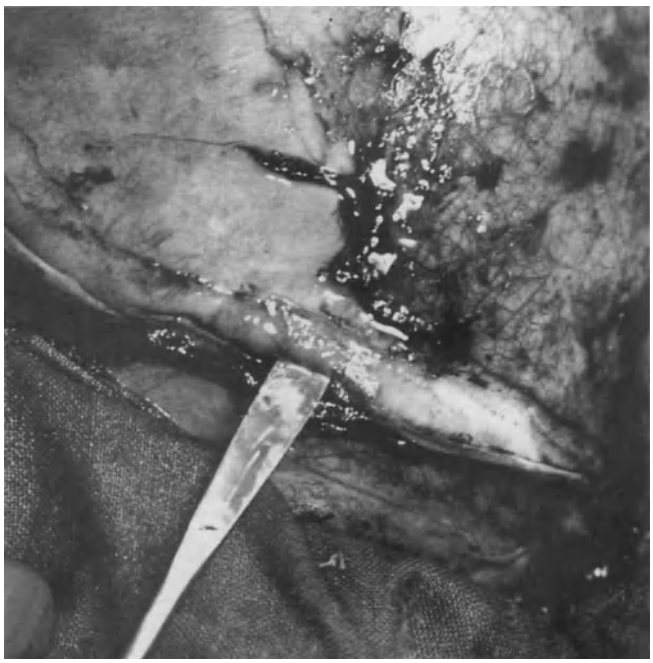
Figure 4.2. Membranous bony plates, seen in transparency, overlying the respective lobes (frontal, parietal, temporal) of the brain. Suturotomy around appropriate bone, therefore, suffices to expose the underlying lobe of the hemisphere.



A



B



C

Figure 4.3. (A) 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 herein illustrated. 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. (B) This is a photograph of the periosteum stripped from the bone at the suture line. (C) The periosteal elevator has been inserted into the epidural space, and is being used to separate the inner layer of the dura from the inner table of the skull.

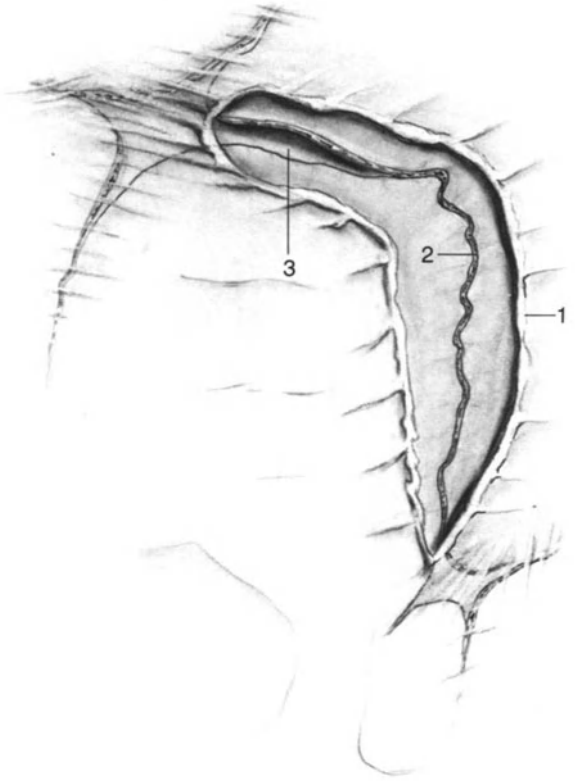


Figure 4.4. The periosteum (*1*) has been stripped from the coronal suture (*2*), and the outer layer of the dura (*3*) from the inner table of the skull.

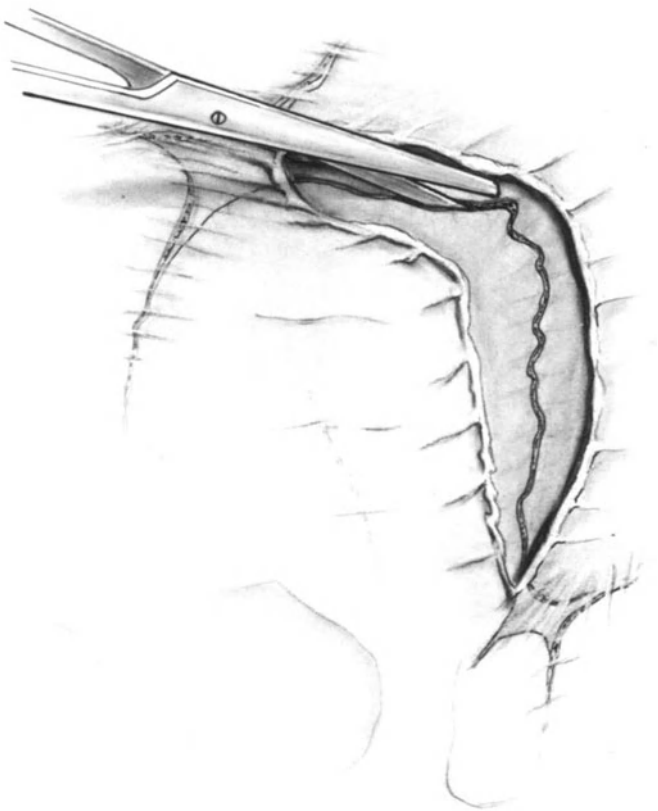
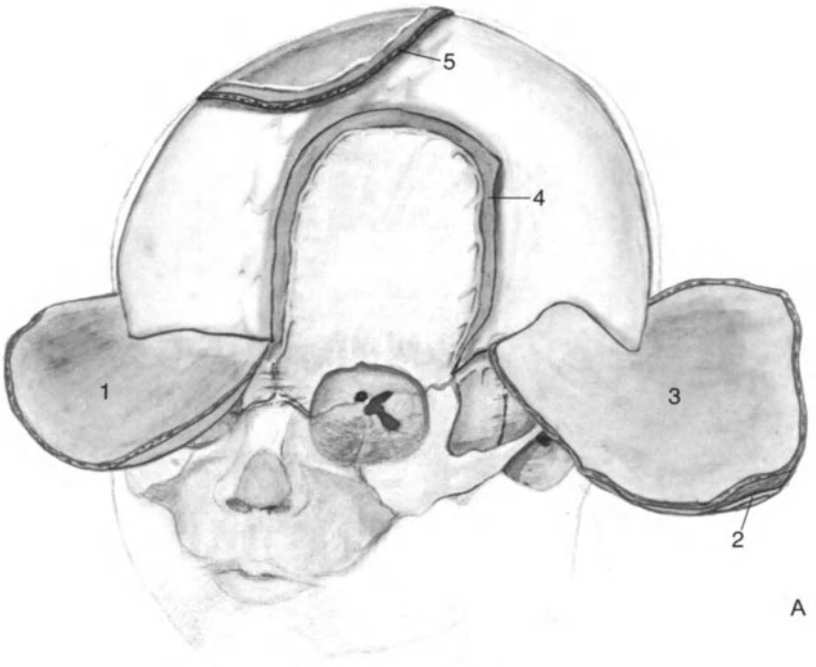


Figure 4.5. The scissors have been inserted so as to cut along the line of junction between the parietal bone and the coronal suture. The blade between the skull and the dura serves as a dissector.



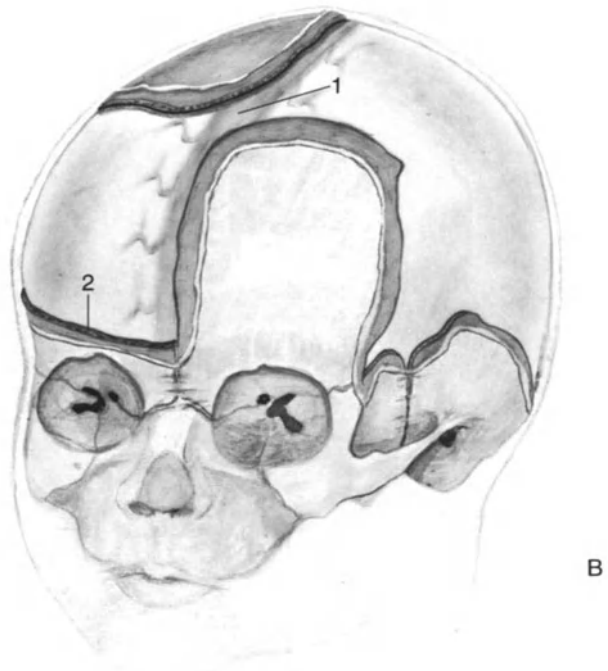


Figure 4.6. (A) The coronal and metopic suturotomies have been performed, reflecting the right frontal bone (*1*) inferiorly along the line extending from the glabella to the zygomatic process of the frontal bone. The periosteum (*2*) remains attached to the outer table of the skull. The left parietal 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.

"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

Chapter 5

Dural Flaps

General Comments (Figure 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 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. 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 fontanels, across a cranial suture, or at the superior sagittal or transverse sinues, since at these points outer layer of dura, cranial suture, and periosteum are one continuous structure in the newborn and infant.

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 is 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.

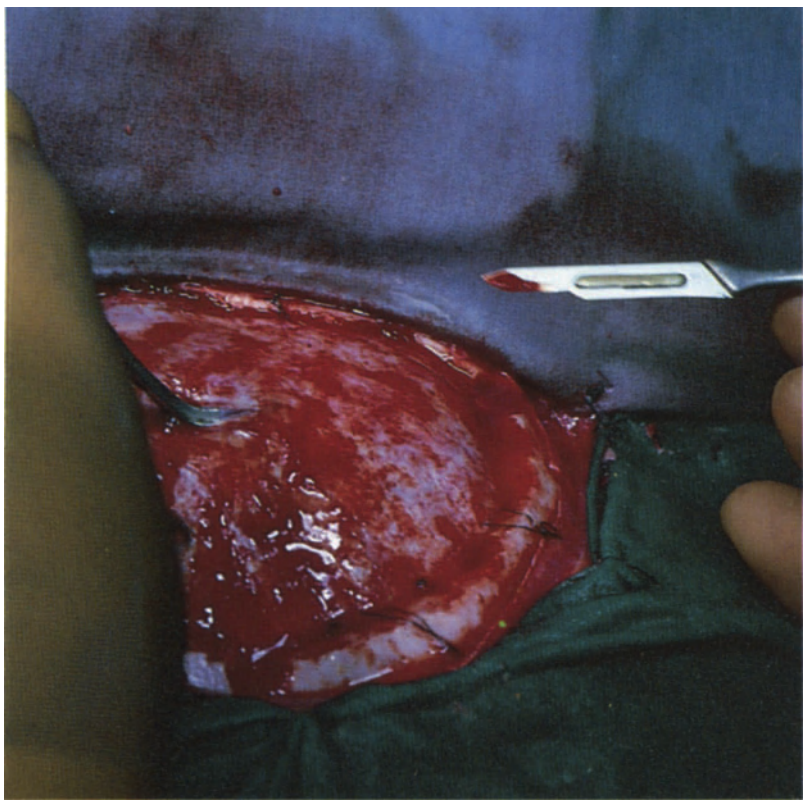
Dural Openings

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.

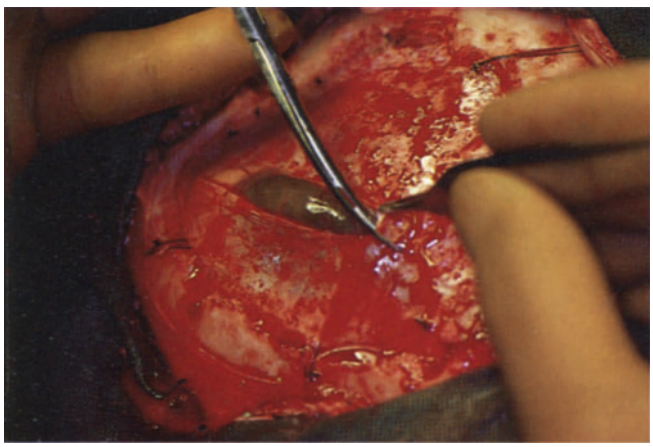
Frontal Dural openings

Medial Frontal Dural Opening (Figure 5.2)

Dural opening and reflection of the medial dural flap for a medial frontal craniotomy is somewhat different from 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



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 opened and the underlying arachnoid 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.

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.

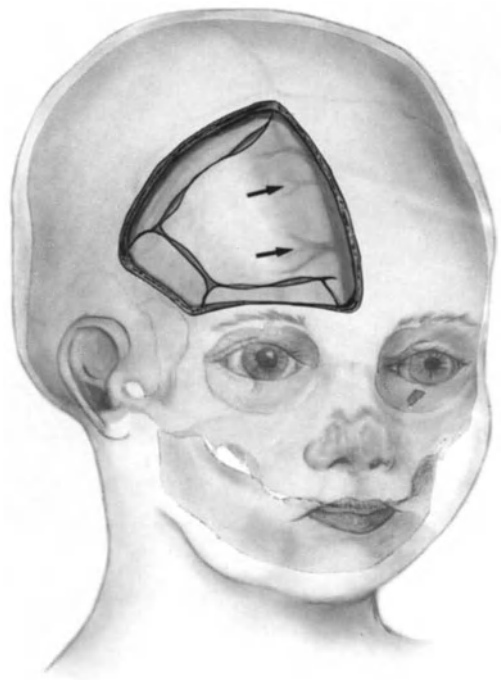
Lateral Frontal Dural Opening

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.

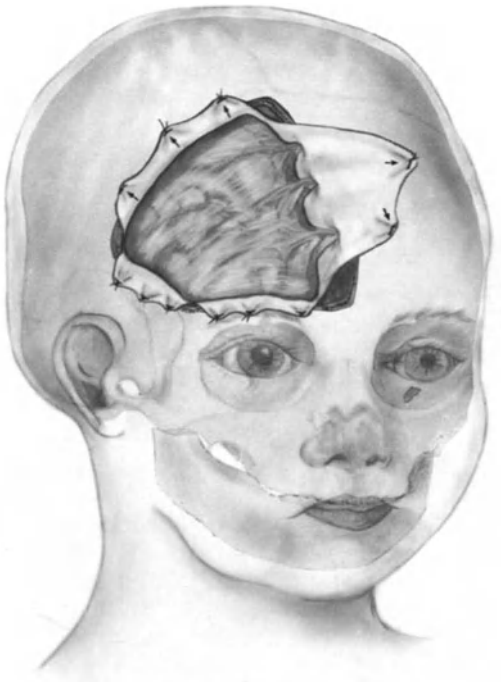
Bifrontal Dural Opening (Figure 5.4)

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 on lateral frontal craniotomy in that trapdoor openings are not used. Rather, a single hockey-stick incision is made on either side, extending from

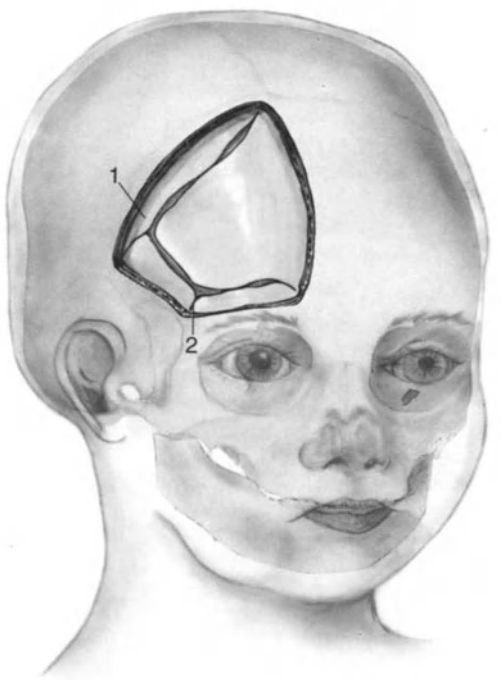


A

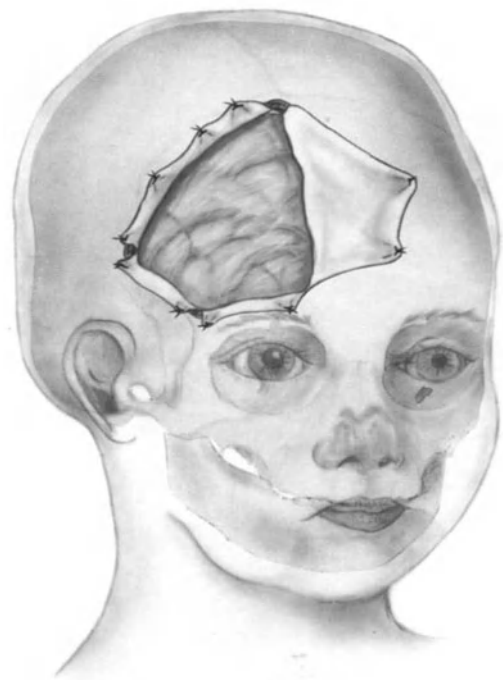


B

Figure 5.2. (A) Medial dural opening. The dura is opened (single trapdoor) 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 that the sutures are placed parallel (arrows) to the cut surfaces of the dura and periosteum. This avoids the suture tearing through these structures when the knot is tied.

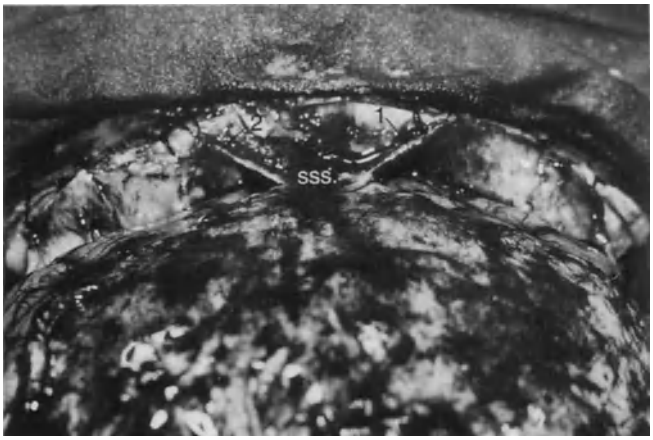


A



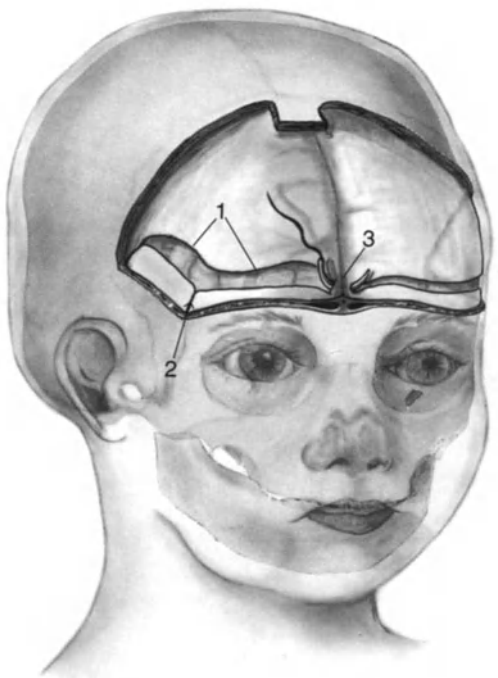
B

Figure 5.3. (A) 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) The single trapdoor and three peripheral edges of dura have been sewn to the periosteum, exposing the brain and securing (and stretching) the dura out of the field.



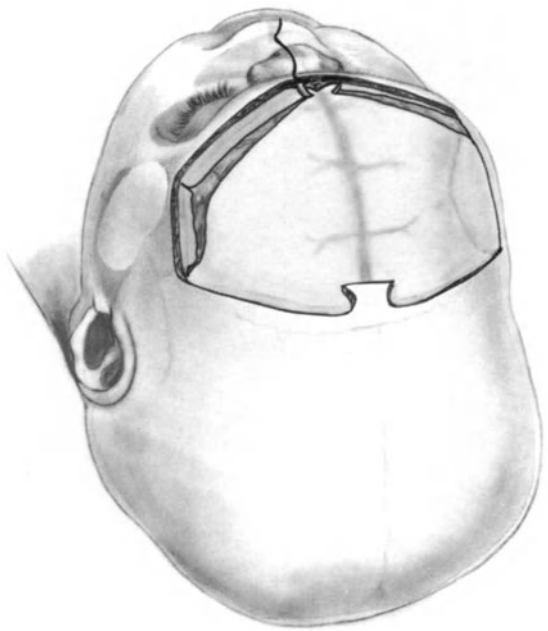
A

B

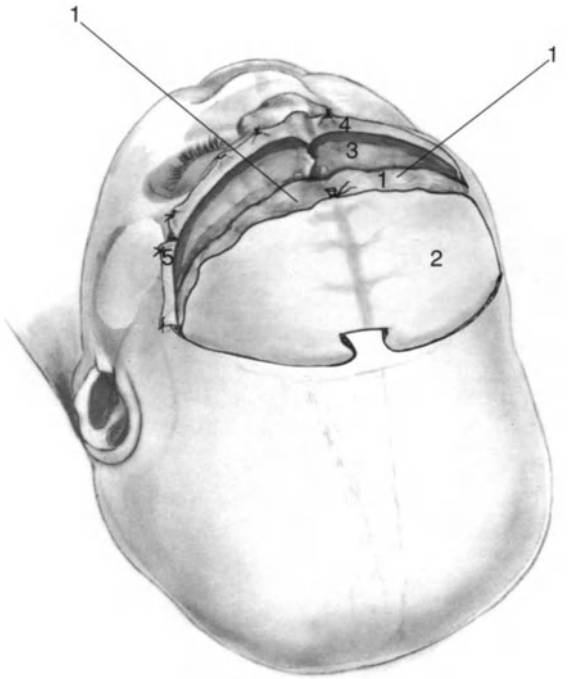


C

Figure 5.4. (A) The bilateral hockey-stick dural opening is photographed from the surgeon's line of vision. The dural opening is extended to the superior sagittal sinus (SSS), but not into it. Both right and left anterior dural falps (*1* and *2*, respectively) have been sewn over the supraorbital ridge to the periosteum, to prevent them from falling continuously into the operative field. (B) Clips have been placed across the SSS on either side. They are angulated in such a manner as to form a diamond. Their hilts are separated from one another by at least 2 mm, permitting one to cut across the dura of the SSS between them. Clips are unreliable in that they may not be successfully extended across the SSS. Also, they tend to slip from the sinus during the operative procedure. Surgicel has been placed over the SSS. (C) 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. (D) 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. (E) 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 peristeum of the orbital rim (*4*) and to that over temporosphenoidal bones (*5*) are thereby prevented from falling continuously into the operative field. (F) Supine position. The SSS has been transected between the two ligatures, and the falx cerebri has been cut from the crista galli. The dural flaps have been sewn to the periosteum, and a telfa placed over the right frontal lobe. Note that the dura (shown on the left) falls away from the orbital roof with the left frontal lobe after the falx cerebri is cut. This is extremely important, since it assures the surgeon that there will be no stretching or tearing of frontal bridging veins, minimizing risk of frontal lobe edema or infarction. The orbital roofs have come into view with the cribriform plate. (G) Right medial frontal dural opening. One can compare the difference in exposure (with regard both to extent of exposure and surgery) between a unilateral and bilateral frontal flap.



D



E

F

G

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 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 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 Openings (Figures 5.5 to 5.8)
The parietal dural openings may be superior parietal, parietotemporal (inferior parietal), biparietal, and parietofrontal.

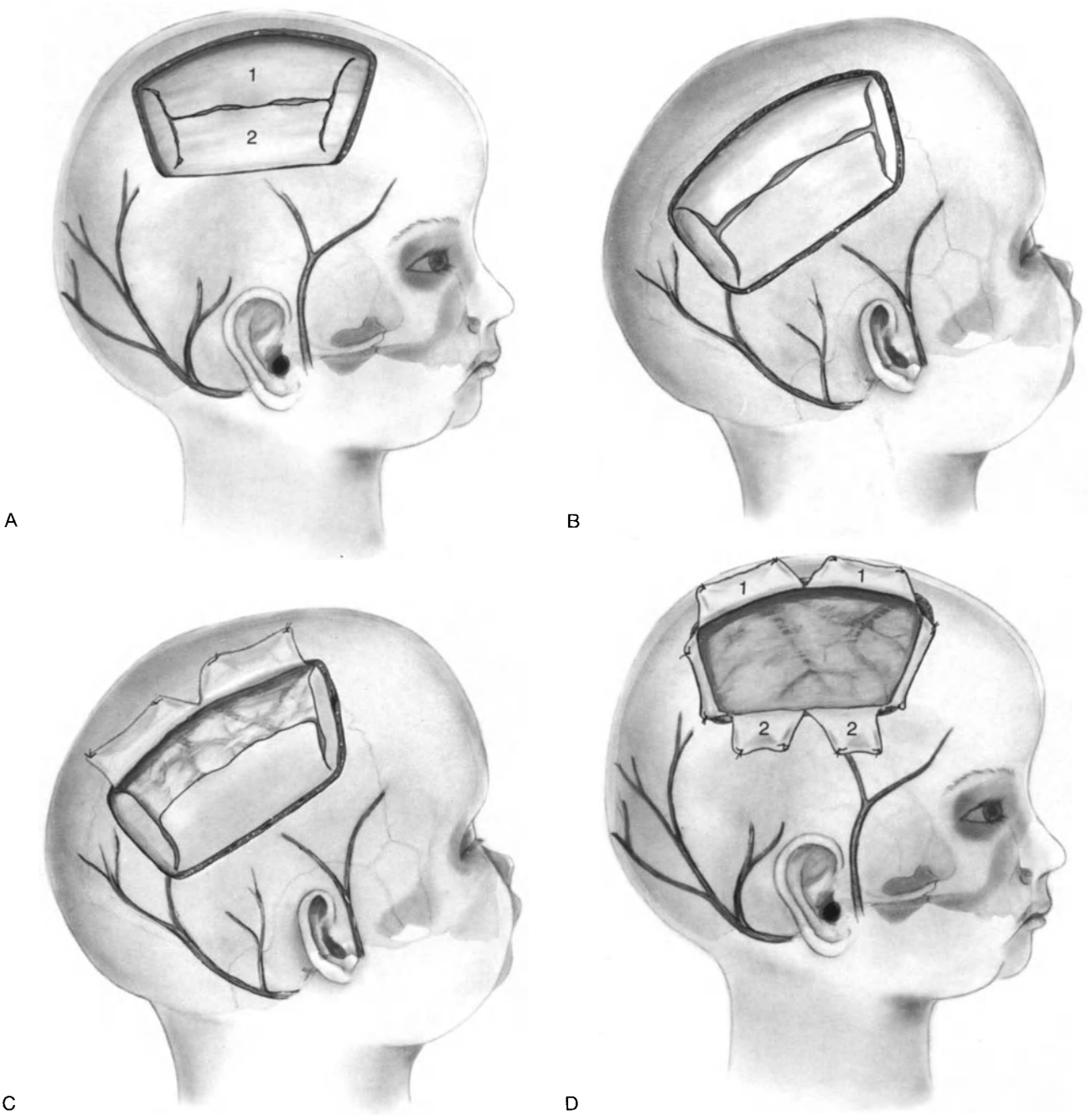


Figure 5.5. (A) Superior quadrilateral parietal flap as seen from straight lateral perspective, in order to visualize the relationship between the posterior extremity of the flap and the parietal eminence. The dura has been incised but not reflected. Note that only the superior portion of the dura (*1*) may be reflected over the superior sagittal sinus without opening the inferior trap door (*2*), or the inferior trap door may be opened for greater access to the parietal lobe. (B) Posterooblique view of the dural opening in a quadrilateral bone flap to illustrate access to the parietal convexity. (C) Posterooblique 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 4 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.

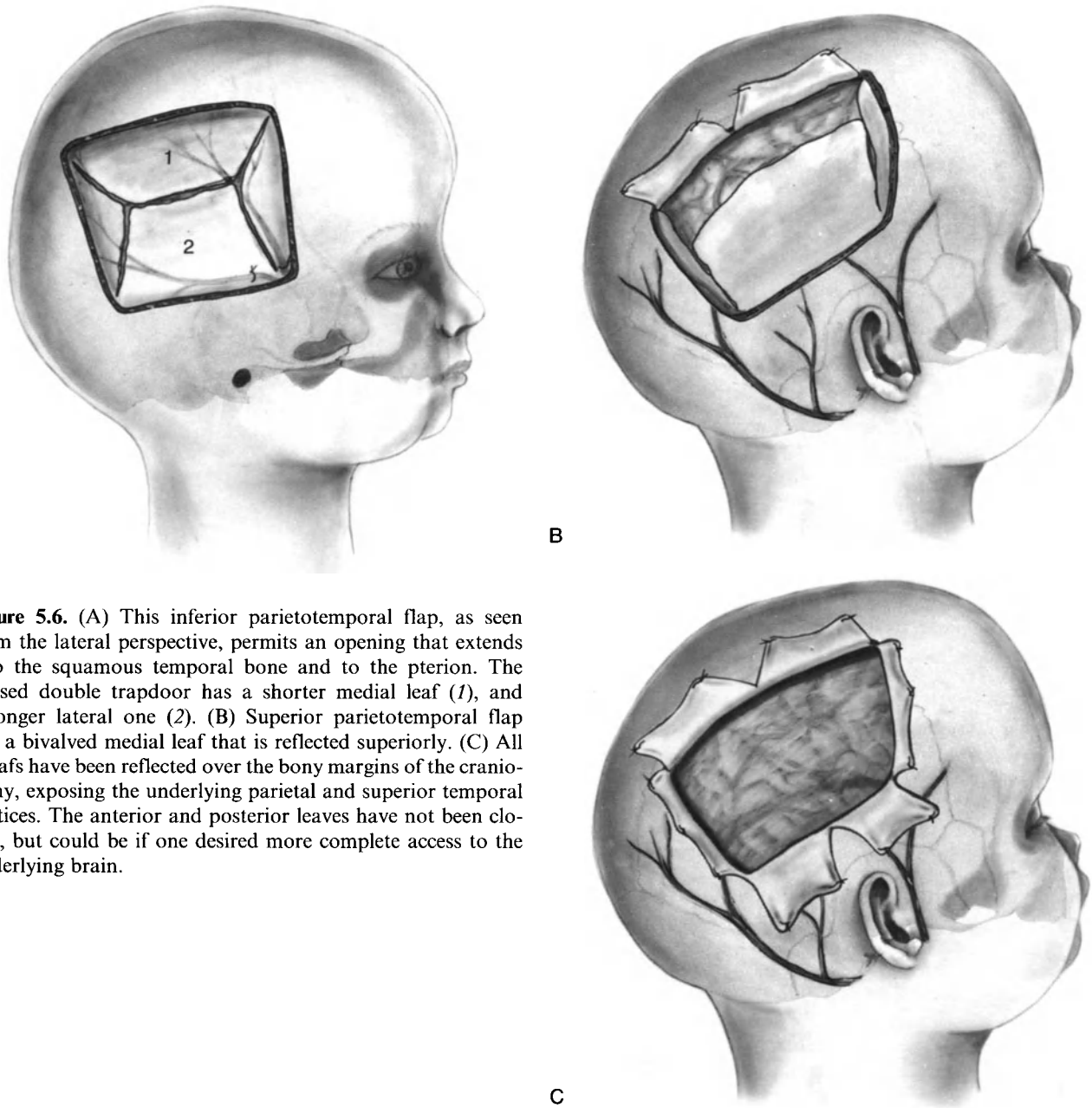


Figure 5.6. (A) This inferior parietotemporal flap, as seen from the lateral perspective, permits an opening that extends into the squamous temporal bone and to the pterion. The incised double trapdoor has a shorter medial leaf (*1*), and a longer lateral one (*2*). (B) Superior parietotemporal flap has a bivalved medial leaf that is reflected superiorly. (C) All 4 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.

Superior Parietal Dural Opening

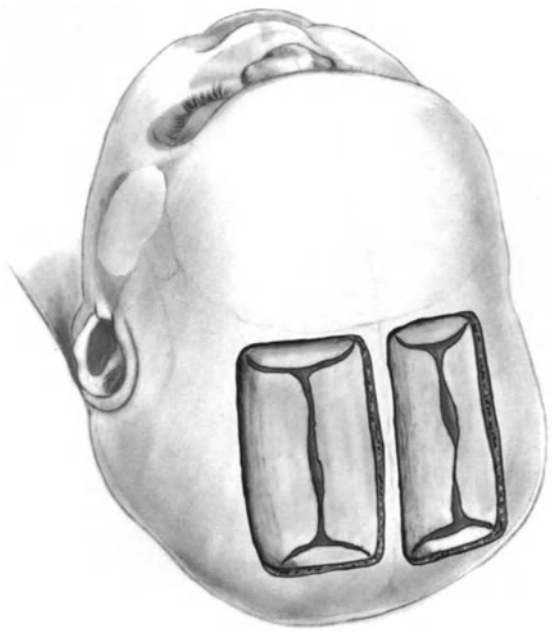
Superior parietal flaps are reflected so as to offer access to the hemispheral convexity, superior sagittal sinus, falx, and corpus callosum. Consequently, an asymmetrical, double trapdoor dural opening is fashioned. This provides adequate exposure of the parietal convexity and sagittal structures, and 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

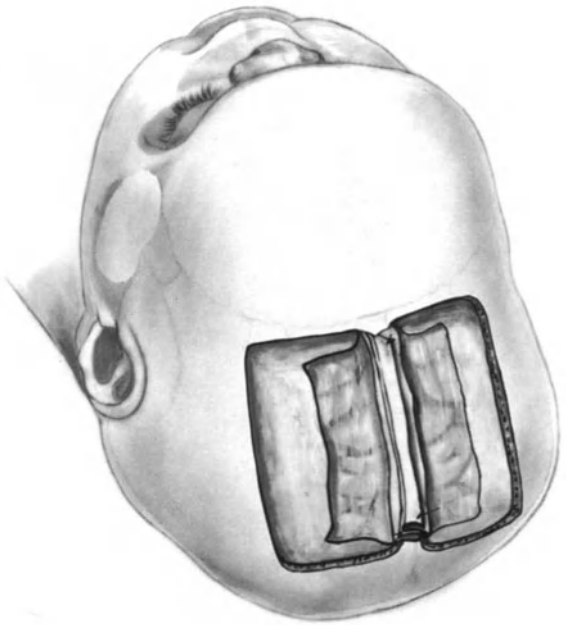
The parietotemporal dural opening is also effected with a double trapdoor, again to expose only the desired area and to permit the option of a more extensive cerebral exposure.

Biparietal Dural Opening

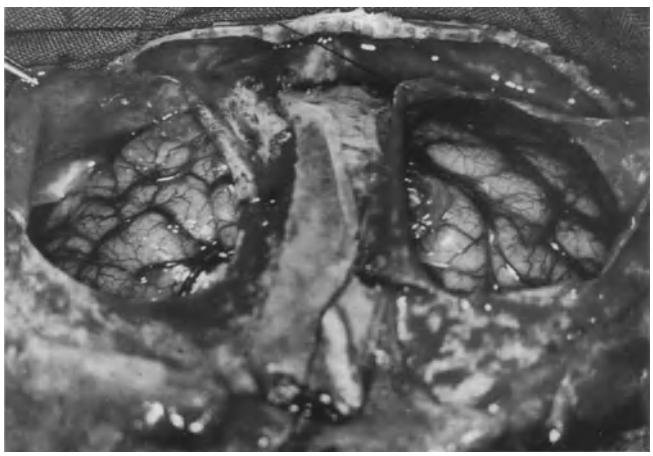
The bilateral dural openings may be either parietal or parietofrontal. They permit access to the convexities of both parietal (or parietofrontal) lobes, to both sides of the falx cerebri, and to the splenium of the corpus callosum. The parietofrontal flap permits access to the genu, as well as the body 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 these choroid plexus papillomas extending



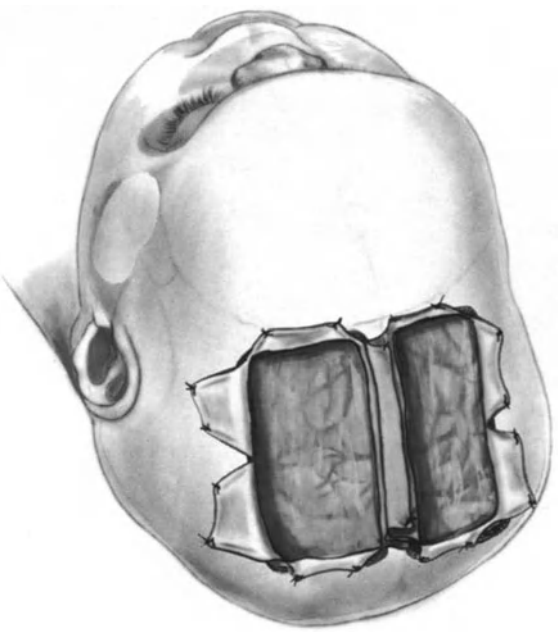
A



B



C



D

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) This photograph of a biparietal dural opening was taken during the performance of a lowering of the superior sagittal sinus (SSS) for chronic subdural effusion. The right medial dural flap has been sewn over the SSS to the dura on the left, which will subsequently be sewn medialward onto the right dural flap. One may appreciate the extensive exposure of the underlying brain this type of dural opening provides. The dural opening has not been completed, so one notes tenting of the superior and lateral flap segments. (D) 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 and sewing both to suspend and protect the SSS sinus when it has.

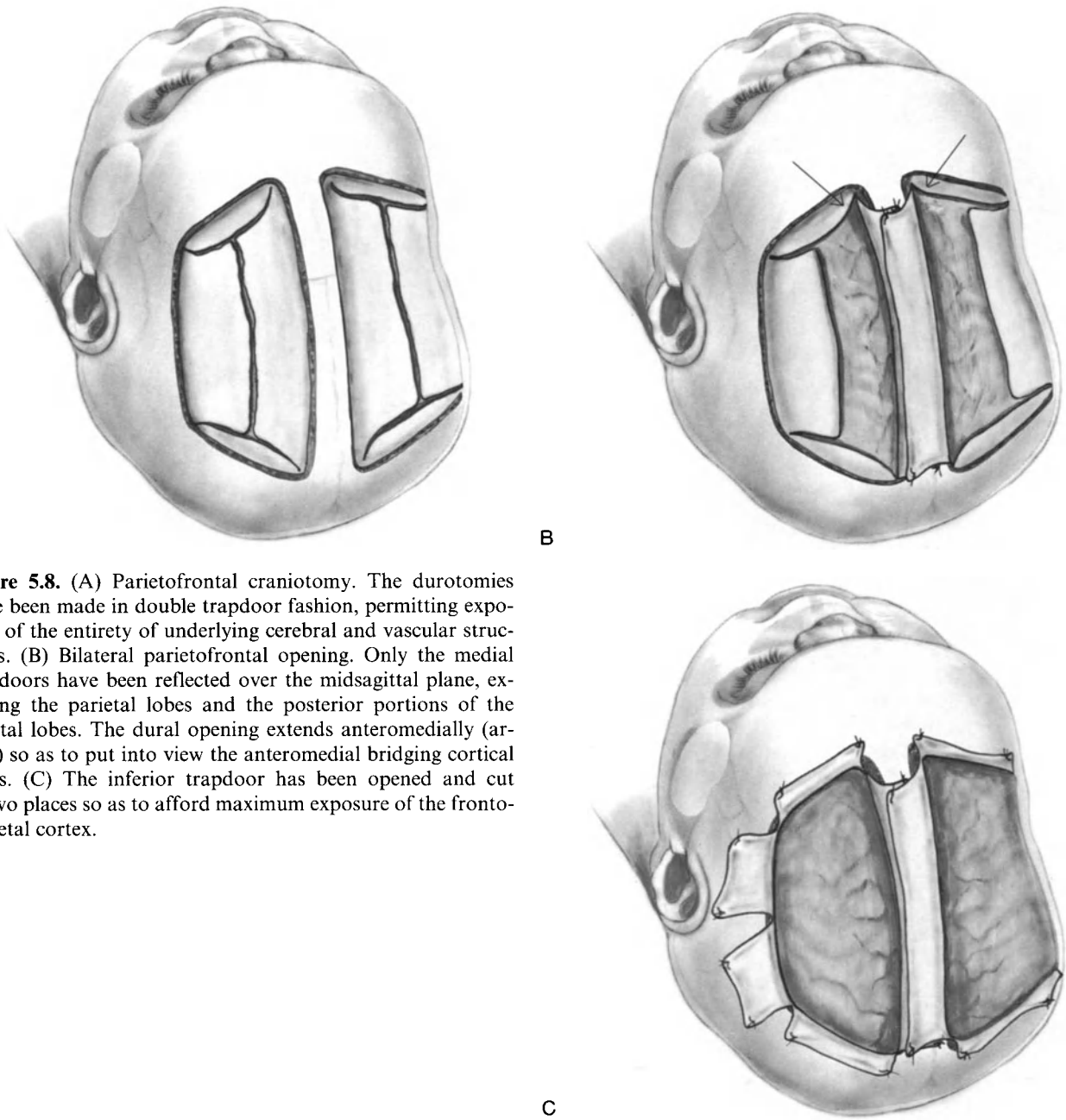


Figure 5.8. (A) Parietofrontal craniotomy. The durotomies have been made in double trapdoor fashion, permitting exposure of the entirety of underlying cerebral and vascular structures. (B) Bilateral parietofrontal opening. Only the medial trapdoors have been reflected over the midsagittal plane, exposing 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.

from the glomus to, and through, the foramen of Monro.

Temporal Dural Openings (Figure 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 supra-

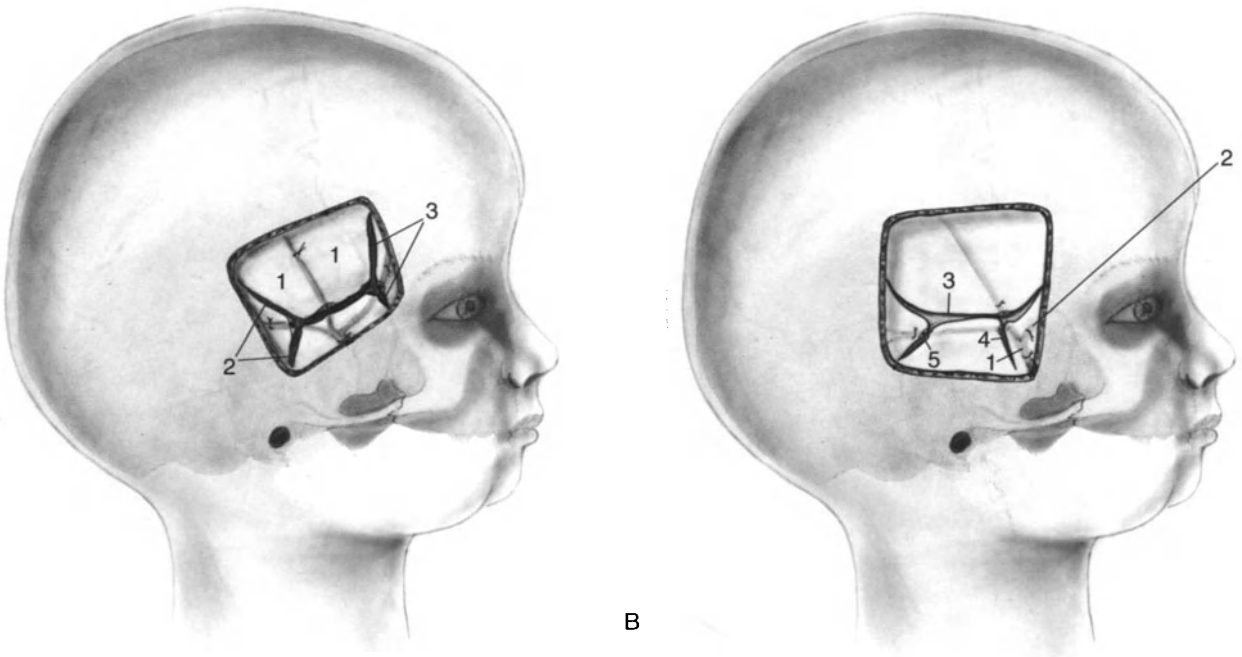


Figure 5.9. (A) The anterior temporal dural incision and reflections are illustrated. The middle meningeal artery is ligated by passing a 4-0 suture through both layers of the dura and then tying them down securely. This prevents bleeding from the dural edges. The dura is then opened by making a semilunar 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 tempo-

marginal 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.

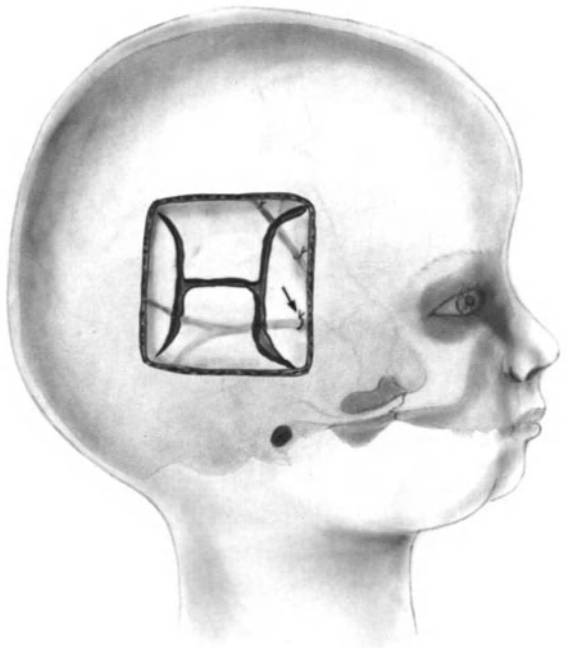
One may choose to cut the dura so as to avoid the middle meningeal artery, but I discourage this because it serves no purpose, permits continuous, nagging bleeding from the dural edges, and increases greatly the risk of an intraoperative epidural hematoma (which may result from tearing the middle meningeal artery as the dura falls from the inner table of the skull).

Occipital Dural Openings (Figures 5.10 and 5.11)

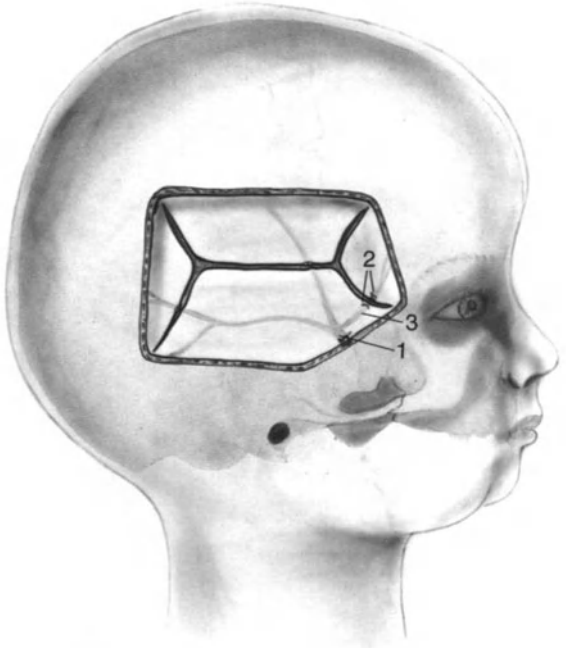
The medial and lateral occipital dural openings permit one, respectively, to work along the junction of the falx and the tentorium to reach the culmen monticuli and the pineal area, and to gain full access to the occipital pole.

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, then cut inferomedially to approximately 2 mm lateral to the torcular Herophili. This allows one to reflect the medial trapdoor over the sagittal plane, and the inferior dural skirt 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.

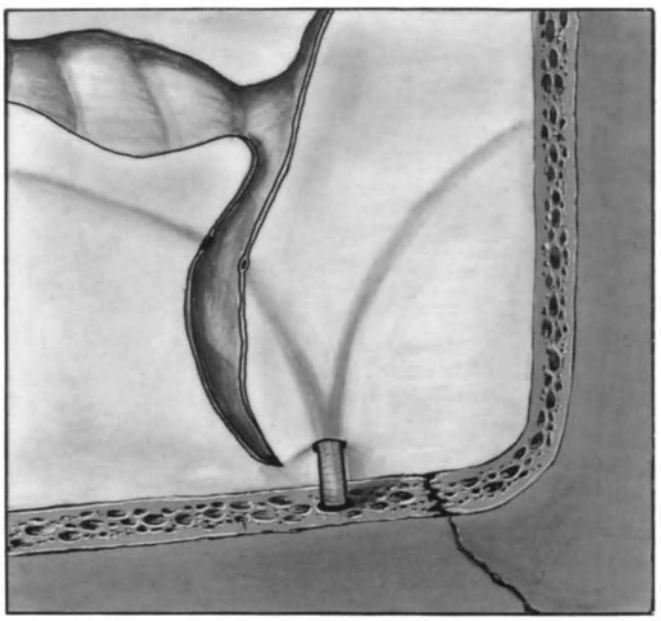


C



D

ral 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) 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.



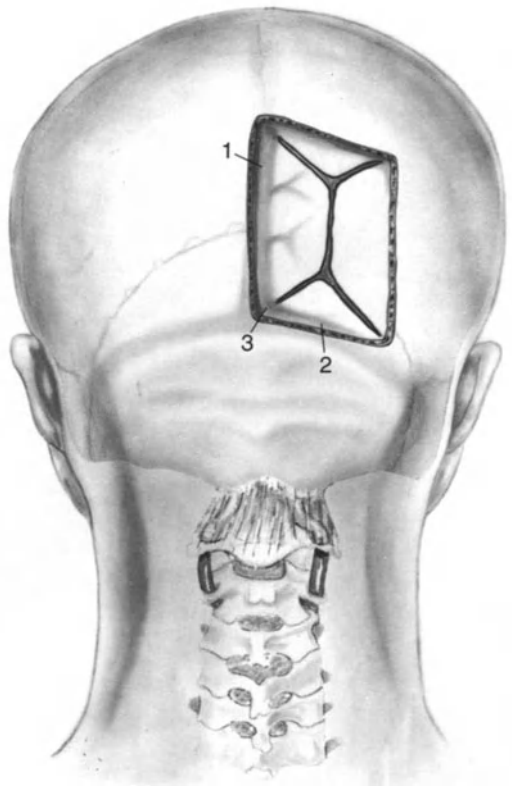
E

Lateral Occipital Dural Opening

The lateral occipital flap is designed exclusively 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.

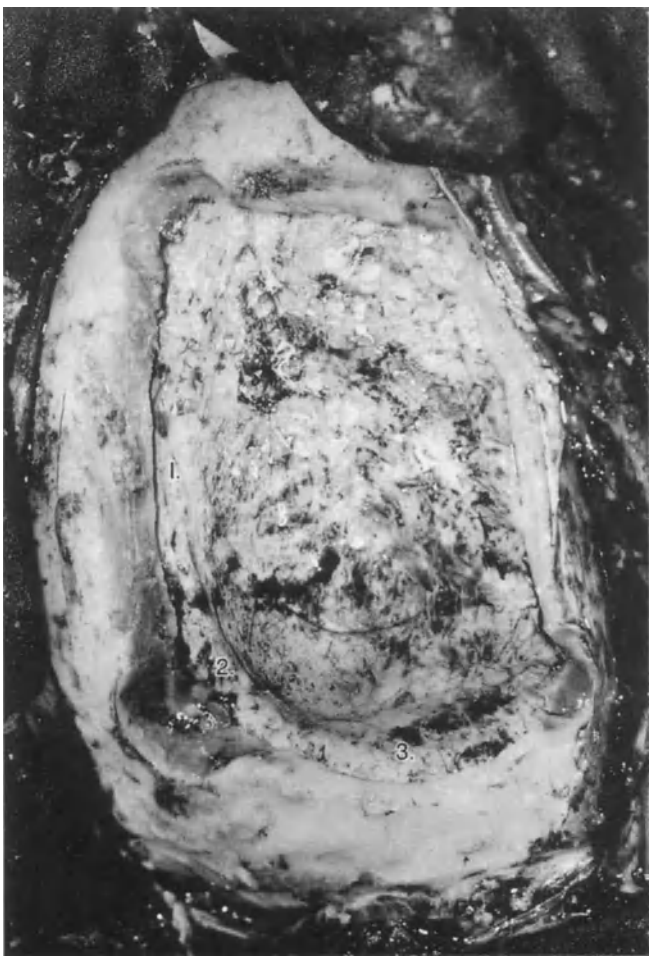
Posterior Fossa: Suboccipital Dural Openings

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 sitting 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 entire 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.

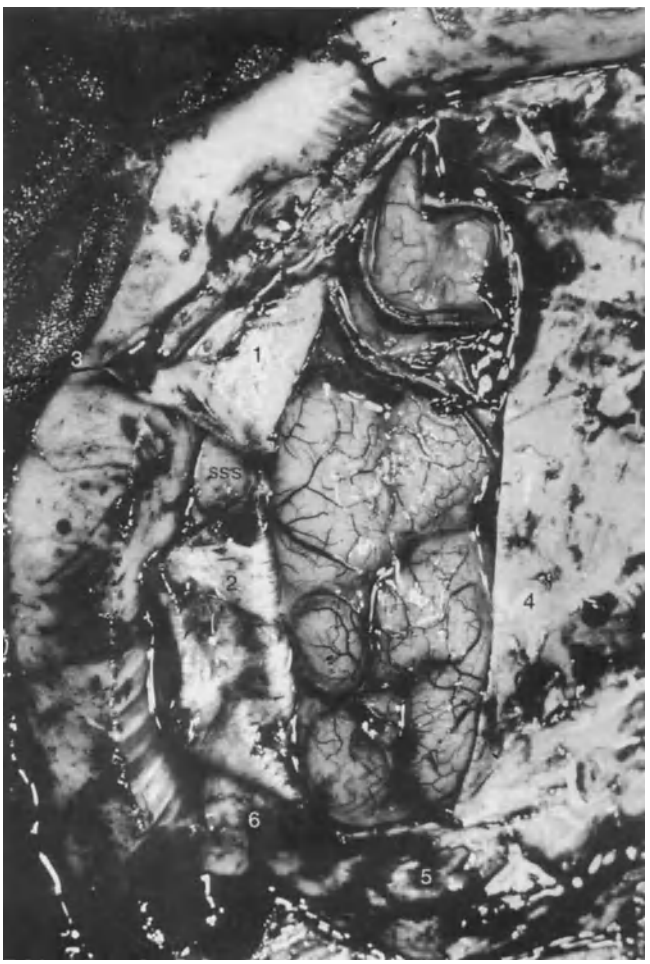


A

Figure 5.10. (A) The dural opening is marked off, permitting reflection of a double trapdoor within the medial occipital craniotomy osteotomy lines. Note that the superior sagittal (*1*) and transverse (*2*) sinuses are within the craniotomy, as is the torcular Herophili (*3*). (B) Dural flaps reflected over the occipital cortex. This photograph of a medial occipital craniotomy illustrates the underlying dura, as well as the exposed superior sagittal sinus (*1*), the torcular Herophili (*2*), and the transverse sinus (*3*). (C) The medial trapdoor of the dural opening has been made, and cleft into superior (*1*) and inferior (*2*) segments, which in turn are being reflected over the superior sagittal sinus at its posterior third. One anchoring suture (*3*) has been placed. Three more will be placed before proceeding to retract the occipital lobe laterally *under* the dura (*4*). Inferiorly, the dura has been opened to the transverse sinus (*5*) and the torcular Herophili has been exposed (*6*).



B



C

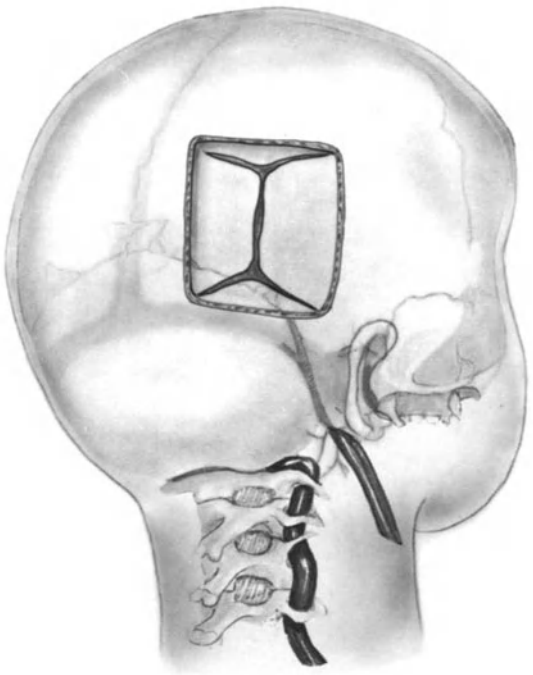


Figure 5.11. The lateral occipital craniotomy exposes dura but neither sinus nor, in most instances, major bridging cortical veins. One must, however, be cautious for there is occasionally a draining vein that goes from the tentorial surface of the occipital lobe into either the tentorium or the transverse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

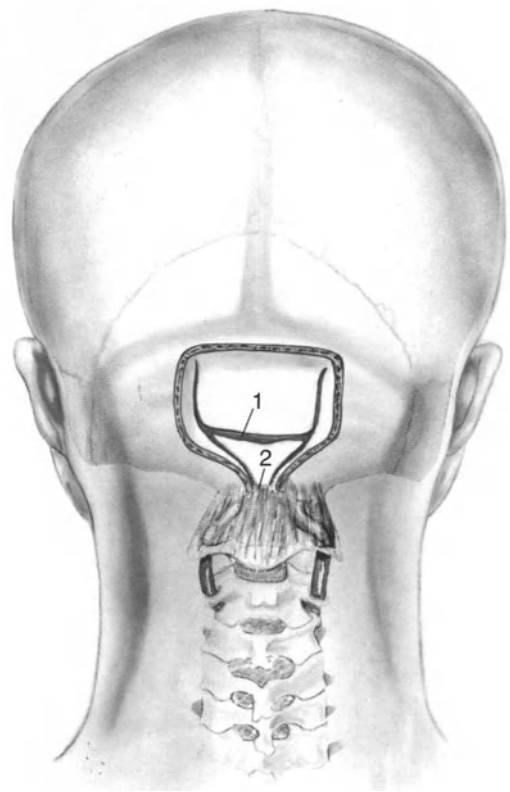
Medial (Midline) Suboccipital Dural Opening (Figures 5.12 to 5.14)

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.

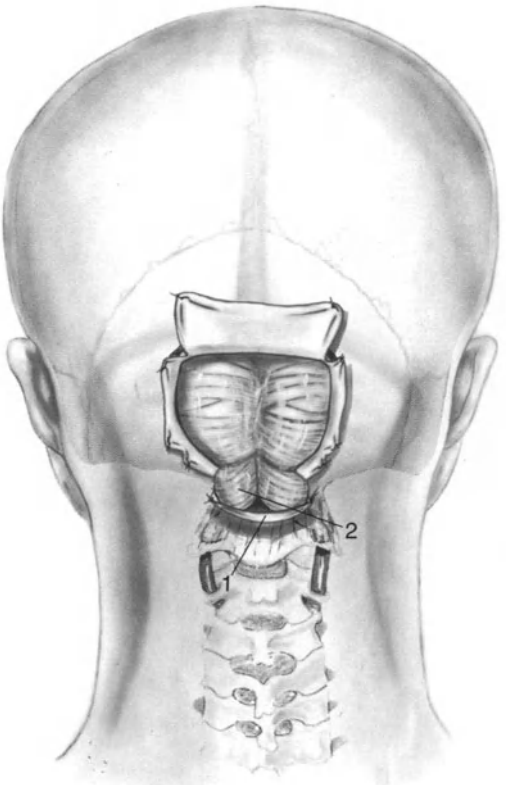
Inferior Cerebellar Triangle Dural Opening

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 atlantooccipital membrane. Thus, for inferior trangle 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.

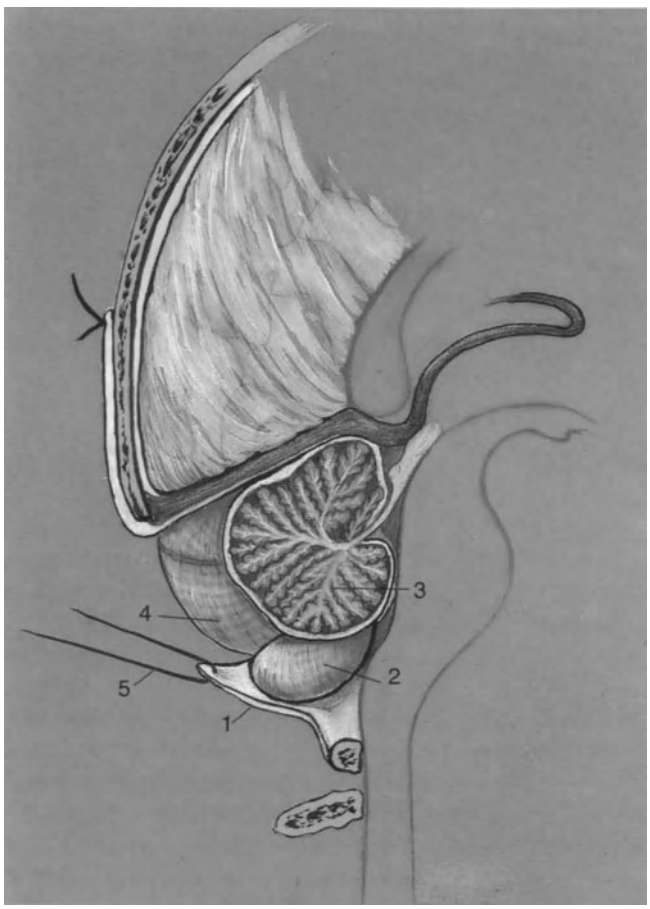
The dura is cut inferomedially from the point of opening (which is best made at approximately the junction of the superior and inferior thirds of the dura ex posed by the suboccipital craniotomy). A cut across the midline from right to left, connecting the two inferomedially coursing durotomy incisions, 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 and cerebellar sinuses. 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 it. Also, and of more immediate danger, opening dural sinuses subjects the child to air embolism. Immediate, or incomplete, closure (often the case when clips are used) of



A



B



C

Figure 5.12. (A) Dural flaps for access to the inferior triangle. The lines of the two U-shaped dural flaps are indicated. They permit 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 hemispheres (*4*). The ligatures suspending it (*5*) are sewn to the erector capiti muscle fascia.

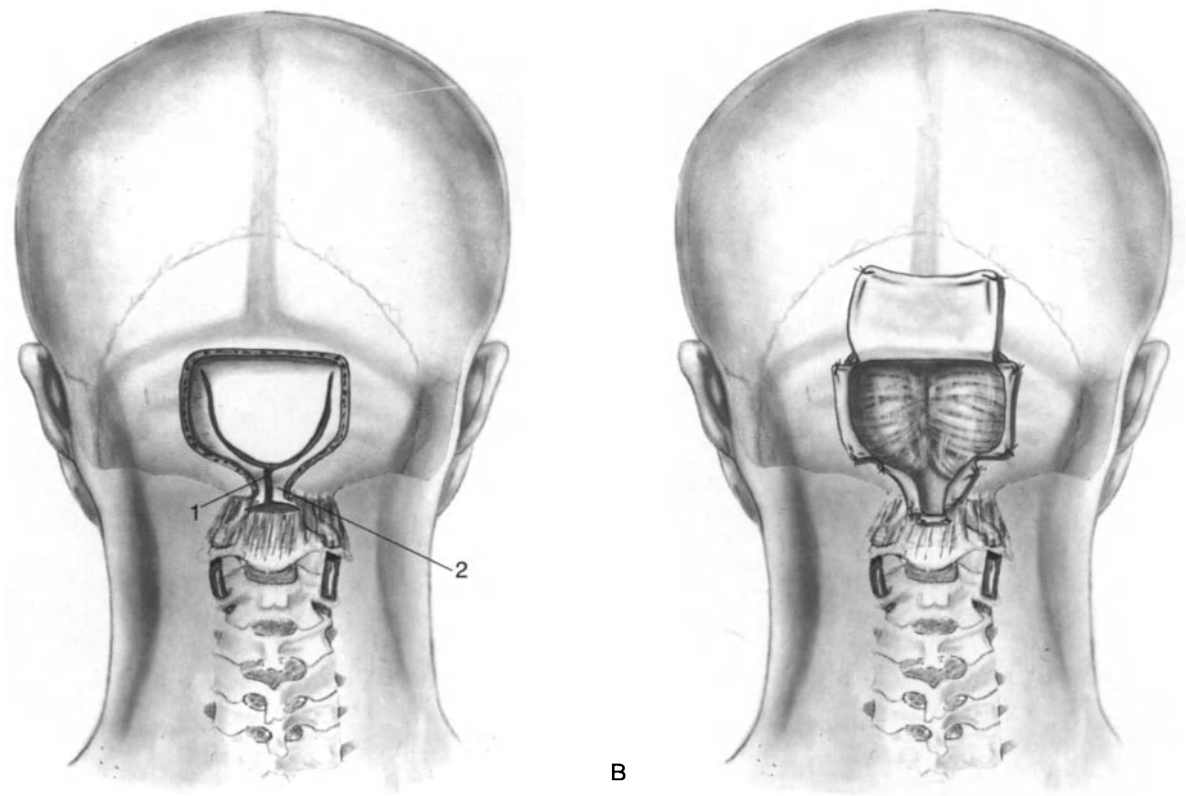


Figure 5.13. (A) This is the recommended durotomy for lesions within, or extending into, the foramen magnum. There is a vertical durotomy (*1*) across the level of the foramen magnum (*2*). (B) The dura is reflected superiorly over the craniotomy edges and inferiorly over the arch of C-1. This exposes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

these dural sinuses may present continued risk of air embolism throughout the operative procedure.

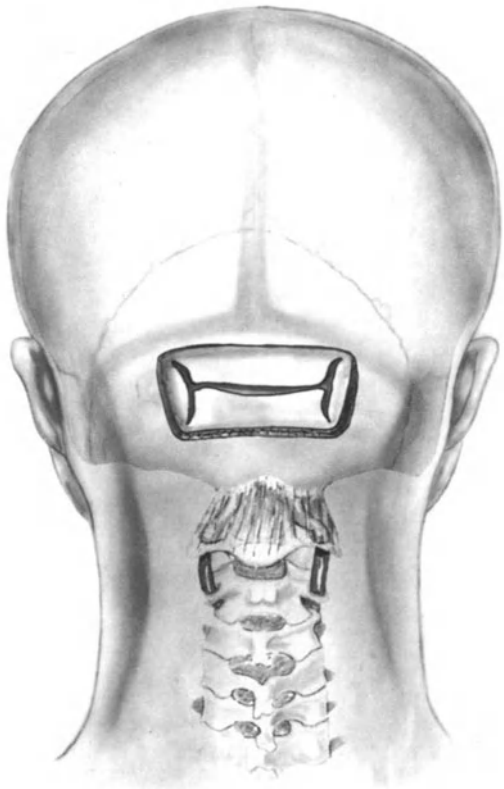
Opening of the dura over *the craniovertebral junction for inferior triangle lesions at the foramen magnum,* entails fashioning a U-shaped dural flap, convexity pointing inferiorly, beginning at the superolateral border on each side of the craniotomy. It should be extended inferomedially to approximately 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 does extend 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 arch of C-1 may then be anchored back into anatomical position at the end of the procedure.

Superior Cerebellar Triangle Dural Opening

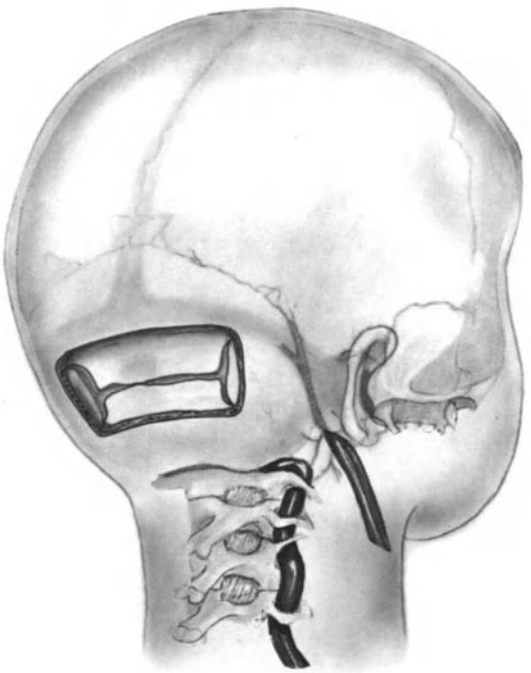
Superior cerebellar triangle exposure and access to the pineal region, through a quadrilateral craniotomy flap, is attained by a double trapdoor durotomy. This permits one to reflect the upper trapdoor superiorly, if the culmen monticuli and pineal region are the target area(s), and the lower trapdoor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

Lateral Suboccipital Dural Opening (Figure 5.15)

For access to the lateral cerebellar hemisphere or the pontocerebellar angle, a double trapdoor durotomy with the superior trap door 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.



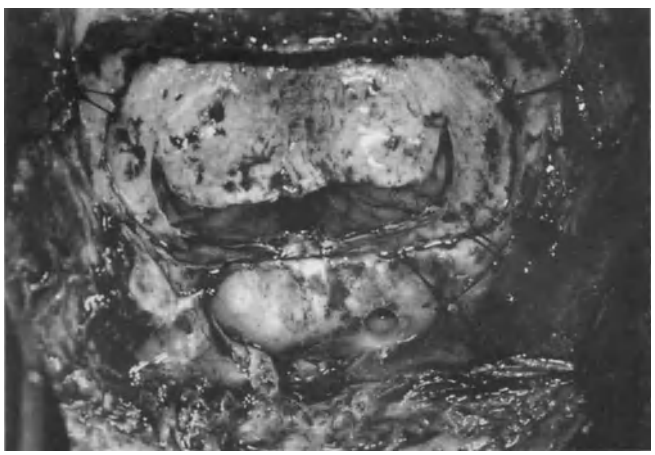
A



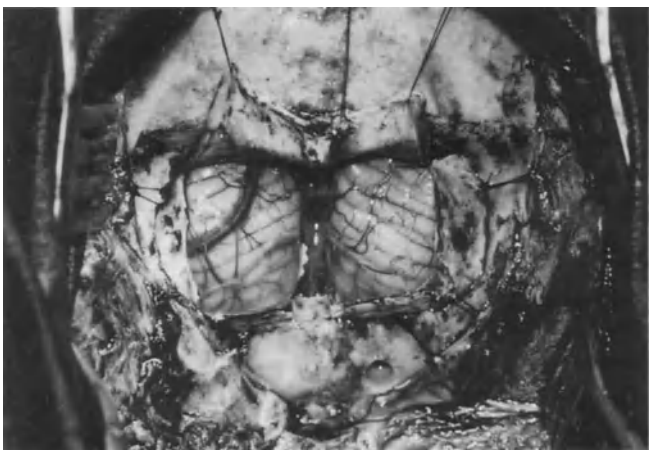
B

C

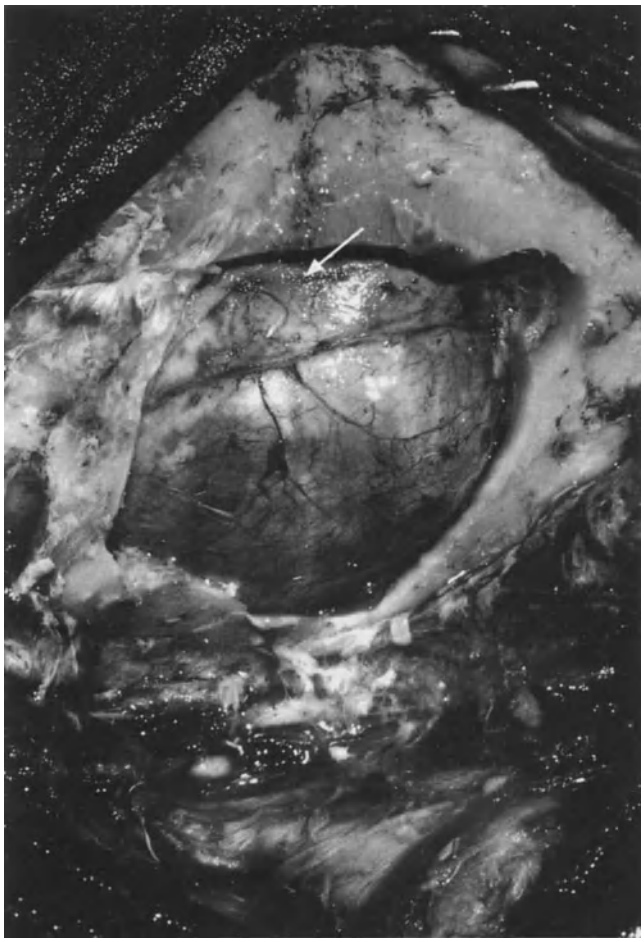
Figure 5.14. The double trapdoor durotomy for access to the superior cerebellar triangle permits the surgeon the option of reflecting only the superior trapdoor, or both. (A) The durotomy lines are depicted for complete exposure of the superior cerebellar triangle. (B) Posterooblique perspective of the double trapdoor durotomy. (C) This photograph illustrates the "hockey-stick" durotomies (*1*) made on either side. Also, one notes the transverse sinus (*2*) indicated along the superior margin of the craniotomy, the dural tack-up sutures (*3*), the drill hole (*4*) made in the sqamous occipital bone (for anchoring the lug of the DeMartell retractor) superolateral to the rim of the foramen magnum (*5*), and the posterior spine of C-1 (6). (D) The horizontal durotomy has been completed. (E) The superior dural flap has been sewn to periosteum, exposing the tentorial surface of the cerebellar hemispheres and the vermis.



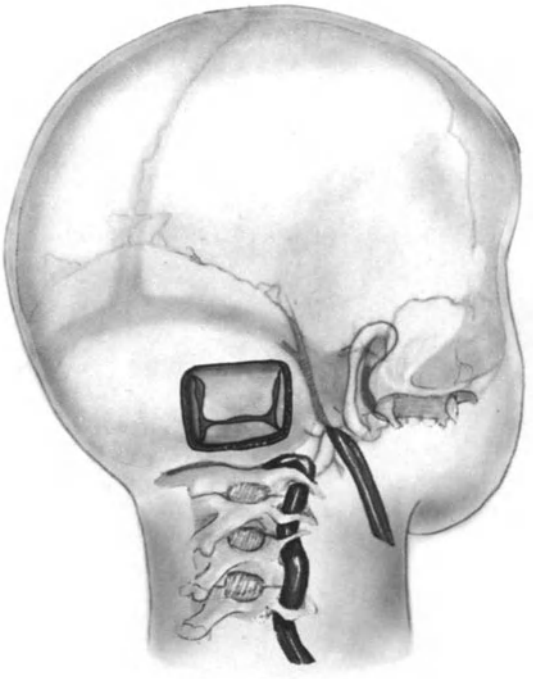
D



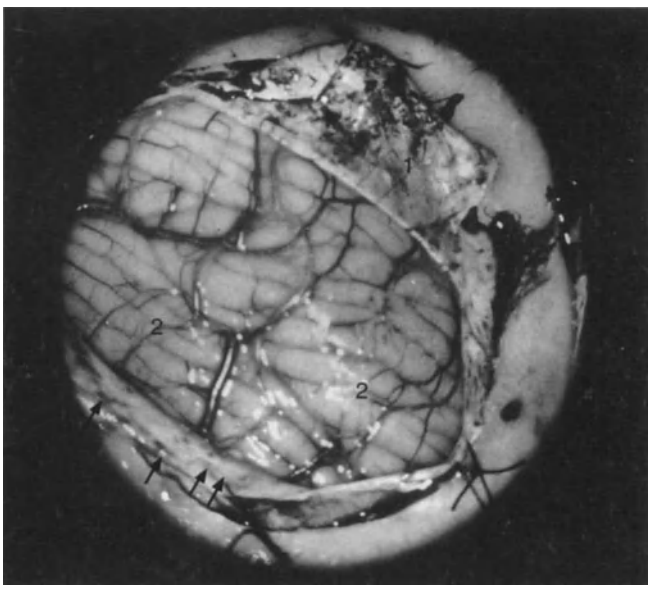
E



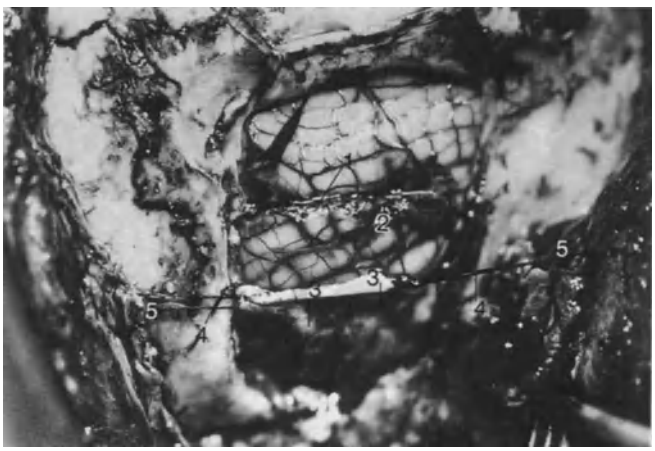
A



B



C



D

Figure 5.15. (A) The dura has been exposed over the most lateral portion of the right cerebellar hemisphere, revealing it to be under extreme pressure (as expressed by its transparency). The transverse sinus (arrow) is exposed. One should expose the transverse sinus only for vascular lesions which are draining either into the lateral surface of the tentorium or the transverse sinus itself. (B) This is an asymmetrical double trapdoor durotomy flap. The transverse sinus has not been exposed. (C) Reflecting the dura over the craniotomy edges exposes the lateral cerebellar surface. (Resection of lateral third of hemisphere, or its elevation, permit exposure of medullopontocerebellar angle). The superior dural flap (*1*) has been reflected, and a dural sling (arrows) has been fashioned to support the inferolateral cerebellar hemisphere (*2*) if one is working on a lesion within the superolateral portion of the cerebellum. (D) A mass within the right lateral cerebellar hemisphere was removed. Note cerebellotomy (*1*), the inferior portion of the right cerebellar hemisphere (*2*), and the dural sling (*3*) tethered from two 4-0 sutures (*4*), which have been anchored to the periosteum on either side (*5*).

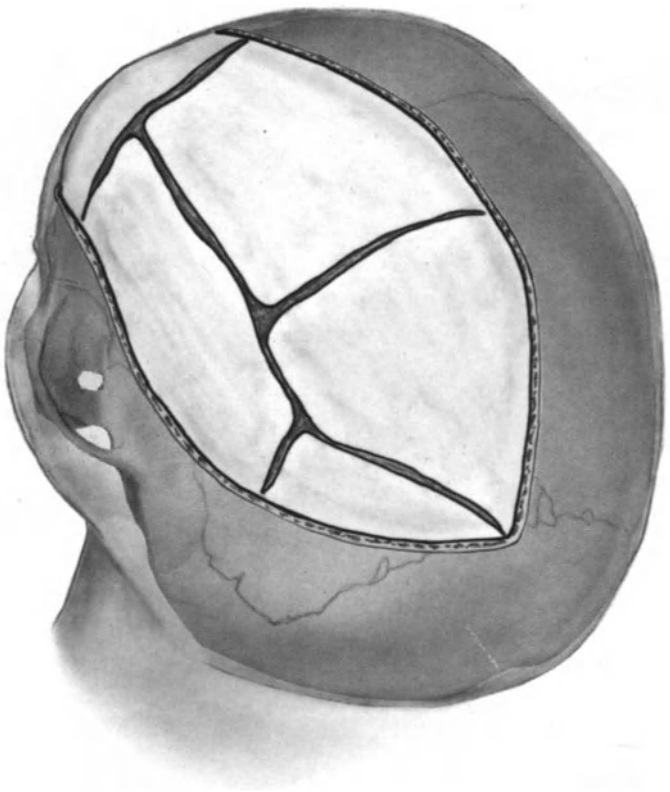


Figure 5.16. Hemispheral dural opening.

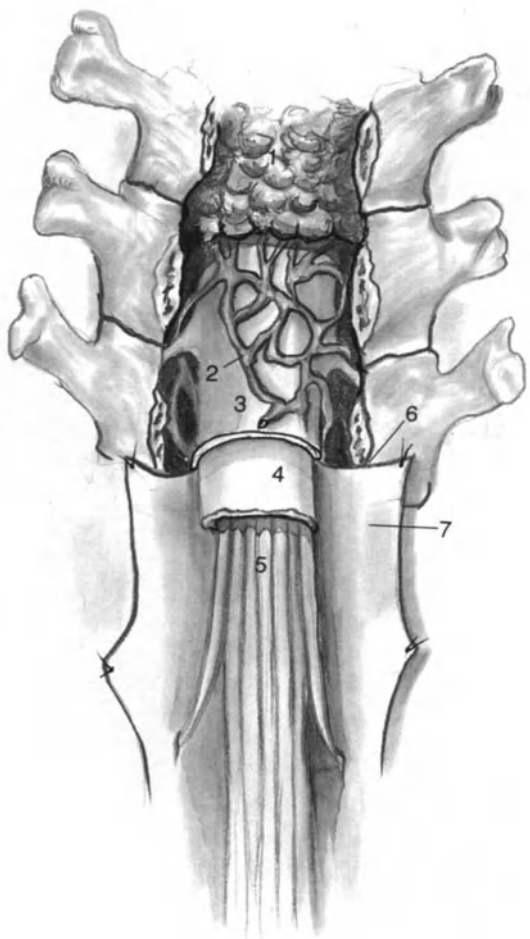


Figure 5.17. The spinal canal contains, from peripheral to central, epidural fat (*1*), epidural venous plexus (*2*), a single layer of dura (*3*), a single arachnoid membrane without compartmentalized subarachnoid chambers (*4*), and then either the spinal cord or the cauda equina (*5*). The dura mater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

Hemispheral Dural Opening (Figure 5.16)
The dural opening for hemispherectomy must be such as to permit one access to the entirety of 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 (Figures 5.17 to 5.20)
The spinal dura mater differs from the cranial dura mater in four surgically significant ways, having

1. a layer of epidural fat between the dura and the bony portion of the spinal canal;
2. a large plexus a epidural veins freely communicating with the intraabdominal 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, fissures; and
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.

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 volume and 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 recommended 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.

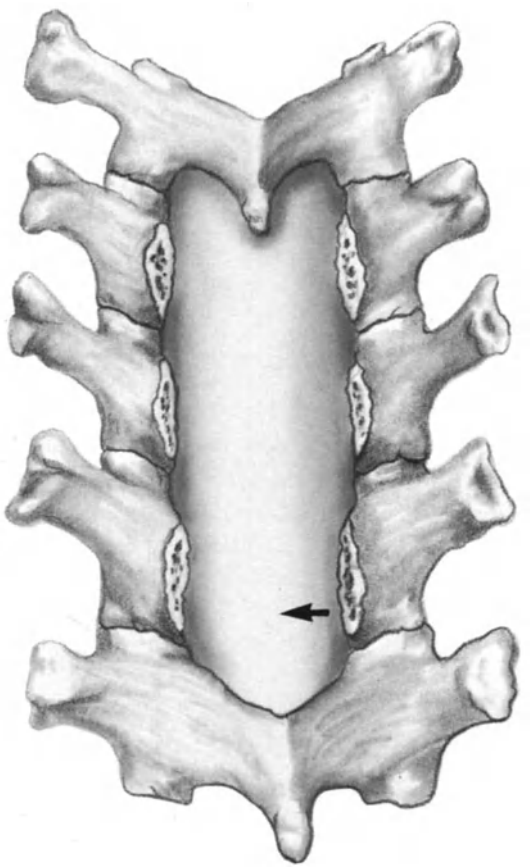


Figure 5.18. The epidural compartment has been cleared of fat and the venous plexus. Expansion of the intradural contents distends the (single layer) dural sac (arrow) to fill the spinal canal after the epidural veins and fat have been removed.

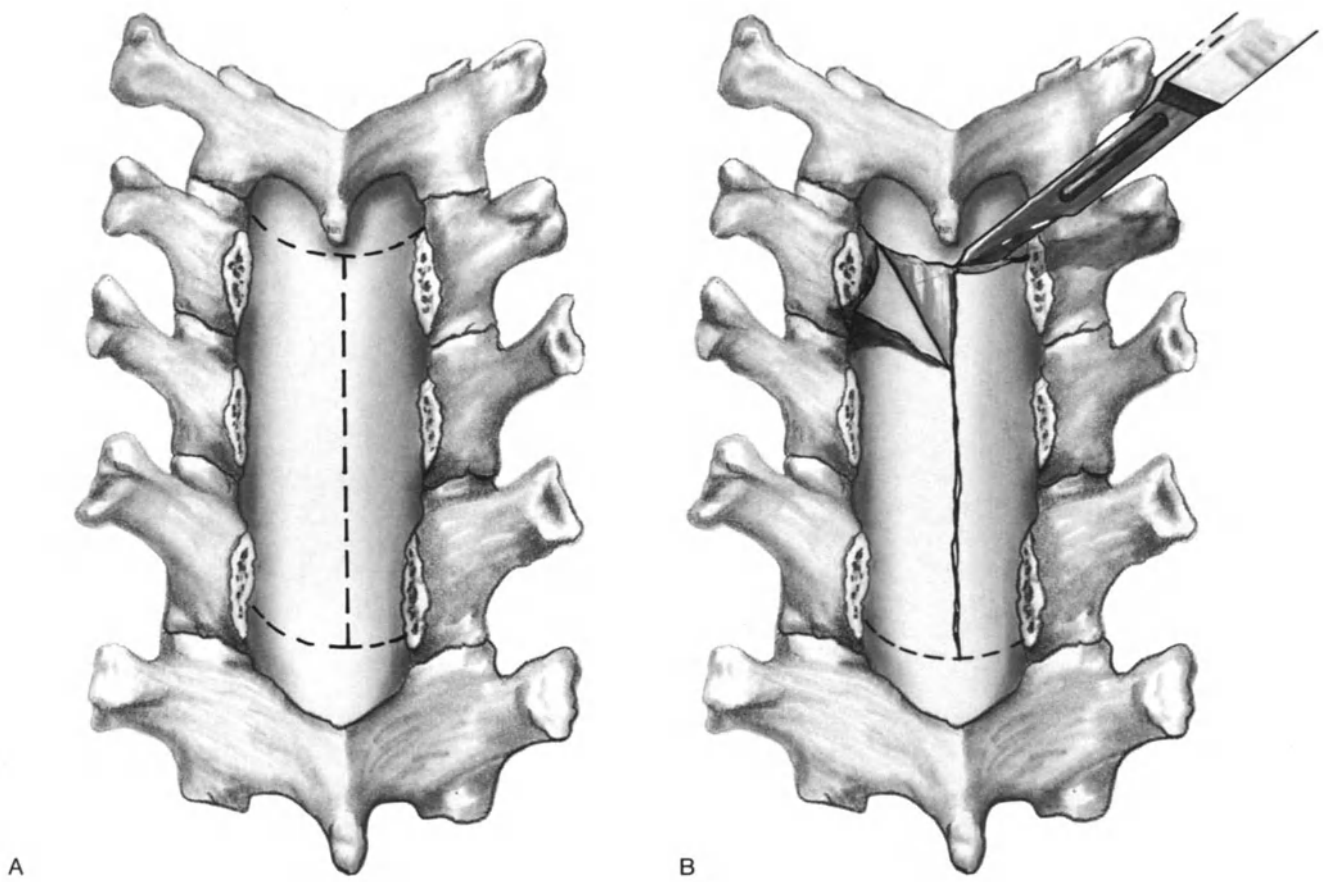
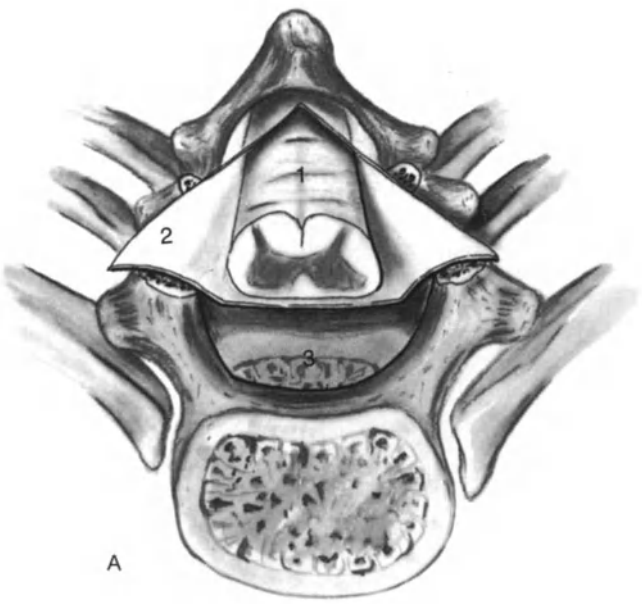
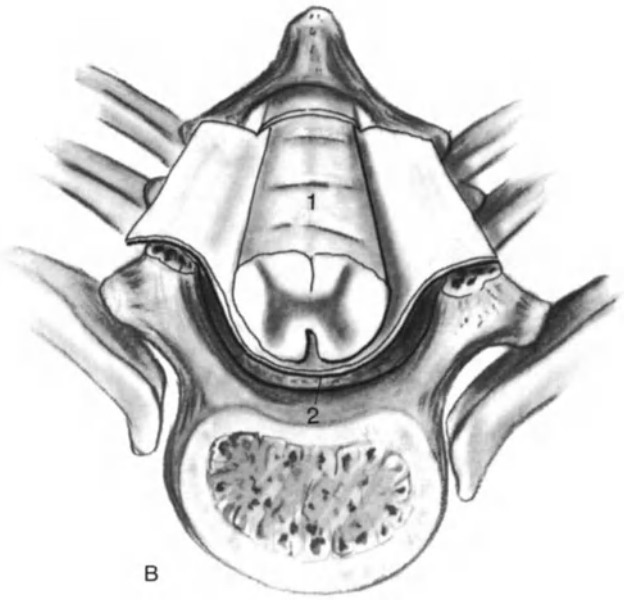
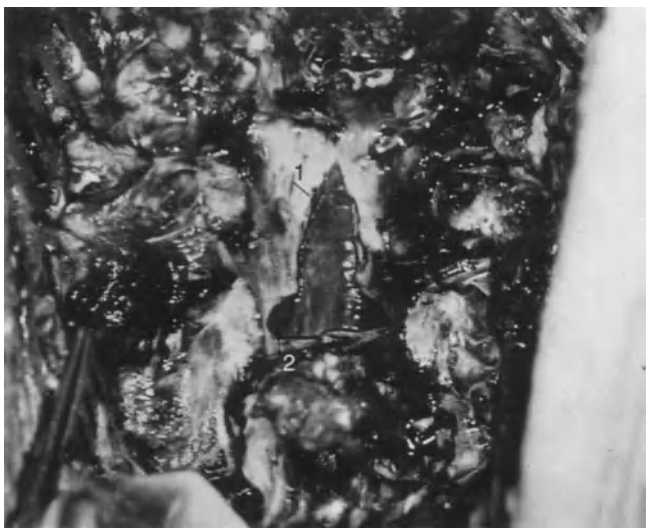


Figure 5.19. (A) The area of dural incision is marked off to show the durotomy lines. (B) The sagittal durotomy has been made and the perpendicular cuts are being extended. One may use either a #15 blade or a tenotomy scissors.Note the lateral reflection of the trapdoor flap on the reader's left, a technique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







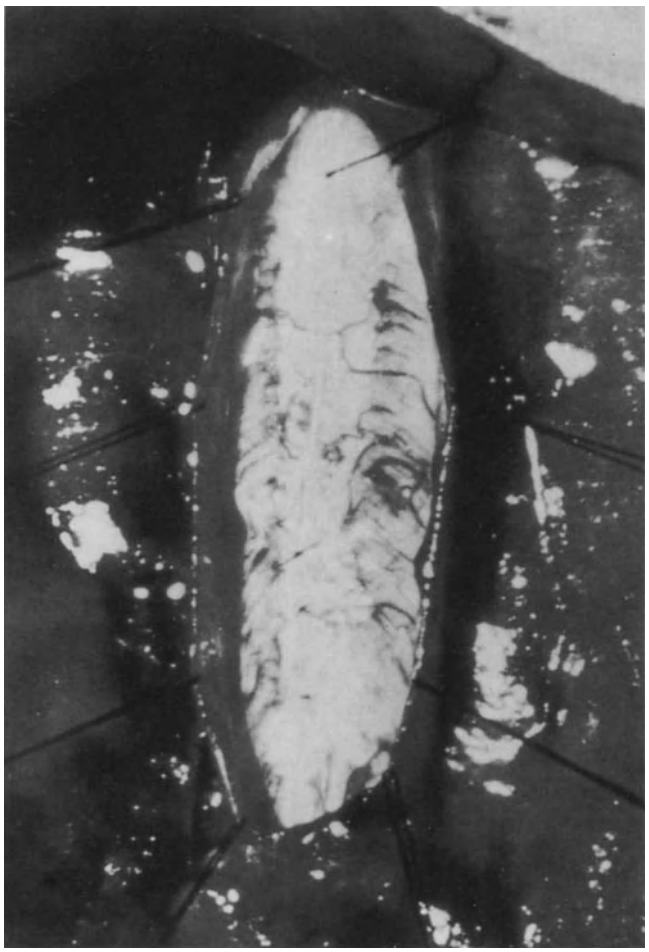


Figure 5.20. (A) Midsagittal durotomy only. A horizontal cut across the ventral aspect of the bony spinal canal, illustrating the suspension of the spinal cord (*1*) by the tented dura (*2*), thus lifting it from the ventral portion of the spinal canal (*3*). This is a potential cause of spinal cord 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*). There is no compression of the spinal cord by stretching dura. (C) Midsagittal (*1*) and circumferential perpendicular (*2*) durotomies made on both the right and left. (D) Incorrect technique for opening and retracting the dura over the spinal cord. Suspension sutures putting traction on the dura and, consequently, causing it to compress the ventral surface of the spinal cord.

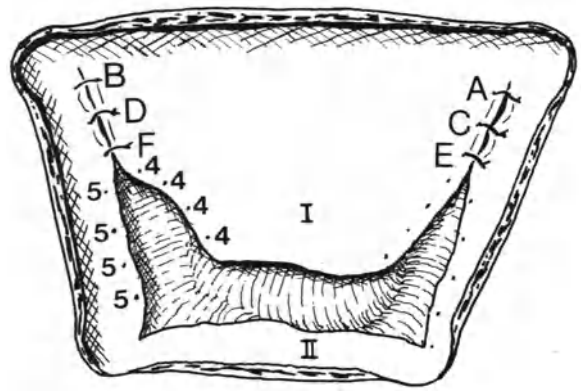


Figure 5.21. Opposite borders of the dural defect are used as initial anchoring sites. The free edge of the dural flap (*I*) is that which is medial to the dural opening, the anchored edge (*II*) is between the dural opening and the bone. Sewing the dura short, along the free edge and long on the anchored edge, with stitches alternating from limb to limb, stretches it out evenly, assuring uniform closure. Here 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 gradural 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.

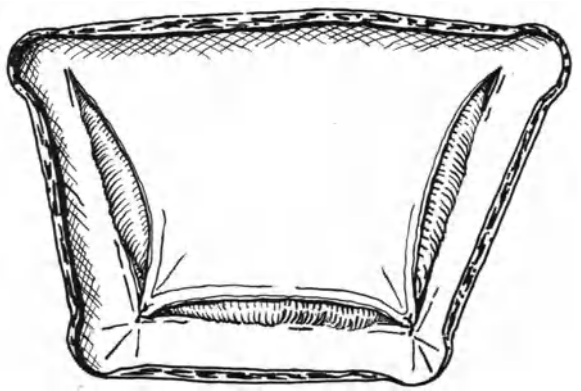


Figure 5.22. Beginning the dural closure by placing sutures at the corners as illustrated here is to be avoided, except in those rare instances when there is an abundant subdural space and the dura is, for all intents and purposes, redundant.

Closure

Cranial Closure

Dural Closure (Figures 5.21 and 5.22)

The dural closure is greatly facilitated when it 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. 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 and permits the brain to bulge through the intervening openings.

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

When it is not possible to close the dura, or doing it demands so 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 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. Erector spinae muscle fascia suffices as a donor source 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 fascia from the 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 erector capitae muscle graft to

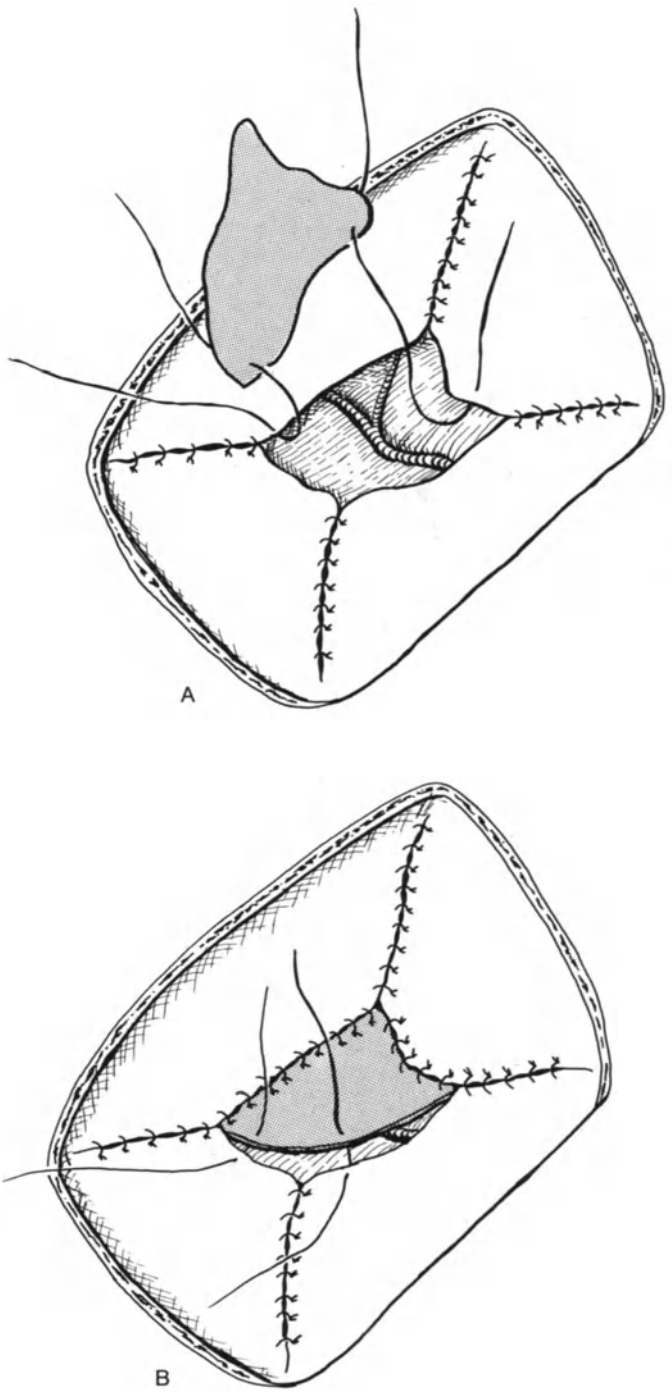


Figure 5.23. (A) Periosteal graft to cover dural defect. Opposite borders of the dural defect are used as initial anchoring sites. (B) 4-0 interrupted sutures are used to attain a watertight closure between graft and dural edges.

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.

Spinal Closure

Arachnoid Closure

The closure of the arachnoid is recommended in spinal cord tumor surgery, and is essential for syringomyelia. It may be closed with either interrupted or continuous 7-0 sutures.

Adequate closure of the arachnoid is guaranteed at the time of opening, since at this time the surgeon has the opportunity to identify it clearly, before placing guide sutures, 7-0 or 8-0, so as to tent it. If the sutures are lain in at 2- or 3-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 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.

References

1. Raimondi AJ: Reflection of a laminar flap for exposure of the spinal canal in children. Clin Neurosurg 25:504–11, 1978.
2. Raimondi AJ, Gutierrez FA: Reconstruction of the posterior vertebral arch and laminotomy for intraspinal surgery. In Ransohoff J (ed): *Modern Technics in Surgery—Neurosurgery*. Mount Kisco, NY, Futura, 1979, pp 10–11.
3. Raimondi AJ, Gutierrez FA, DiRocco C: Laminotomy and total reconstruction of the posterior spinal arch for spinal canal surgery in childhood. J Neurosurg 45:555–60, 1976.

"What is history, but a fable agreed upon!"

NAPOLEON BONAPARTE
Comments

Chapter 6

Cerebral Retraction

Cistern Openings (Figures 6.1 to 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 cerebral spinal 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 hemispheral 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),

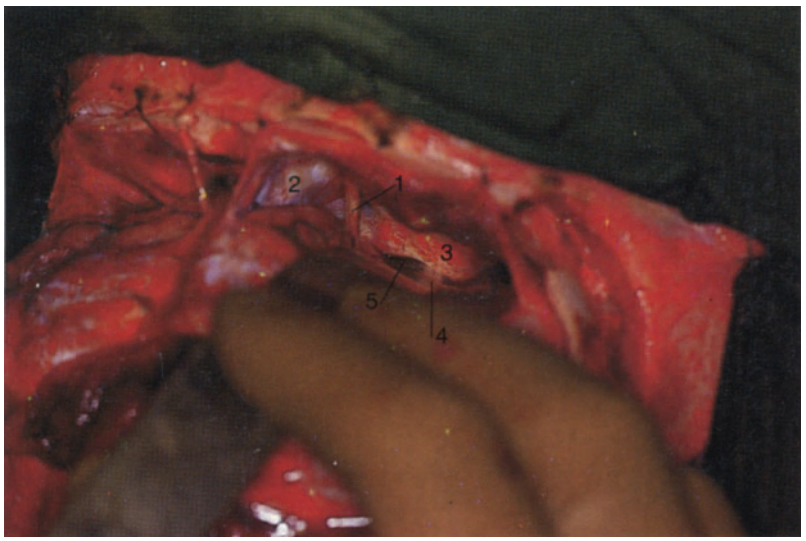


Figure 6.1. Olfactory, prechiasmatic, and optic cisterns. The dura has been opened and the right frontal lobe is being retracted, exposing the right olfactory nerve (*1*) after the cistern has been opened with bipolar forceps and the arachnoid incised. The crista galli (*2*), anterior clinoid (*3*), 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*.

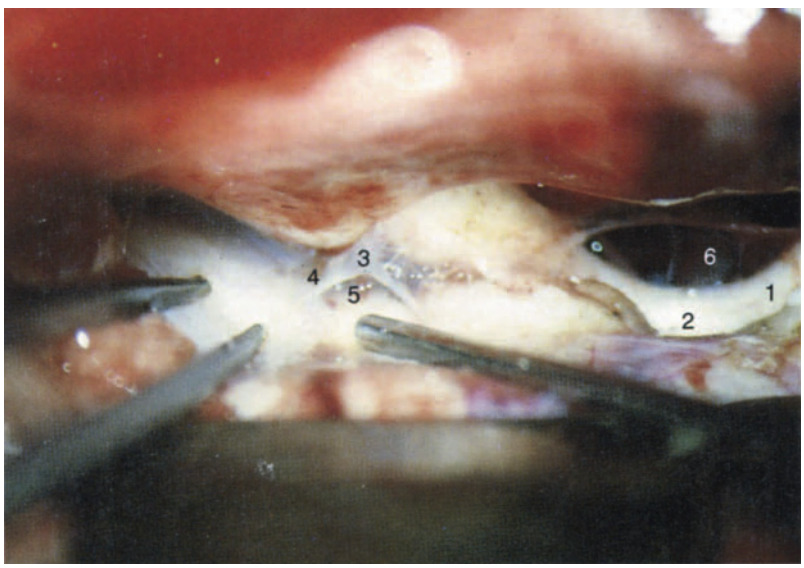


Figure 6.2. The right optic nerve (*1*), optic chiasm (*2*), and arachnoid covering the left optic nerve (*3*) are well shown, as is the arachnoid bridging from the left optic nerve to the left temporal lobe (*4*). This arachnoid, along the interval between the optic nerve and the temporal lobe, has 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.

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.

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 egrees 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

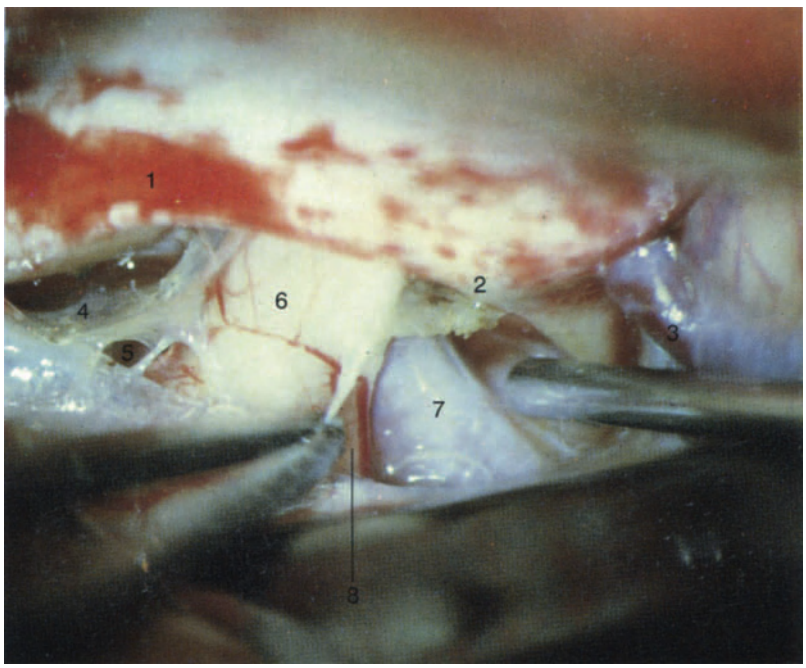


Figure 6.3. Interopticocarotid cistern. The planum sphenoidale (*1*), anterior clinoid (*2*), and sphenoparietal vein (*3*) are shown. The arachnoid from over the *prechiasmatic cistern* (*4*) has been opened (*5*), permitting removal of the arachnoid from over the right optic nerve (*6*) and the internal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

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.

Ambient Cistern Lesions

Ambient cistern lesions (medial temporooccipital 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 interpeduncular 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

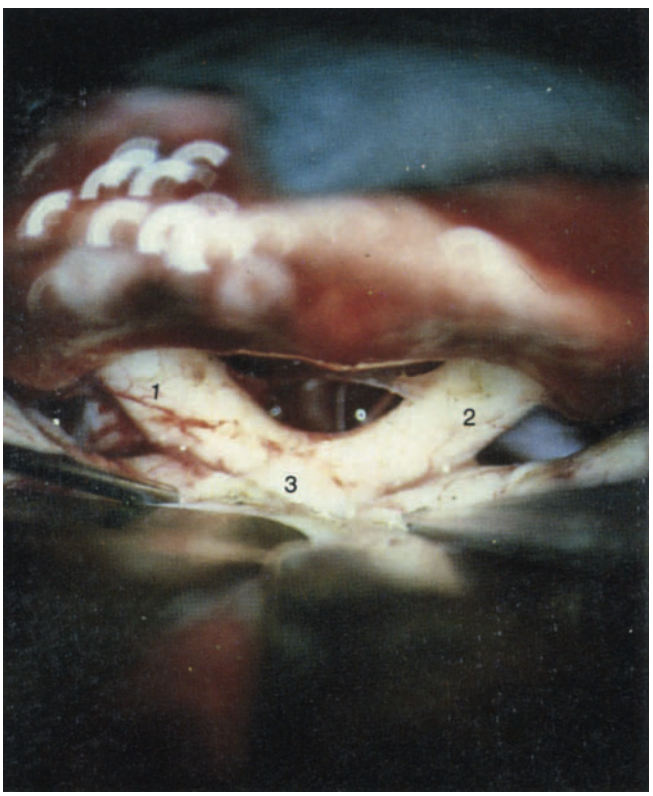


Figure 6.4. Chiasmatic cistern. Both optic nerves (*1, 2*) are exposed after the *prechiasmatic and optic cisterns* have been opened, allowing one to expose the optic chiasm (*3*) after lifting the arachnoid from over it.

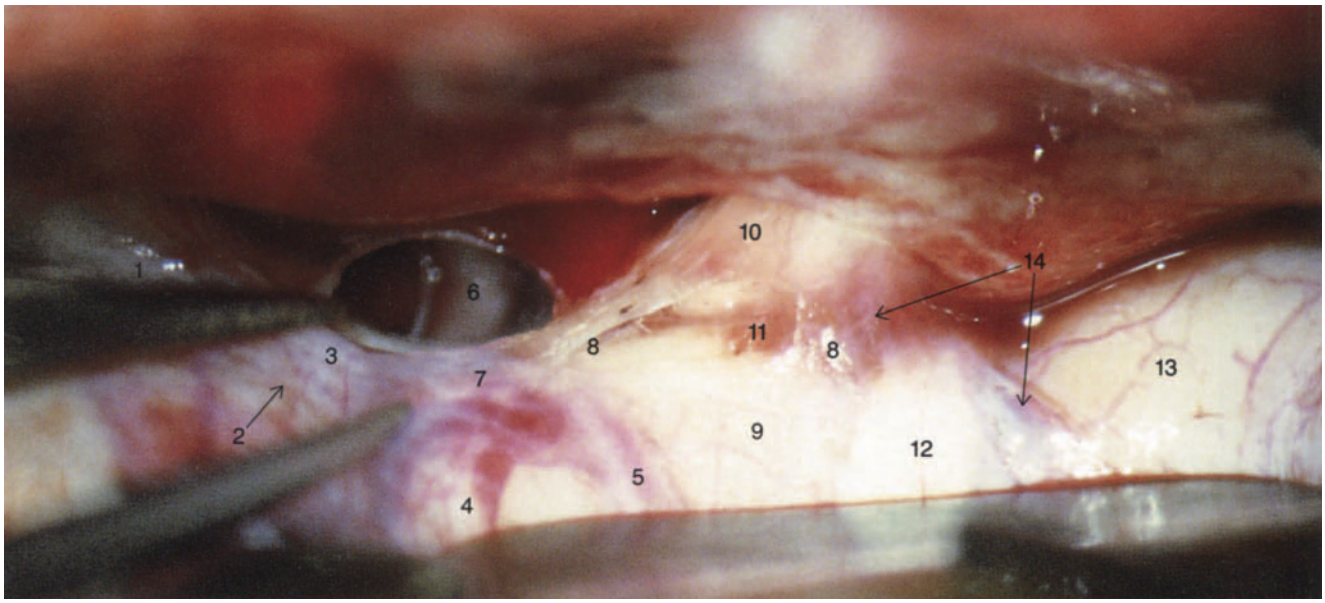


Figure 6.5. The left optic nerve (*1*), olfactory nerve (*2*), and gyrus rectus (*3*) are exposed, as are the right gyrus rectus (*4*) and right olfactory nerve (*5*). The *prechiasmatic cistern* has been opened, exposing the pituitary stalk (6). The arachnoid bridging from the left gyrus rectus to the right gyrus rectus (*7*), at the tip of the sucker, is enclosing the *interhemispheral 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*).

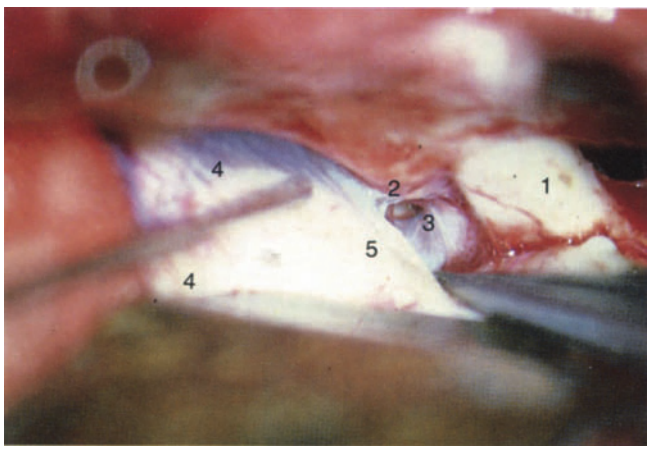


Figure 6.6. The left optic nerve (*1*) and anterior clinoid (*2*) are visible. The arachnoid has been taken from the lateral surface of the left internal carotid artery (*3*) but not from the posterior orbital gyrus of the left frontal lobe (*4*). One sees the arachnoid over the *Sylvian fissure* (*5*) 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.

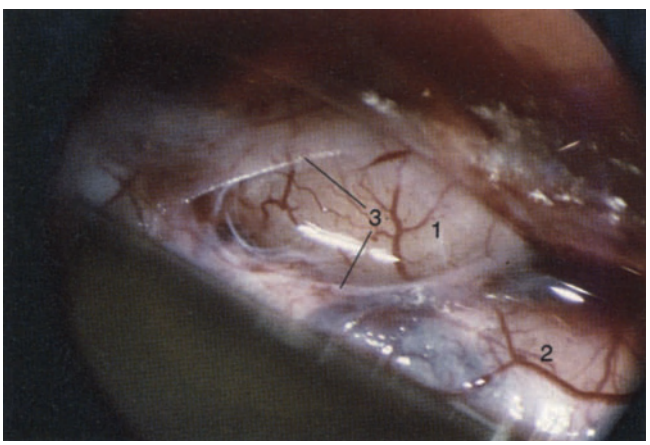


Figure 6.7. The lateral Sylvian fissure. The superior medial surface of the left temporal lobe (*1*) is not covered by arachnoid, but the inferior lateral surface of the left frontal lobe (*2*) is. The arachnoid covering the lateral third of the Sylvian fissure has been opened so one may see its edges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

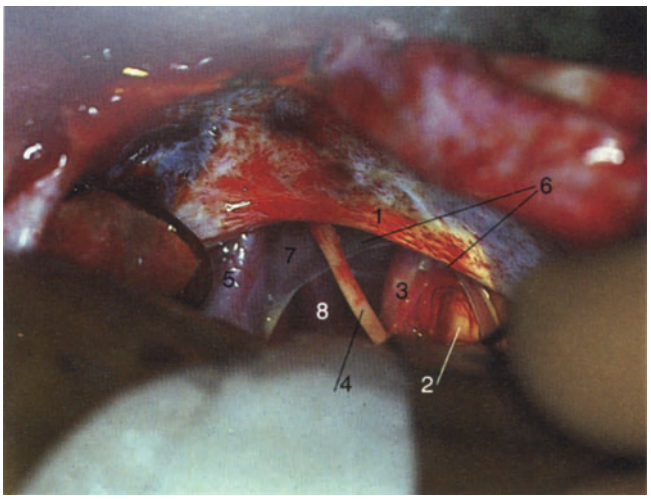


Figure 6.8. Ambient and interpeduncular cisterns. The tentorial edge (*1*), pons (*2*), basilar artery (*3*), III cranial nerve (*4*), and internal carotid artery (*5*), are well visualized. The *ambient cistern* runs along the tentorial edge (*6*). Lillequist's membrane (*7*) has been opened, allowing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

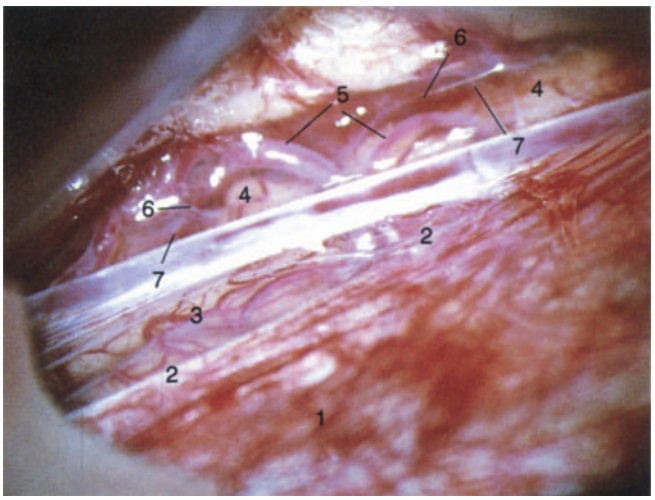
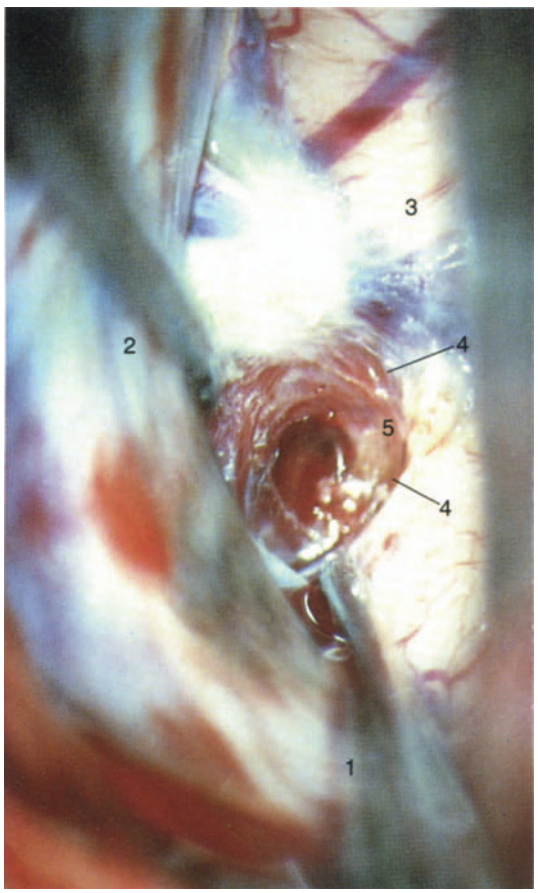


Figure 6.9. Pericallosal cistern. The falx cerebri (*1*), inferior longitudinal sinus (*2*), supracallosal gyrus (*3*), corpus callosum (*4*), and pericallosal artery (*5*) are all identifiable. The *pericallosal cistern* (*6*) is located between the corpus callosum and the arachnoid (*7*), and contains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



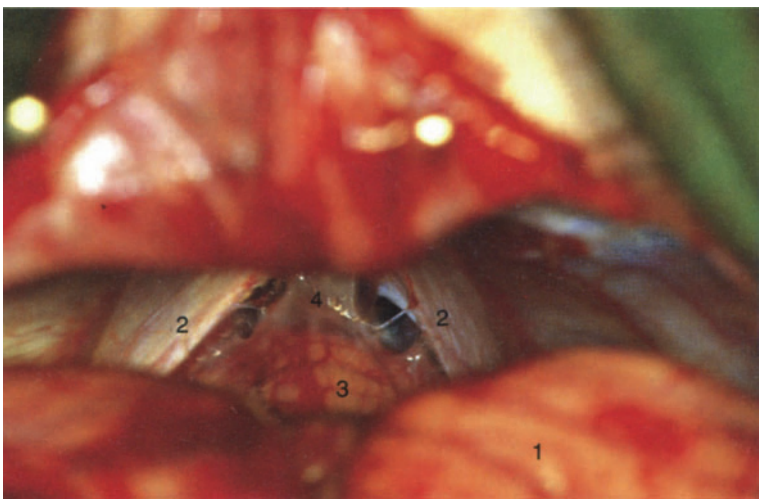


Figure 6.10. (A) Supracerebellar cistern. This child has a pineal tumor. The tentorium (*1*), falx cerebri (*2*), and isthmus of the hippocampus (*3*), have been exposed, and the arachnoid (*4*) along the posterior surface of the *supracerebellar cistern* opened, exposing the tumor (*5*) which occupied 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*).

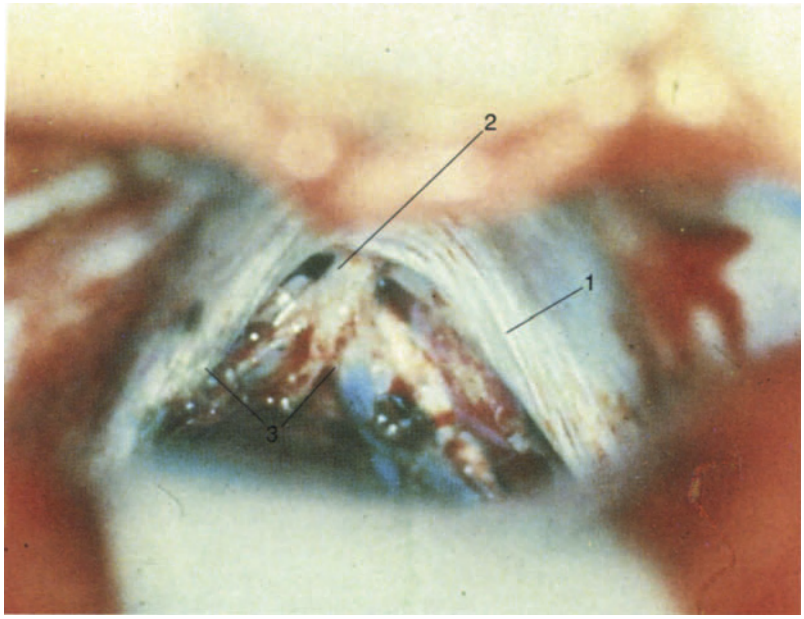


Figure 6.11. Quadrigeminal cistern. The tentorial opening (*1*) has been exposed and the culmen monticuli of the cerebellar vermis retracted downward, after the posterior and anterior arachnoidal membranes of the *supracerebellar cistern* had been coagulated and opened. This exposed the great vein of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

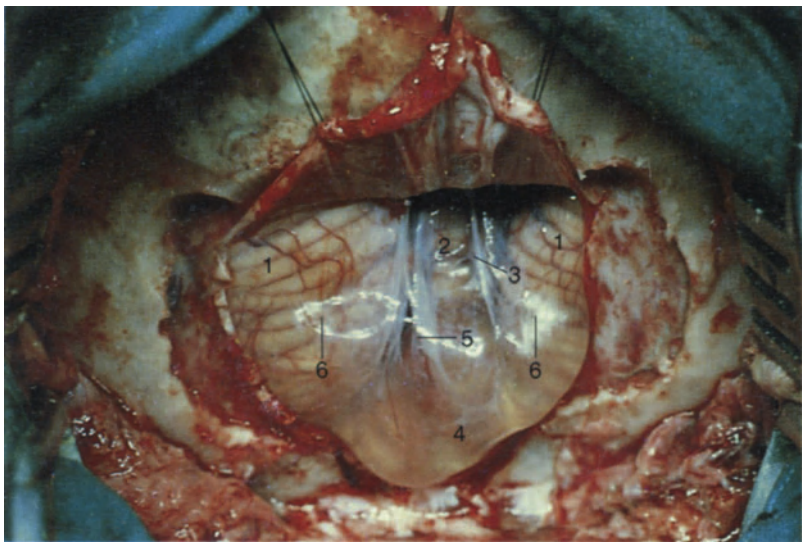


Figure 6.12. Cisterna magna. The cerebellar hemispheres (*1*) and cerebellar vermis (*2*), as well as the inferior vermian veins (*3*) may be seen beneath the arachnoid of the *cisterna magna* (*4*). This cistern has been opened (*5*) but most of the cerebrospinal fluid remains within it, permitting 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*).

and along the parasagittal route! If one approaches the corpus callosum with the cerebral hemisphere inferior and the falx cerebri superior, he does, indeed, gain great exposure advantages in that the cerebral hemisphere follows gravity. A remarkable space opens between the parasagittal surface of the parietal lobe 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.

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, 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

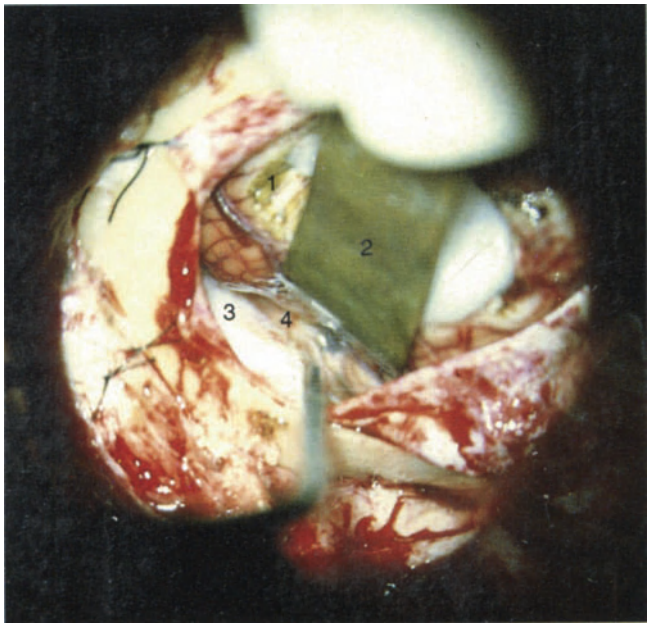


Figure 6.13. Pontocerebellar cistern. The left cerebellar hemisphere (*1*) has been elevated superomedially by a spatula (*2*), exposing the dura over the petrous apex (*3*) and the left *pontocerebellar cistern* (*4*) at the point of entry of the lateral recess of the IV ventricle.

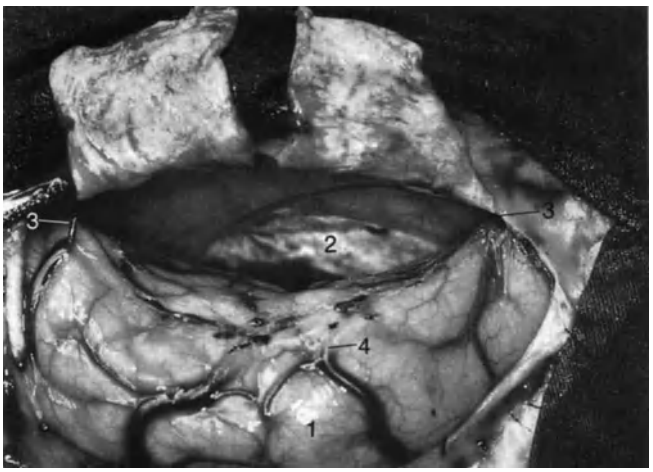


Figure 6.14. Disadvantages of gravitational retraction for pineal tumors. Notice the stretching of bridging veins. This child's pineal tumor was operated along the parasagittal route, with the child's head positioned so that the right parietal lobe (*1*) was recumbent. The retraction, of course, is greatly 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 particularly unacceptable, 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.

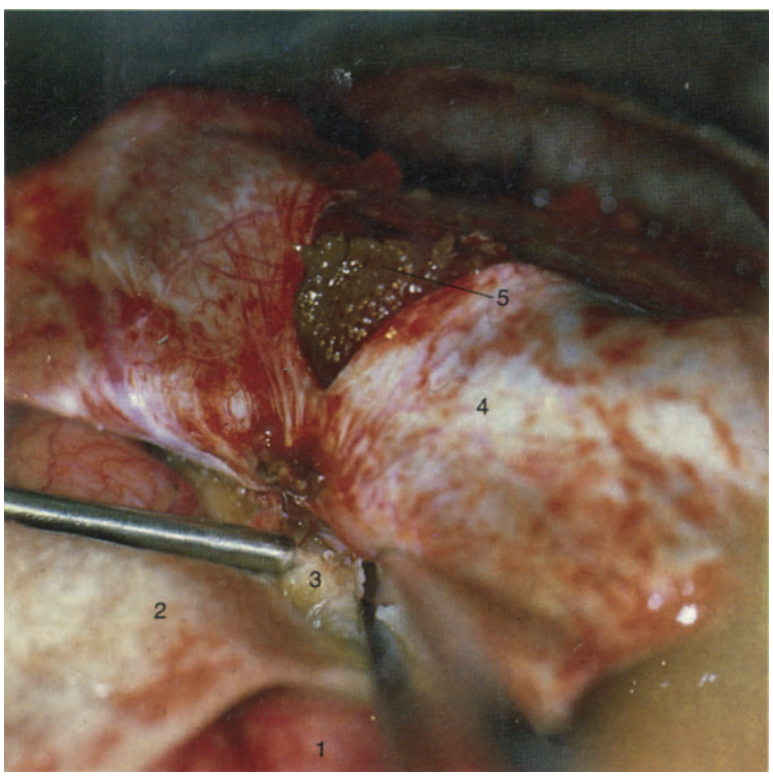


Figure 6.15. Separation of parietal lobe from arachnoidal granulations. This child, who also had a right parasagittal approach to his pineal tumor, was positioned with his left parietal lobe recumbent. Consequently, retraction of the right parietal lobe entailed elevating it, thus eliminating unwanted 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 superior sagittal sinus from compressive occlusion.

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, the wide-bladed De Martel 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 lesion is now completed (Fig. 6.16). 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 fornices!

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 contrast 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% 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 atlantooccipital 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 the pontocerebellar cistern superolaterally.

Use of Cotton Fluffies and Telfa

(Figures 6.17 to 6.19)

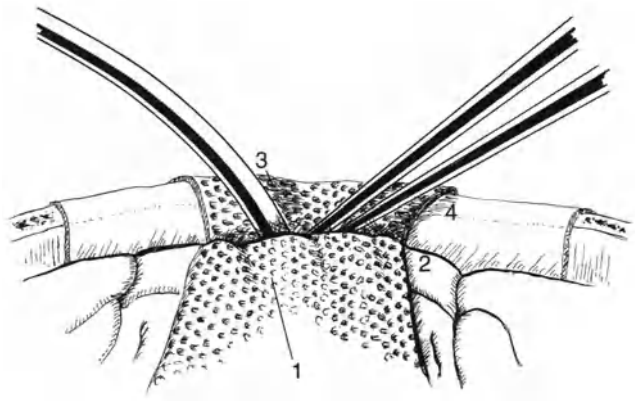
Cotton fluffies, prepared simply by soaking absorbent cotton in saline after it has been fashioned into the desired size and form, provide excellent protection during the exposure of such vital and delicate structures as the cranial nerves, normal or pathological vasculature, the intraventricular (ependymal)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

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!

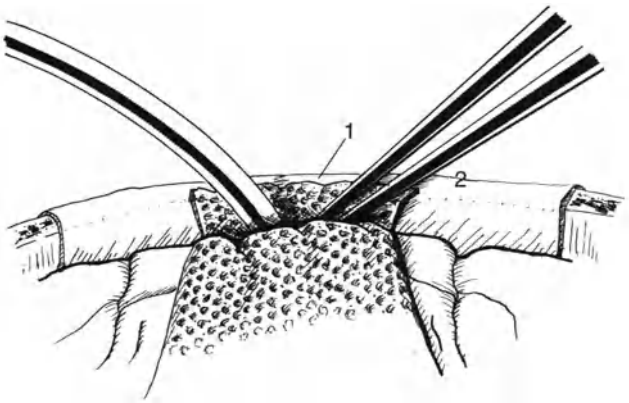
Telfa, cut to the desired form and length, moistened, and then lain over the surface of the brain, provides an excellent means by which proper retraction may be attained and the brain protected either from compression or inadvertent glancing blows or cuts. The telfa is applied directly to the surface of the brain. Sliding it, moist or dry, over the brain is 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 6.16. The retracted parietal lobe permits exposure of the falx cerebri (*1*) down to the pericallosal cistern (*2*). Splitting the corpus callosum (*3*) exposes the pineal tumor (*4*), located between the roof of the III ventricle and the inferior surface of the body of the corpus callosum.

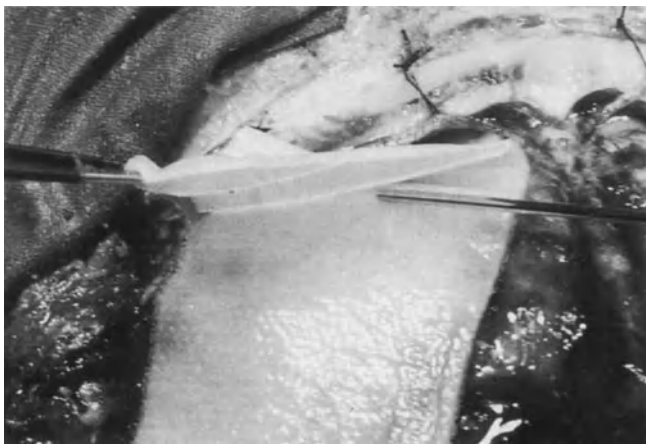
Figure 6.17. Cotton fluffies should be pulled into the desired form and then soaked immediately before applying them to the brain or a bleeding vessel. They should not be left soaking in a basin prior to use, for this makes them dense.



A



B



C

D

E

Figure 6.18. (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, causes minute subarachnoid or intrapial hemorrhages. (E) The telfa has been lain over the frontal pole, exposing the dura over the orbital roof.

Figure 6.19. (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 (Figure 6.20)

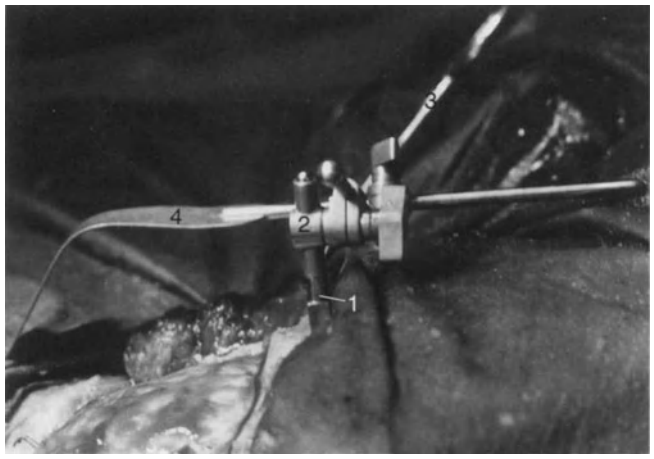
Self-retaining retractors are essential to the proper performance of many neurosurgical procedures. They allow the surgeon to perform microsurgical procedures without the need of an assistant. It is physically impossible for an assistant to hold them steadily because of the extremely small working area 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, 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 advised 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, *with 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%."[5]

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 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 reexpand 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 minutes every 5–10 minutes.



A

B

C

Figure 6.20. (A) Mounted De Martel self-retaining retractor, illustrating the mounting lug (*1*), universal joint (*2*), extension rod (*3*), retractor blade (*4*). (B) Allowing the frontal lobes to gravitate from the anterior fossa, before proceeding to apply the retractor and blades to the telfa-covered brain minimizes considerably the risks of cerebral damage. In this photograph, the frontal lobes (covered with telfa) have fallen from the dura and frontal bone. (C) Application of two self-retaining retractor blades, parallel to one another, over telfa placed on the frontal lobes, permits one to expose the entire anterior fossa, parasellar area, and pterional regions. Minimal, if any, retraction is necessary if one patiently aspirates cerebrospinal fluid from the basal cisterns and allows time for gravity to permit the frontal lobes to settle.

References

1. Magendie F: Mémoire sur liquide qui se trouve dans la crane et l'épine de l'homme et des animaux vertèbres. J Physiol Exper Pathol 7:1–17, 1827.
2. Key EAH, Retzius G: Studien in der Anatomie des Nervensystems und des Bindegewebes. Stockholm, Samson och Wallin, 1875–1876.
3. Donaghy RMP, Numoto M, Wallman LJ, et al.: Pressure measurement beneath retractors for protection of delicate tissues. Am J Surg 123:429–31, 1972.
4. Albin MS, Bunegin L, Bennett MH, et al.: Clinical and experimental brain retraction pressure monitoring. Acta Neurol Scand 56(suppl 64):522–3, 1977.
5. Yokoh A, Sugita K, Kobayashi S: Intermittent versus continuous brain retraction — An experimental study. J Neurosurg 58:918–23, 1983.
6. Balani DK, Pathak SN, Sriramachari S: Pathology and pathogenesis of experimental extradural cerebral compression. Indian J Med Res 74:438–61, 1981.
7. Ishii S, Hayner R, Kelly WA: Studies of cerebral swelling. II. Experimental cerebral swelling produced by supra-tentorial extradural compression. J Neurosurg 16:162–6, 1959.
8. Miller DJ, Slanek AF, Langfitt TW: Cerebral blood flow regulation during experimental brain compression. J Neurosurg 39:186–96, 1973.
9. Numoto M, Donaghy RMP: Effects of local pressure on cortical electrical activity and cortical vessels in the dog. J Neurosurg 33:381–7, 1970.
10. Rosenorn J, Diemer NH: Reductions of regional cerebral blood flow during brain retraction pressure in the rat. J Neurosurg 56:826–9, 1982.
11. Yamagouchi M, Shirakata S, Yamasaki S, et al: Ischemic brain edema and compression brain edema. Water content, blood brain barrier and circulation. Stroke 7:77–83, 1976.

"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 MACALAY
Horatius. Lays of Ancient Rome

Chapter 7

Cerebrotomy

Exposure of intraparenchymal or intraventricular tumors necessitates opening the cerebrum (cerebrotomy). Classically, there are two techniques for performing cerebrotomy: (1) going through a gyrus; (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 damage, just as retraction of a sulcal cerebrotomy may cause tearing of sulcal vessels and also 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.

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 grey 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 places, its destruction in others. 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 five 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 ineffi-

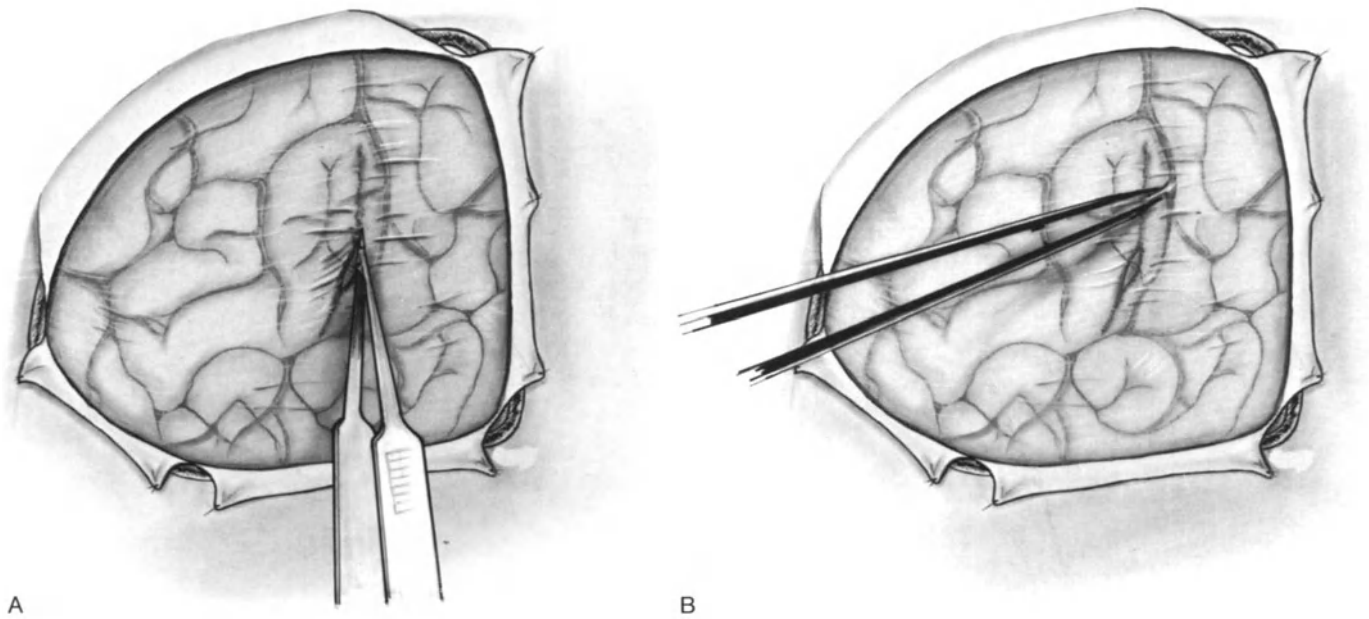


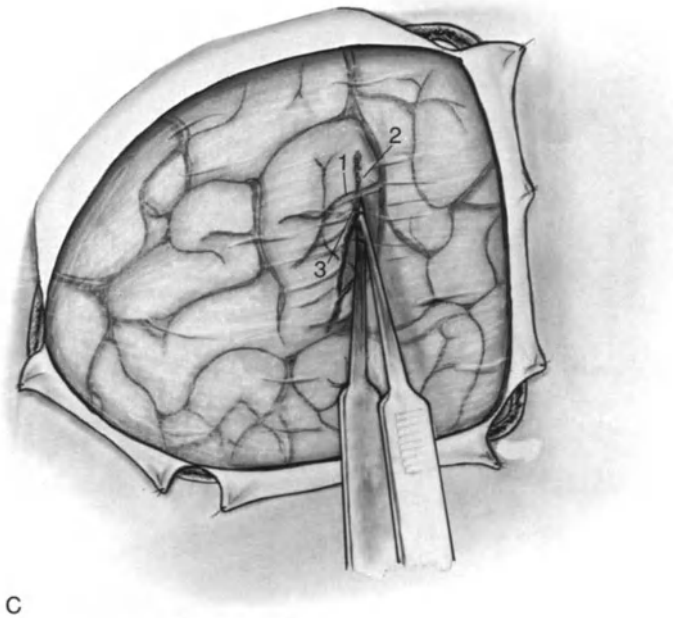
Figure 7.1. Gyral cerebrotomy. (A) After the arachnoid and adjacent microvasculature have been coagulated over the full extent of the cerebrotomy incision, and a bipolar forceps has been used to "burn" an opening in the arachnoid, microscissors are inserted into the opening so as to take only the arachnoid 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. (E) Adjacent gyri (*1*) are being opened for the cerebrotomy. The arachnoid and underlying cortical surface (*2*) have been coagulated as has a sulcal vein. (F) The coagulated sulcal vein is being transected with microscissors.

cient. One reaches the desired depth more quickly and efficiently by using two spatulas as described later in this chapter.

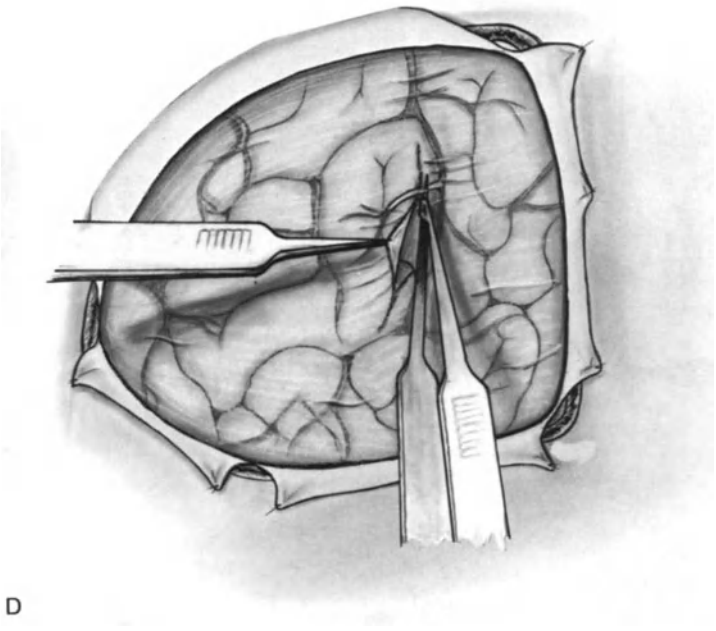
Gyral Cerebrotomy (Figure 7.1)

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 fluffly cottons to dry the field) for uncoagulated microvasculature.

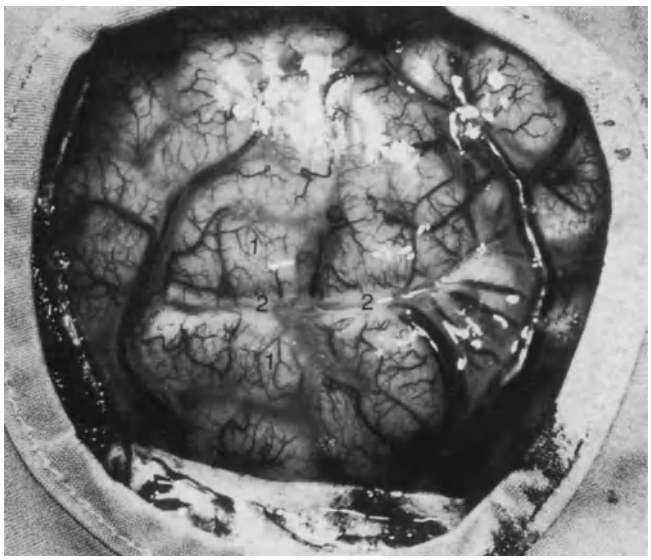
Pia-arachnoid and cortical vessels, arteries or veins, running perpendicular to the line of cerebrotomy should be coagulated and cut. This technique for transecting vessels, especially arteries, prevents one from cutting a vessel only partially, an event which results in bleeding that is particularly 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.



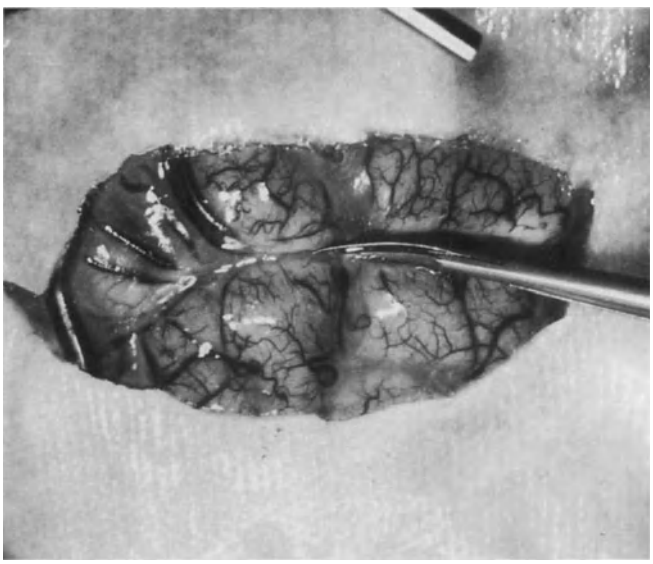
C



D



E

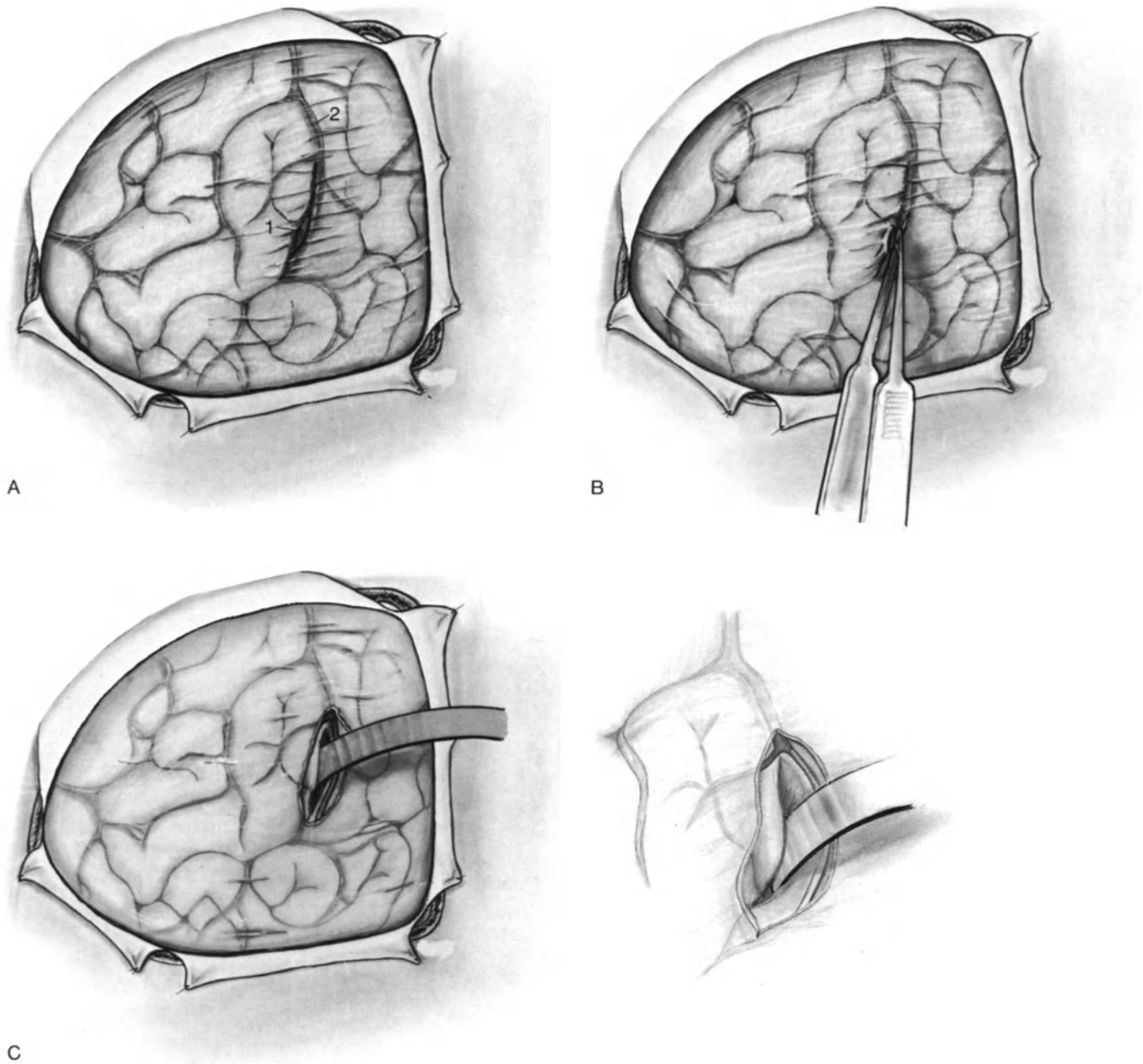


F

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 (Figure 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 across 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 coagulation 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 (Figure 7.3)

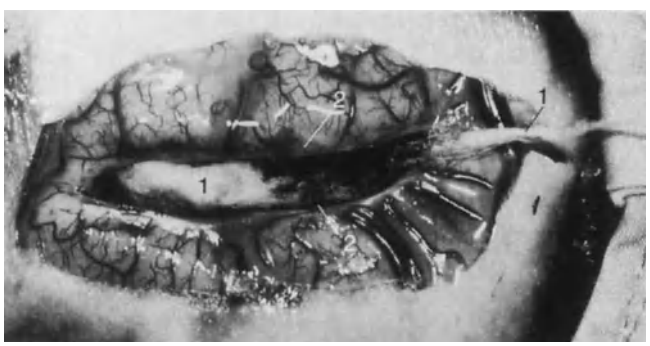
The small vessels at the very depth of the sulcus or gyrus are coagulated individually. Generally, they withdraw and may be pulled apart with the bipolar forceps since they are tiny. Occasionally, however, it may become necessary to cut them with the microscissors. In either event one should coagulate completely all vessels at the depth of the sulcus. Then, he may insert a long, thin fluffy cotton 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 (Figures 7.4 and 7.5)

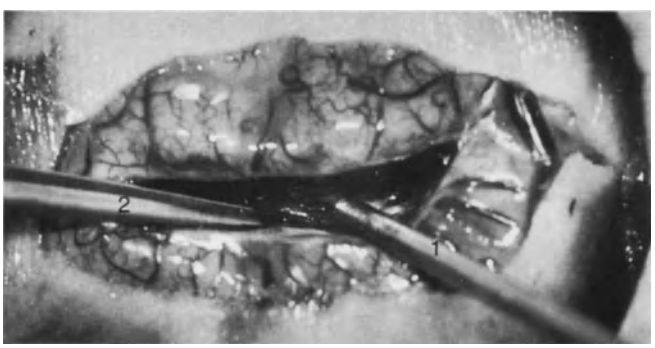
In extending the cerebrotomy down to either a deep-seated lesion or the intraventricular compartment, one need no longer worry about bleeding: there is no vascu-

D

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 sulcal microvasculature have been coagulated and cut, the malleable thin-bladed spatula is inserted to perform the dissection down to the white matter. (D) An extensive, circumferential, sulcal cerebrotomy (in which it was necessary to transect a cortical vein), illustrating the opening to the junction of grey and white matter. The Penfield #1 dissector is retracting the cortical cerebrotomy.



A



B

Figure 7.3. (A) Whether a gyral or sulcal cortical cerebrotomy is performed, the bleeders within the cortex are managed in identically the same way. A small fluffy cotton (*1*) is inserted into the depth of the cerebrotomy (*2*) to identify bleeding vessels large enough to coagulate, and to occlude those so small that they will stop spontaneously. (B) After the bleeding vessel has been identified, it is drawn into the microsuction (*1*) and then occluded with the bipolar forceps (*2*).

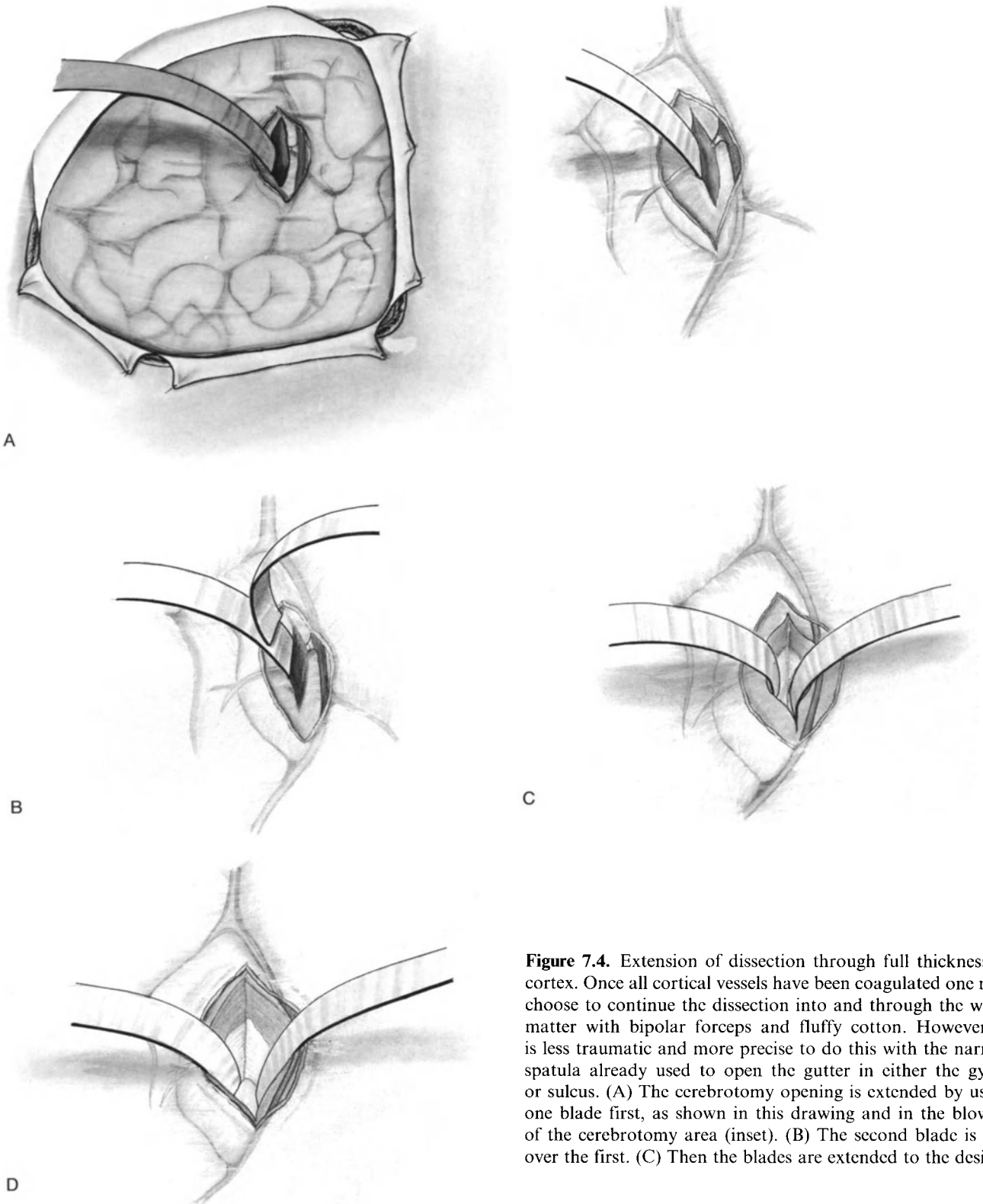
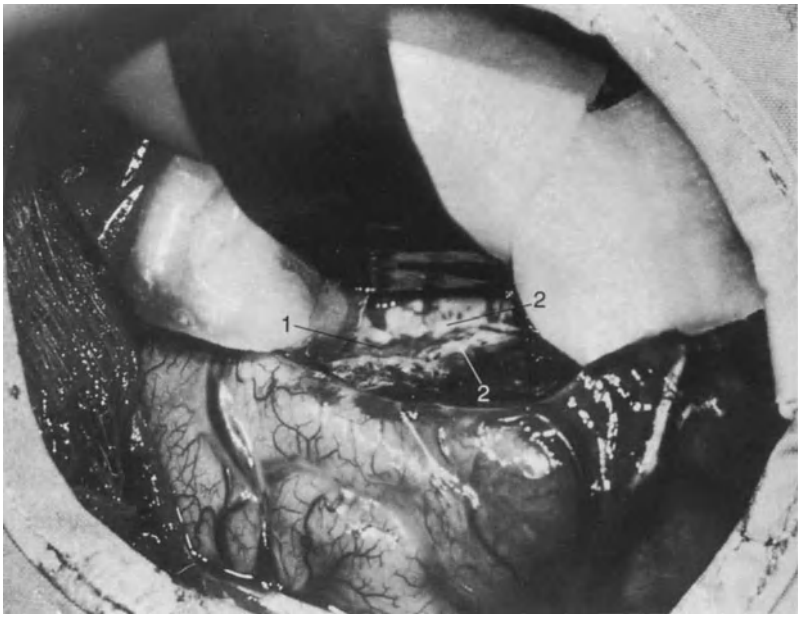
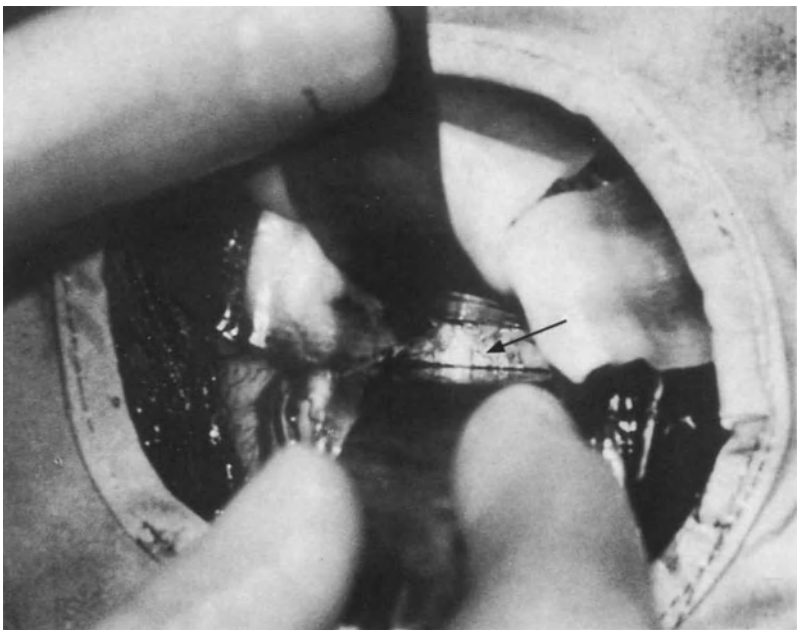


Figure 7.4. Extension of dissection through full thickness of cortex. Once all cortical vessels have been coagulated one may choose to continue the dissection into and through the white matter with bipolar forceps and fluffy cotton. However, it is less traumatic and more precise to do this with the narrow 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



E



F

depth and the parenclyma 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. (E) The superior spatula has reached the ventricular wall (*1*), which is covered on either side by white matter (*2*). (F) The second spatula has been brought into place and the subependymal covering (arrow) of the periventricular parenchyma exposed.

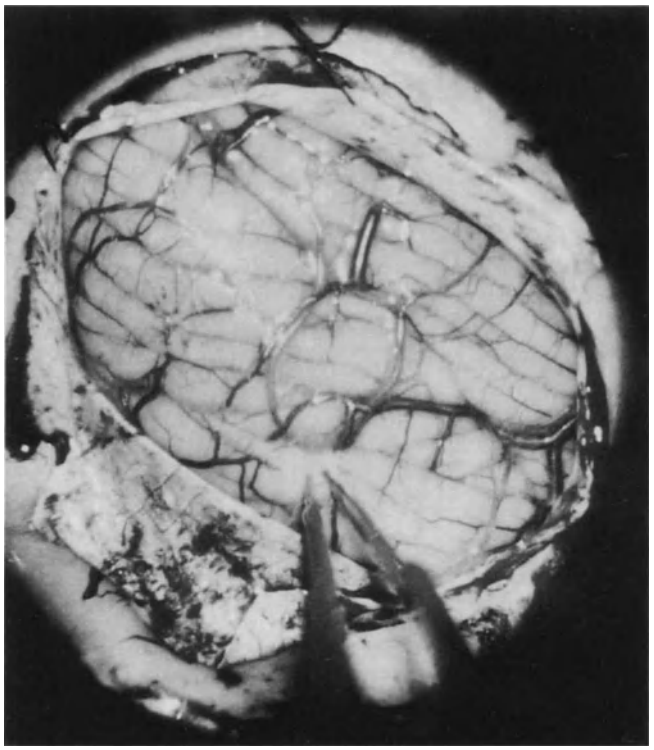


Figure 7.5. Cerebellotomy. The technique for cerebellotomy is identical to that used for the cerebrotomy, though it is not possible to place the cortical cerebrotomy precisely either in a sulcus or over a gyrus.

lature within the white matter. Consequently, it is 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 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. They are inserted either into the ventricle or over the surface of an intraparenchymal lesion.

"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

Chapter 8

Cerebral Resection

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 $1\frac{1}{2}$ to 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 a pituitary biopsy forceps, and the bed irrigated with saline and then filled with a fluffy cotton. The bleeding is stopped with bipolar cautery, Avitene, or fluffy cotton and aspiration. One should wait approximately ten minutes 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, and only after, the vascular structures have all been exposed and coagulated, may one 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 internal 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 posterior 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 di-

rection 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 best to place the line of incision, consequently, 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 best transected with spatulas, 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 spatulas; it is not as precise as microscissors in cutting arachnoid; it is not as reliable as 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 spatulas, and it does not preclude the use of spatulas, since one must retract the already incised parenchyma lest it flop back into the field.*

Frontal Lobectomy (Figure 8.1)

The bifurcation of the internal carotid artery is identified by retracting posteriorly this frontal lobe from the orbital 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 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, basel 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 the 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 of 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 stage 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 he identifies them. This allows entrance into that portion of the insular cortex beneath the olfactory, triangular, and frontal operculae (all segments of the frontal lobe), permitting the surgeon to identify 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 that nourish the insula, the parietal operculum, the temporal operculum, and the anterior portion of 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. They should be left adherent to the arachnoid. This 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 clipped and transected, cutting the arachnoid with them.

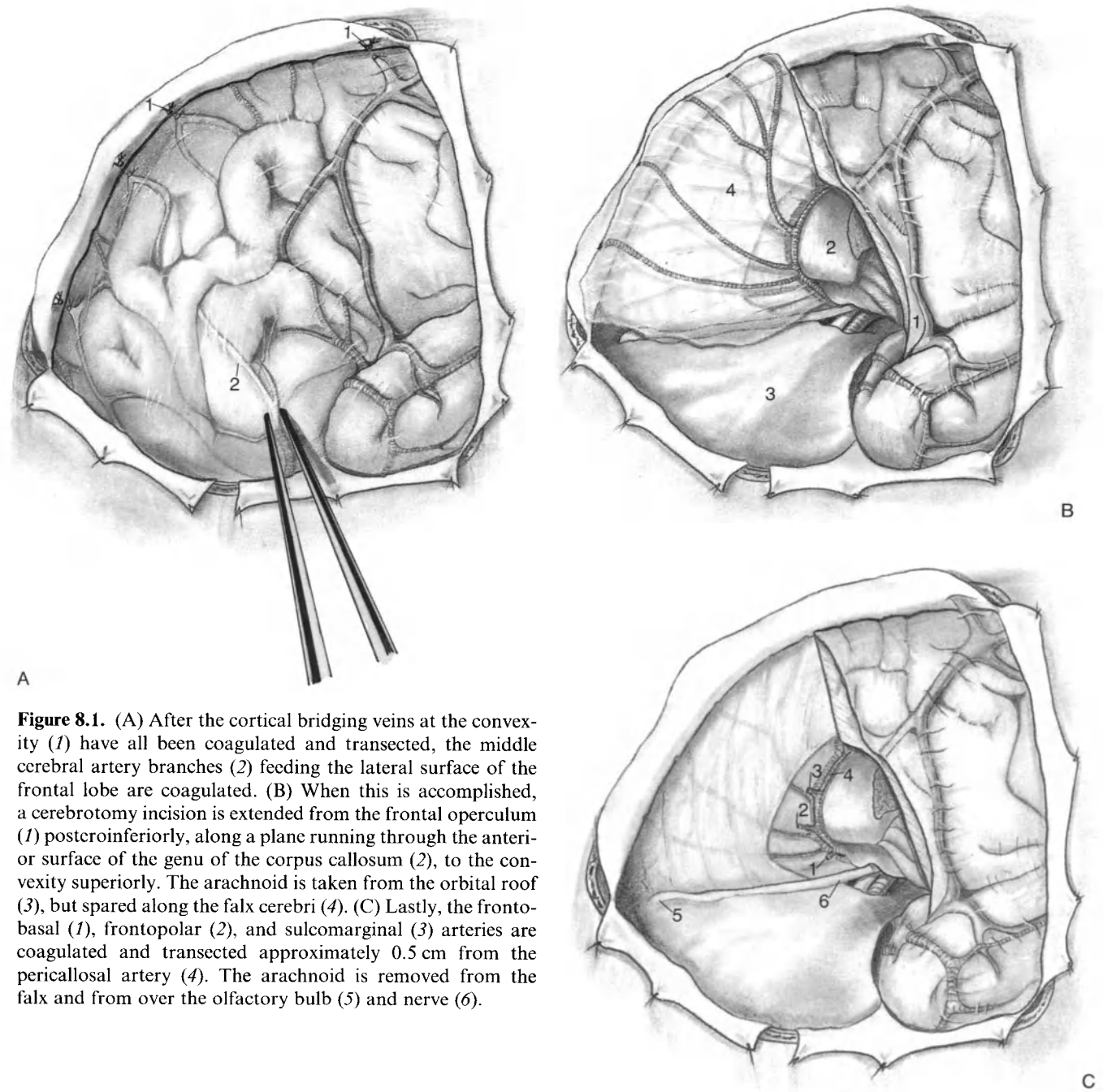


Figure 8.1. (A) After the cortical bridging veins at the convexity (*1*) have all been coagulated and transected, the middle cerebral artery branches (*2*) feeding the lateral surface of the frontal lobe are coagulated. (B) When this is accomplished, a cerebrotomy incision is extended from the frontal 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*).

Temporal Lobectomy (Figure 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 sulcus and, thus, gain entrance into 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 into the Sylvian fissure, and then over the temporal operculum and surface of that portion of the temporal lobe which borders upon the parietal lobe, one coagulates and transects the superior temporal 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 this 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 choroid plexus is identified resting within the choroidal fissure. It is coagulated at its junction with the glomus and transected, entering the ambient cistern. The coagulation of the choroid plexus is then extended anteriorly along the choroidal fissure until the anterior choroidal artery is identified, penetrating the most anterior portion of the temporal horn through the choroidal 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. This permits the surgeon to remove 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 insula and the trigone, looking at the glomus of the choroid plexus.

Occipital Lobectomy (Figure 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 at a time, extending first from inferolateral to superolateral and then from superolateral to superomedial, be-

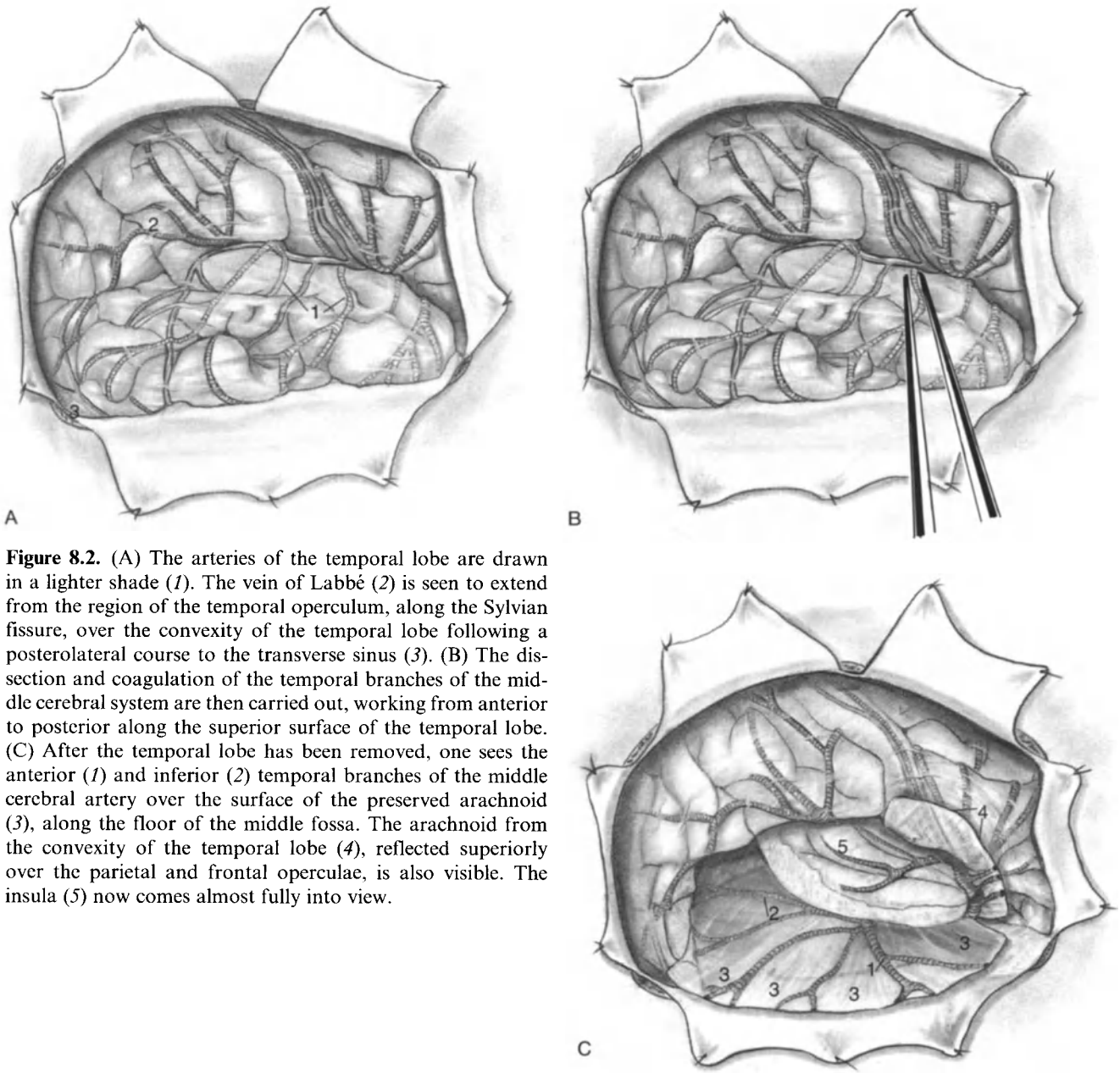


Figure 8.2. (A) The arteries of the temporal lobe are drawn in a lighter shade (*1*). The vein of Labbé (*2*) is seen to extend from the region of the temporal operculum, along the Sylvian fissure, over the convexity of the temporal lobe following a posterolateral course to the transverse sinus (*3*). (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 preserved 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.

fore 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 (Figures 8.4 and 8.5)

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 trap-door fashion and the bridging cortical veins, which extend from the superior portion of the cerebellar hemi-

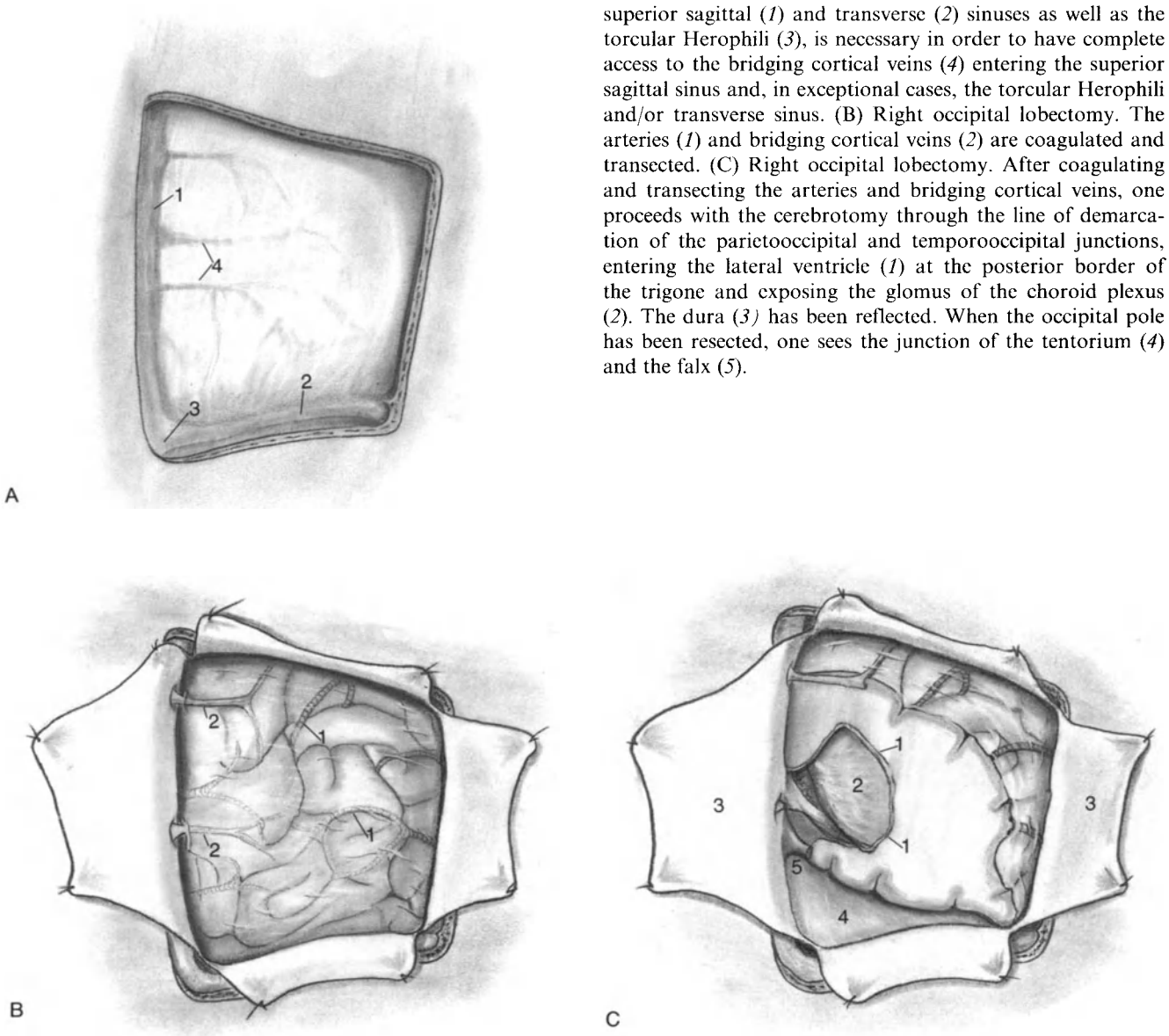


Figure 8.3. (A) Right occipital craniotomy. Exposure of the superior sagittal (*1*) and transverse (*2*) sinuses as well as the torcular Herophili (*3*), is necessary in order to have complete access to the bridging cortical veins (*4*) entering the superior sagittal sinus and, in exceptional cases, 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 parietooccipital and temporooccipital 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*).

sphere 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 then proceed fron 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 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 (Fig. 8.5). This may be done with great ease using the handheld CO_2 laser, or the micromanipulator if one has already brought the operating microscope into the field.

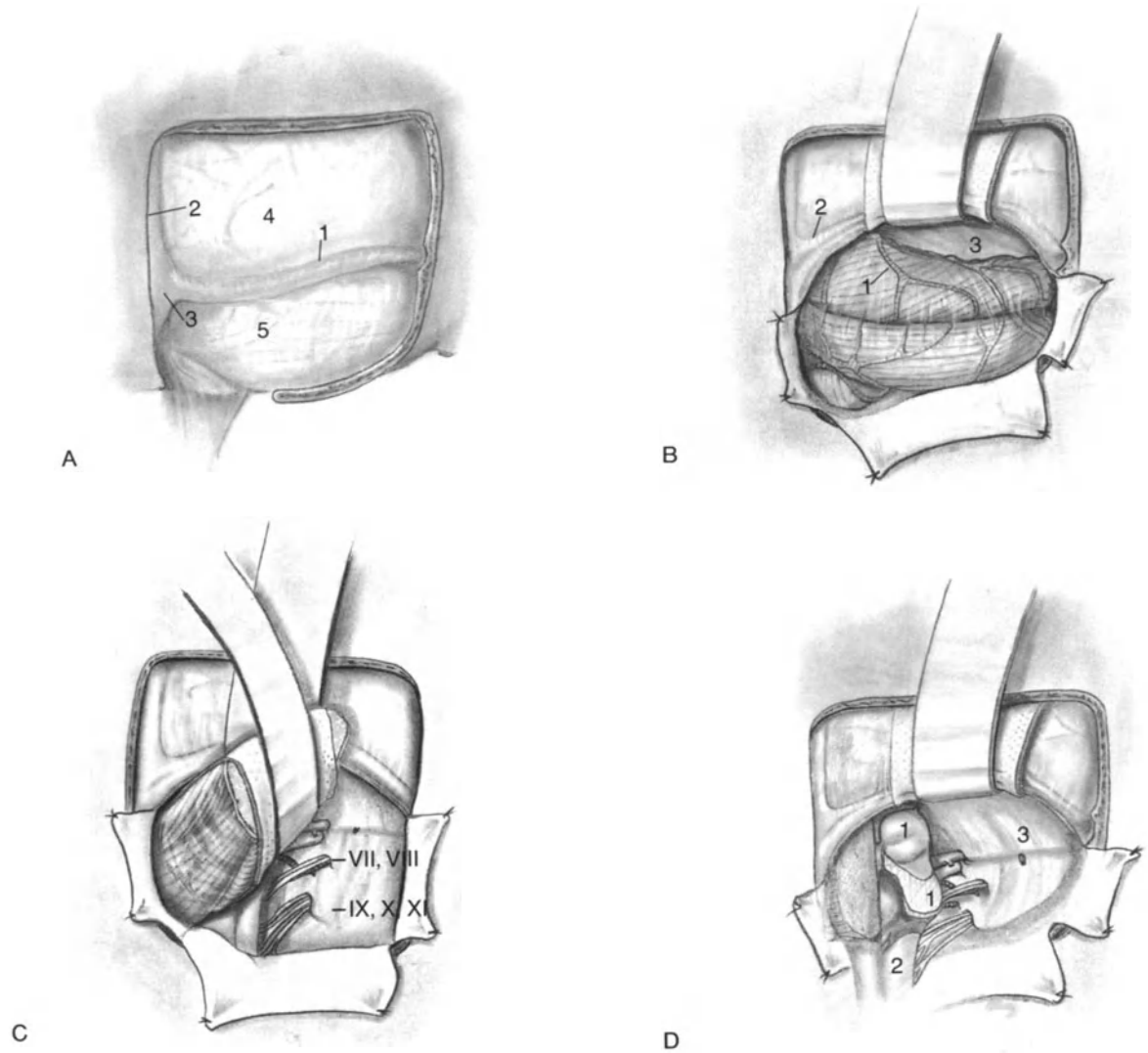


Figure 8.4. Cerebellar lobectomy. (A) The craniotomy should permit visualization of the transverse sinus (*1*), the superior sagittal sinus (*2*), the torcular Herophili (*3*), the occipital lobe (*4*), and the cerebellar hemisphere (*5*). (B) After the superior bridging veins (*1*) coursing from the 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 under surface of the tentorium (*3*) is also exposed.

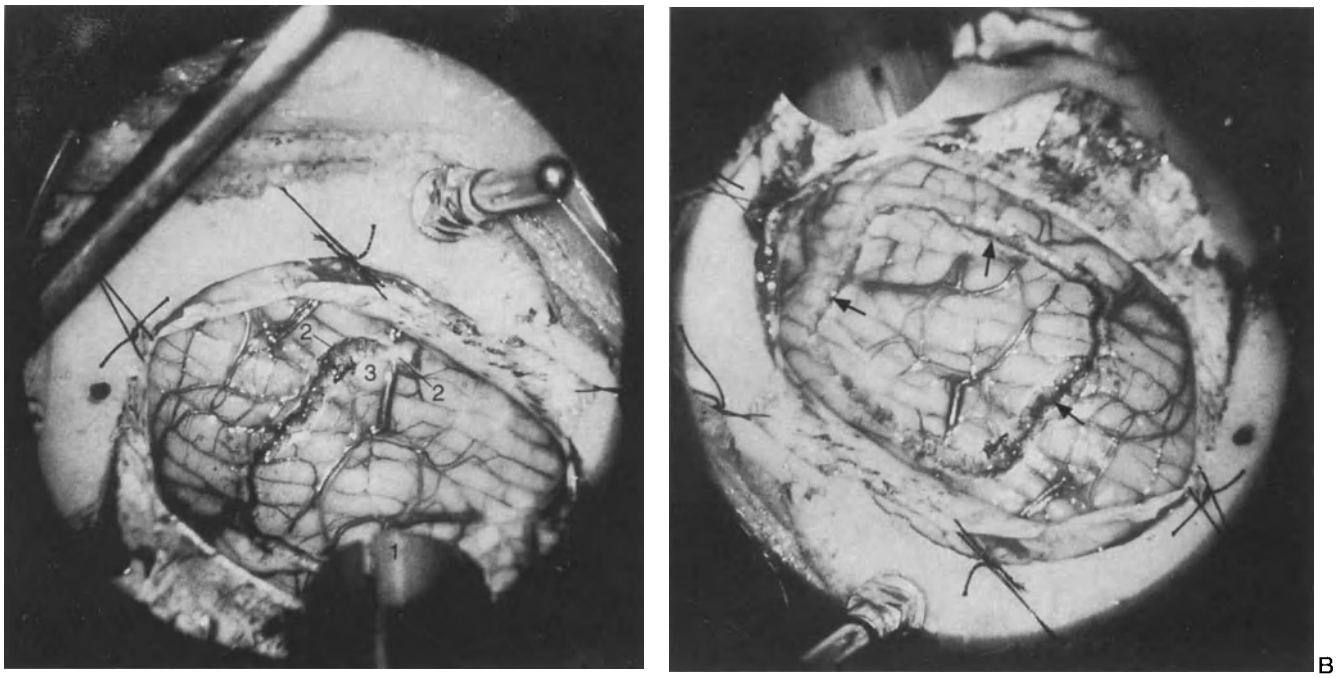


Figure 8.5. (A) The hand-held laser (*1*) is being used to perform a block resection of a 2-cm surface area of the lateral cerebellar hemisphere for access to a solid astrocytoma. The larger (>3 mm) vessels have already been coagulated with the bipolar forceps (*2*), but the smaller surface vessels (*3*) are effectively coagulated with CO_2 laser in a defocused beam. (B) The completed cerebellotomy (arrows).

Hemispherectomy (Figure 8.6)

Exposure of the entire hemisphere is essential for the performance 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 performance 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 choroidal arteries are the only remaining arterial 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 choroidal 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

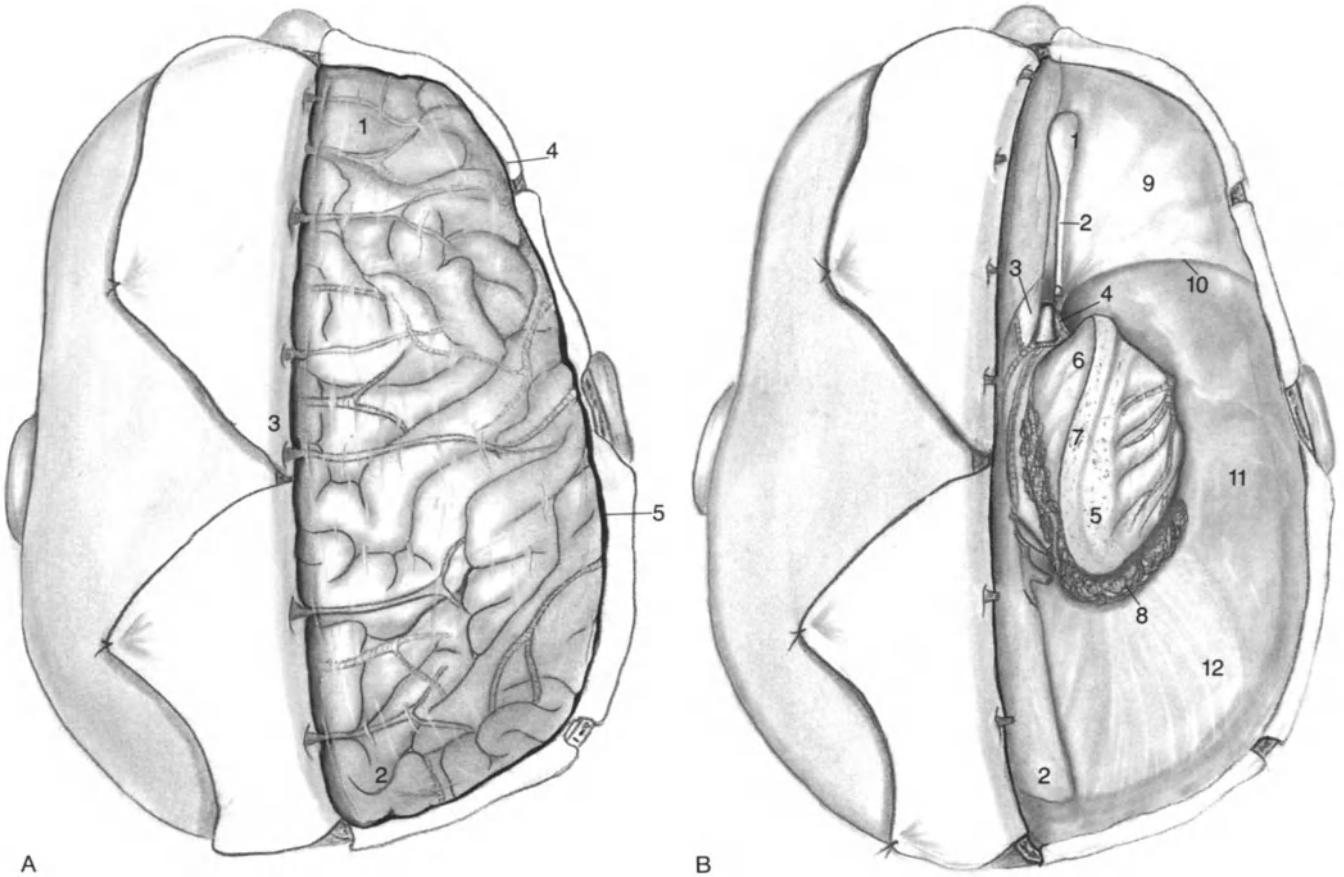


Figure 8.6. (A) When the craniotomy for a hemispherectomy has been completed, one has full visualization of the hemisphere from the frontal (*1*) to the occipital (*2*) poles, and from the superior sagittal sinus (*3*) to the lateral surfaces of the anterior (*4*) and middle fossae (*5*). (B) After removal 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*) all are 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.

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 devascularizes 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 the isthmus of the hippocampus at the posterolateral surface of the thalamus. The freed cerebral hemisphere may be lifted from the right hemicranium.

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

HOMER
The Iliad

Chapter 9

Hemostasis

Techniques for Stopping Bleeding

The technique for stopping bleeding varies not only from artery to vein to sinus, but also with the location of the artery or vein (cortical, sulcal, cisternal, etc.) and the nature of the sinus (sagittal, cavernous).

Use of Cotton Fluffies (Figure 9.1)
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 a 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. 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 three or five minutes to remove the dried fluffy with a 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
(Figure 9.2)
If this does not suffice to stop the bleeding, one uses the same technique, but applies either Gelfoam, Surgical, or Avitene,* applying one of them 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, Avitene. This is the inverse order of effectiveness. In fact, Gelfoam may be forgotten, Surgicel is only 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

* Avitene: microfibrillar collagen hemostat.

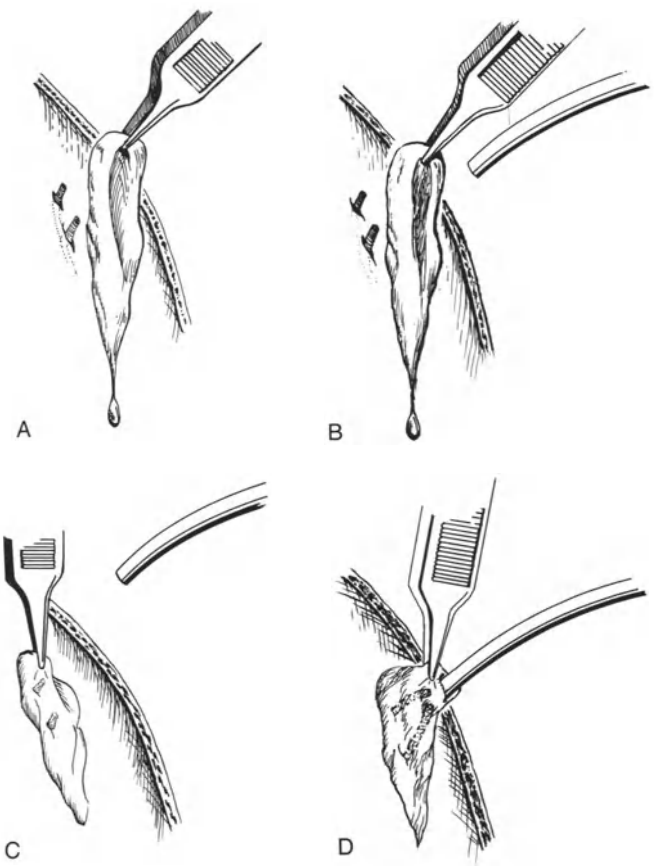


Figure 9.1. Use of fluffy cotton. (A) The soaked fluffy is brought to the bleeding surface. (B) The suction is brought into the field, but not applied to the fluffy. (C) The fluffy is pressed firmly against the bleeders. (D) The fluffy is sucked dry until it becomes glistening white, and then the forceps is released but the suction is held in place.

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 put 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 fluff of 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 do adhere snuggly 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 or dura (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 bone and periosteum, hence the high incidence of subperiosteal and epidural (diploic or inner table of skull) hematoma.

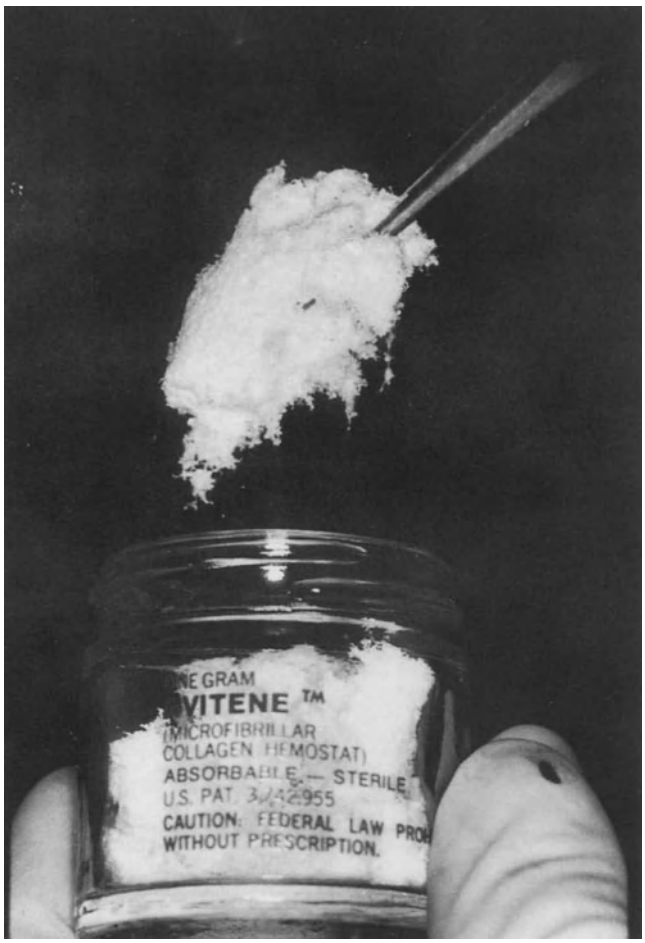
Bone Surface Bleeding

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.

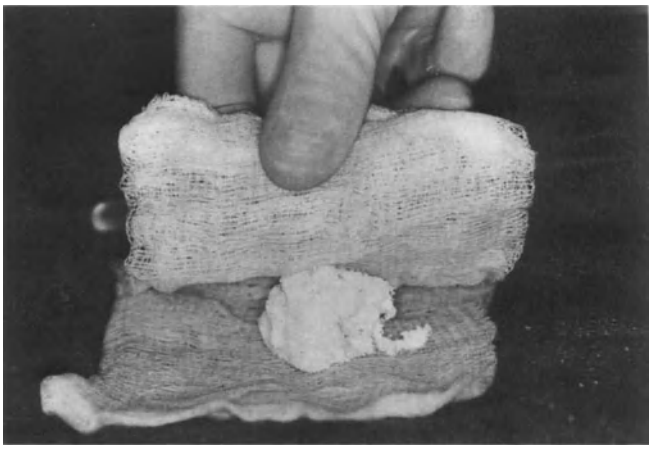
Bone bleeding is readily stopped with bone wax. It is immediate and permanent. However, the wax must be warm, so that it may be applied readily, without the need to exert force, so that it may penetrate the vascular channels of the smooth surfaces of the outer or inner tables. These tables are richly 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.

Diploic Bleeding (Figures 9.3 and 9.4)

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

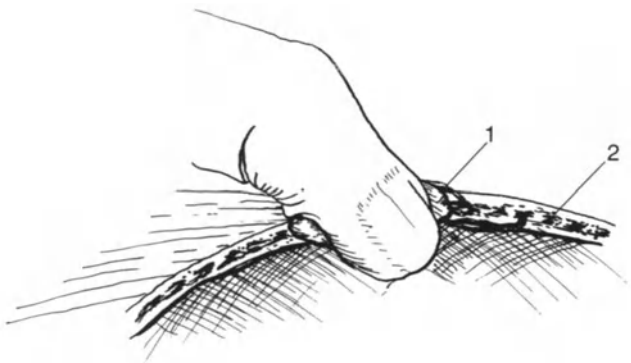


A

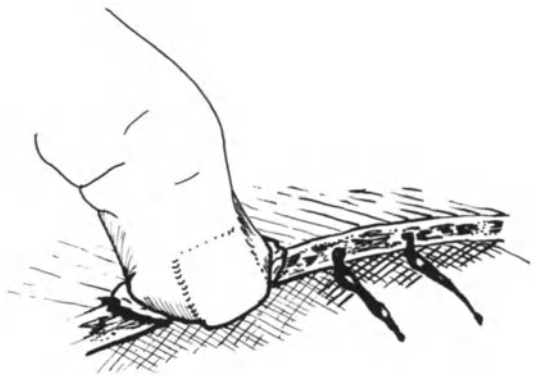


B

Figure 9.2. Use of Avitene. (A) The loosely packed masses of Avitene may be lifted directly from the sterile container. (B) If one wishes, the mass may be compressed within a soaked sponge.



A



B

Figure 9.3. 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 finger tip is pressed perpendicularly against the diploë as it passes beyond dural surface and inner table of the skull. This prevents wax from being squeezed between dura and skull.

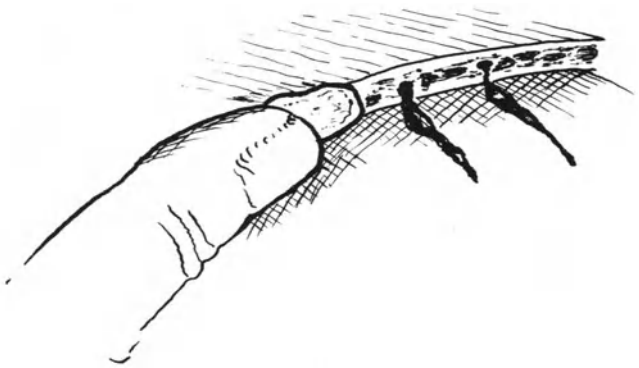
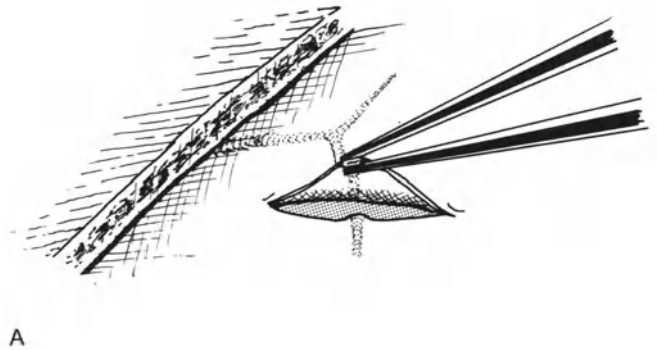


Figure 9.4. Incorrect application of bone wax. When the compressing finger is guided along the surface of the diploë, wax is jammed between dura and inner table, separating one from the other and adding to the degree of skull bleeding.



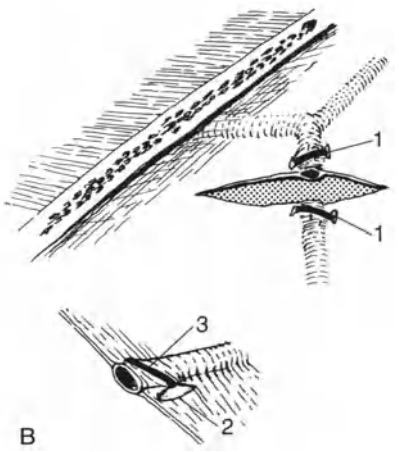


Figure 9.5. Dural arterial bleeders. (A) The smaller ones are coagulated between bipolar forceps, with the surgeon taking care to pick up the full circumference of the transected artery in both blades of the forceps. (B) Larger dural bleeders require clipping or ligature. Here, a hemoclip has been applied 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.

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 the edge of the bone as it is withdrawn, pressing the soft wax into the diploë. If the finger is parallel to the edge of the bone, or angled 10° away 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. This causes serious problems.

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), 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 (Figures 9.5 and 9.6)

The smaller arterial dural bleeders may be coagulated 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 (Figures 9.7 to 9.11)

Dural sinuses must be ligated if they are to be tran-

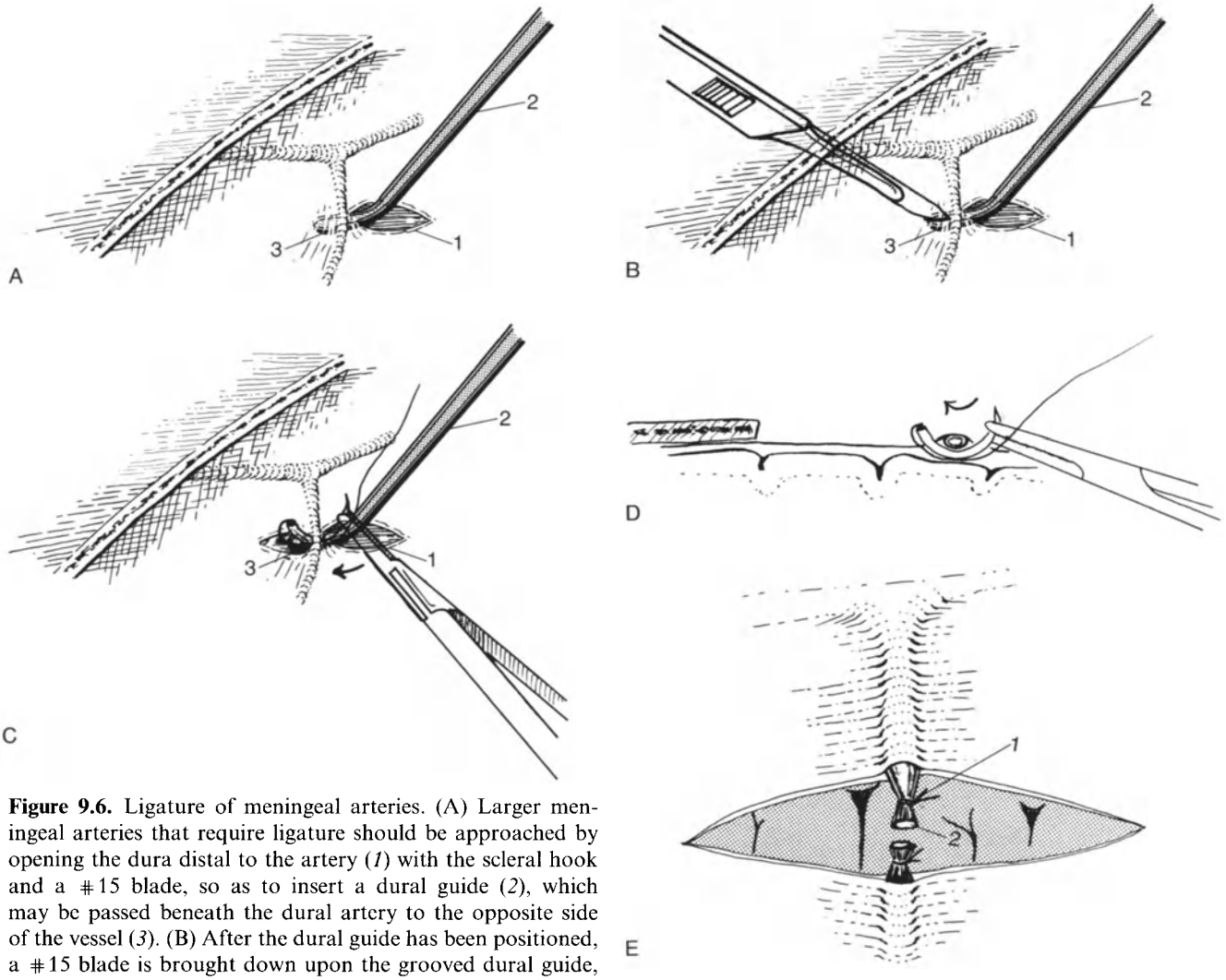


Figure 9.6. Ligature of meningeal arteries. (A) Larger meningeal arteries that require ligature should be approached by opening the dura distal to the artery (*1*) with the scleral hook and a #15 blade, so as to insert a dural guide (*2*), which may be passed beneath the dural artery to the opposite 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 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*).

sected! 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 "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 open-

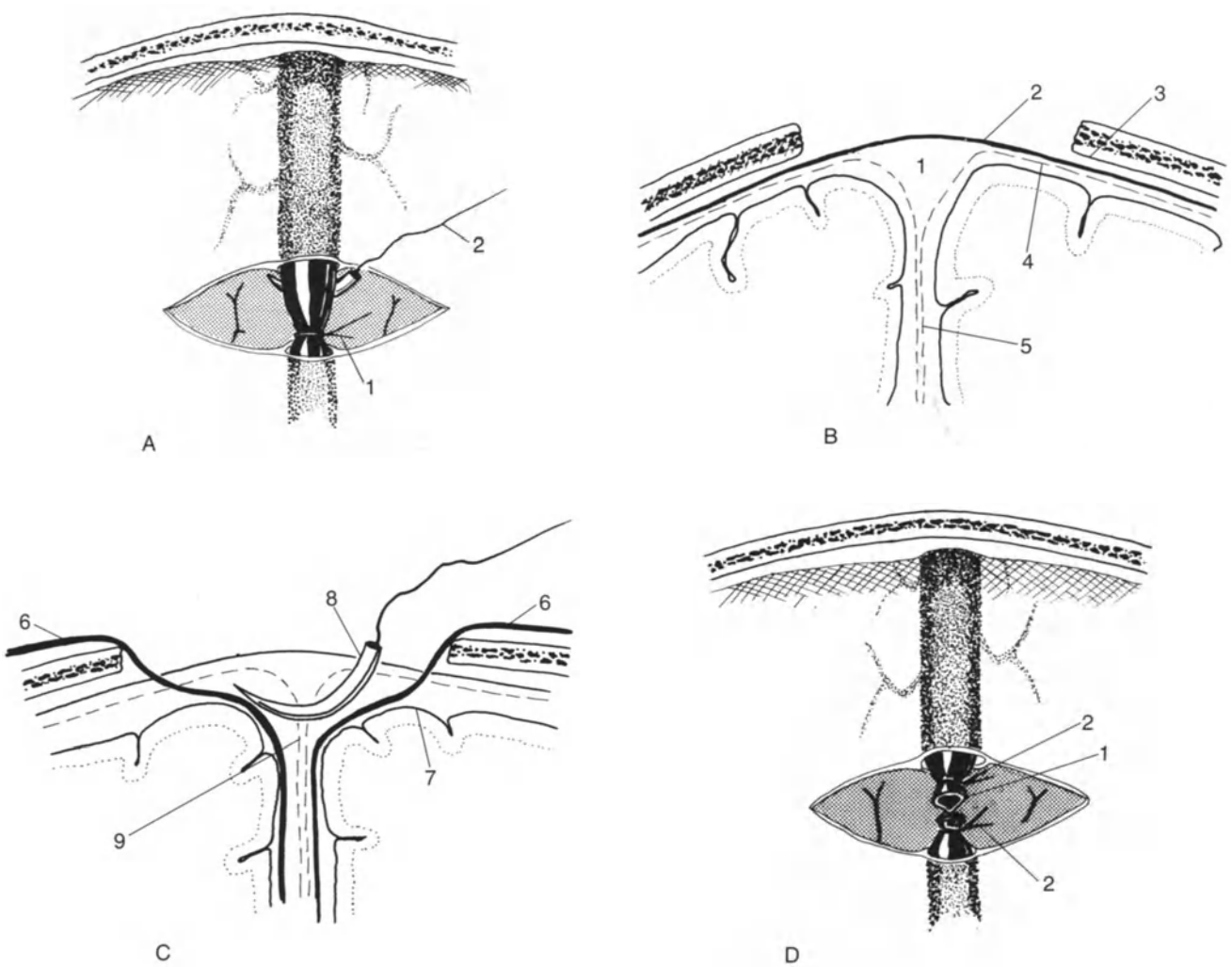


Figure 9.7. Ligature of dural sinus. (A) After the dura has been opened to the very edge of the dural sinus and either the falx or tentorium cerebri identified and exposed bilaterally, a 3–0 or 2–0 suture is passed beneath the apex of the dural sinus. The ligature is tied down (*1*). A second ligature is passed about 3 to 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 superior sagittal sinus (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.

ing. 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 to 10 minutes. 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.

Cortical Bleeding (Figure 9.12)

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 touches 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 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 great 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 stready, not snapping, motion. This avoids slippage of one blade of the clip over the other. Removal may be complicated 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 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

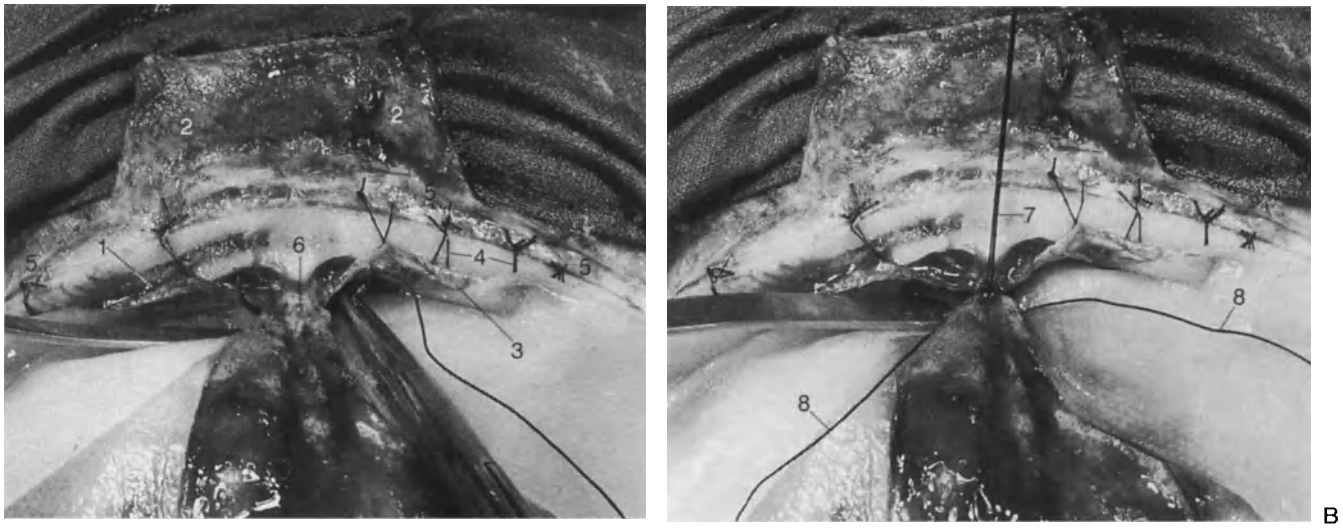
Cortical veins, sulcal veins, and cisternal or bridging veins require different techniques for rapid and precise coagulation and transection.

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.

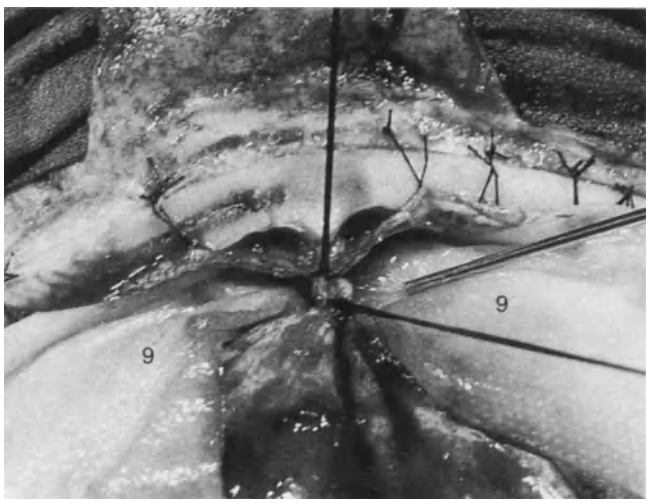
Sulcal Veins

Sulcal veins, on the other hand, fish freely within a small amount of cerebrospinal fluid in the subarachnoid



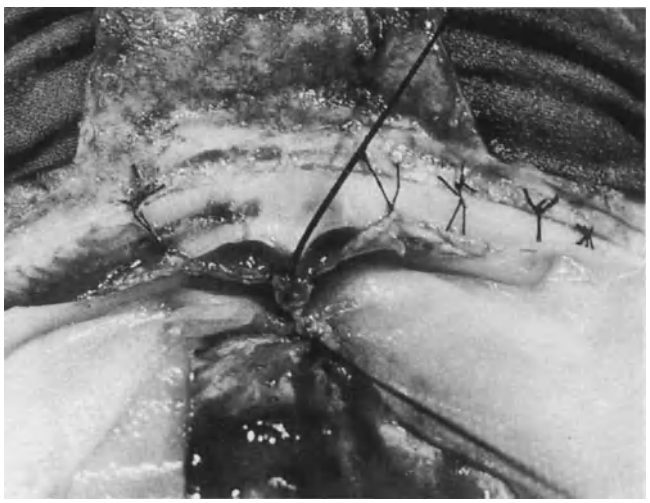
A B

Figure 9.8. The various steps in double ligature and transection of the superior sagittal sinus (SSS) at the crista galli. (A) In this series one also notes that the left frontal air sinus (*1*) is already developed, so a flap of periosteum (*2*) was prepared to wall it off from the epidural space at the time of the closure. The dura (*3*) is tacked (*4*) to the periosteum (*5*) before proceeding to ligating the SSS (*6*). (B) One ligature (*7*) has already been tied, and the other (*8*) passed through the falx cerebri immediately beneath the SSS. (C) Both ligatures have been tied and telfa (*9*) lain over the frontal lobes. (D) A #15 blade is used to cut across the ligatured SSS, as gentle traction is exerted upon the ligatures. (E) The SSS has been transected. (F) The falx cerebri (*10*) is identified and then cut, with either tenotomy or Metzenbaum scissors, from its attachment to the crista galli, freeing dura and SSS so that they may follow the cerebral hemispheres. The dural insertion onto the crista galli is then coagulated with bipolar forceps. (G) Once the falx cerebri is separated from its attachment to the crista galli, the dura (*1*) follows the frontal lobes (*2*) as these latter settle inferiorly.

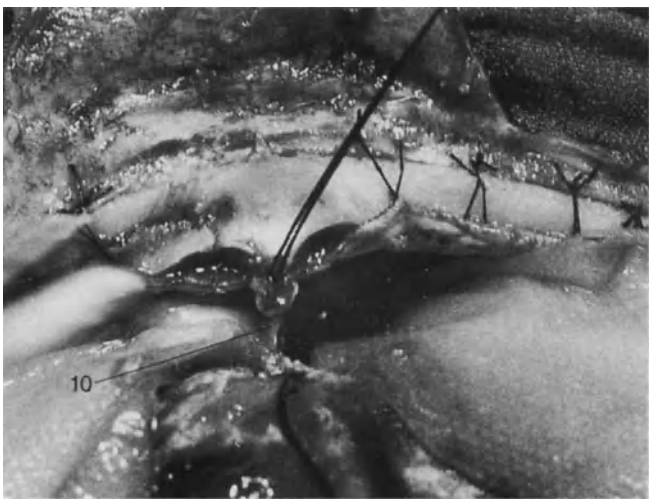


C

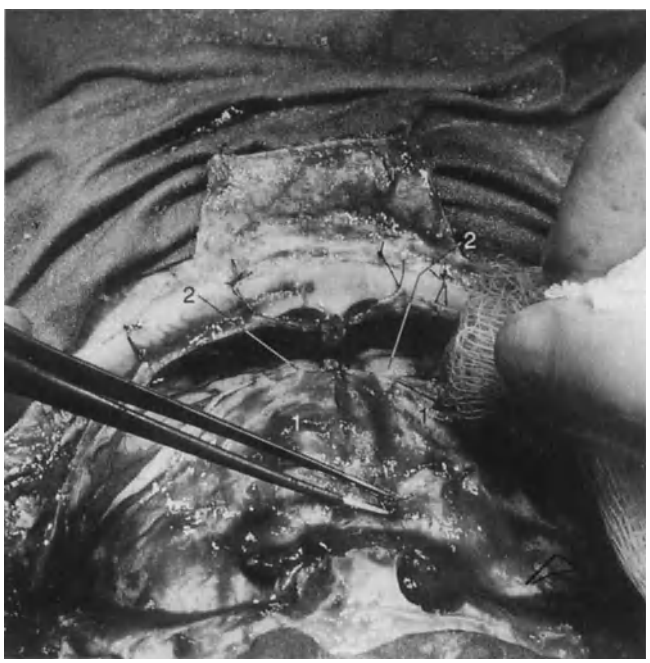
D



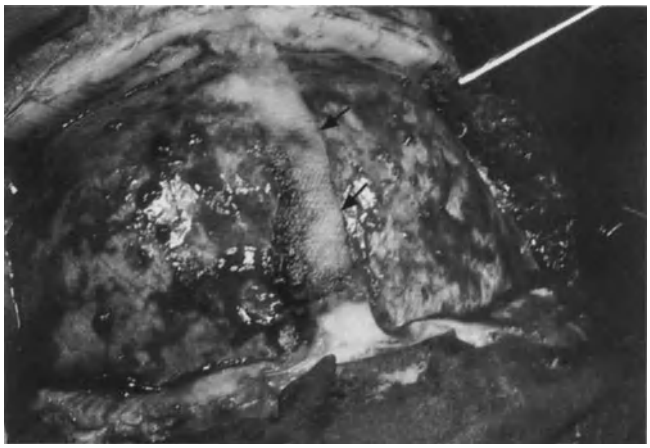
E



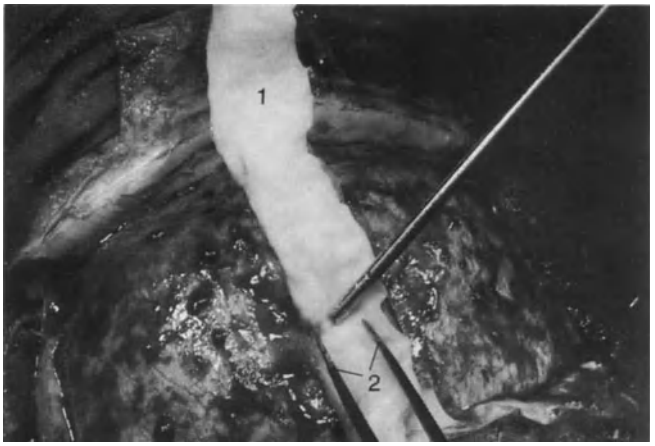
F



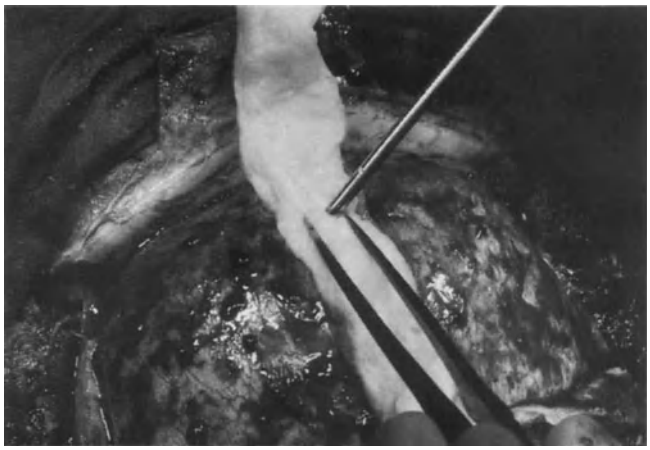
G



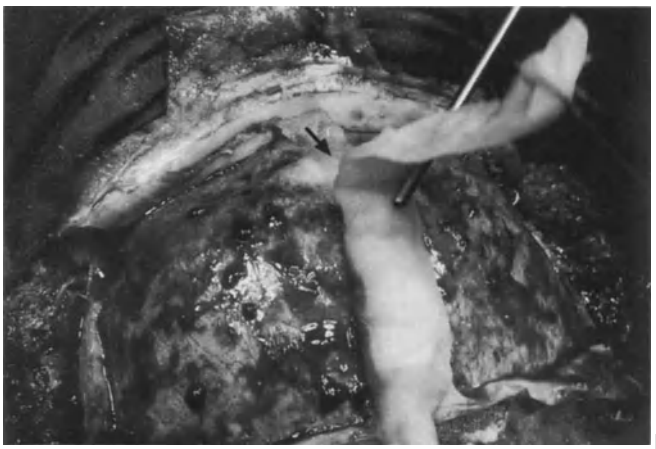
A



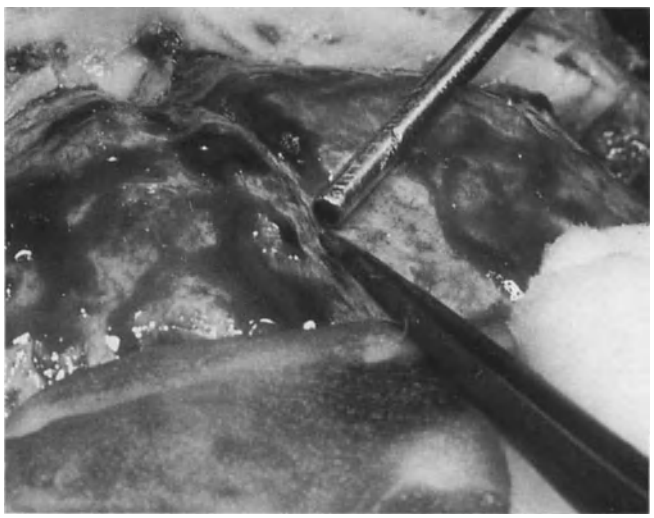
B



C



D



E

Figure 9.9 Repair of sinus: sandwiches of Surgicel and cotton fluffies. (A) A Surgicel "sandwich" (arrows) has been lain over the oozing SSS. (B) Soaked fluffy (*1*) is compressed over the "sandwich" and is being held firmly against it with spread bipolar forceps (*2*), taking care to lay them parallel to the sinus. (C) The soaked fluffy is sucked dry, applying the sucker between the flatly placed blades of the bipolar forceps. (D) Once the bleeding has stopped, the dried fluffy is removed by *rolling* it away from the underlying "sandwich." This allows the surgeon to assure adherence of the Surgicel (or Avitene) to the underlying sinus (arrow). (E) Small sinus bleeders may be stopped with a bipolar forceps by sucking the blood directly from the bleeding site and coagulating the rent with the forceps laid, blades slightly opened, over it.

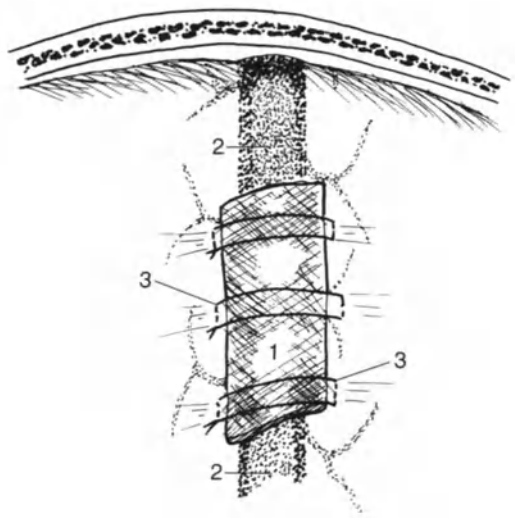


Figure 9.10. A Surgicel-Avitene "sandwich" (*1*) has been laid over the oozing or bleeding sinus (*2*) and then anchored into place with sutures passing over it from the dura on one side to that on the other (*3*).

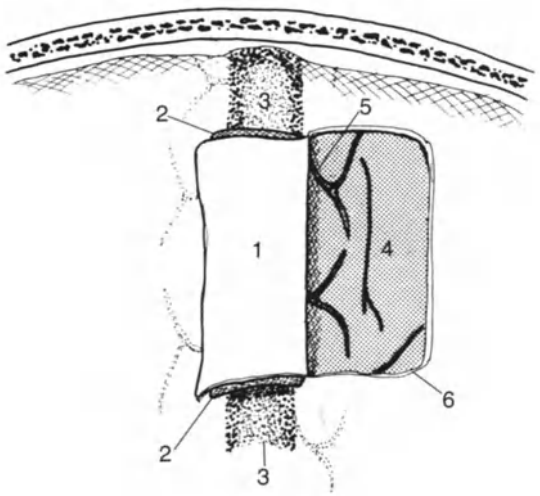
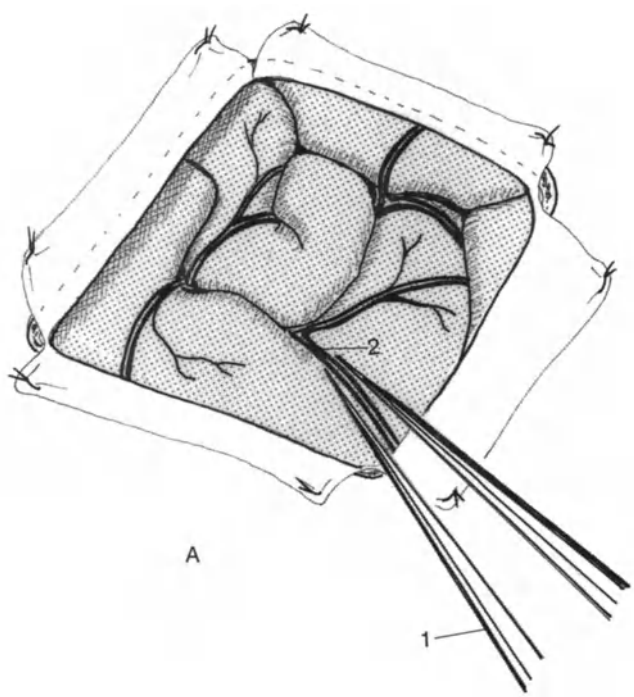


Figure 9.11. In this illustration, a dural flap (*1*) has been sewn over an underlying Surgicel-Avitene "sandwich" (*2*), compressing it against the sinus (*3*). The underlying cortex (*4*), cortical bridging veins (*5*), and edge of dural donor site (*6*) are illustrated.



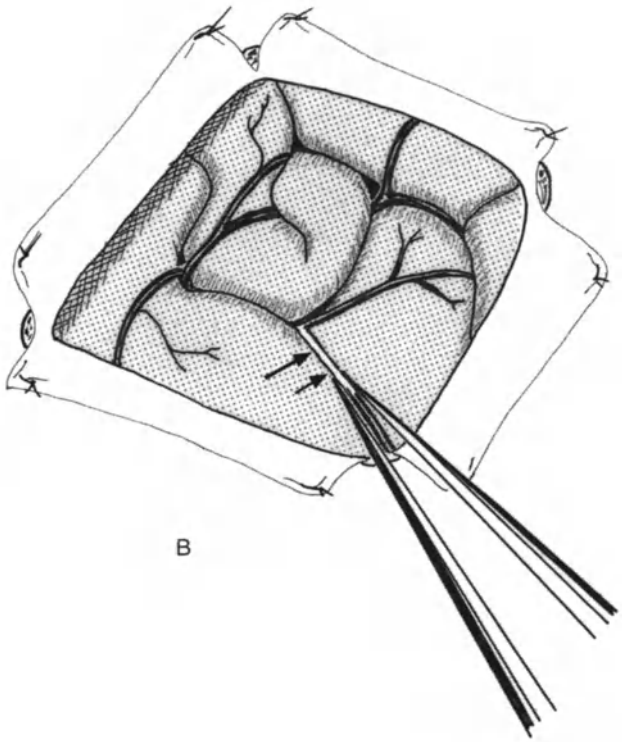


Figure 9.12. (A) The forceps (*1*) are placed parallel to the axis of the vessel (*2*), without being closed completely. (B) The cortical vessel is coagulated over a 3- to 5-mm length (arrows) until it is converted into a yellowish band. This coagulated strip should then be transected and each stump coagulated until the edges have sealed.

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 vein 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 to 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.

Cortical Bridging Veins (Figures 9.13. to 9.15)

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. 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 surface rather than run the risk of attempting the coagulation either over the main segment of 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 to 6 mm, the constriction of the vein should be 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 to 5 mm. Once the vein is completely occluded it may be transected by using microscissors.

Choroid Plexus (Figure 9.16)

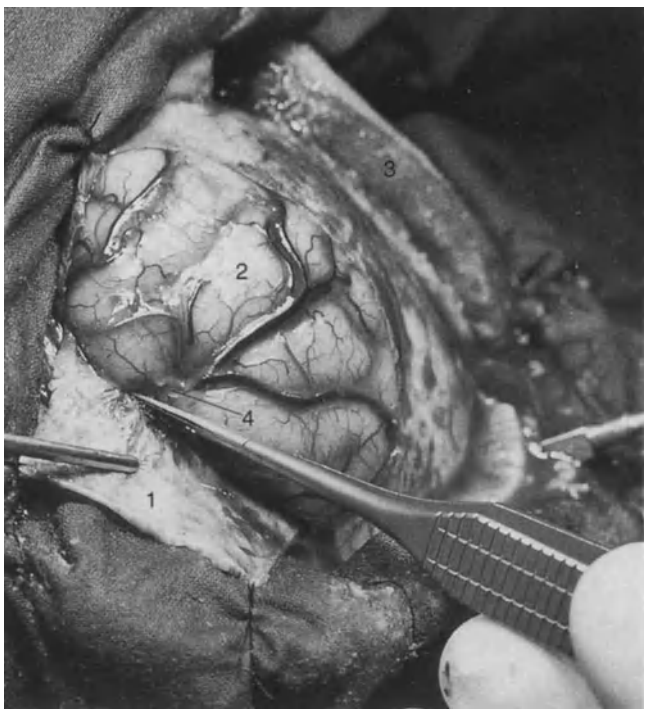
Choroidal tissue may be safely dissected from some portions of the ventricular surface, as in the area of adhesion of the tela choroidea of the III ventricle, along the surface of the thalamus at the junction between lateral and III ventricle.

This is not true for the 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 cerebral 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.

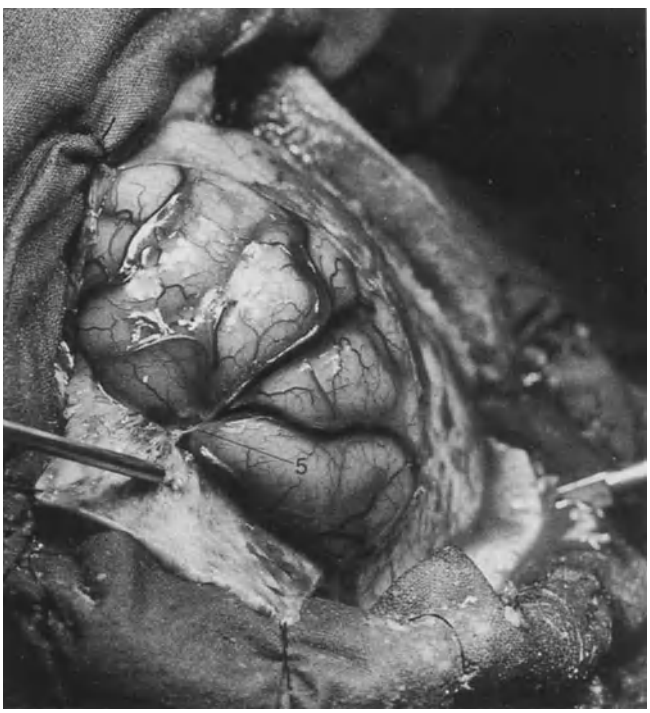
Bleeding from the surface of the choroid plexus is readily stopped by applying soaked (cotton) fluffies and sucking them dry once they have been firmly pressed against the bleeding site, or by coagulating the plexus. Coagulation of the plexus offers no unusual problems. It is performed by applying the 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

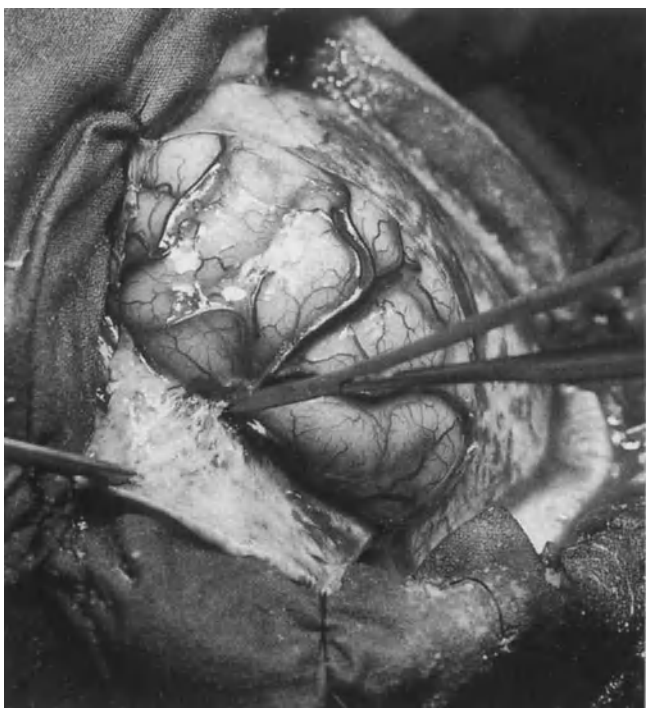
Stopping bleeding from tissue calls for a different technique from that used in coagulating, clipping, or the



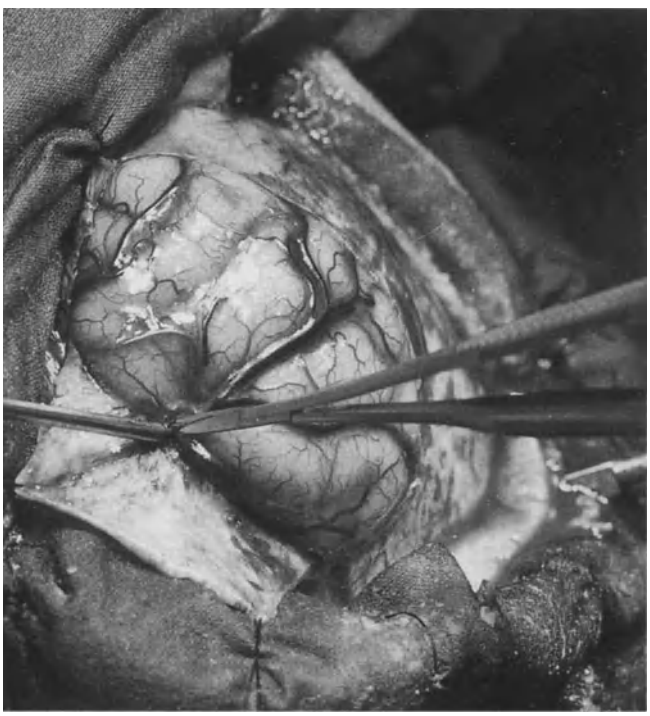
A



B

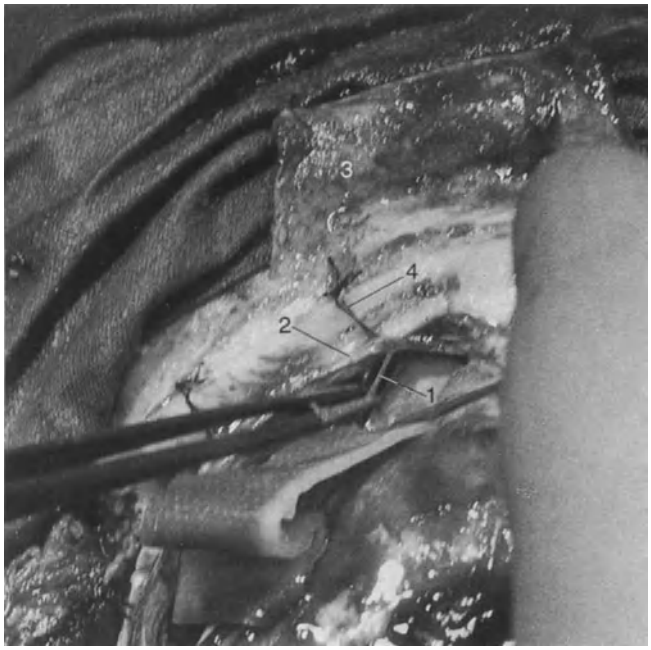


C



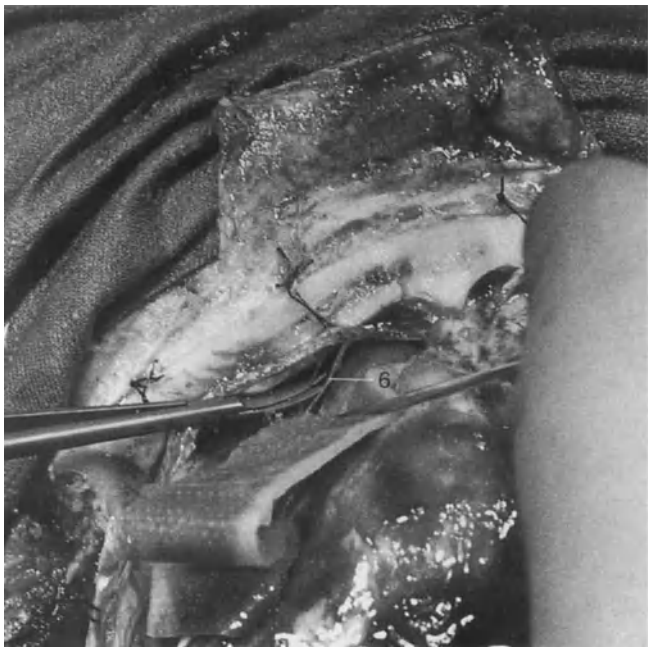
D

Figure 9.13. 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 bridging cortical vein (*5*) has been coagulated, converted into a fibrous band. (C) Microscissors are used to transect the coagulated bridging cortical vein. The blades are placed on either side of the vein. (D) Then 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.



A

B



C

Figure 9.14. Coagulation and transection of a frontopolar bridging cortical vein. (A) The vein (*1*) has the forceps blades touching its surface. Note the frontal air sinus (*2*) and the dural flap (*3*), as well as the tackup sutures (4). (B) The bridging vein has been coagulated into a fibrous band (*5*). (C) Microscissors, curved, cut the coagulated vein at its center (*6*). One must always see both tips of the scissor blades while placing them *and* when cutting!!

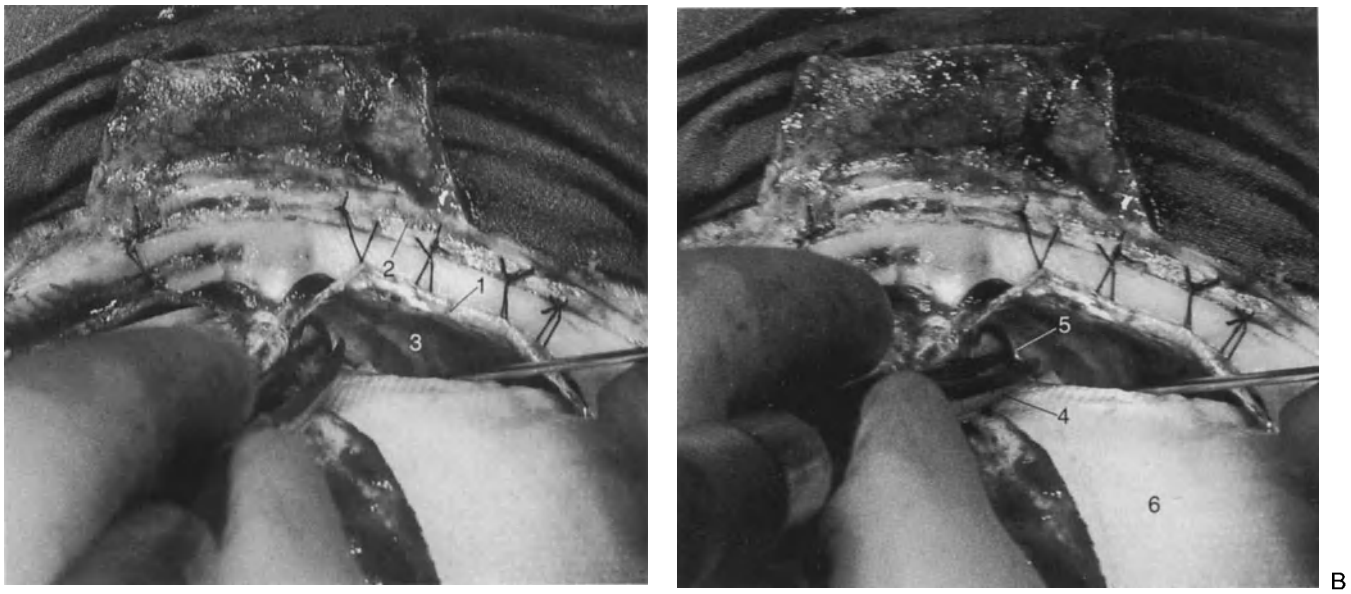


Figure 9.15. Coagulation of a bridging frontobasal vein. (A) The dura (*1*) has been sewn to the periosteum (*2*), and the most anterior portion of the anterior fossa (*3*) is visible. (B) the biopolar forceps (*4*) are applied to the surface of the bridging frontobasal vein (*5*). Telfa (*6*) protects the surface of the frontal lobe.

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 (Figures 9.17 and 9.18)
Bleeding from the Galea may be stopped by using a continuous interlocking full-thickness skin suture preoperatively, running it approximately 1 to 1.5 cm distal to the planned suture line (as already described). This has the disadvantage of also occluding the Galeal vessels. Consequently, at the time of closure Galeal vessels must be identified and coagulated. If this is not done, they may retract out of site and only temporarily occlude themselves with an intraluminal clot, which may subsequently come free and cause hemorrhage.

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

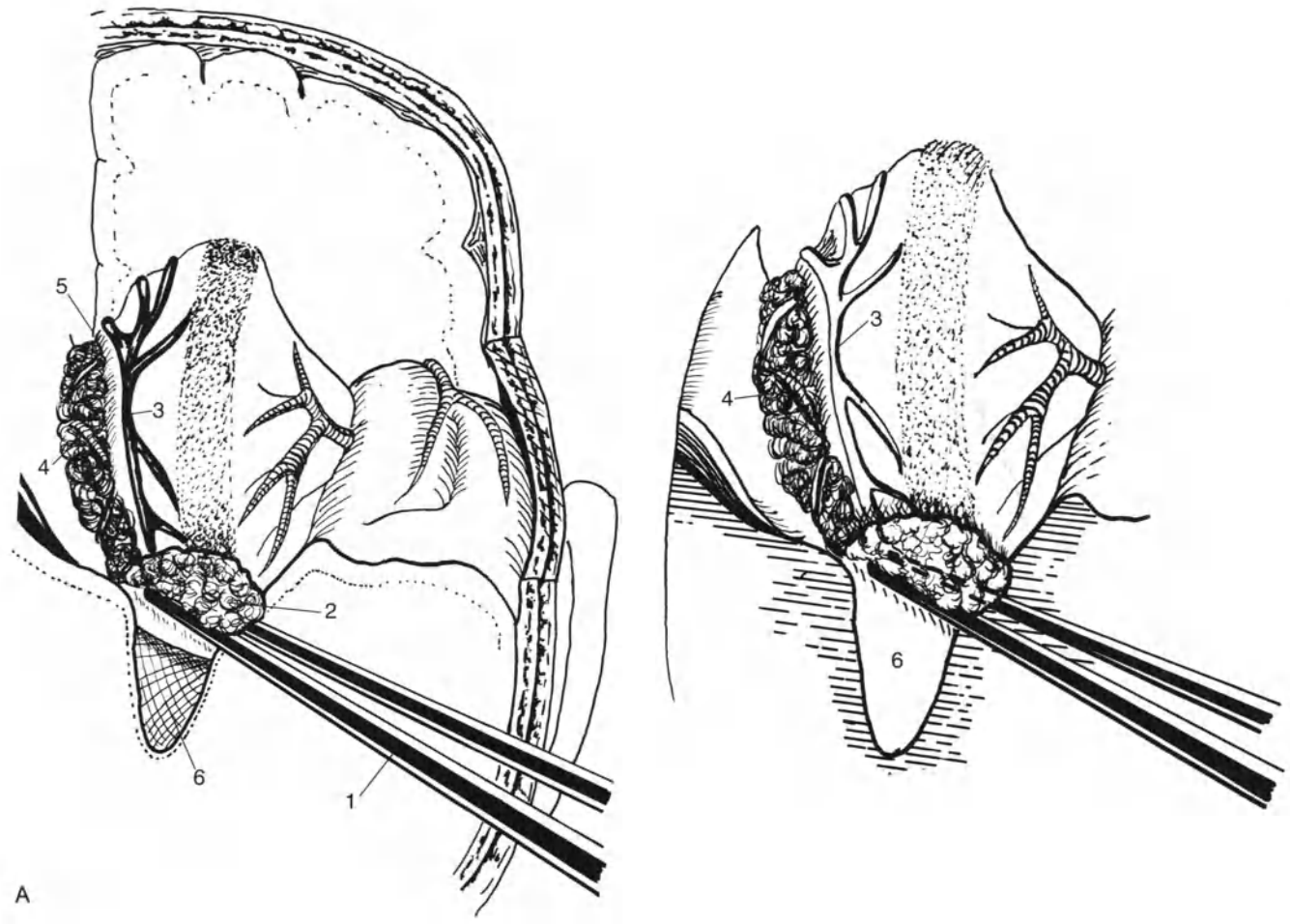


Figure 9.16. Coagulation of choroid plexus. (A) The forceps (*1*) are applied to the surface of the glomus (*2*) and the coagulation begun. One avoids coagulating the terminal vein (*3*) running in the terminal sulcus. If the surgeon chooses, he may extend the coagulation along the entire choroid plexus within the lateral ventricle (*4*), up to the foramen of Monro

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.

Muscle Bleeding

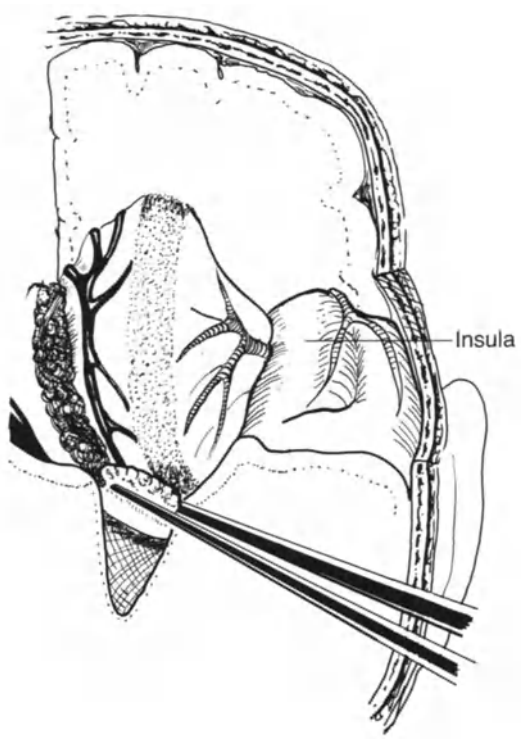
Muscle bleeding is generally stopped by gentle compression, no more. However, especially in the erector capiti group, large arterial or venous bleeders retract deeply into the muscle belly, rendering identification and coagulation tedious and difficult. Those bleeders that have retracted into the erector capiti and erector cervicis muscle groups must be searched out, found, and coagulated. This is not true for temporalis muscle bleeders, since the temporalis, a bipenniform muscle, has 3 fascial planes entrapping the 2 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.

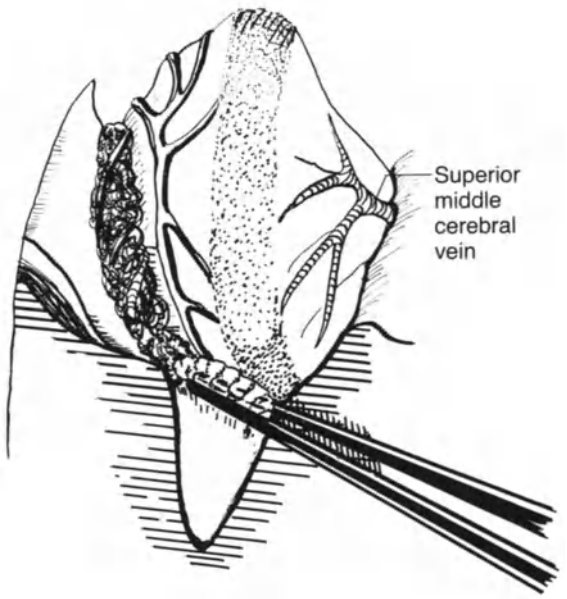
Parenchymal Bleeding

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



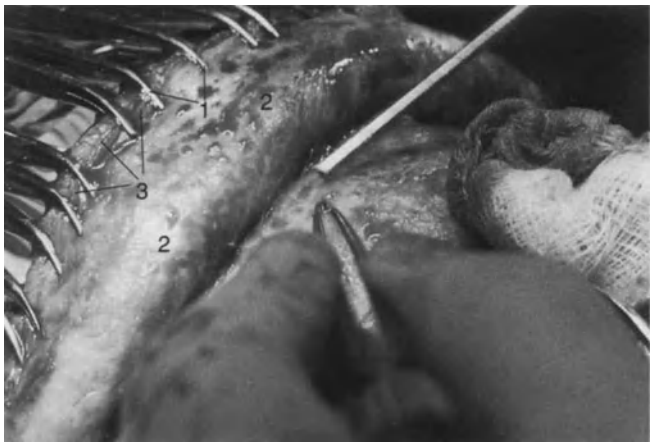


B

(*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 snugly the choroid plexus between them. The coagulation is continued until the bleeding stops, and a plane of coagulated tissue may be identified for sectioning with microscissors.

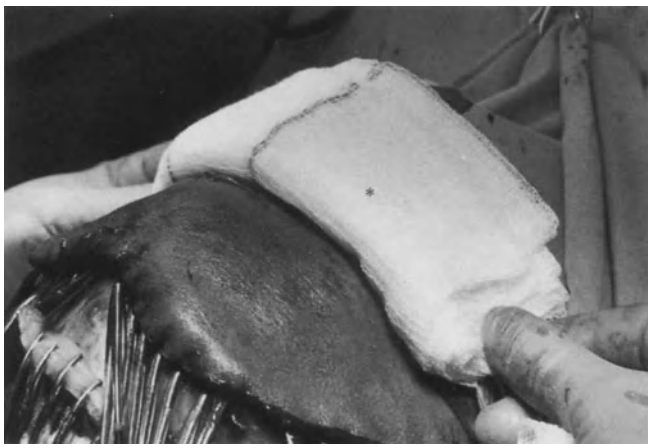
raw, bleeding, tumor surface. Consequently, identification of bridging vessels from the surrounding tissue to the neoplasm should be undertaken by skirting the circumference of the neoplasm, using fluffy and telfa to dissect parenchyma from neoplasm, and coagulating the bridging vessels as one proceeds. 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 subependymal 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.

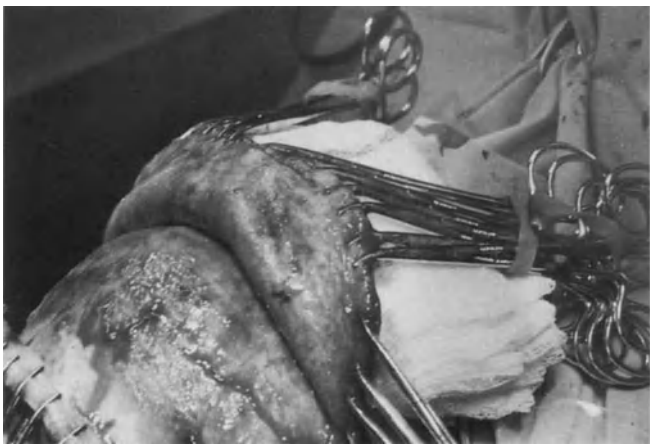


A

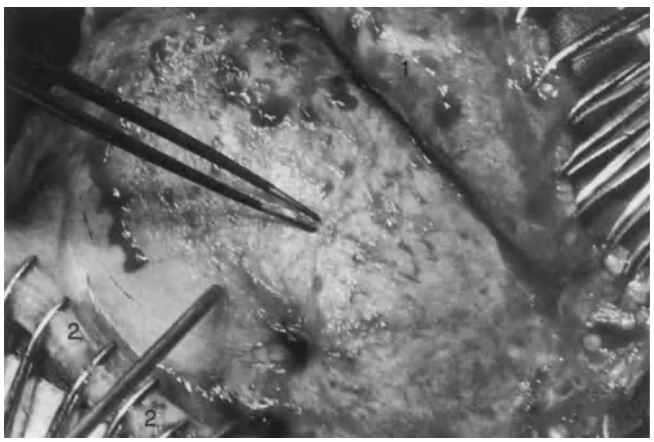
B



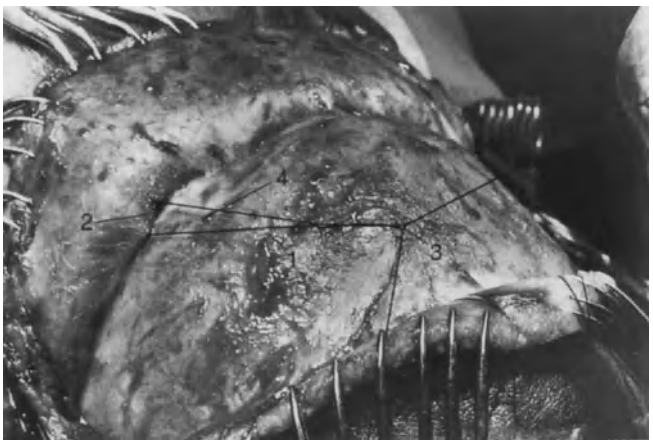
C



D

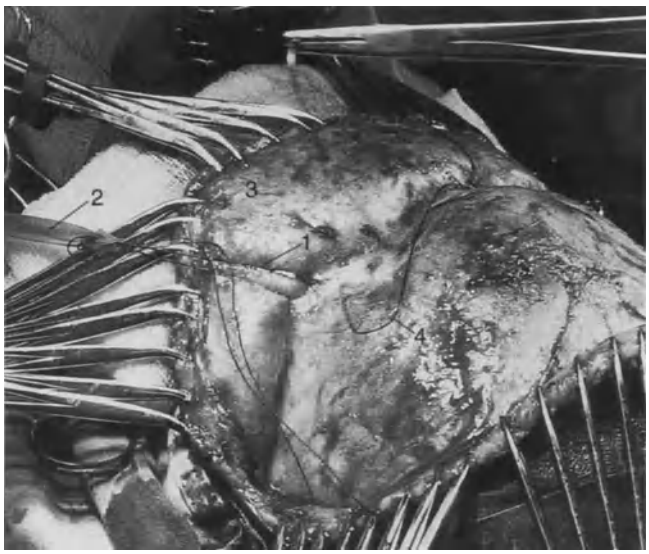


E

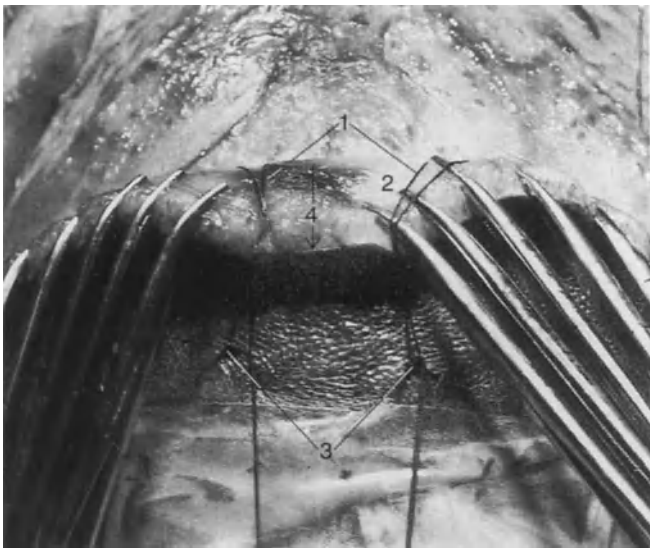


F

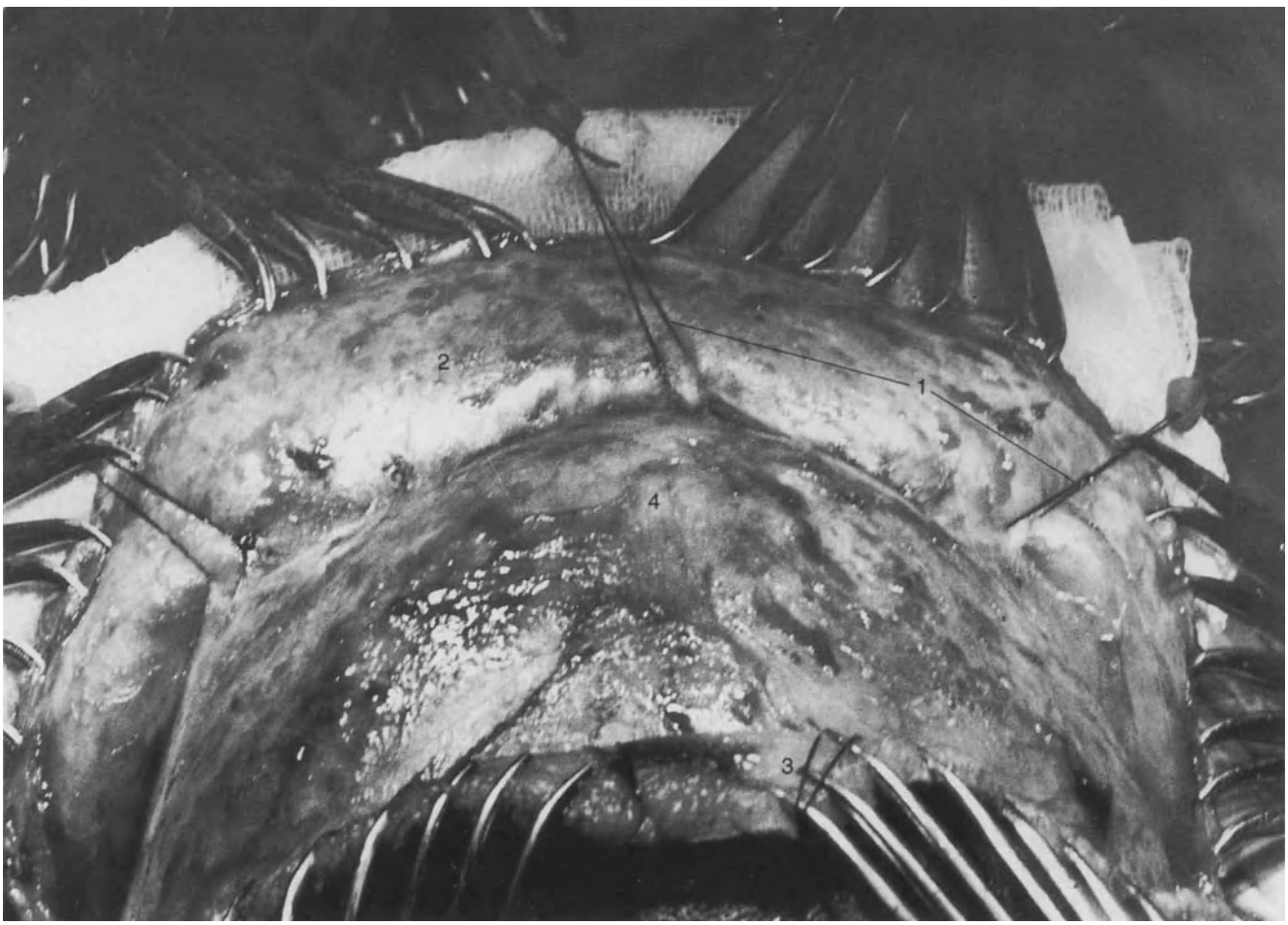
Figure 9.17. Galea. (A) Fine-toothed clips (*1*) have been applied to the cut Galea (*2*) along the edge (*3*) of the scalp flap. (B) The cutaneous surface of the scalp flap is now ready to be folded over itself. (C) A roll of sponges (*) are laid along the supraorbital ridge. (D) Then the scalp flap is rolled over it, resulting both in compression of small scalp vessels and prevention of scalp necrosis. (E) After bleeding from both edges (*1, 2*) of the reflected Galea has been stopped, the periosteal bleeders are coagulated. (F) Retention sutures (*1*) are placed into the dense connective tissue and Galea (*2*), and then a "dead knot" is tied (*3*), thus leaving a loop in the suture (*4*) through which a rubber band may be passed. (G) The retention suture (*1*) has been engaged by a rubber band (*2*), stretching the pedunculated scalp flap (*3*). A second retention suture (*4*) has been placed, but not tied. (H) Retention sutures (*1*) are passed through the Galea into the dense connective tissue of the anchored flap (*2*) and then downward through the scalp (*3*). When tied down, the anchored edge of the flap is retracted inferiorly (*4*). (I) The retention sutures (*1*) sewn into the pedunculated flap (*2*) and the anchored flap (*3*), when retracted in opposite directions, expose the entirety of the frontal bone (*4*).



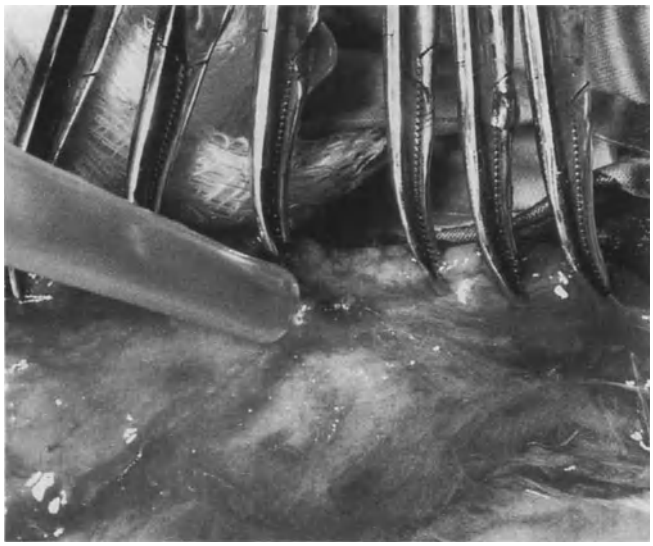
G



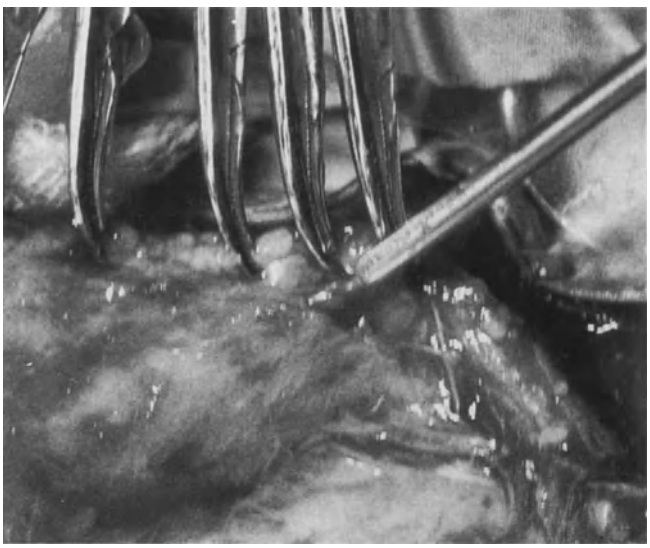
H



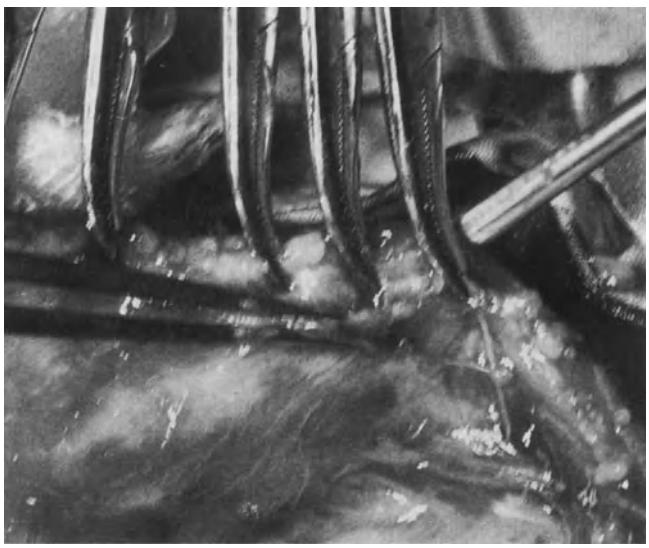
I



A



B



C

Figure 9.18. Identification and coagulation of bleeders along the undersurface of the Galea. (A) The bleeder is identified, and its location confirmed by the use of irrigation. (B) The bleeding vessel is aspirated into the sucker. (C) The sucker is removed after the bipolar forceps have been used to occlude the vessel. If there is no bleeding, the coagulation is activated and the bleeding vessel occluded.

"Hands, hold you poison or grapes?"

DYLAN THOMAS—
Ears in the Turrets Hear

Chapter 10

Tumors: Approach and Removal

Bone Tumors

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 are invariably 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 osteoma rarely grows from the inner table of the skull into the intracranial compartment, almost invariably expands from the outer table of the skull as a relatively smooth, broad-based, bony-hard excresence 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, as fibrous dysplasia, involve all three layers of the skull and may be tender to touch. The histologic 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 purely radiologic. Therefore, once a bony "tumor" is diagnosed, one makes a decision to operate or to observe. The following description relates to operative observations and management.

Dermoid and Epidermoid Tumors (Figure 10.1)
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 eroded. 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 burr tip. Once the thin, osseous, outer table of the skull has been removed, the granulomatous tissue may be taken away either with a curet 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.

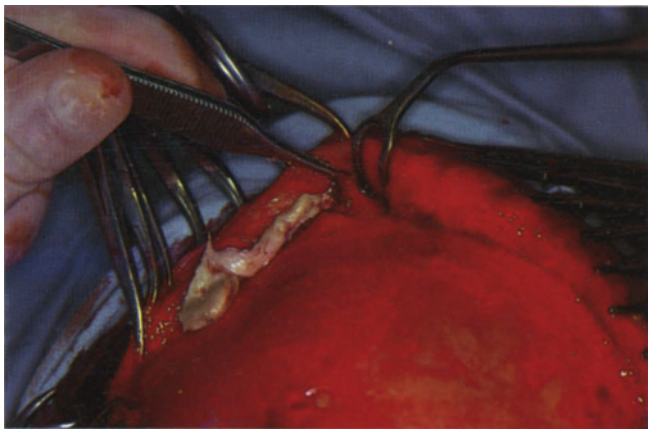


Figure 10.1. This dermoid tumor, growing within the region of the pterion, consisted of fibrocollagenous tissue which was semisolid. When the extruding material herein shown was removed, a cartilaginous, intradiploic, portion was excised *in toto*.

Fibrous Dysplasia

There is no curative surgical treatment for fibrous dysplasia when it is polyostotic. However, the unsightly excrescences that often extend across suture lines should be removed with the use of a high-speed burr, brushing the protuberances away until a smooth contour results. 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. Monostatic 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 burr 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! 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

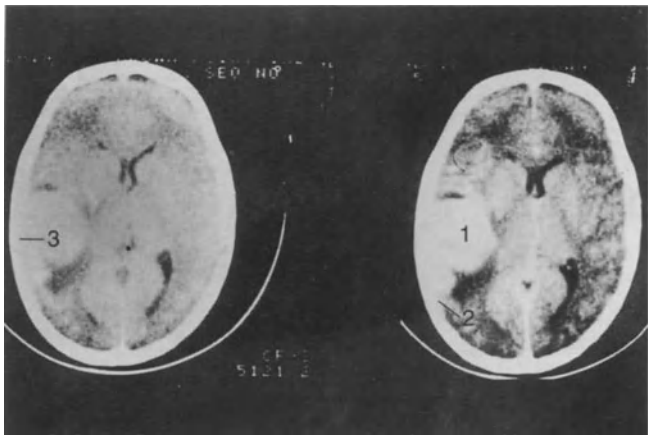
The excresence of the osteoma is simply shaved away with a burr, until the normal contour of the skull has been restored. It is not necessary to perform a craniectomy, though doing so offers no particular disadvantages: no bone needs to be grafted if the defect is small, and rib is excellent as donor tissue if the defect is large.

Dural Sarcoma (Figure 10.2)

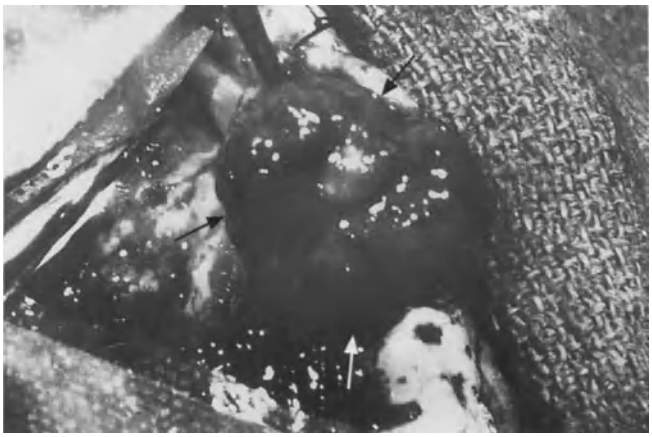
Though the meningioma does not occur in the newborn or infant, and is extremely rare in the toddler and juvenile, except in the occasional child suffering neurofibromatosis, the dural sarcoma is, in the infant and toddler, relatively common, occurring as often as the primitive neuroectodermal tumor (PNET). On computed transmission tomography (CTT) 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, since their biological activity is not reflected by histological appearance. Therefore, the surgical approach should consist of resection of 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. The CO_2 laser is ideal, providing it is used with the beam defocused, at 15 to 20 W, to vaporize the tumor without cutting through or around it. If the beam is focused, it will cut into the myriad of vessels this tumor holds, resulting in unacceptable bleeding. Individual bleeding areas should be stopped immediately so that the laser may be used effectively, without the formation of a layer of carbonized blood.

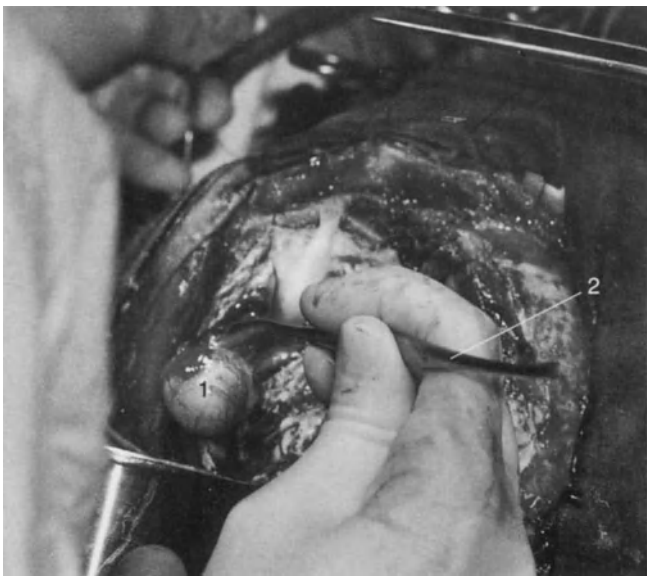
Since resection of the tumor entails resection of parenchyma and dura, one finds himself exposing bone. This complicates the closure and increases greatly the risk of cerebrospinal fluid fistulae, since it is impossible to attain a dural closure. In addition to this, the bone flap may be approximated only over a portion of the remaining cerebral tissue, so that the only barrier between subarachnoid spaces and the environment is the skin. Periosteal grafts are ineffective, so *fascia lata* grafts may be used if extensive defects remain. 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. Postoper-



A



B



C

Figure. 10.2. Dural sarcoma. (A) The CTT study reveals an enhancing lesion (*1*) originating from the dura (*2*) with thickening of the overlying calvarium (*3*). The mass extends deeply into the cerebral parenchyma, and is characterized by a relatively well-delimited border. (B) Operation on an invasive dural sarcoma (arrows), extending into the cerebral parenchyma. It was necessary to use tenotomy scissors for the dissection because of the thickness of the tumor. (C) This dural sarcoma (*1*), on the other hand, was so well encapsulated that it could be removed with a Penfield #2 dissector (*2*).

ative roentgen therapy should not be begun until there is complete healing of the skin and no indication of cerebrospinal fluid leak.

Orbital Tumors

General Comments: Periorbital Tumors

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 masses expanding around the orbit and impinging upon it, which may, as the mucocele, result in diminished orbital volume, are fibrous dysplasia, edentulous maxillary cyst, adamantinoma papillary tumors of the paranasal region.

The intraorbital tumors which a neurosurgeon may encounter are *neurofibroma* (especially the plexiform type), *retinoblastoma, rhabdomyosarcoma, lacrimal gland tumors,* the *hemangioma* (which, incidentally, is the second most common cause for unilateral exophthalmos), and *dermoid* and *epidermoid tumors.*

The most superficial tumors, and the most common, are the *dermoid* and *epidermoid tumors* (and the *lacrimal gland tumors*, which they closely resemble in appearance and location). They are located in the superolateral portions of the orbits, at times deepening the lacrimal 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

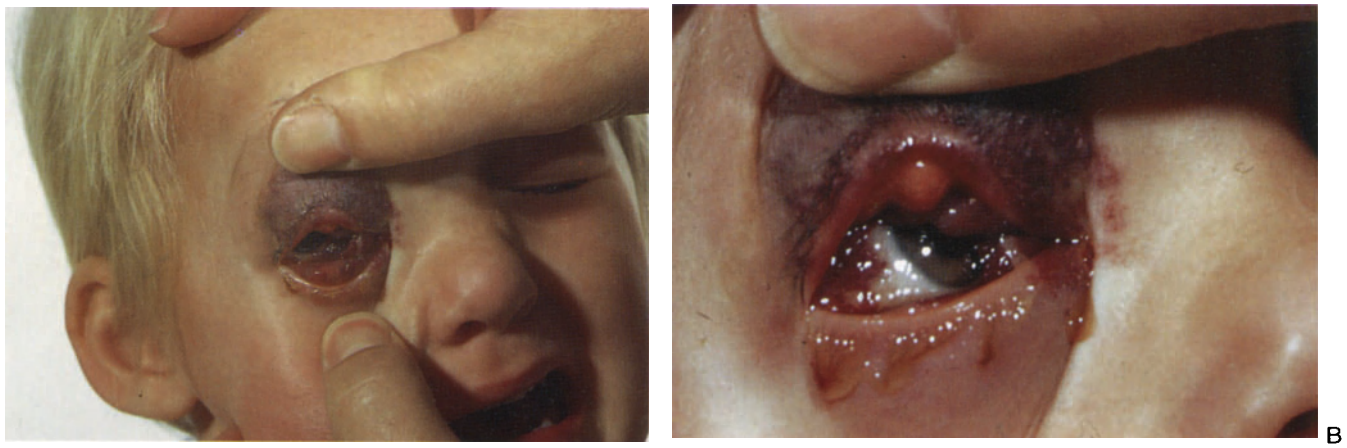


Figure 10.3. (A) The hemangioma has caused an hemorrhagic conjunctivitis, so that the eye is nonfunctional. (B) This is a closeup shot of the conjunctiva and sclera, illustrating the weeping hemangioma.

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 observed for a prolonged period of time, to see whether they disappear spontaneously before attempting to operate upon them. Others (meningioma, dermoid, epidermoid, hamartoma) should be operated 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 compresing structures related to vision, is not indicated.

Hemangioma (Fig. 10.3), the most common primary intraorbital tumor, is very prominent in younger children, most often disappears spontaneously, tends to invade the adnexa of the orbit and the lids, and commonly causes orbital enlargement and proptosis. These tumors often originate primarily within the muscular cone, diffusely spread throughout the contents of the cone, 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 extraordinarily difficult to accomplish without producing unacceptable deficits, and is to be discouraged unless the tumor progresses in volume (as the child ages) and threatens vision. 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 orbit and periorbital region. They are not resectable, and are extraordinarily 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 entirely without definable confines.

Retinoblastoma 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 a very invasive tumor, extending uniformly into the orbital adnexa, eroding and destroying bone. Neither of these tumors is operable.

Neuroblastoma may actually present clinically as a primary orbital tumor, when, in fact, the orbital lesion is a metastasis.

Orbital Tumors

Intracranial Approach to Orbital Tumors (Figure 10.4)

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. In fact, 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 bones should allow 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 should be carried out slowly and gently, since no cerebrospinal fluid is being aspirated. If this dissection appears difficult or if the surgeon wishes to assure minimal retraction of the frontal lobe, then either Lasix or mannitol may be given to provide temporary diminution of cerebral volume.

The orbital roof and supraorbital rim are then removed in the manner already described. 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 always difficult because of the abundant amounts of fatty tissue, the volume of the globe, the rapidly tapering bony confines of the orbit itself, and the unpredictability of displacement of intraorbital arteries and veins. The use, consequently, of self-retaining retractors is mandatory if one wishes 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. It is important to remember the course of the optic nerve in the orbit. Tumors involving or invading muscle, adnexa, or bone are highly malignant and, consequently, nonresectable.

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 cleavage between the tumor surface and neural or adnexal elements. Modern technique allows for the use of the laser to vaporize 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 should be covered with telfa strips before the laser micromanipulator is used, with settings of 4–6 W and intermittent beam. Planes of cleavage may be dissected more precisely if the laser is used and tumor traction is minimized. Retraction, however, is mandatory, so one must apply self-retaining retractors. The DeMartel unit is ideal.

Anterior Cone Tumors (Figure 10.5)

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 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, and because both techniques provide progressively greater exposure of the area as the tumor diminishes in volume. As the tumor is taken from the surface of the globe, one may follow this latter structure 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.

Nerve Sheath Tumors (Figure 10.6)

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 entirely 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 of a given point of the nerve only to be reflected back into it by the nerve sheath.

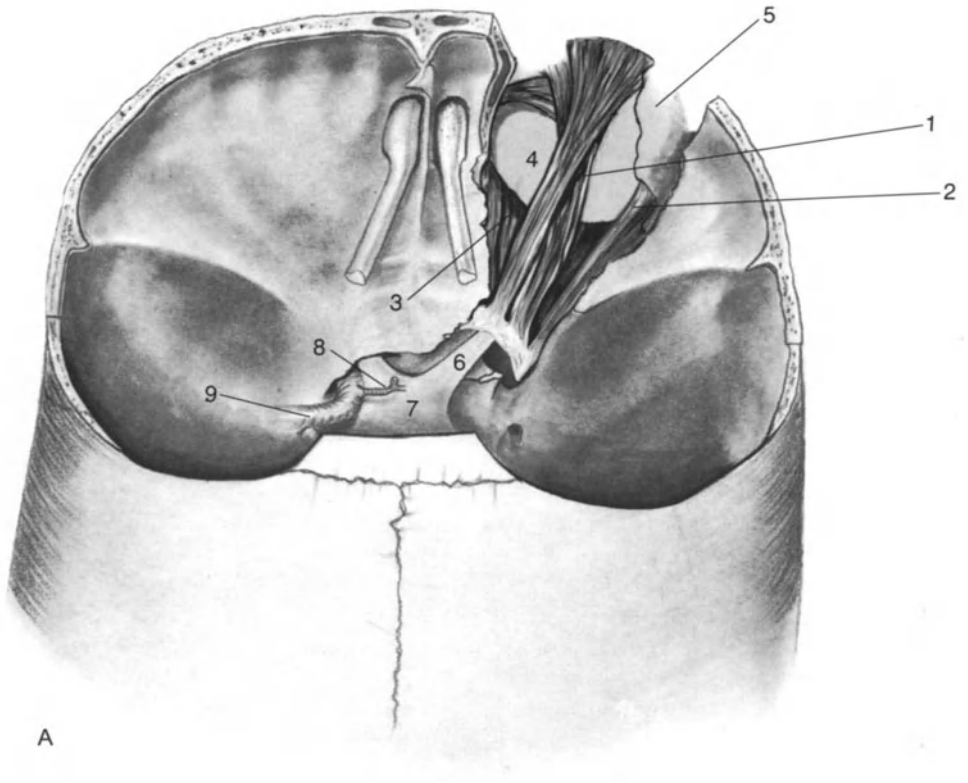
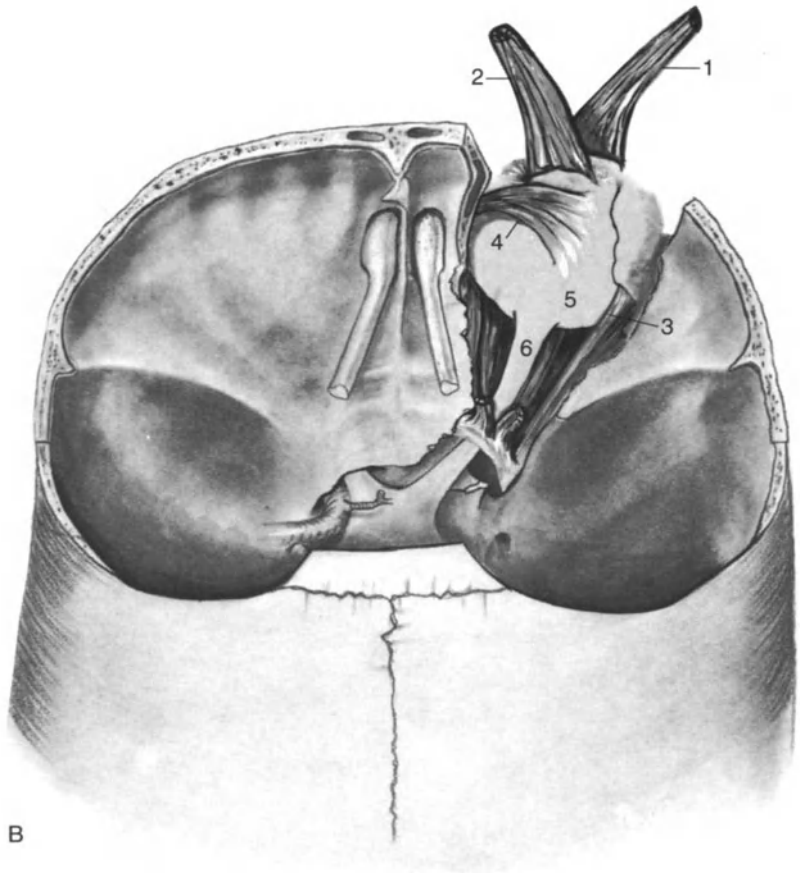
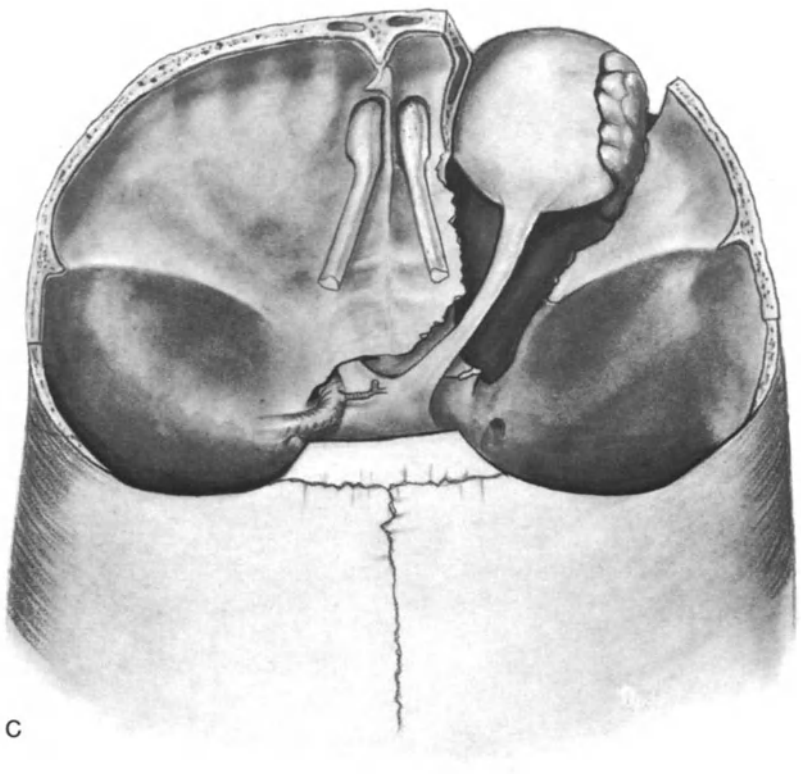


Figure 10.4. (A) The orbital roof and supraorbital rim have been removed, exposing the superior (*1*), lateral (*2*), and medial (*3*) rectus muscles, as well as the globe (*4*) and lachrymal gland (*5*). The optic nerve (*6*) and chiasm (*7*), as well as the anterior (*8*) and middle (*9*) cerebral arteries are included for anatomical orientation. (B) Transecting the levator palpebrae (*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.



B



C

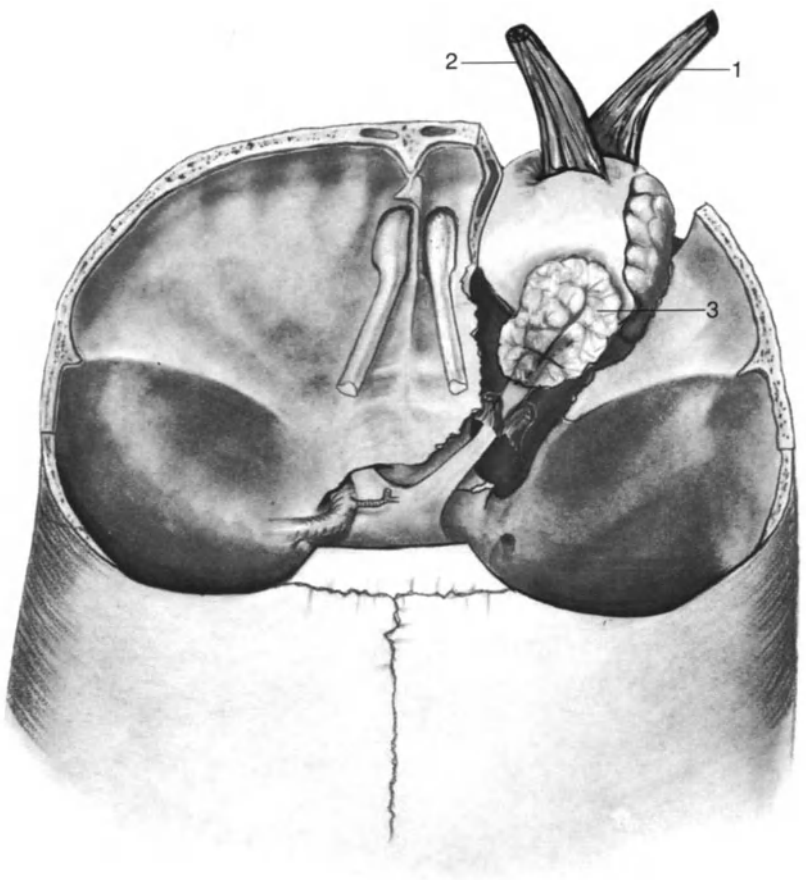


Figure 10.5. The tumor is nestled within the base of the cone, between the globe and the optic nerve. Reflection of the levator (*1*) and superior rectus (*2*) muscles exposes completely the tumor (*3*) within the anterior portion of the cone. The intraorbital fatty tissue has been removed.

Incision of this sheath with a #11 blade gives egress to tumor tissue, allowing the surgeon to inspect it before 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.

Optic Nerve Tumor (Figure 10.7 and 10.8)
If the tumor is entirely 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 obviously incompatible with vision, so the only indication for excision is an unsightly exophthalmos 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 since preoperative orbital computed transmission tomograms (CTT) will already have revealed that the optic foramen is normal. An enlarged optic foramen on computed transmission tomograms (CTT) and metrizamide cisternography will already have informed the surgeon whether there is extension of tumor into the subarachnoid portion of the optic nerve.

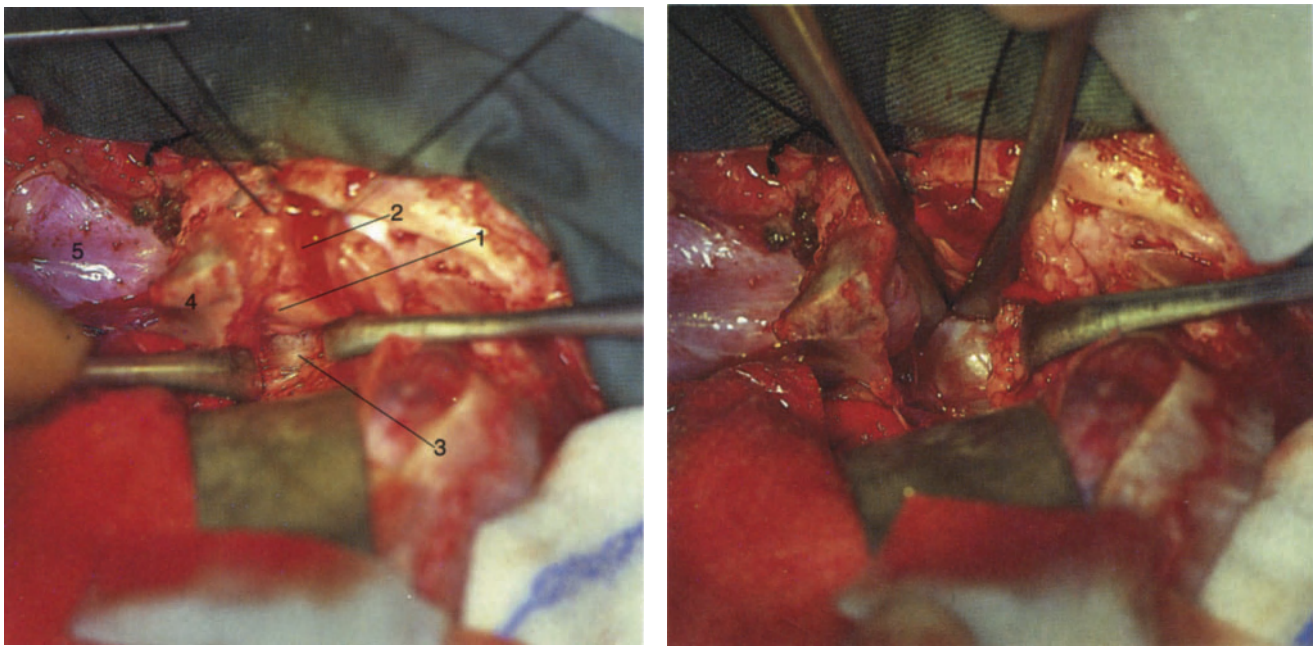
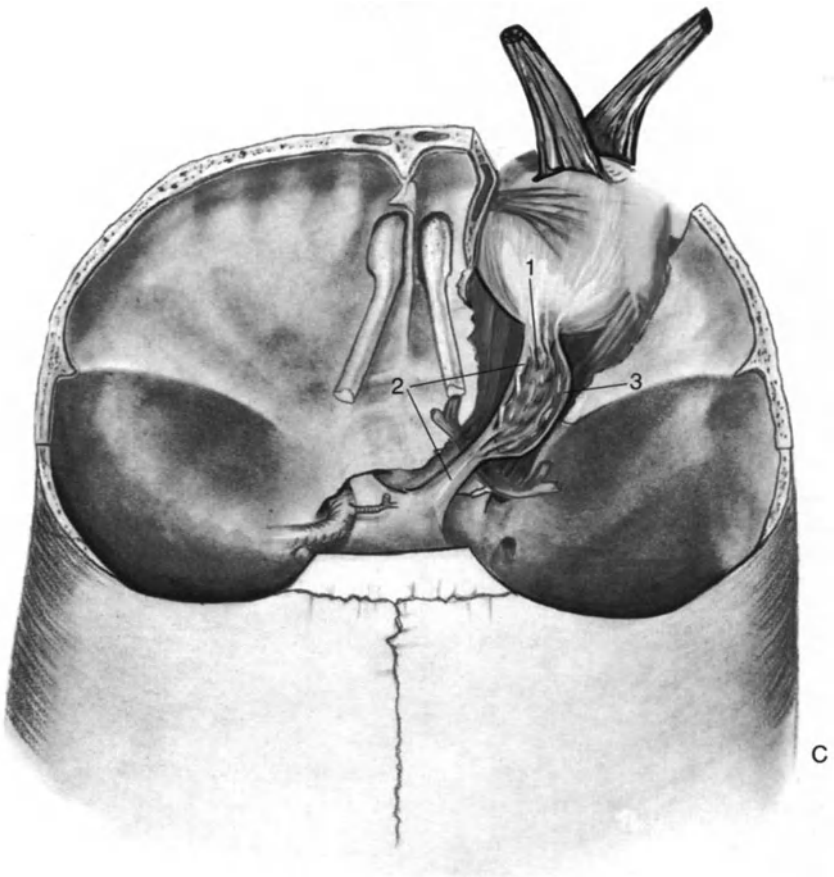


Figure 10.6. (A) This child had a meningioma growing from the optic nerve sheath. The globe (*1*) has been exposed after the levator palpebrae and superior rectus muscles (*2*) were transected and reflected anteriorly. The widened optic nerve (*3*) is exposed between the two Penfield dissectors. It is 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 zyomatic 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*).



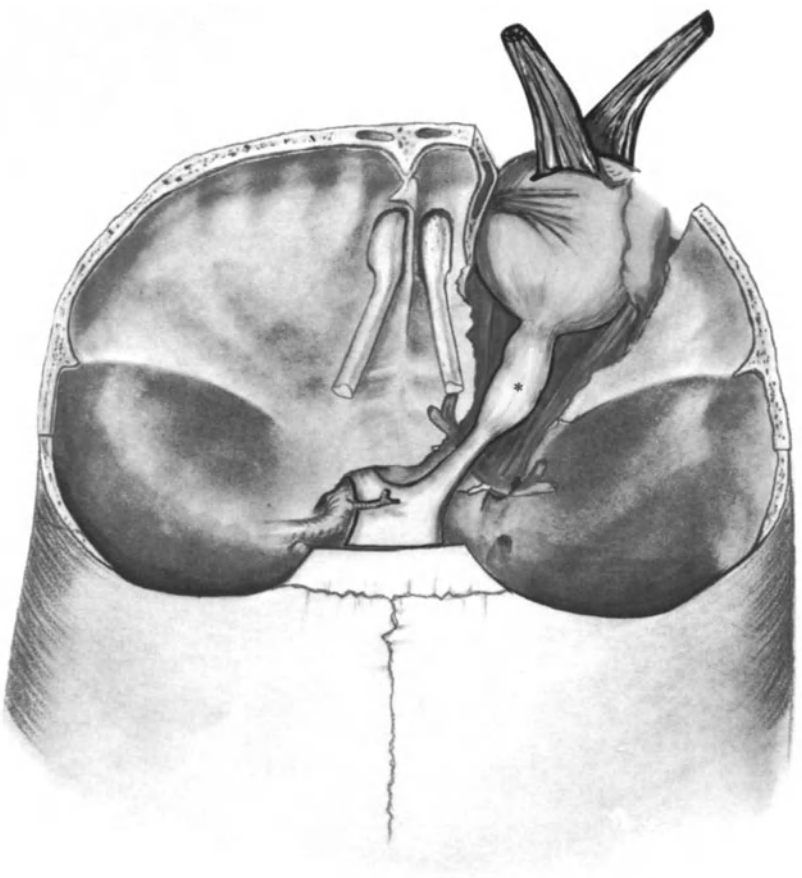


Figure 10.7. An optic nerve glioma (*) growing entirely within the intraorbital portion of the optic nerve does not extend through the optic foramen.

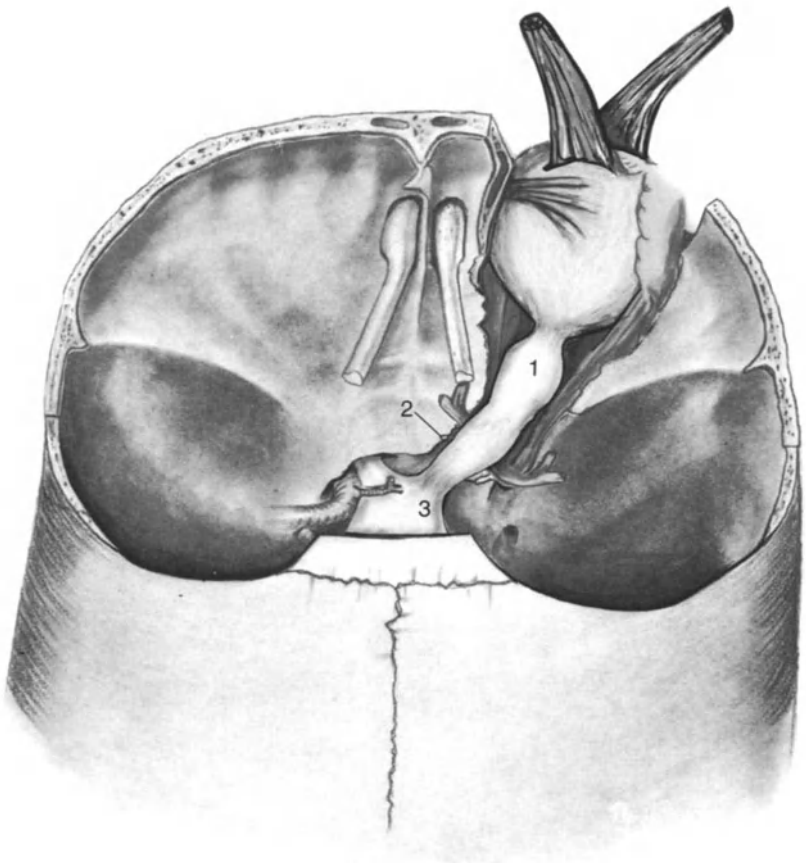


Figure 10.8. An optic glioma (*1*) extending through the optic foramen (*2*) and up the optic chiasm (*3*).

Hemispheral Tumors

Solid Hemispheral Tumors

The solid hemispheral tumors in childhood range from the extraordinarily vascular ependymoma and optic pathway glioma, which extends into the temporal lobe from the optic tracts, to the totally avascular hamartoma and astrocytoma diffusum. Different from the adult, who may have a highly vascular tumor which does not show vascular pattern or stain on the angiogram, the child's solid tumor vascularity may invariably be predicted with angiography. Identical to the adult, CTT characteristics permit no insight into the vascularity of the tumor nor does enhancement on CTT studies offer incontrovertible proof that the lesion is neoplastic.

The cerebral hemisphere glioma has a very bad prognosis: only 20% survive more than five 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.

Highly Vascular Hemispheral Tumors

(Figures 10.9 and 10.10)

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 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 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 cerebral incision before isolating and occluding the feeding vessels is ill-advised.

The craniotomy and dural opening selected for a highly vascular hemispheral tumor should be such as to provide access to all main-trunk arterial and venous systems 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 temporoparietooccipital 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. Any other approach results in an unacceptable 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.

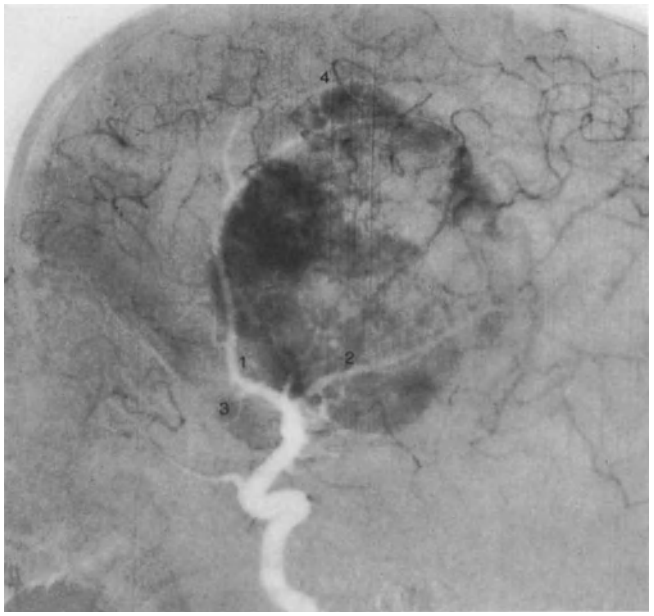
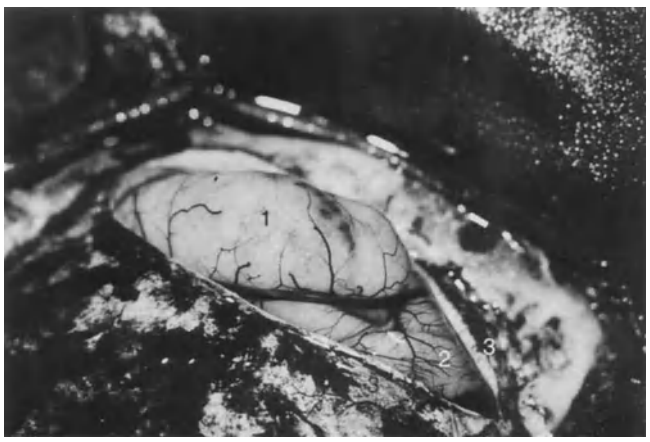
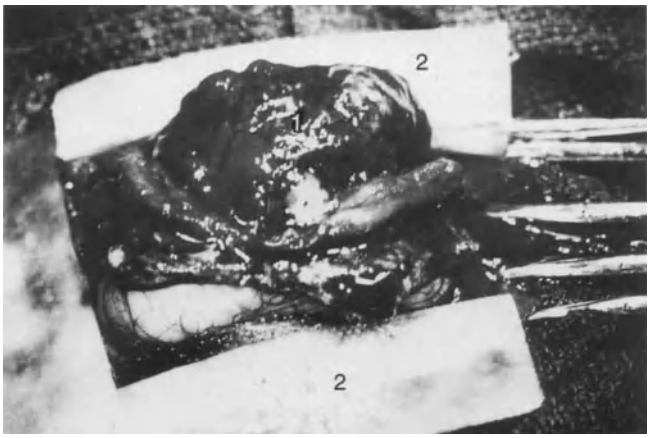


Figure 10.9. Solid vascular hemispheral tumor. This study reveals the superimposition of the anterior (*1*), middle (*2*), frontobasal (*3*), and sulcomarginal (*4*) arteries upon the tumor stain, allowing one to appreciate extension of the tumor within the rostrum, genu, and body of the corpus callosum as well as into the septum pellucidum and III ventricle.

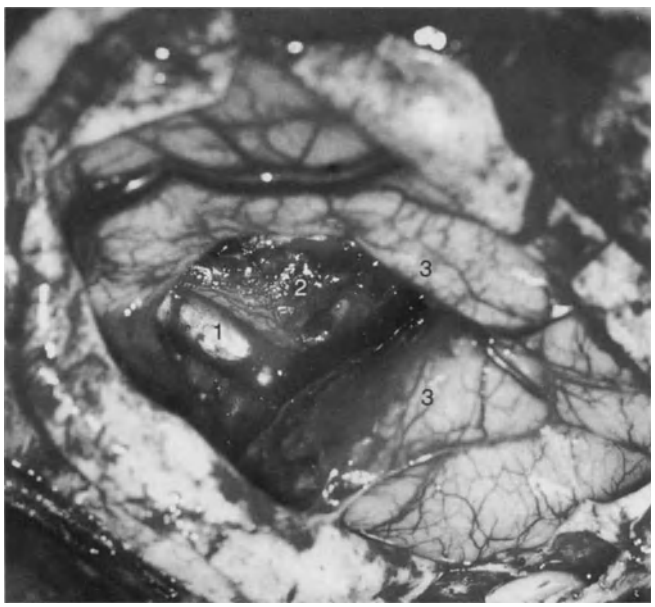
Figure 10.10. This highly vascularized, well-circumscribed mass was resected by working around the periphery of the tumor, at its confine with normal cerebral tissue, after having occluded the main feeding vessels. The bridging vascular structures between the tumor and surrounding tissue were coagulated and transected as the dissection proceeded, freeing the tumor (*1*) completely from the surrounding cortex and white matter (*2*).



A



B



C

Figure 10.11. (A) A relatively well-circumscribed avascular tumor. The neoplasm has invaded the gray matter, and there is a clear line of demarcation between tumor (*1*) and apparently normal gray matter (*2*). The dural opening has allowed the remarkably swollen underlying neoplastic brain tissue to deliver itself, whereas the normal brain remains beneath the level of the dura mater (*3*). (B) Performing a cerebrotomy over the *excrescences* of neoplastic parenchyma permits the delivery of a relatively well-circumscribed primitive neuroectodermal tumor (*1*). Cotton telfas (*2*) are used to separate the tumor from the surrounding brain. (C) After the tumor has been completely removed, its bed (*1*) is dried by irrigation and insertion of relatively large cotton fluffies (*2*). Residual tumor, which may be readily identified as distinct from surrounding normal tissue (*3*), is resected with laser, providing clean edges.

Different from a lobectomy or hemispherectomy, one does not take the venous channels prior to performing 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, gray matter, white matter, through an extensive 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 all of 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, the tentorium. When the vascularity of the neoplastic cerebral parenchyma is such as to reveal a uniform granular blush with a paucity of vascular lakes, and without arteriovenous shunts or neoplastic vessels, one may resect the tumor in a complete and bloodless manner by using the laser. This permits vaporization of the neoplastic cells and coagulation of the microvasculature.

Avascular Hemispheral Tumors

(Figures 10.11 and 10.12)

Avascular intraparenchymal tumors are removed either with the cerebrotomy or lobectomy techniques, depending upon the size and extent of the mass. It is unusual

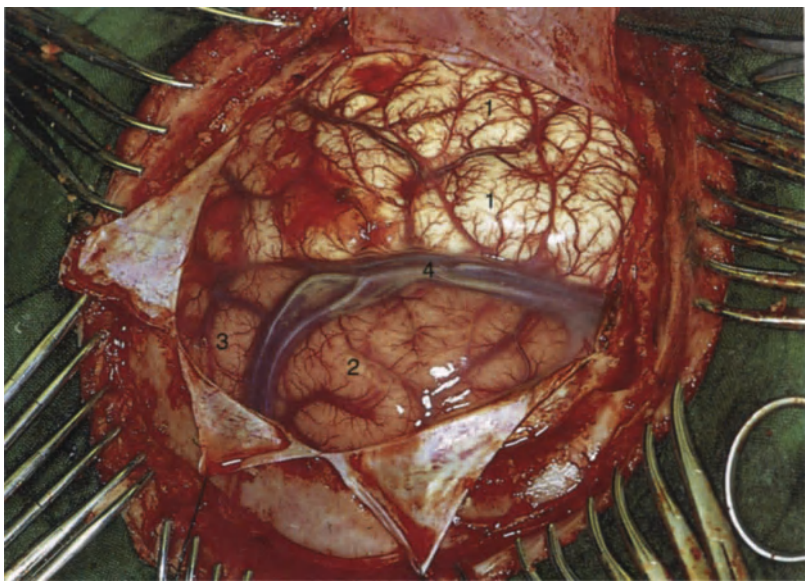


Figure 10.12. This is an interesting and unusual case: hamartoma (*1*) of the temporal lobe. In this, the surgeon's view of the hamartoma of the left temporal lobe, one notes the frontal operculum (*2*), the parietal operculum (*3*) and the exit of the vein of Trolard (*4*) from the insular region. A complete temporal lobectomy was performed.

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, 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, smaller tumors. One must not resect parietal lobe, basal ganglia, or thalamic tumors. Roentgen therapy is the treatment of choice for tumors in these structures since it is associated with a minimum morbidity, resulting in a far better quality of survival.

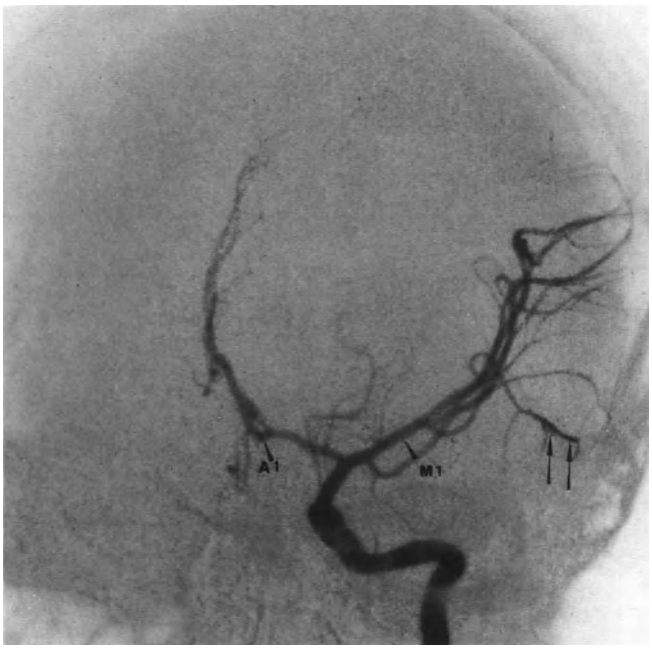
Cystic Hemispheral Tumors (Figure 10.13)
Cystic tumors of the hemispheres may be approached along the convexities, since vasculature is not a problem and one need not concern himself with isolating the main trunks of the carotid or terminal basilar systems. These tumors also assume enormous proportions 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 cystic fluid, and the nodule.

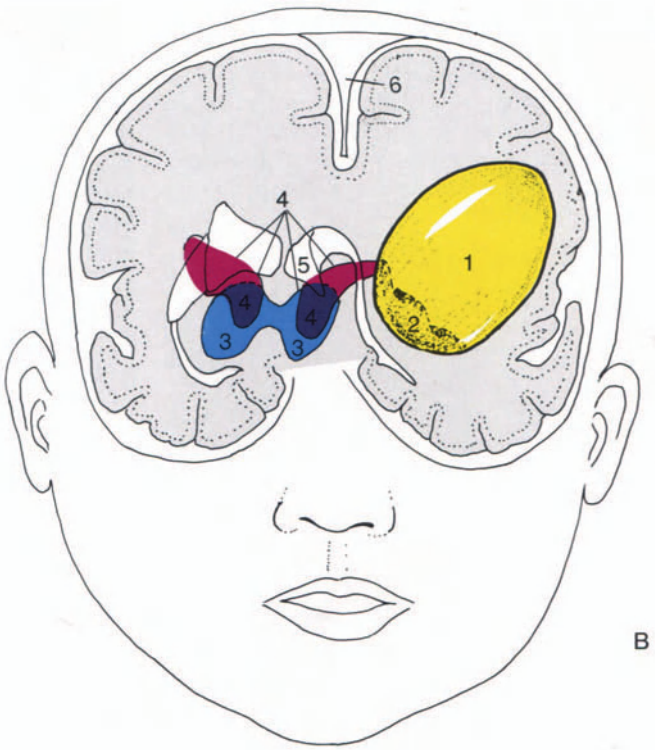
Capsule (Figures 10.14 and 10.15)
After the cerebrotomy has been performed and the surgeon comes upon the capsule, he should dissect along its surface but only enough to provide himself adequate surface area to enter the cavity and drain the fluid. *En bloc* resection is discouraged. Most glioma and 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 on and parallel to the capsule, to separate gliotic tissue from capsule. Slight compression upon the cystic mass by the tip 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 over 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 between telfa and tumor, and so on.



A



B

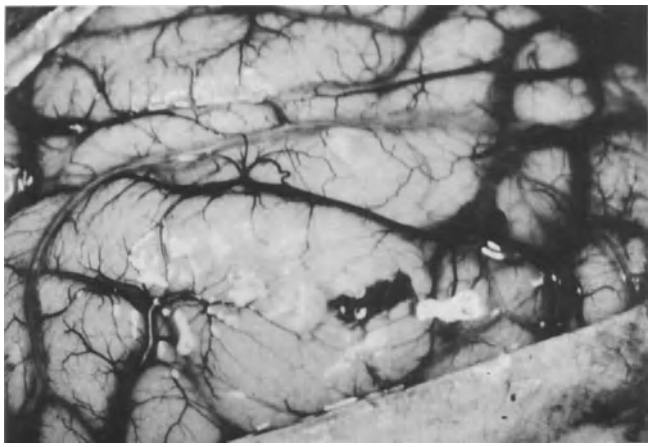
Figure 10.13. (A) This cystic astrocytoma is located in the frontal lobe, anterior to the central (Rolandic) sulcus and superior to the Sylvian sulcus. The A-1 portion of the anterior cerebral artery is displaced posteriorly, as is the M-1 portion of the middle cerebral artery. The tumor nodule is displacing 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*).

Cystic Fluid

The cystic fluid is never crystal clear. It ranges in color from yellow through amber to chocolate brown. 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, 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.

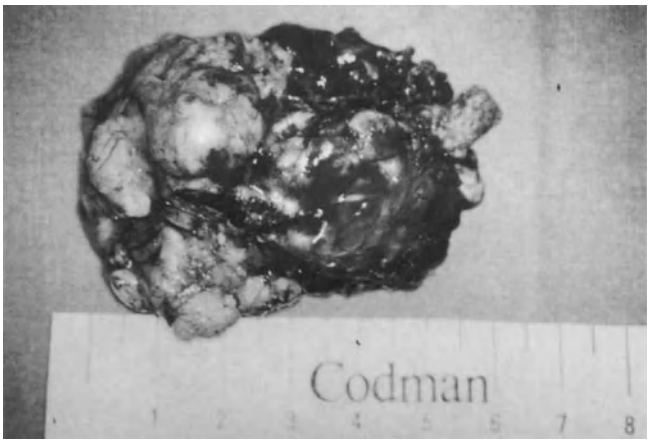
Nodules

Nodular masses within cystic tumors are not common. When present, they may be avascular (the majority) or vascular (the minority). The nodules are best removed by coagulation or, preferably, vaporization with laser, whether they are vascular or avascular. Either technique destroys the nodule without subjecting the surrounding brain to the stretching damage, which is unavoidable when the nodule is pulled away with a pituitary forceps or a sucker. If laser is not available, a 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



A

B



C

Figure 10.14. 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.

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.

Ventricular Tumors

Apart from the choroid plexus papilloma—and the *extraordinarily* rare meningioma—intraventricular tumors are either *ependymal* or *glial* in origin, with the ependymal tumors taking origin from the ependymal lining of the ventricles and the glial tumors 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 high incidence of meningioma in children with Von Recklinghausen's disease.

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 ex-

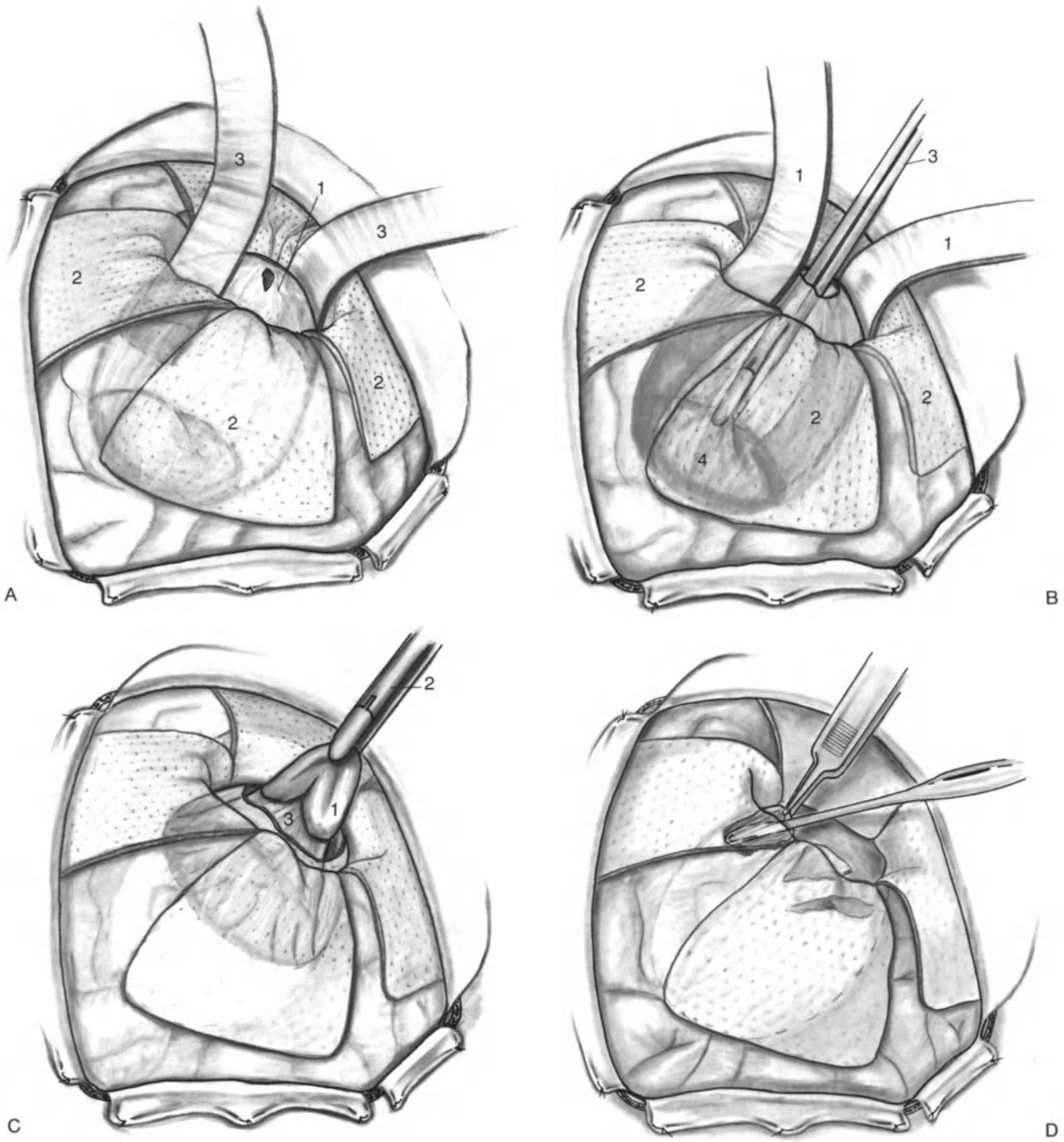


Figure 10.15. (A) The cerebrotomy has been performed and the surface of the tumor exposed, so that its dome (*1*) delivers itself through the opening. Telfa strips (*2*) are laid over the capsule, separating it from the surrounding brain. Spatulae (*3*) may be insinuated along the tumor surface after the cyst has been opened and the fluid drained. (B) The 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. (C) 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 laser to vaporize the tumor nodule and capsule is much preferable! (D) A Penfield #4 dissector may be used to separate the frayed portions of the cyst capsule from the surrounding tissue.

ample, may expand within the entire 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 ependymona entails different surgical and microsurgical technique for total removal than a papilloma expanding at the trigone and occluding 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 qudrigeminal plate and pushing into the posterior fossa. Consequently, preoperative evaluation of intraventricular tumors is extraordinarily 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 extends into the cerebral parenchyma.

Lateral Ventricle Tumors: General
(Figures 10.16 to 10.19)

Conceptualization of lateral ventricular tumors is facilitated greatly if one first envisions the craniofacial region from 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. 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, 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. In fact, one really never should split more than a third of it!

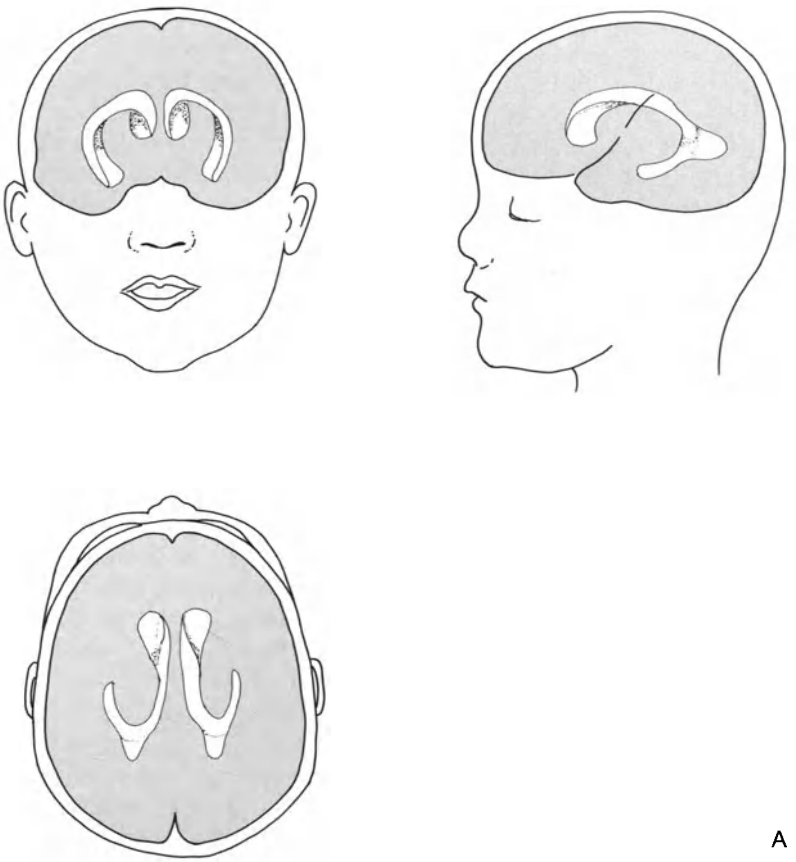
When a lateral ventricle tumor obstructs the foramen of Monro, there ensues remarkable dilation of the homolateral ventricle, with shift to the opposite side, resulting from the combined pressure vectors of the tumor and the asymmetrical hydrocephalus. Though this combination of forces aggravates drastically, and acutely, the clinical situation, it provides the surgeon with an enlarged ventricular chamber through which he may work in resecting the tumor. 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 demands great caution in avoiding 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 their tendency to be highly vascularized and to grow along 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 that extend into the brain substance change in consistency, but not in vascularity.

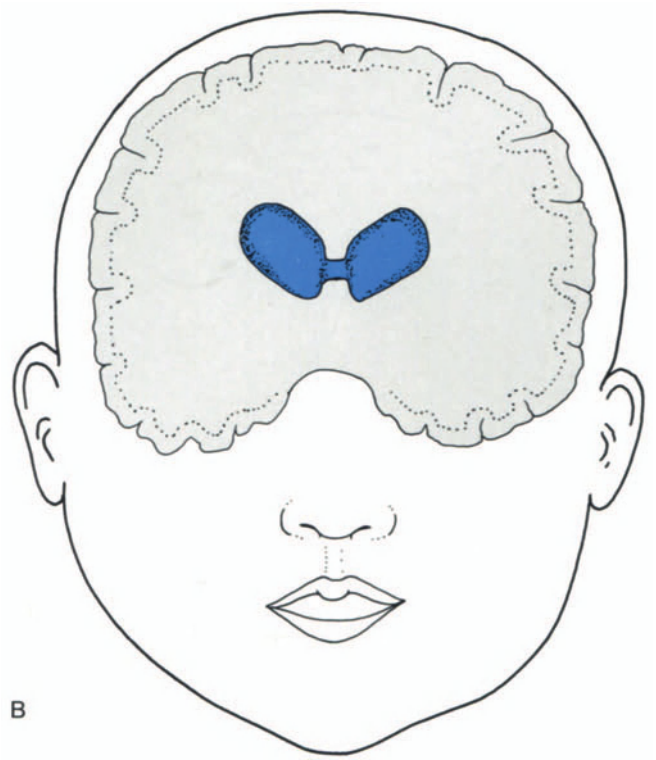
One of the most important surgical considerations in this tumor is the matter of identifying precisely the area of attachment, since the ependymoma tends to have a rather sessile base from which it extends into the cerebral parenchyma, sulci, and cisterns. Therefore, one is confronted with a tumor which 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 gradual increase in intracranial pressure is accepted well by the child until the *midline syndrome*[1] becomes evident. Consequently, the tumor may grow from one lateral ventricle to the foramen of Monro and into the III ventricle. On occasion, it may actually extend through the contralateral foramen of Monro, to fungate within the opposite lateral ventricle, resulting in a tumor mass which assumes the form of the ventricular system: *plastic ependymoma*. No ef-

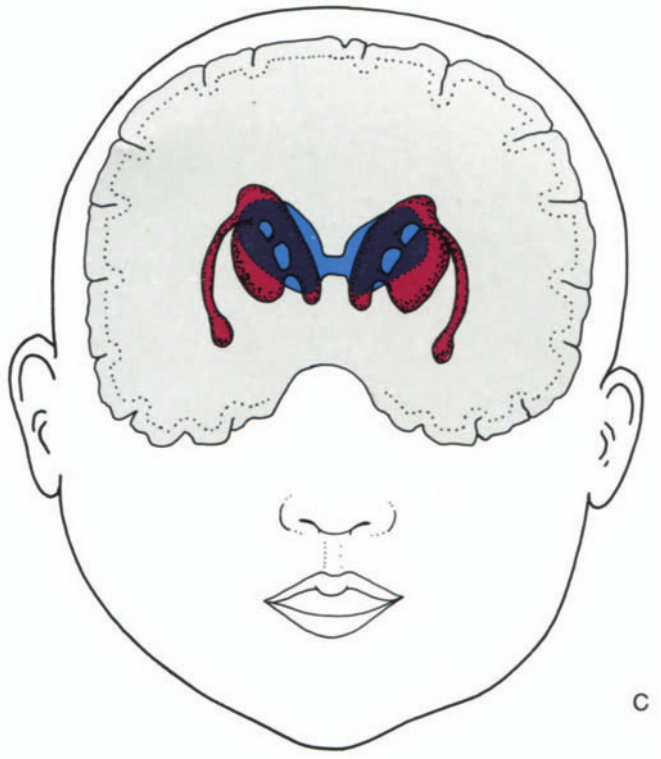


A

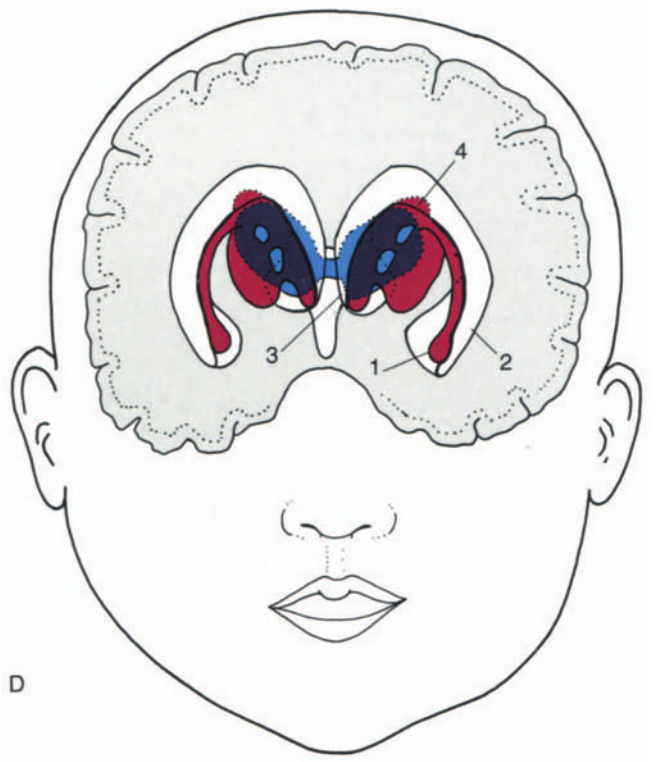
Figure 10.16. In evaluating the location, extent, and complicating ventricular dilation in a child with an intraventricular tumor, it is best to conceptualize the craniocerebrum in anterior-posterior, axial, and lateral projections. (A) The lateral ventricles are drawn in three basic projections, allowing the reader the opportunity to study the general size and spatial orientation of each ventricle in each of these projections. (B) The thalami have been drawn in, to allow appreciation of their location at the most inferomedial portions of the cerebral hemispheres. (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.



B



C



D

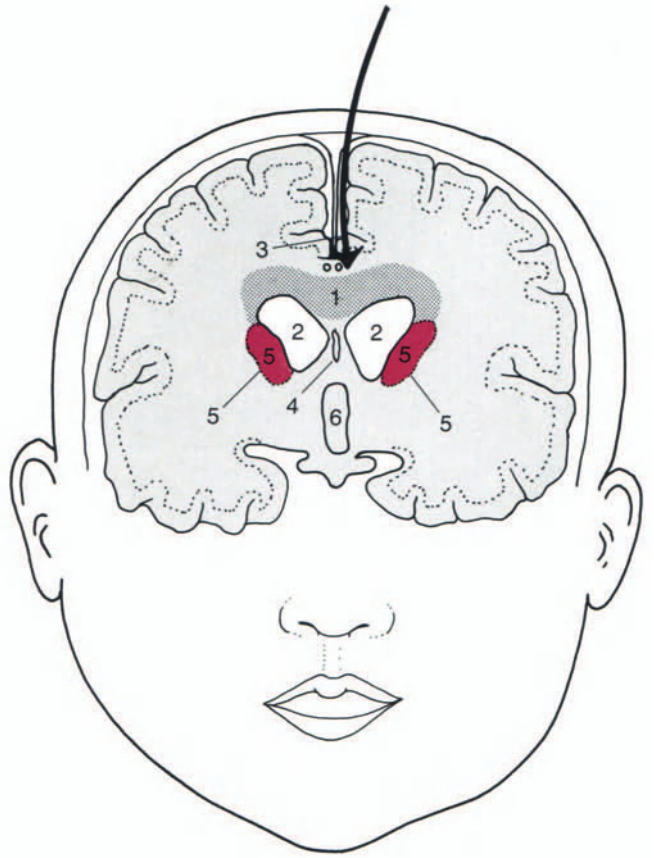


Figure 10.17. How dissection along the parasagittal surface of the corpus callosum (*1*) allows one to enter the body, portion 3, of the lateral ventricle (*2*). For orientation purposes, the falx cerebri (*3*), septum pellucidum (*4*), head of the caudate nucleus (*5*), and III ventricle (*6*) are also labeled. The slightly curved arrow indicates the parasagittal approach to the corpus callosum and ventricular system.

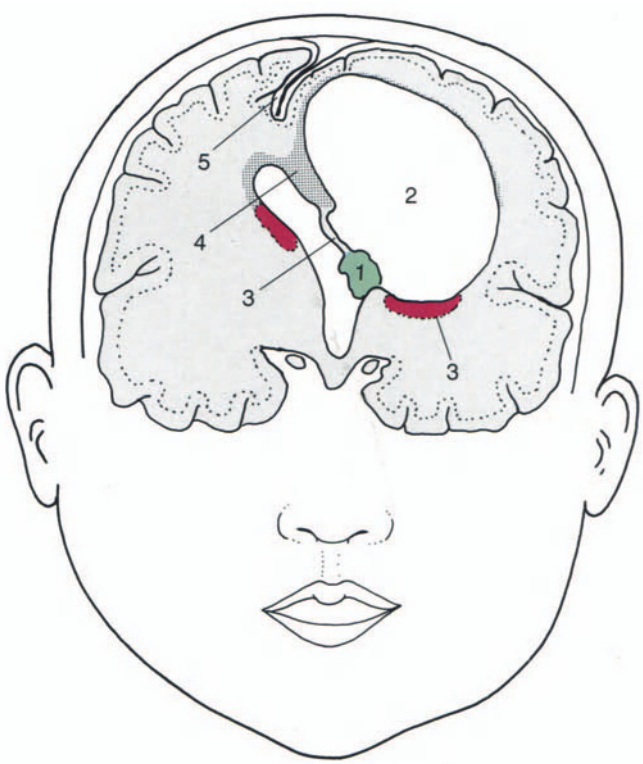


Figure 10.18. The obstructing lesion (*1*), at the foramen of Monro, dams the cerebrospinal fluid in the lateral ventricle, causing it to dilate (*2*) and resulting in a shift of the septum pellucidum (*3*) and corpus callosum (*4*) to the opposite side. In children, the interhemispheral fissure (*5*) is often shifted because the falx tends to be hypoplastic.

fort 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 uncovered unprotected).

Preferably, the tumor resection is performed with the laser. Before beginning to "lase" the tumor, 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 vaporize the tumor with the laser, starting at 5 or 6 W, pulsed, and progressing gradually to the most effective wattage and continuous wave. The vaporization 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. Any bleeders should be immediately coagulated, otherwise blood covers the surface of the tumor and is carbonized by the laser. This diminishes the vaporizing effect of the laser.

Because of the biologically malignant nature of ependymoma that borders on the ventricular or subarach-

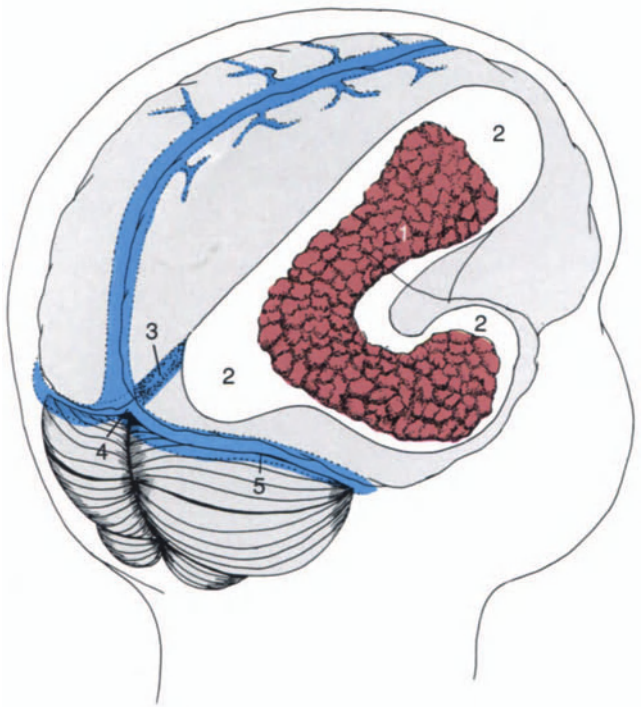


Figure 10.19. An intraventricular tumor, in this case a choroid plexus papilloma (*1*), causing dilation of the entire lateral ventricle (*2*). For orientation purposes, the straight sinus (*3*), torcular Herophili (*4*), and transverse sinus (*5*) are illustrated.

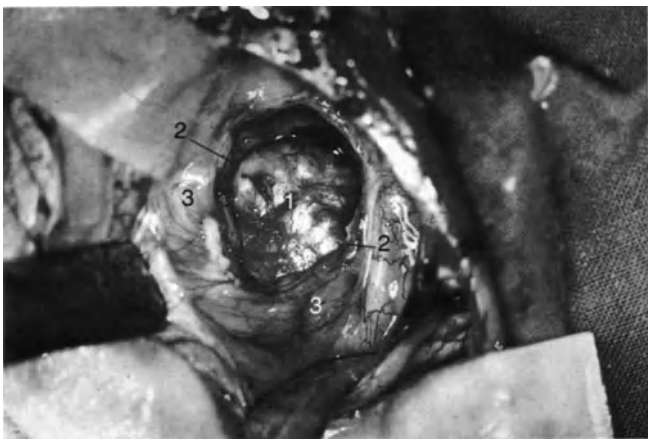


Figure 10.20. Giant subependymal astrocytoma, which occupied the entirety of the left lateral ventricle. The tumor (*1*) is seen through the cerebrotomy opening (*2*), allowing one to appreciate both its mass and density, which were most likely responsible for the gliotic appearance of the overlying cerebral hemisphere (*3*).

noid surfaces, irrespective of its histological appearance, excessive efforts to attain a complete resection are to be avoided. The surgeon does best by limiting himself to as complete a resection as possible, given the limitations of controlling the bleeding and following the tumor into the brain substance. Therefore, the temptation to resect an ependymoma *en bloc* should be resisted: the tumor is so soft, so friable, so lobulated that it is impossible to accomplish this, and the expense of the effort is an excessive blood loss.

Glial Tumors (Figure 10.20)

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 the superolateral wall of the ventricle or the septum pellucidum. Such tumors behave in a manner 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, greyish 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 get the incorrect impression that the tumor is, in essence, a deformed tumefaction of the ventricular wall.

These are benign tumors, they seldom invade the basal ganglia or thalamus, and they are not radiosensitive; consequently, the treatment is surgical. It should be directed to 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 contralateral ventricle would aggravate the transfalcical shift, and 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 laser represents the ideal instrument for removal of intraventricular glioma, techniques using sharp dissection, suction, or cautery loops are acceptable only when a laser is unavailable. Sharp dissection is awkward and imprecise within the ventricles, aspiration 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 laser permits the surgeon precise dissection 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 then 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 toward his line of vision. Bringing the medial surface superior is not advisable, since it puts the base of the tumor on the stretch and augments the risk of tearing subependymal veins.

With the pedicle of the tumor visualized completely 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 any 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

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. 10.18). This approach affords excellent exposure of both lateral ventricles, permits 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 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 compliment 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

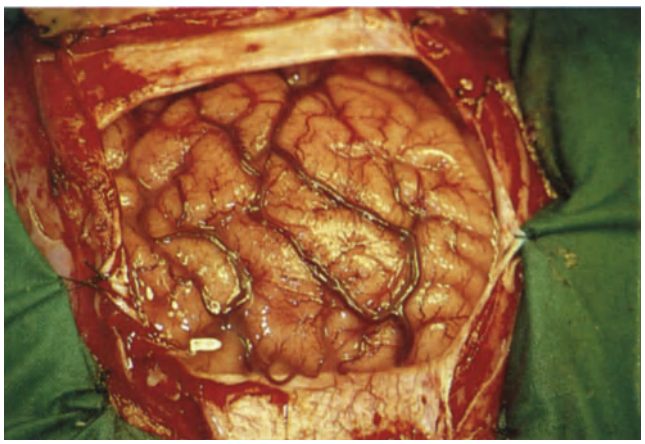


Figure 10.21. Cortical adhesive arachnoiditis (secondary to extraordinarily high CSF protein) in a child with a choroid plexus papilloma of the lateral ventricle. The granular appearance of the arachnoid over the right hemisphere is quite typical of this entity.

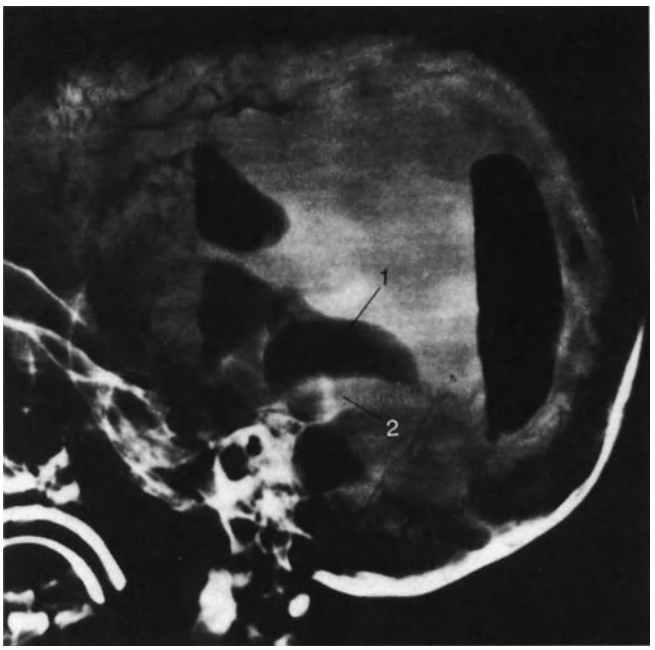


Figure 10.22. Multiple chambers within the ventricular system. A pneumoencephalogram performed (in 1966) on a female child with a papilloma at the trigone on the right, revcals a pattern suggestive of porencephaly with hydrocephalus. The cyst (*1*) over the surface of the tumor (*2*) is clearly shown. This pneumogram is shown here only to illustrate the formation of cavities which have the appearance of porencephaly. It is archaic as a diagnostic study.

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 cost because of the permanent and totally disabling intellectual consequences!!

Removal of these tumors is carried out by using the laser. 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

The asymmetrical hydrocephalus (Fig. 10.21) 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.

Choroid Plexus Papilloma

The choroid plexus papilloma (Figs. 10.22–10.25) 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,

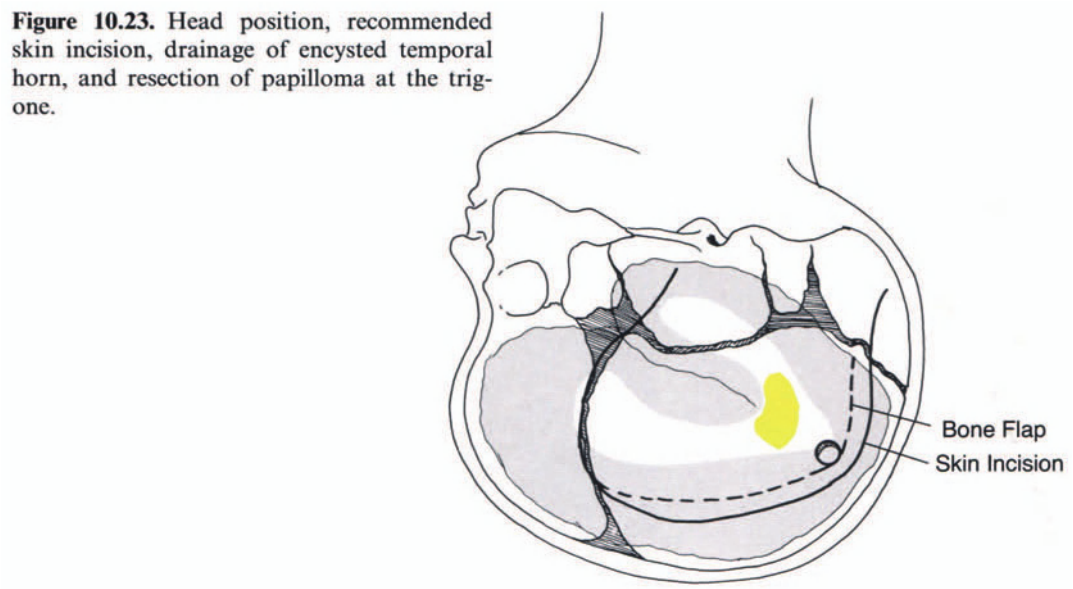
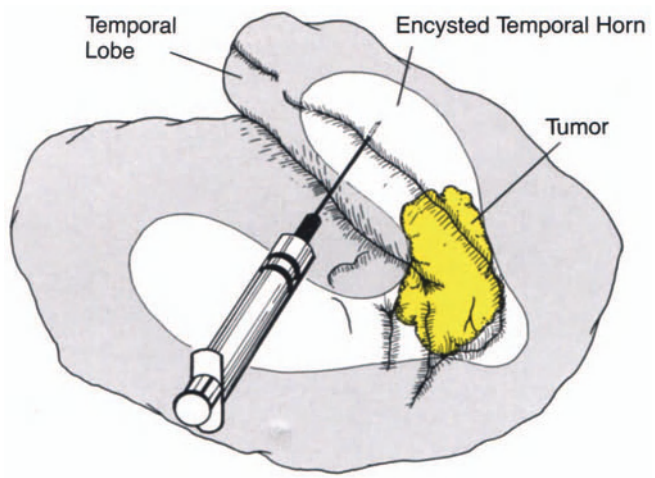
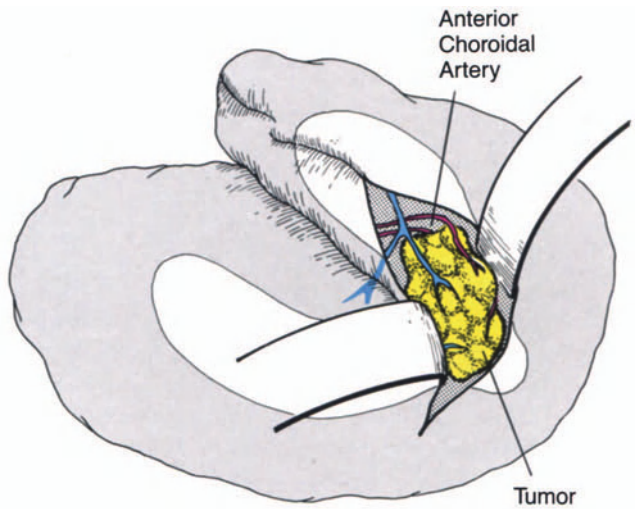


Figure 10.23. Head position, recommended skin incision, drainage of encysted temporal horn, and resection of papilloma at the trigone.





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 invariably 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 cc CSF). A linear cerebrotomy, 3 to 4 cm, within a sulcus located behind the angular gyrus provides generous access to the trigone. The cerebrotomy is best made in the most flattened pathologic portion of the cortex, which is overlying the tumor. It is not necessary to resect a cone of cerebral tissue, since a long linear incision provides more than adequate exposure of the ventricular system immediately over the trigone tumor mass. 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 complete hemostasis of this highly vascular area at the time of cortical incision.

The dissection through the white matter is best performed with two spatulae, thereby minimizing the area of cerebrotomy and maximizing the depth of opening. Laser cerebrotomy may be performed, but it is neither more efficient nor easier. 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.

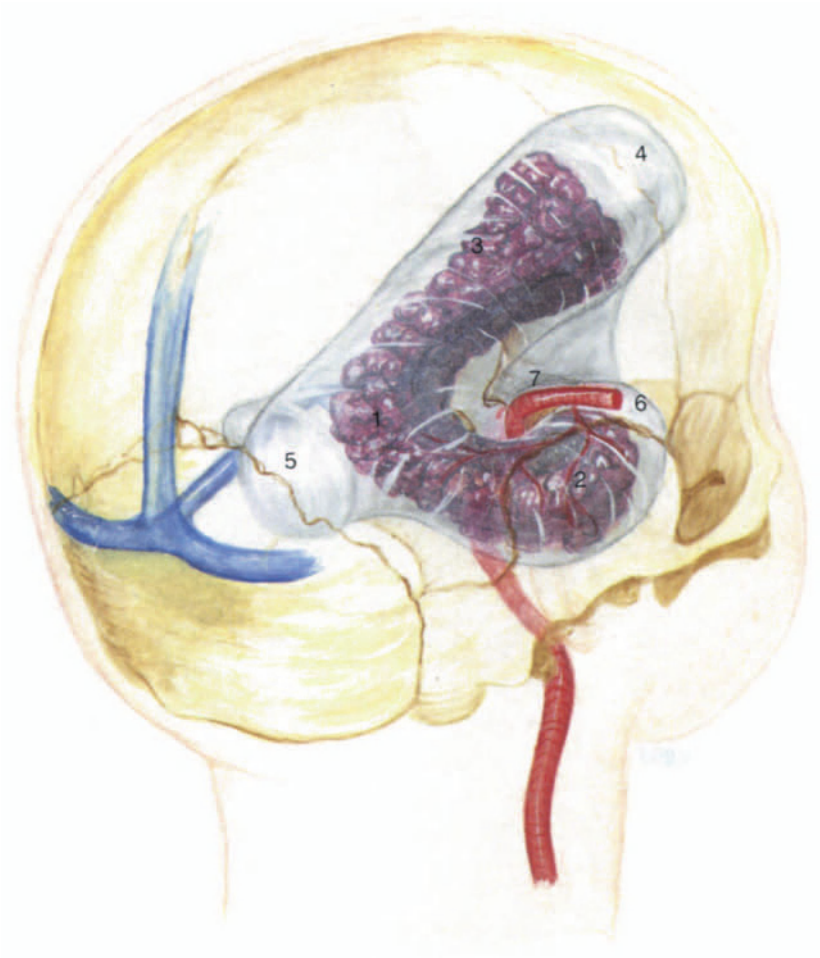
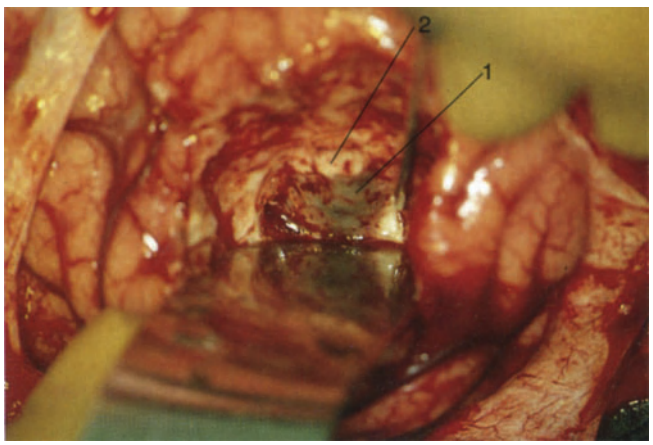


Figure 10.24. This lateral ventricle papilloma occupies the trigone (*1*), temporal horn (*2*), and body (*3*) of the lateral ventricle. The frontal (*4*), occipital (*5*), and temporal (*6*) horns are encysted. The middle cerebral system (*7*) is flattened between expanding frontal and temporal horns. 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 anterior septal veins.

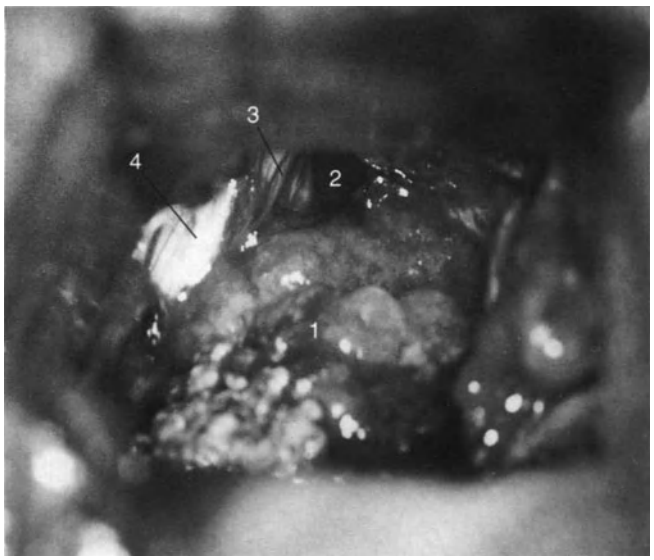
Cotton fluffies are pulled into the desired size and form, and then freshly soaked in normal saline at body temperature, prior to being used to dissect the surface of the tumor from the surrounding ependyma and parenchyma, with which the tumor may have established continuity or adhesions. Telfas are 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 of 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), vaporized *in situ* with a laser, 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 in light of introduction of the laser into the surgical armamentarium and the remarkable increase in efficiency of the thermocautery units.

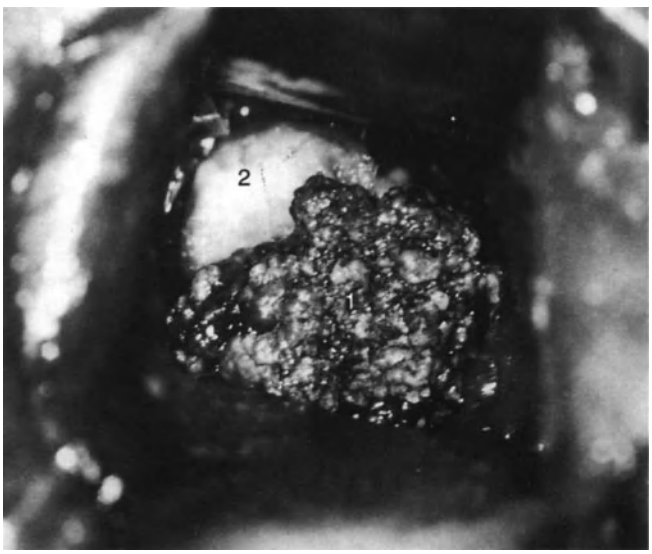
Linear cerebrotomy through the encysted temporal horn permits ample access to the tumor pedicle, and to the feeding branches of the anterior choroidal arter-



A



B



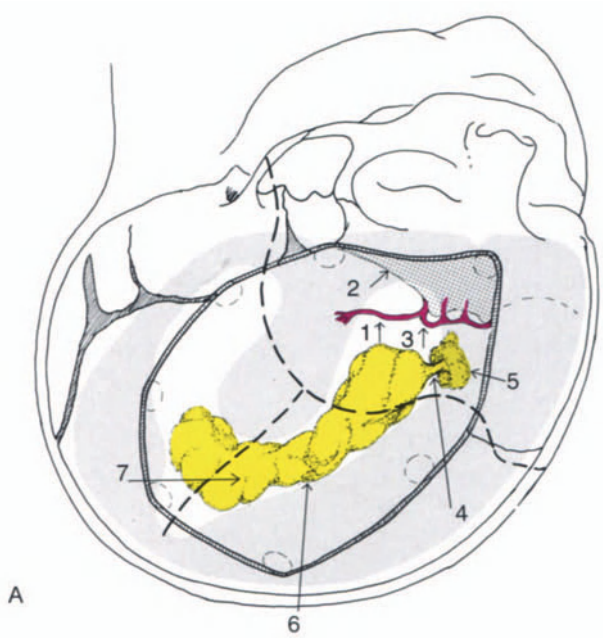
C

Figure 10.25. (A) After the cerebrotomy has been made and the dissection carried through the white mater, the spatulae are positioned at either extremity of the cerebrotomy, exposing the ependyma (*1*) and a thin rim of white mater surrounding it (*2*). (B) Opening the enpendyma with either bipolar forceps or laser at the trigone brings the papilloma (*1*) immediately into view. The mass occupies the entirety of the opening into the lateral ventricle so that its exposure presents no problems. Note the foramen of Monro (*2*), the body of the fornix (*3*), and the ependyma (*4*). (C) This is the characteristic appearance of papilloma as it is being vaporized with the laser. The tumor mass (*1*) is being elevated by a fluffy cotton (*2*), which has been lain beneath it both to compress the blood supply and protect the underlying ventricular surface from errant laser sweeps or deflections.

ies, 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 coagulated, 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. The draining veins enter the subependymal system, penetrate the choroidal fissure to enter the quadrigeminal and Galenic systems. They may also extend downward to enter the supraculminate system. In either event, they are best dealt with within the lateral ventricle where they 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 *en bloc* removal of the tumor mass, it is essential to wait for approximately 10 minutes after filling the ventricle with normal saline so as to be certain that there is no active bleeding, or oozing. One should inspect the entire lateral ventricle, especially the foramen of Monro, to assure himself that there is no tumor 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 purposes and for the benefit of those confronted with the need to resect a choroid plexus papilloma but without thermocautery of 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 to be avoided in such a vascular tumor as this! If one notes that 8 W are not enough,



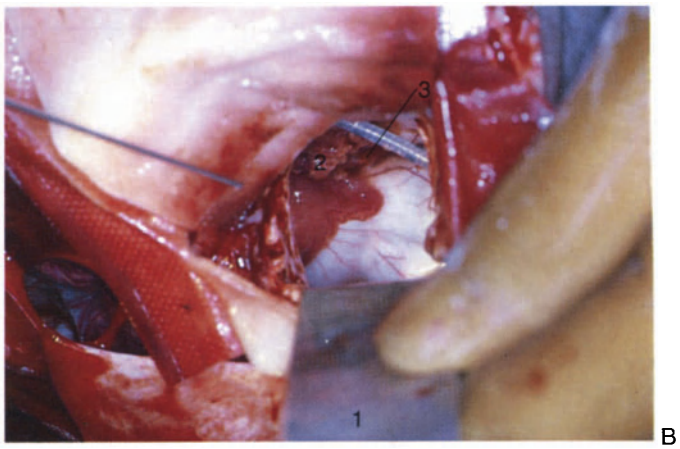


Figure 10.26. (A) Recommended position, bone flap, lines of dissection, and extent of tumor in papillomas entering the III ventricle through the foramen of Monro. The middle cerebral artery (*1*), lesser wing of the sphenoid (*2*), anterior cerebral artery (*3*), tumor extending through the foramen of 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.

he may increase to 12–15 W, but not beyond this because the friability and vascularity of the papilloma are such as to render working at higher voltage unsafe.

Lateral Ventricle Choroid Plexus Papilloma

The lateral ventricle (Figs. 10.26–10.28) 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 callosum 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 to 4 cm in length is made behind the angular gyrus and the tumor is isolated prior to sectioning its vascular pedicle. Vaporization or bipolar coagulation of the tumor facilitate its isolation and identification 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 adhesion (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

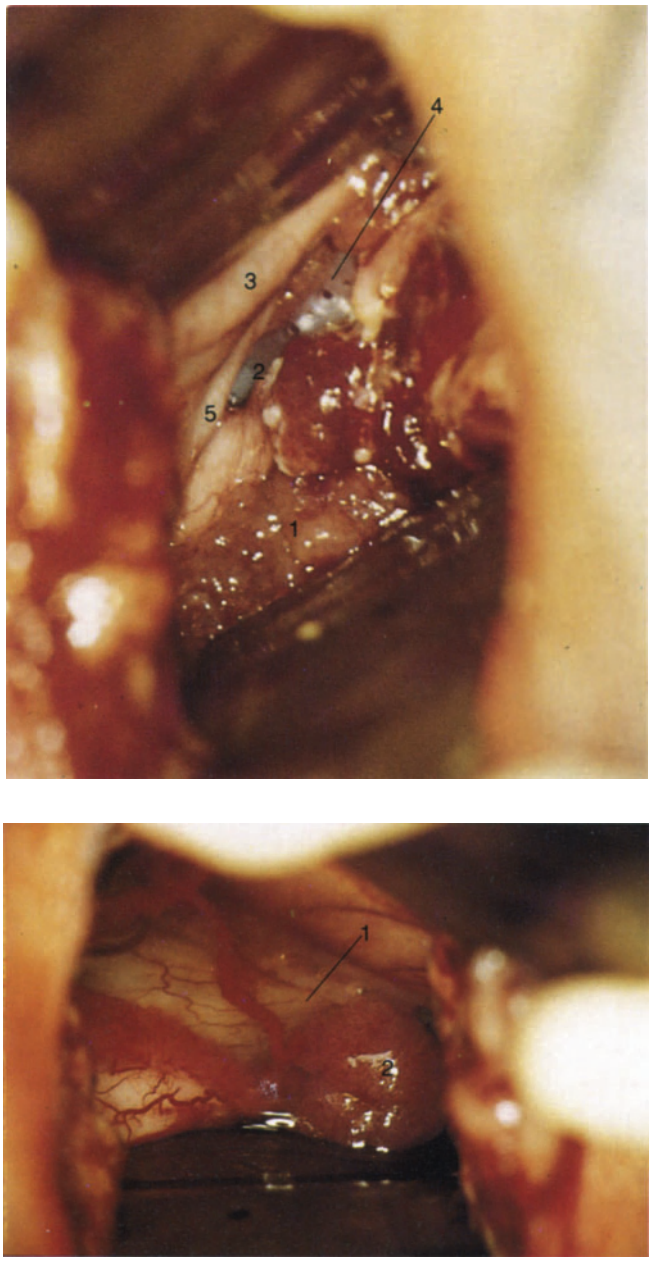


Figure 10.27. Technique and exposure for opening the septointerthalamic portal to the III ventricle: specifically, the attachment of the *lamina affixa* to the thalamus at the terminal sulcus, the line along which the choroid plexus also runs. Brushing the choroid plexus from the surface of the thalamus, 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 ventricle (*2*) to well up into the operative field, since there is now direct communication between the lateral ventricle and the III ventricle.

III ventricle, and which has the advantage of avoiding dissection along the columns of the fornix, entails stripping the tela choroidea of the roof of the III ventricle from its insertion onto the superior surface of the thalamus at 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, he must first assure himself 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 from its extension into the III ventricle, and subsequent removal of the tumor in two (lateral and III ventricular) portions, rather than one. It is not wise to attempt to lift the tumor from the lateral and III ventricles as one piece: the lateral ventricular mass "blinds" the surgeon to the vascular supply of the III ventricle mass.

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

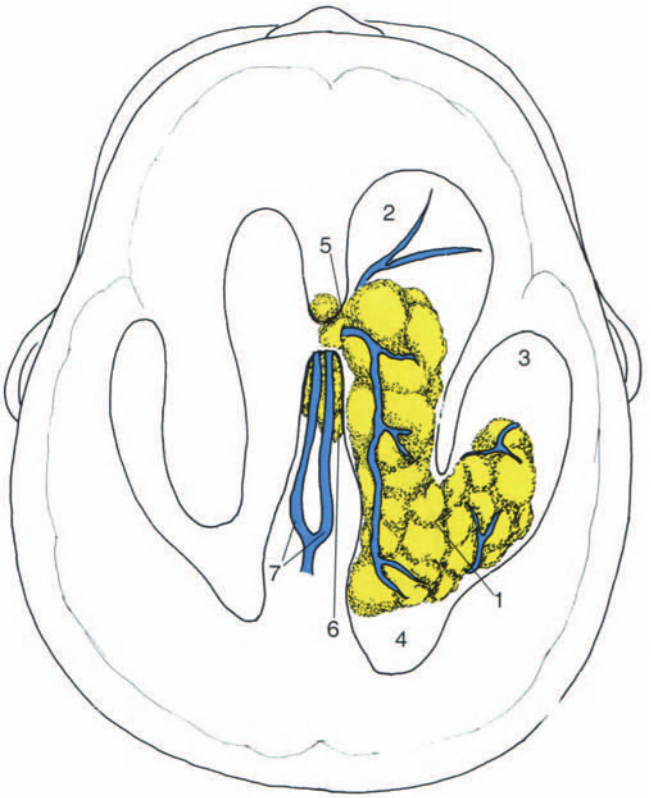


Figure 10.28. Diagrammatic representation of full axial view of tumor occupying the trigone (*1*), encysting the frontal (*2*), temporal (*3*), and occipital (*4*) horns, and extending through the foramen of Monro (*5*) into the III ventricle (*6*). The veins draining the tumor enter the internal cerebral vein at the foramen of Monro. Tumor in the III ventricle elevates the tela choroidea and deforms both internal cerebral veins (*7*).

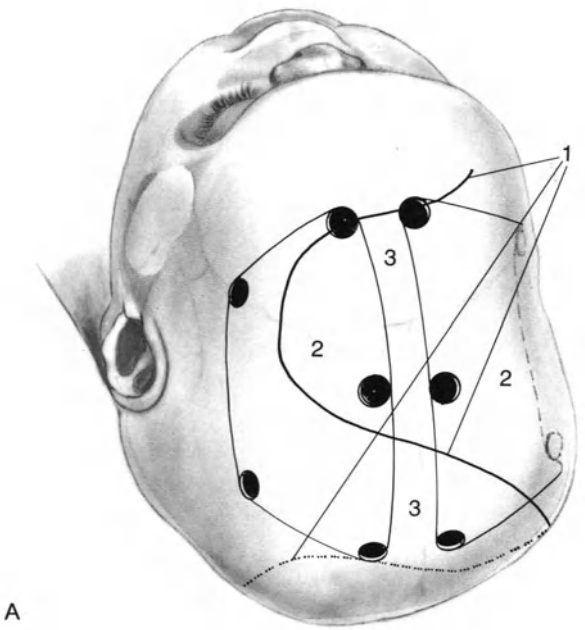
the floor of the III ventricle. When this is done, gently, and with soaking wet telfas, one avoids damaging the vascular or nuclear structures bordering upon the III ventricle and assures freeing of the intraventricular mass.

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 supraoptic recess. If the tumor has lobulated inferiorly, and 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 as far inferiorly as 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.

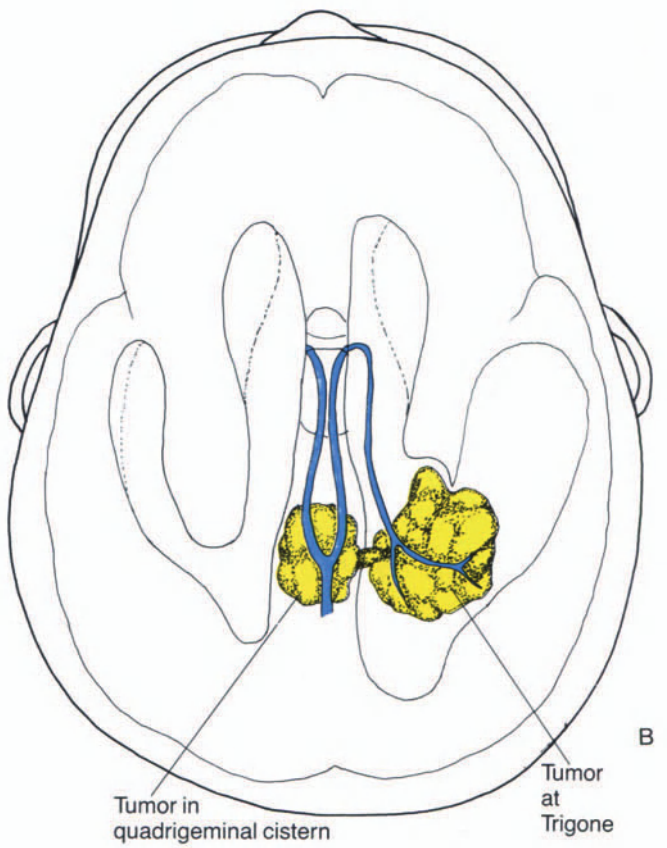
Lateral Ventricle Papilloma

Lateral ventricle papillomas (Fig. 10.29) may extend through the *choroidal fissure* into the *quadrigeminal cistern* and, at times, even into the *contralateral ventricle*. Those papillomas extending directly medialward through the choroid fissure, and into the quadrigeminal cistern, may either expand "dumbell 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 commissure 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.

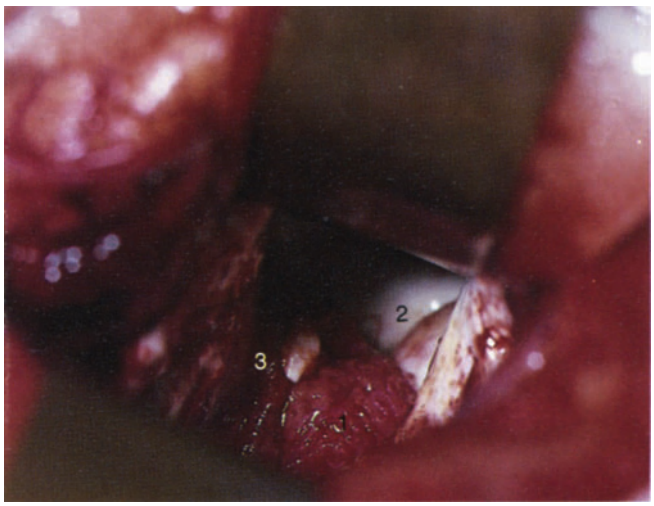
The tumor is fed through the medial and lateral branches of the posterior choroidal arteries on both sides: quadrigeminal and inferior-retrosplenial arteries. It drains into the internal cerebral veins (lesser Galenic system), the veins of Rosenthal, the greater Galenic sys-



A



B



C

Figure 10.29. (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 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 supra pineal 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.

tem, the supraculminate 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 after 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, diminishes 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 depending upon whether it has extended, dumbell fashion, only into the quadrigeminal cistern or has entered the contralateral ventricle.

Again, it is advisable to wait approximately 10 minutes 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 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, almost invariably grow superiorly into the aqueduct, and 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 in infancy and early childhood is located almost directly 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 expand superior to the roof of the III ventricle, in which case they are approached as roof of III ventricle tumors; remain within the pineal and quadrigeminal cisterns, in which case they are approached as posterior III ventricle tumors; or, they may 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 (Figs. 10.30–10.31) 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 valecula and cisterna magna inferomedially. Access to the cisterna magna, valecula, 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

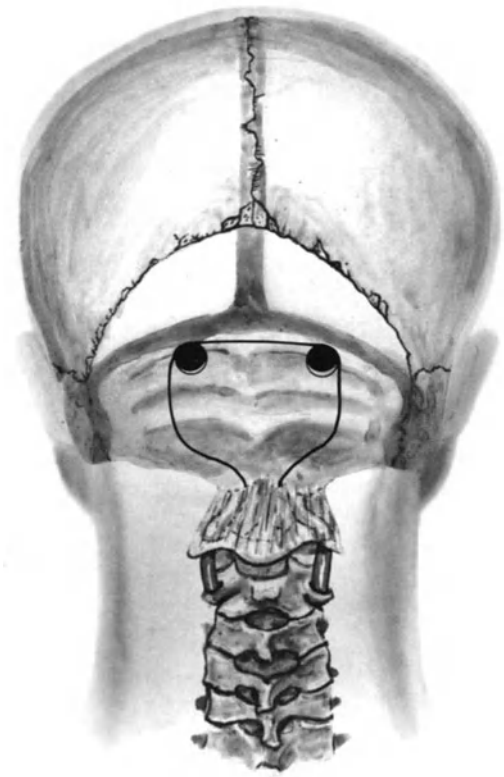


Figure 10.30. Inferior suboccipital craniotomy as viewed in a straight posteroanterior line, illustrating the burr holes and osteotomy lines outlining the free (squamous occipital) bone flap.

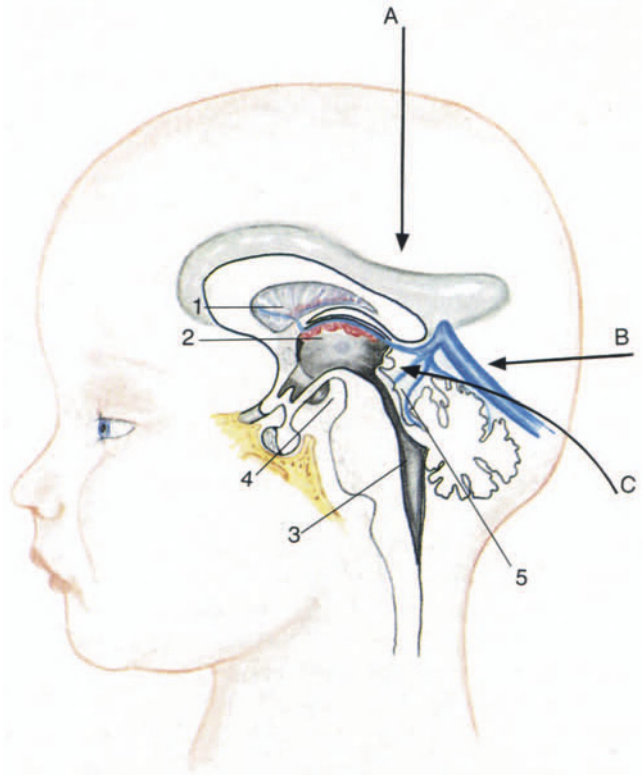


Figure 10.31. The midline ventricular system within the skull of a child. The septum pellucidum (*1*) is located anterosuperior to the III ventricle (*2*), which, in turn, is located anterosuperiorly to the IV ventricle (*3*). The very large basal (*4*) and quadrigeminal (*5*) cisterns surround the brainstem, which enters the cerebral hemispheres at almost a right angle. The three approaches, parasagittal (A), occipital (B), and infratentorial/supracerebellar (C) are indicated. This illustration clarifies the selection of surgical approach, which is based upon the direction of displacement of the deep venous structures.

paramedian structures, located at the point where the pontocerebellar junction meets 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 occpital 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 (Figure 10.32)

The III ventricle is extraordinarily diverse from an anatomical point of view, delicately situated at the inferomedial aspect of each hemisphere, wedged between the thalami and mesencephalon, and kept 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, 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 its inferior surface. If this were to be projected from the axial perspective,

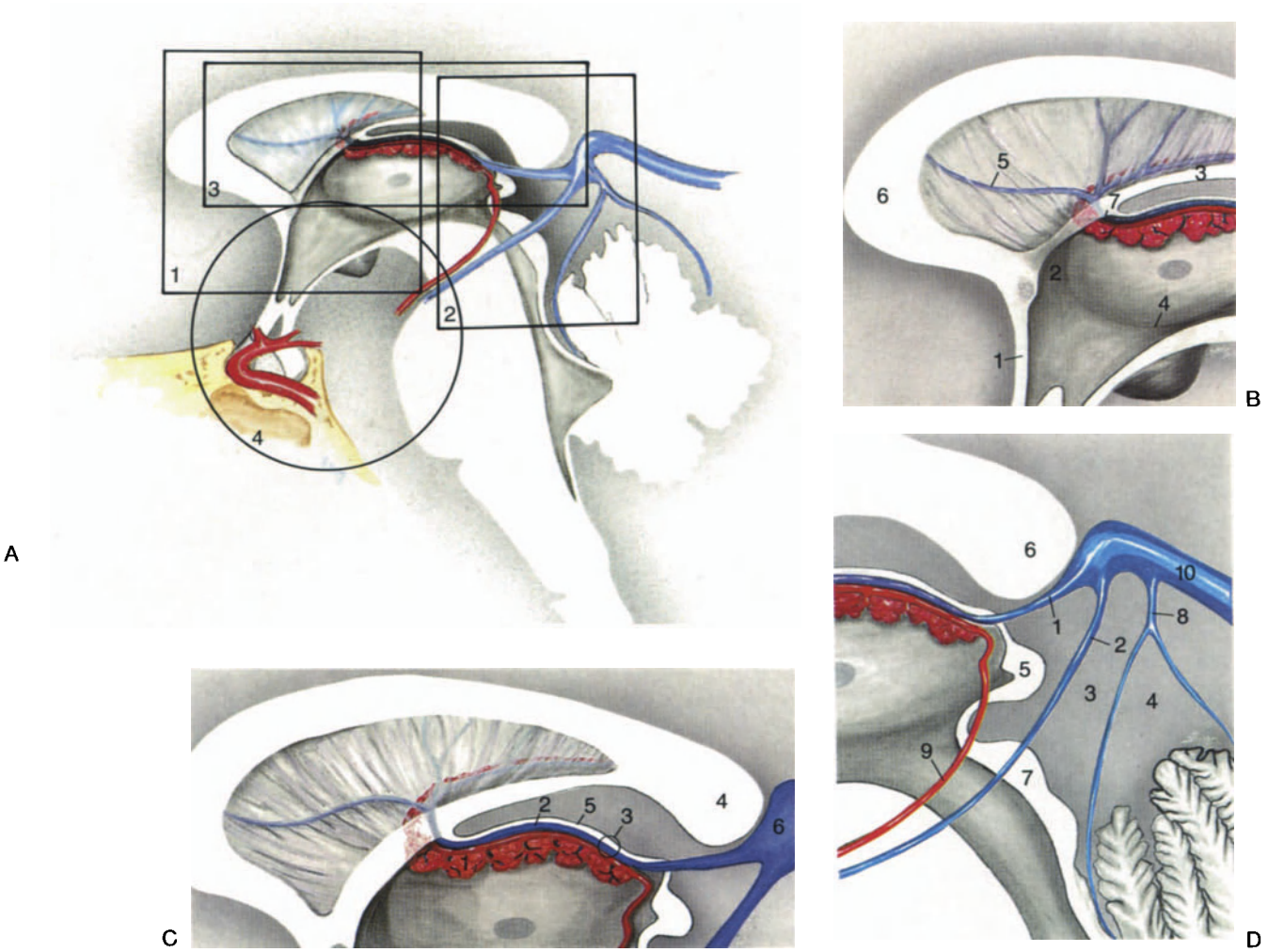


Figure 10.32. (A) One may reasonably divide the III ventricle into anterior (*1*), posterior (*2*), and superior (*3*) portions. This distinction is rational since the differences in these three sections of the III ventricle are both anatomical and functional, despite the fact that there is overlap between the superior and anterior portions anterosuperiorly, and the superior and posterior portions 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.

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 of 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, 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 structures within the roof may be damaged, then consideration should be given to entering the III ventricle through the lateral ventricle: separating the choroid plexus along the terminal sulcus as described for papillomas extending into the III from the lateral ventricle. This establishes an open communication between lateral and III ventricles.

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, translaminar 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.

Anterior III Ventricle Tumors

Anterior III ventricular tumors, consequently, are best approached through a right 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 by a transtentorial or transcallosal approach to the pineal region. The exception is those which expand primarily within the superior cerebellar cistern, displacing the great vein of Galen upward. In these cases, 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 (Figure 10.33)

The *subependymal glioma* may grow from the lamina terminalis. This is an extraordinarily 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*, and "relatively" is used here (in childhood) in a most generous sense. 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.

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 Penfield #4 dissector, running this latter instrument within the space between the rim of the foramen of Monro and tumor capsule. It should be directed from

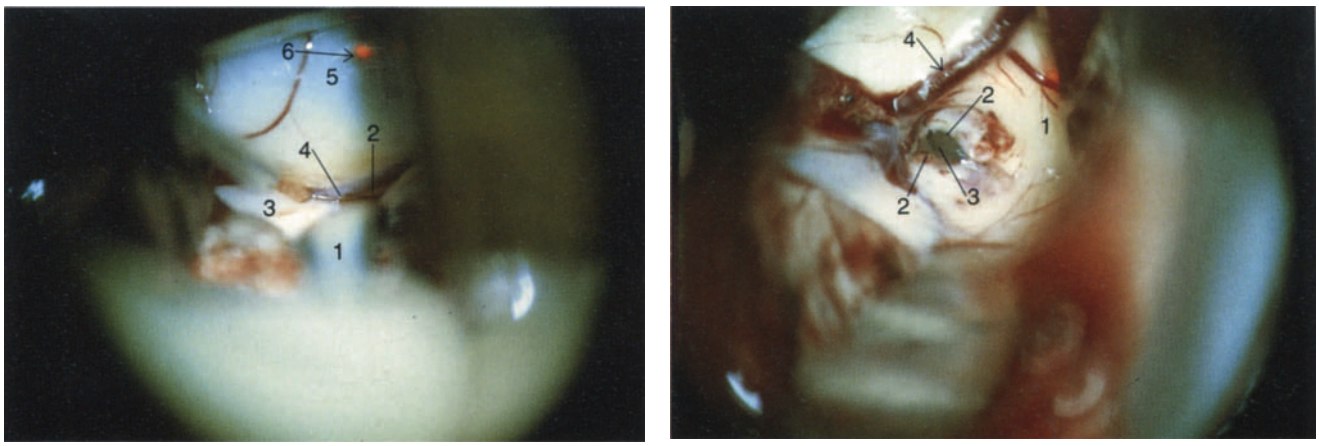


Figure 10.33. Transventricular approach to III ventricle colloid cyst. It does not matter whether one enters the lateral ventricles through the middle frontal convolution or along the parasagittal route and through the corpus callosum. In fact, the parasagittal route is quite easy in the child, though 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.

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. 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 cyst without opening it. If it is opened, inadvertently or intentionally, one may drain the contents and then work his way around the collapsed capsule, freeing it from the surrounding ependyma as he goes.

Tumors of the Root of the III Ventricle(Figure 10.34)
The only tumor which may be considered to grow from the roof of the III ventricle in childhood is the *choroid plexus papilloma.* Although maningiomas, theoretically, may occur here they do not grow in childhood and, for all intents and purposes, there is no clinical or surgical difference between meningioma 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 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.

The choroid plexus papilloma of the III ventricle is approached by using the septointerthalamic 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 (Fig. 10.35), using a medial parietal bone flap and parasagittal access to the corpus callosum, entails splitting of the body of the corpus

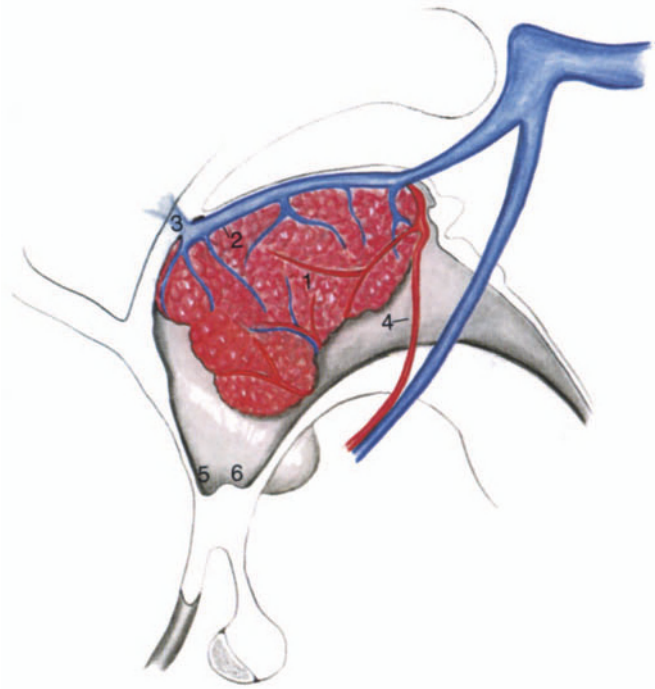


Figure 10.34. The papilloma (*1*) elevates the entirety of the internal cerebral vein (*2*) from the foramen of Monro to the suprapineal recess. The tumor always extends into the foramen of Monro (*3*). The major feeding vessel is the medial posterior choroidal artery (*4*). Posteroinferiorly, the mass may plug the aqueduct. It generally dilates the inferior III ventricle, deforming the supraoptic (*5*) and infundibular (*6*) recesses.

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 is split, the strawberry-red multilobulated tumor and the flattened internal cerebral veins immediately come into view. The tela choroidea should be cut 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 together as it is delivered. Once all of the tumor 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 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—lest one coagulate the internal cerebral vein—venous bleeding. After the tumor has been lifted from the III, its bed should be identified and then covered with Avitene so as to assure complete hemostasis.

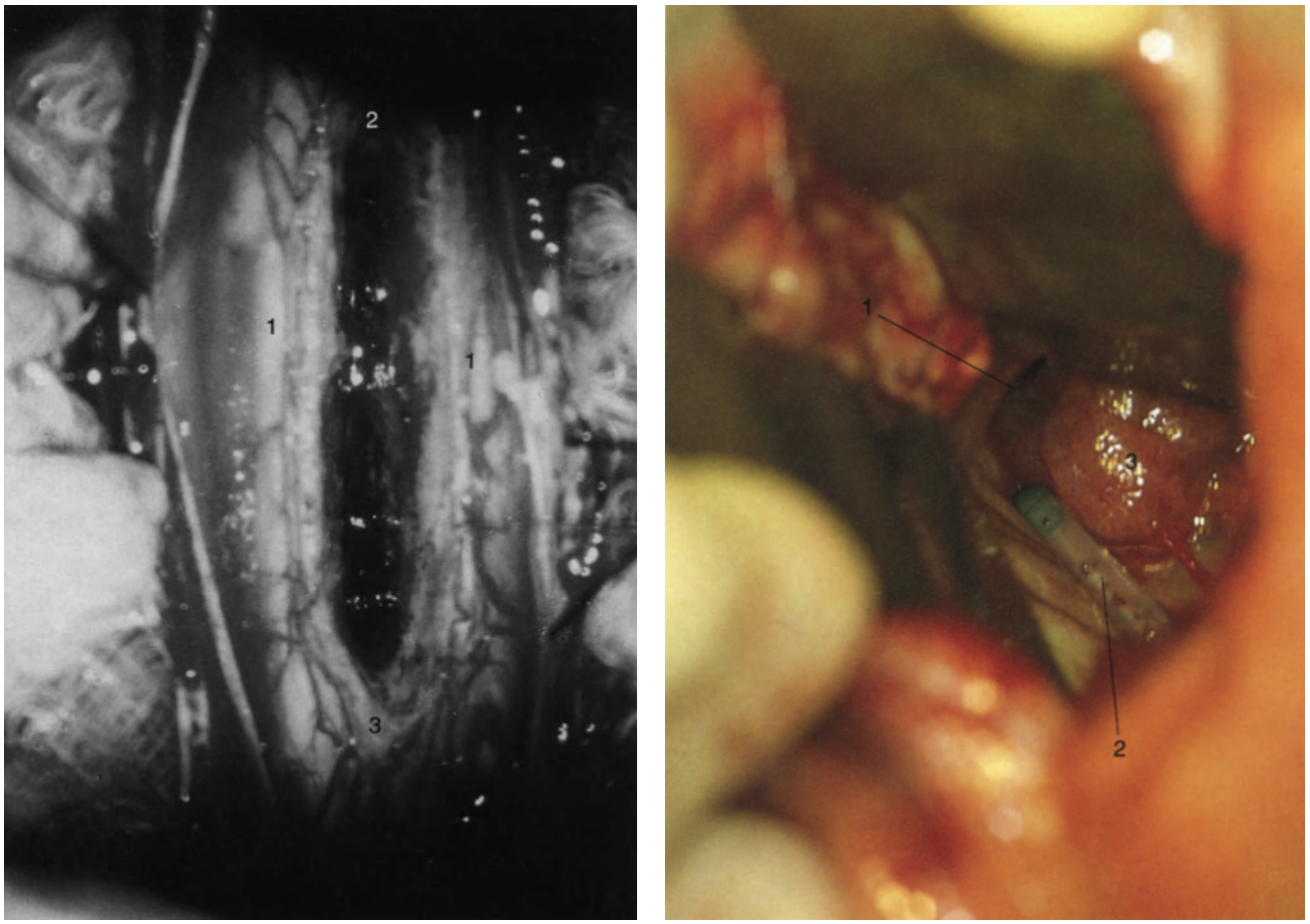
Transforaminal Approach
to Superior III Ventricle Tumors

If the transforaminal (transparenchymal, transventricular) (Fig. 10.36) 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 choroidal 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.

A great deal of confusion has resulted from the term pinealoma, first used by Krabbé[2] in 1923 and then adopted and disseminated by Cushing[3] in 1927 and Bailey[4] in 1948. Krabbé's definition of pinealoma was histological. The majority of tumors within the pineal region are not tumors of the pineal gland. In this text,



A B

Figure 10.35. The transcallosal approach to the III ventricle, presenting fewer difficulties than in the adult because of the smallness both of bridging cortical veins and Pacchionian granules, does require a more tedious dissection, transection of the corpus callosum, retraction of the internal cerebral veins located in the tela choroidea of the III ventricle. (A) The corpus callosum (*1*) is highly vascularized. A laser incision has been made in the body of the corpus callosum, extending from the posterior margin of the genu (*2*) to the splenium (*3*). (B) The split corpus callosum has been retracted and the roof of the III ventricle (*1*) opened. Since the septum pellucidum was also opened, the proximal end of the shunt tubing (*2*) is seen between the tela choroidea and a lobule of the III ventricle papilloma (*3*).

consequently, posterior III ventricular-pineal tumors will be described from a purely anatomical point of view as regards surgical technique.

One should, preferably, begin a discussion of "pineal tumors" with the description of the anatomical 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, surgical approach to tumors of the pineal region, and natural history of treated and untreated tumors in this area.

The same degree of sophistication of thought has recently been applied by Rubinstein[5], 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, entodermal yolk-sac tumors
2. Pineal cell tumors: pineocytoma, pineoblastoma
3. Glial tumors: astrocytoma, glioblastoma, oligodendroglioma, ependymoma
4. Other tumors: mesenchymal origin tumors such as meningioma, rhabdomyosarcoma, 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

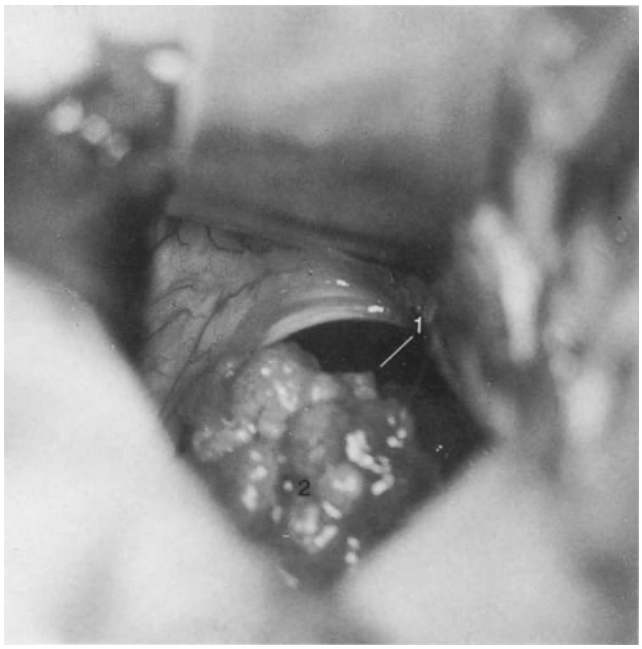


Figure 10.36. The transforaminal (transparenchymal, transventricular) approach to the foramen of Monro (*1*) also provides access to papillomas (*2*) within the III ventricle. It, as the transcallosal approach, is less desirable than the transventricular/septointerthalamic approach previously described. The disadvantage of this (transforaminal) approach to the tumor is that the tumor is on the other side of the foramen of Monro, behind the anterior septal vein and beneath the internal cerebral veins. Consequently, one may encounter bleeding behind the tumor which he is unable to control. Also, unfortunately, the use of the laser for transforaminal removal of III ventricle papillomas is much more complicated and dangerous than in the septointerthalamic exposure, which permits one to retract the internal cerebral veins. The reason for this is that for the transforaminal approach, the surgeon always has the tumor on the other side of normal structures. Through a frontal cerebrotomy, the right lateral ventricle is entered and the foramen of Monro identified. The III ventricle papilloma is plugging the foramen of Monro.

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 entire tumor mass before being able to identify with precision the histological nature. It is worthy of note that craniopharyngioma has been reported to occur in the pineal region. *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 entites. In addition to this, one notes rather remarkable effectiveness of treating the complicating bi- or triventricular hydrocephalus, successful results of radiation therpy, 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 probabilities 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 or removed completely, but whether direct surgical attack of the tumor is preferable to Roentgen therapy and, if so, whether the parasagittal, occipital, or suboccipital approach should be used. In the past, the decision to operate or radiate 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 interest of the patient to radiate the lesion rather than to operate. The present state of the art concerning the use of such biological markers as human chorionic gonadotropin, carcinoembryonic antigen, and alpha-fetoprotein as indicators of germ cell tumors is such that one may proceed directly with Roentgen therapy if cerebrospinal fluid analysis reveals the presence of one or more of these markers. Though positive cerebrospinal fluid cytology studies for malignant cells do not permit the precise identification of the specific tumor type, they do 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 is necessary when the cerebrospinal fluid studies are negative.

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 entirety 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, and
4. inferiorly, along the collicular plate and invading or displacing the lobulus centralis and culmen monticuli of the cerebellar vermis.

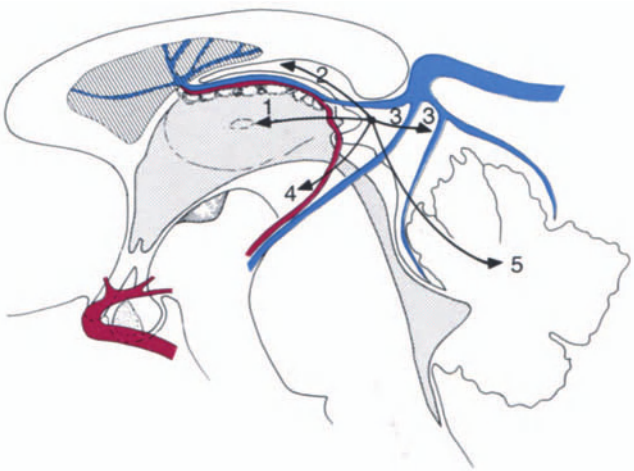


Figure 10.37. Routes of pineal tumor expansion and invasion (*1*) into the III ventricle; (*2*) between the splenium of the corpus callosum and the roof of the III ventricle; (*3*) into the quadrigeminal and supracerebellar cisterns; (*4*) into the brainstem; (*5*) along the anterior medullary velum and into the cerebellar vermis.

Intraparenchymal extension into the pulvinar of the thalamus, the tectum, or the hippocampus is not unusual. Similarly, multidirectional extension into, for example, the infrasplenial and trigone areas, of the III ventricle and quadrigeminal cistern, are common observations. One may expect a pineal tumor to be intraventricular, intracisternal, intraparenchymal, or any combination of the three (Fig. 10.37).

Metrizamide cisternography, with the use of the computed transmission tomography (CTT) scanner, puts into relief the anatomical interrelationships between the midbrain and the cisterns surrounding it (Fig. 10.38). These figures are quite illustrative of the many considerations which surgery of tumors of the pineal region oblige the surgeon to make.

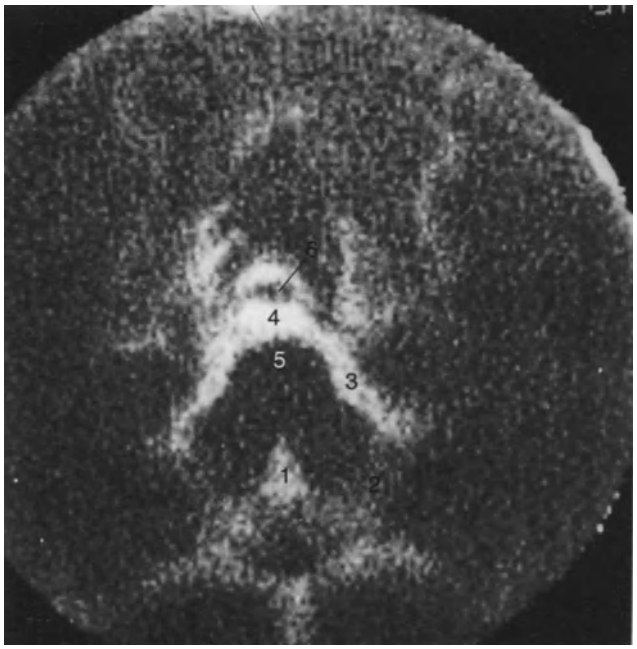
It is an accepted fact that presently there is no difficulty whatsoever diagnosing the presence of a pineal tumor or outlining its size and extent, since sophisticated CTT studies reveal the presence and distribution of calcification, the form and volume of the neoplasm, and the extent of peritumoral edema.

These data *do not* provide the surgeon with adequate information to plan the most suitable approach for direct surgical attack of the neoplasm, since they give no information concerning location and identification of specific venous (Galenic and supraculminate) and arterial (medial and posterior choroidal) systems. Cerebral angiography provides the surgeon with the necessary information for the identification of these vessels and the determination of their displacement (Fig. 10.39). In addition to this, it reveals information concerning the association of teratomatous lesions with arteriovenous malformations in the pineal region (Fig. 10.40).

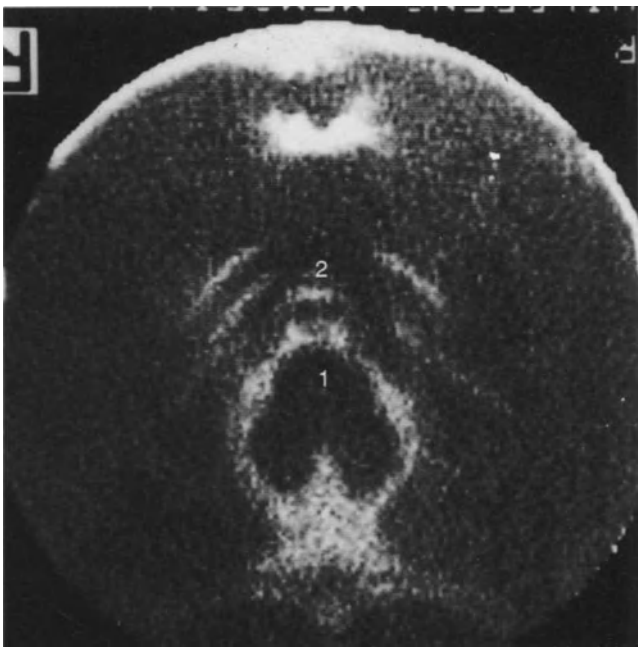
There has been considerable literature published concerning the three basic approaches to tumors of the pineal region: the parasagittal/transcallosal approach of Dandy[6], the occipital/transtentorial approach of Poppen[7] and Jamieson[8], and the suboccipital/supracerebellar approach of Krause[9]. It is my 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 vein of Galen in order to biopsy or resect a pineal tumor.

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 with laser vaporization, or 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.

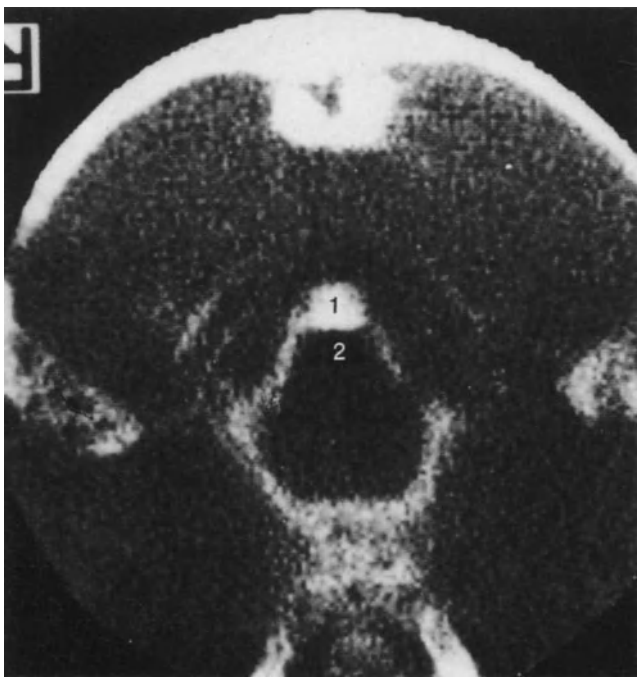
If resection of a benign tumor proceeds smoothly,



A



B



C

Figure 10.38. (A) Metrizamide cisternogram outlining the midbrain and pineal region. One notes the interpeduncular (*1*), crural (*2*), ambient (*3*) and quadrigeminal (*4*) cisterns, as well as the upper midbrain (*5*) and the pineal gland (*6*). (B) In addition to the cisterns demonstrated in A, one notes the outline of the mesencephalon (*1*) and better visualization of the pineal gland (*2*). (C) The quadrigeminal cistern (*1*) posterior to the pineal gland, over the inferior colliculi (*2*), is completely filled.

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.

Intra(III)ventricular Pineal Tumors: Suboccipital, Supracerebellar Approach
(Figures 10.41 to 10.43)
The technique for removal of intra(III)ventricular pineal tumors is identical to that recommended for removal of choroid plexus papilloma of the III ventricle. The only specific considerations for the pineal tumor have to do with its vascularity, since this tumor does take 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 when it is not completely neoplastic, in order to occlude individual feeding vessels to the tumor mass.

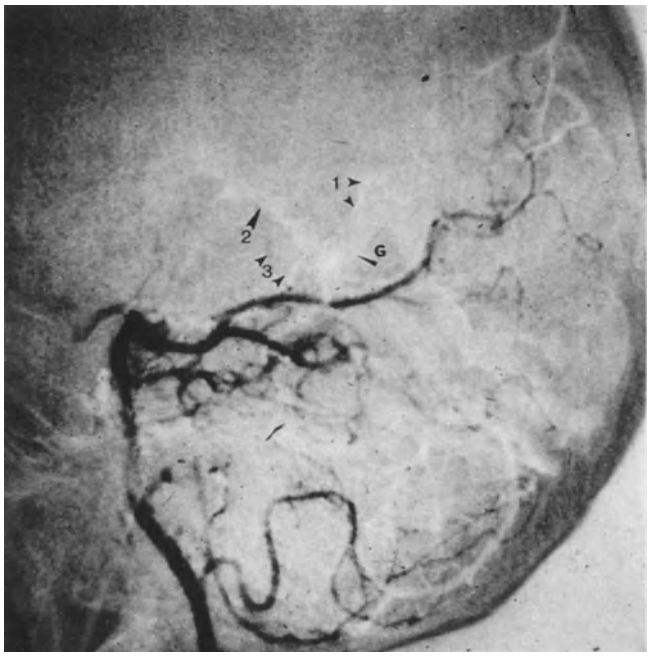


Figure 10.39. A double superimposition subtraction print illustrating arterial and venous systems. The vein of Galen (*G*) is normal but it appears that a posterior ventricular vein (*1*) is stretched, deformed, and displaced posteriorly, and that the venous stain within the roof of the III ventricle (*2*) bulges superiorly. The medial posterior choroidal artery (*3*) is not deformed.

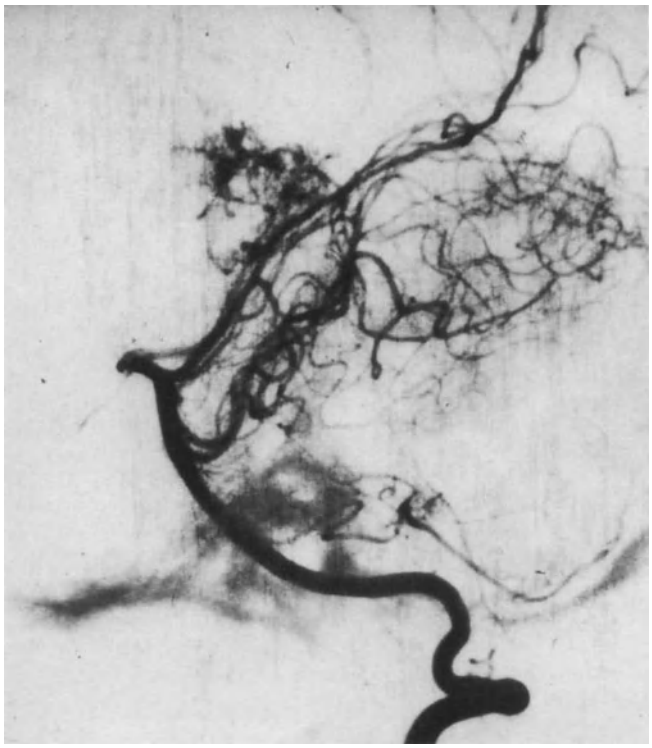


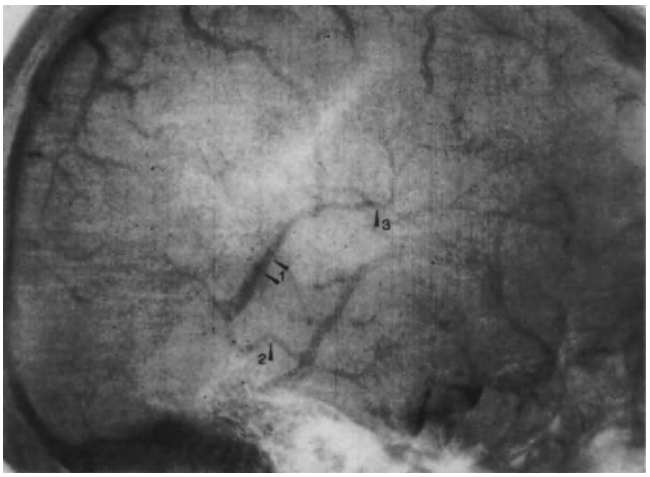
Figure 10.40. A 9-year-old boy with a teratoma in the pineal region. Note the vascular anomaly in the pineal region. The boy had a teratoma occupying the III ventricle, the quadrigeminal and supracerebellar recesses, and the trigones of both lateral ventricles.

When cerebral 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 invariably 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 he may not predict with confidence whether both foramina of Monro are patent.

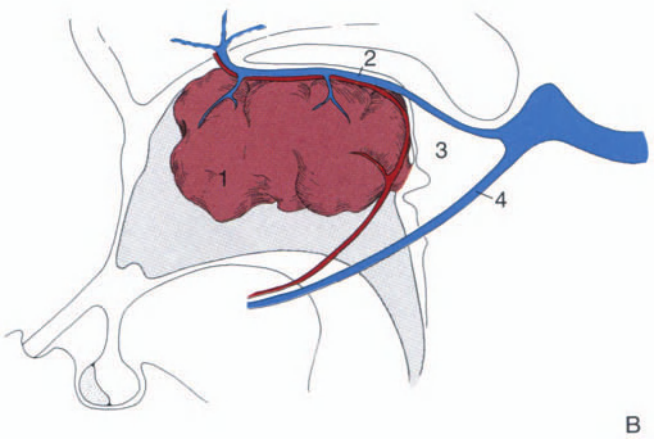
Elevation of the internal cerebral vein, as revealed by cerebral angiography, secondary to superior displacement by a mass within the III ventricle, suggests the desirability of avoiding a parasagittal/transcallosal approach to the tumor, since the surgeon would be obliged to work around the two internal cerebral veins in order to gain access to the inferiorly located tumor mass. Suboccipital craniotomy permits an infratentorial/supracerebellar approach to the pineal region, and excellent exposure both of 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 atlantooccipital 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 hemispheres. It is convenient to place a self-retaining retractor over the cerebellar hemispheres, both for 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 efficient, rapid, and convenient. If used, however, care must be taken to identify the anatomy of the area precisely, lest the supraculminate vein be inadvertently 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 should take note of the fact that the splenium of the corpus callosum may be draped around the (posterior III ventricular) pineal



A



B

Figure 10.41. (A) When the pineal tumor extends into the posterior portion of the III ventricle and expands primarily beneath the internal cerebral veins, it causes two very characteristic venographic changes: elevation of the internal cerebral vein, at times converting its posterior portion into an almost rectilinear structure (*1*); and stretched deformity of the vein of Rosenthal, resulting in a hooklike transformation of that portion of this vein, which is located within the confines of the ambient and quadrigeminal cisterns (*2*). Further expansion of the tumor mass within the III ventricle allows it to extend anteriorly beyond the venous angle, elevating this structure (*3*), and occluding the foramen of Monro. (B) The intraventricular pineal tumor (*1*) elevates the internal cerebral vein (*2*), allowing one to conceptualize the advantages of working through the quadrigeminal cistern (*3*) and between the veins of Rosenthal (*4*).

tumor so that the neoplasm itself 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 completely the posterior aspect of the neoplasm, permitting identification of the feeding vessels.

It is best to resect the tumor by working initially at 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 of the tumor along its border with the tentorial edges, gently freeing fibrinous adhesions, coagulating and transecting more dense or vascular bridges. The tentorial edges should be freed completely. Once this is accomplished, the superior cerebellar, internal occipital, and posterior choroidal arteries are separated from the surface of the tumor. It is not possible, nor advisable, to attempt to free the arterial and venous structures independently. Rather, each vascular structure along the tentorial edge, within the posteroinferiormost portion of 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 single 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 tear. 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 should be left alone.

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, should be used to hold the splenium upward, so that one may follow the dome of the tumor into the III ventricle, which it invariably 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 should be coagulated with bipolar forceps. Microscissors are used to cut along the line of coagulation. If the neoplasm is continuous with the brachium of

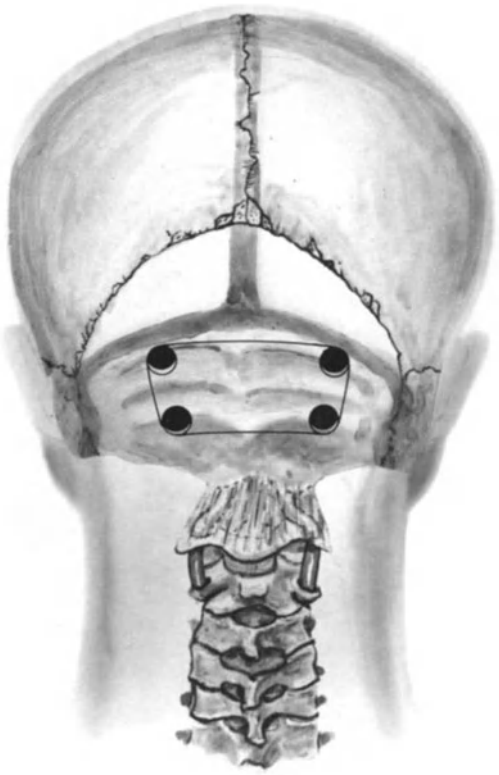


Figure 10.42. Craniotomy for suboccipital/supracerebellar approach to pineal tumors. A craniectomy is not recommended.

the superior colliculi, the resection must be stopped at least 2 mm superior to this structure.

Freeing the tumor entirely 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.

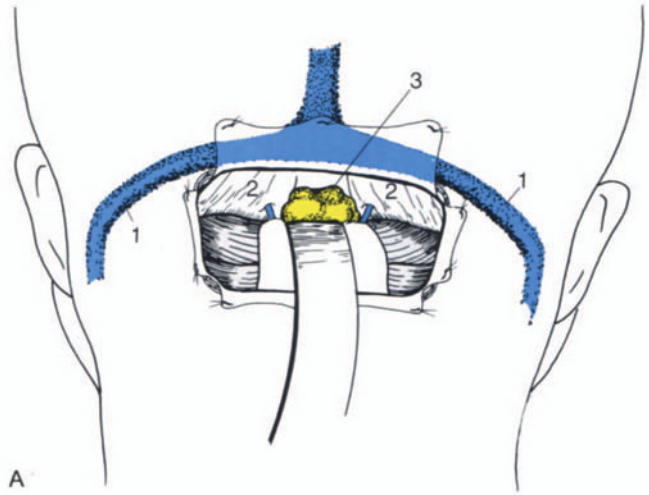
One may now direct himself to removing the inferior pole of the tumor. This is generally not a difficult procedure, but may prove to be impossible if the tumor has invaded the culmen monticuli. Its resection should be effected with the same technique as used for debulking the mass and removing its superior pole, though one may exert gentle traction on the inferior pole in order both to elevate it from the underlying superior cerebellar vermis and identify bridging vessels, which are to be coagulated and transected.

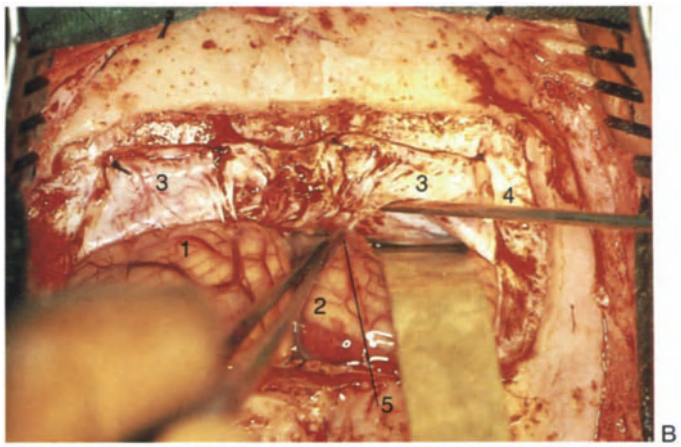
After the entire mass has been removed one should see the III ventricle, the centrencephalic venous system, the superior cerebellar and internal occipital arteries, the collicular plate, the inferior portion of the splenium, and the medial surfaces of the isthmus of the hippocampus.

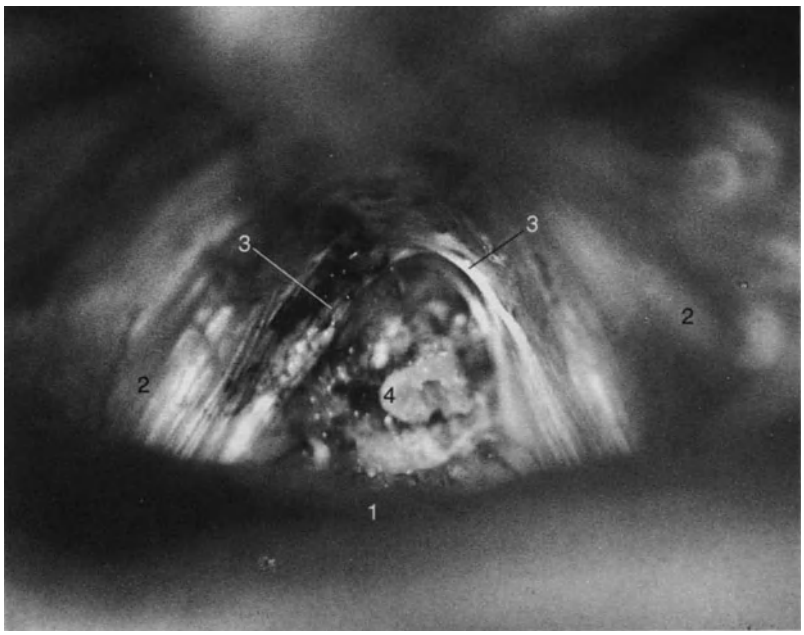
Superior to III Ventricle Pineal Tumors: Parasagittal Approach (Figures 10.44 to 10.46)

When the internal cerebral vein and the great vein of Galen are depressed, this is indicative of primary extension of the tumor mass into the interval between the body of the corpus callosum and the roof of the III ventricle. 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







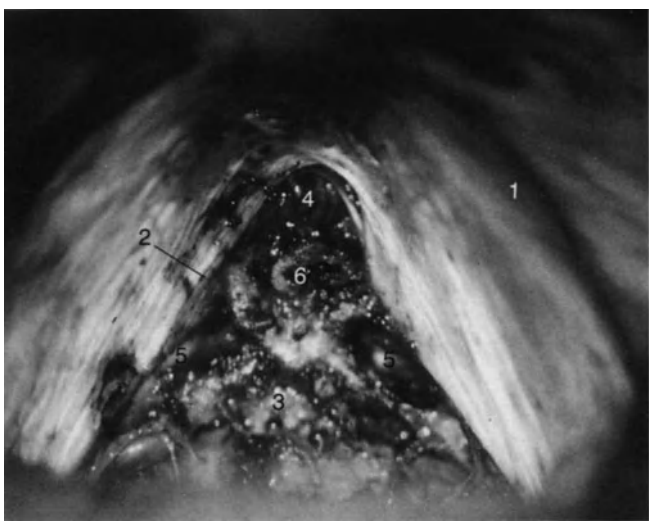
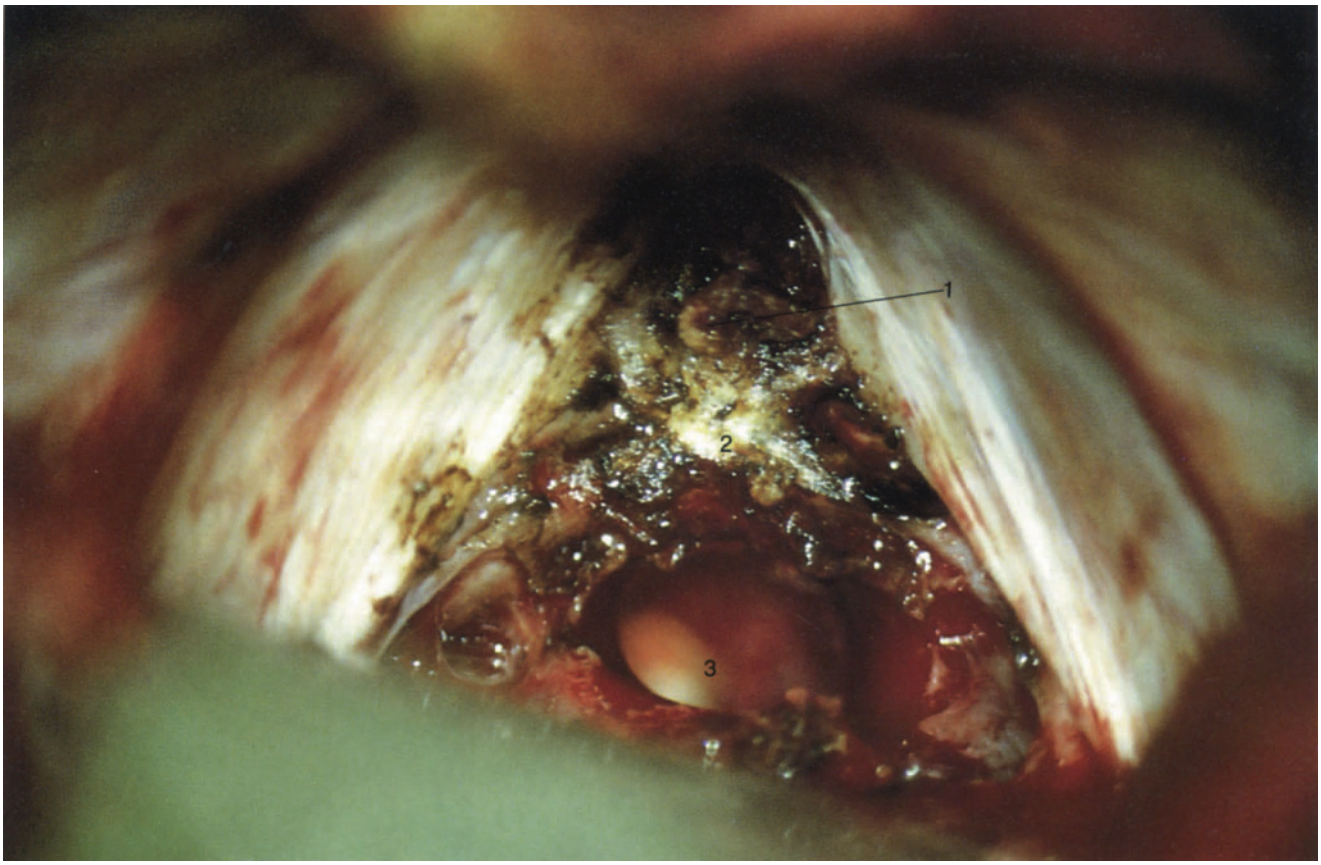
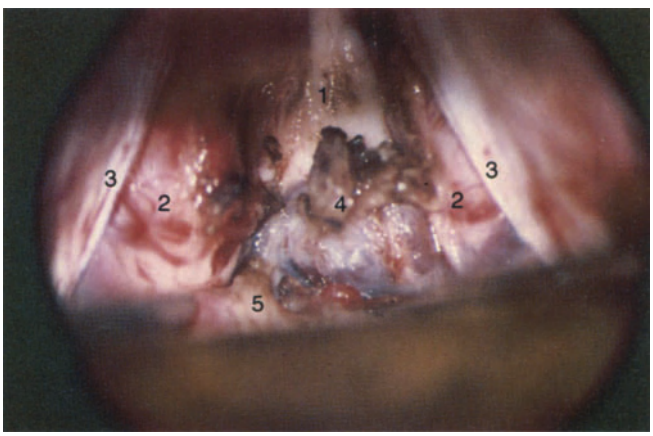


Figure 10.43. (A) The infratentorial/supracerebellar approach. Note that the dura is opened all the way to the edge of the transverse sinus (*1*) in order to give complete exposure of the inferior surface of the tentorium (*2*), exposing tumor (*3*) within the tentorial opening. (B) The cerebellar hemisphere (*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. (C) After the bridging veins from the superior surfaces of the cerebellar hemispheres and vermis to the transverse sinus and tentorium have all been coagulated and transected, a large telfa is lain over the surface of the cerebellum (*1*). This exposes the cerebellar surface of the tentorium (*2*) bilaterally, the tentorial ring (*3*), and, after the arachnoid of the superior cerebellar cistern has been re- ►

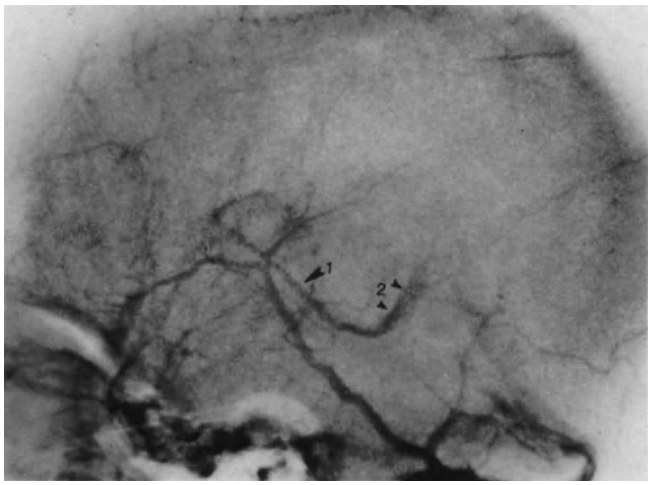


E

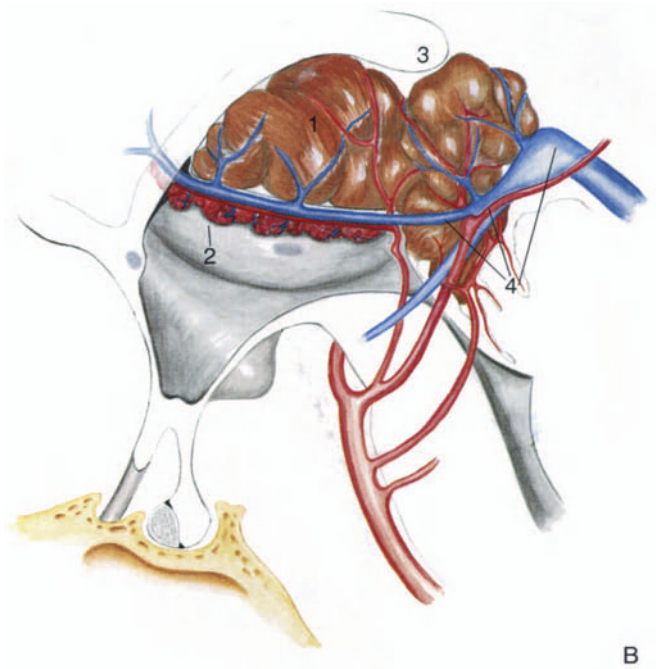
sected, the pineal tumor (*4*). (D) The cerebellar surface of the tentorium (*1*) and the tentorial opening (*2*) are shown in this photograph taken through the operating microscope. Most of the tumor has been vaporized with a CO_2 laser, though small fragments (*3*) remain beneath the most posterior portion of the great vein of Galen (*4*), between the two veins of Rosenthal (*5*). One sees, also, neoplastic pineal gland immediately *anterior* and *superior* to the great vein of Galen, projected *beneath* (*6*) the vein of Galen because the operating microscope and camera cannot "see" in three dimensions. (E) 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. (F) 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*).



F



A



B

Figure 10.44. (A) Extension of pineal tumor between the roof of the III ventricle and the corpus callosum. The internal cerebral vein (*1*) and great vein of Galen (*2*) are displaced inferiorly and posteriorly, as indicated by the directions of the long and short arrows. (B) The 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 centrencephalic venous system (*4*).

the body of the corpus callosum. Splitting of the body (*not the entirety*) of the corpus callosum exposes the tumor mass 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, he may encounter lobules of tumor extending superiorly, over the great vein of Galen, and inferiorly, wedged between the internal cerebral and basal veins. Since it is in this interval that the pineal gland normally is located, 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, however, the bleeding may be stopped with use of Avitene and fluffy cottons. This event 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 at a time, until he has the impression that he has removed all the tumor 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.

III Ventricle Pineal Tumors: Posterior and Inferior Occipital/Transtentorial Approach
(Figures 10.47 and 10.48)

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 of the vermis, 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.

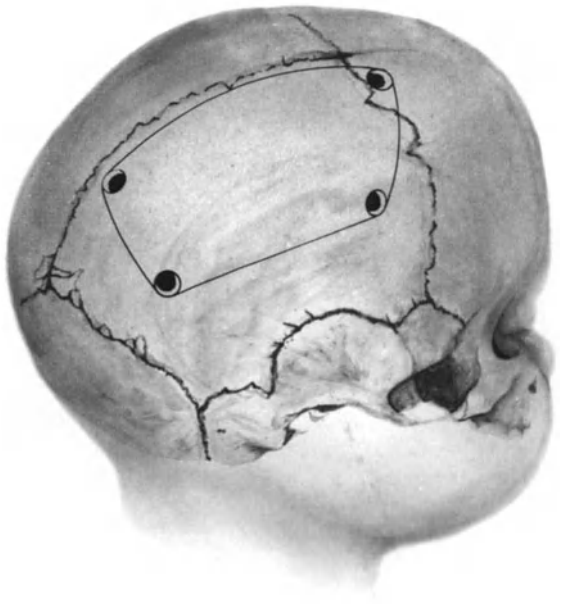


Figure 10.45. Placement of burr holes for frontoparietal craniotomy for approach to tumors located between roof of the III ventricle and the corpus callosum. This provides excellent exposure of the tumor and access to the anterior and posterior portions of the roof of the III ventricle, without need to operate around the internal cerebral veins for access to the tumor.

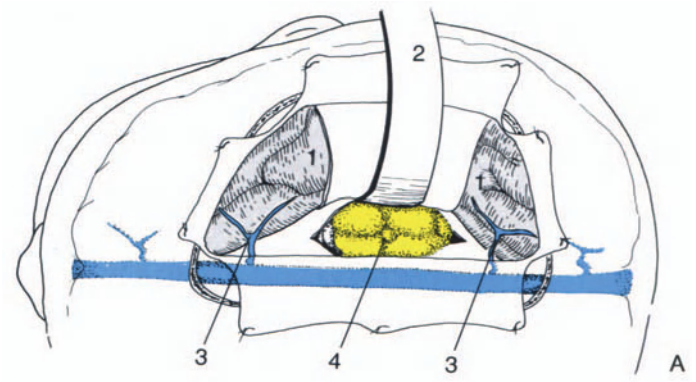
An occipital flap, exposing both the superior sagittal and the transverse sinuses, provides the surgeon adequate space to perform a wide incision of the tentorium cerebelli and exposure of falx cerebri as well as the supra- and infratentorial areas. The tentorium cerebelli should be coagulated on a line which runs obliquely from the transverse sinus to the tentorial edge, using the microbipolar forceps, with the blades separated by a distance of 1 to 2 mm, and applying a drop of water so as to coagulate as completely as possible the surface of the tentorium. The line of coagulation should never 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 technique, 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 splenium, the collicular plate, the isthmus of the hippocampus, and the superior portion of the cerebellar vermis must all be inspected and reinspected. At the end of the procedure, however, one does not have a complete view into the III ventricle: his line of sight permits him to see only the posterior portion of the floor of the III ventricle and the iter to the aqueduct.

Fourth Ventricular Tumors

(Figures 10.49 to 10.51)

The term "IV ventricular tumors" is intended to describe those tumors which occupy completely and primarily, but by no means exclusively, the IV ventricle.



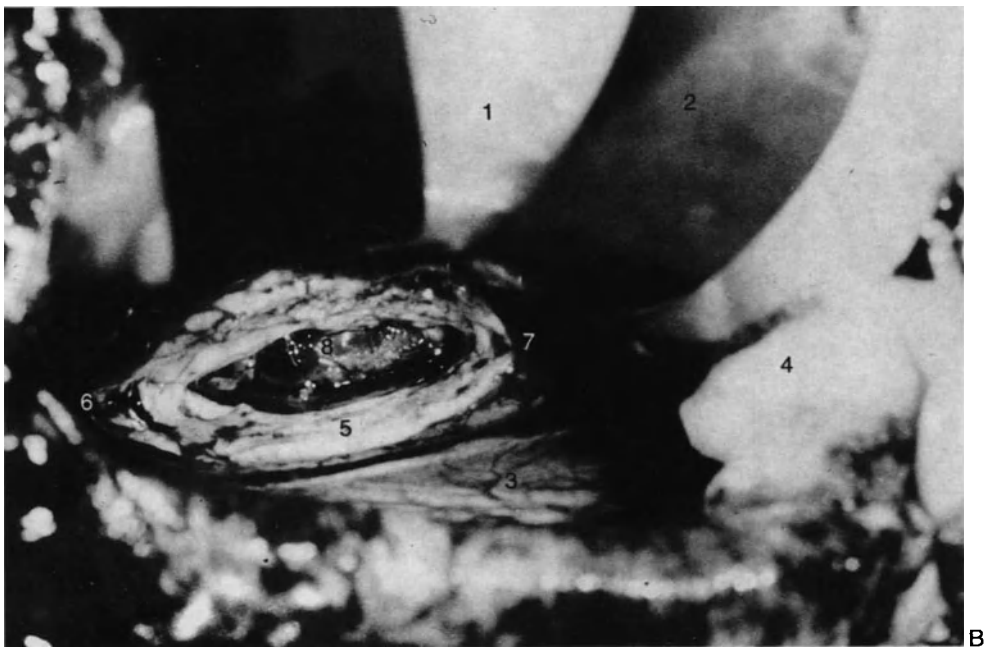
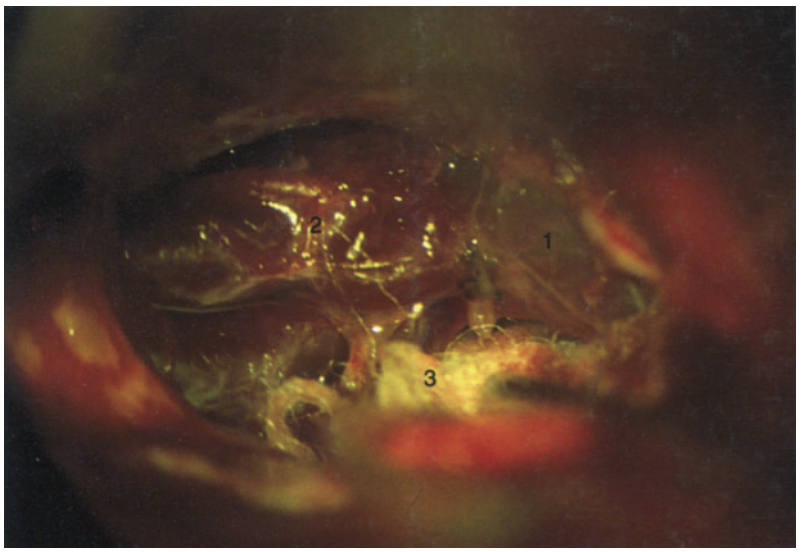
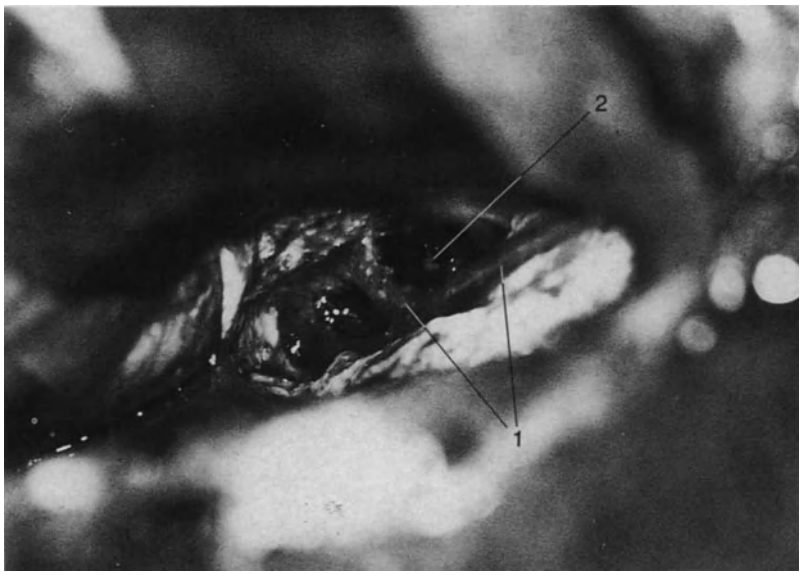


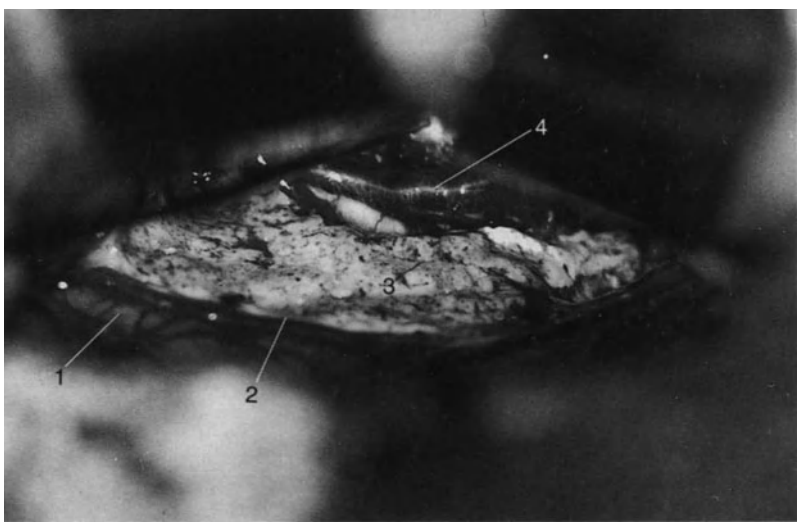
Figure 10.46. (A) Diagrammatic representation of the parasagittal approach to pineal tumor. The child is operated supine with the head turned entirely to the left. The parietal lobe (*1*) is elevated with a self-retaining retractor (*2*). The purpose for elevating the parietal lobe rather than allowing gravity to retract it out of the field (which one could accomplish simply by turning the head to the right), is to give the surgeon complete control over the degree of retraction. If gravity is used, uncontrollable traction of the bridging cortical veins results and the patient may suffer cerebral infarction from venous stasis. This technique permits one to control the degree of stretch upon the bridging cortical veins (*3*) as the tumor (*4*) is exposed. (B) The parietal lobe has been covered with telfa (*1*) and retracted superiorly, using two De Martel blades (*2*). One sees the falx cerebri (*3*) and a fluffy cotton (*4*), which is covering a bridging cortical vein in order to keep it moist and protect it. The body of the corpus callosum (*5*) has been split, but the section of the commissure has extended neither toward the genu (*6*) nor the splenium (*7*). The tumor (*8*) is seen bulging through the opening in the body of the corpus callosum. (C) 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. (D) All of the tumor has been removed, exposing the underlying veins (*1*) within the roof and the cavity of the III ventricle (*2*). (E) The cingulate gyrus (*1*), pericallosal artery (*2*), partially incised body of the corpus callosum (*3*), and internal cerebral vein (*4*) are now back in normal anatomical juxtaposition. ►



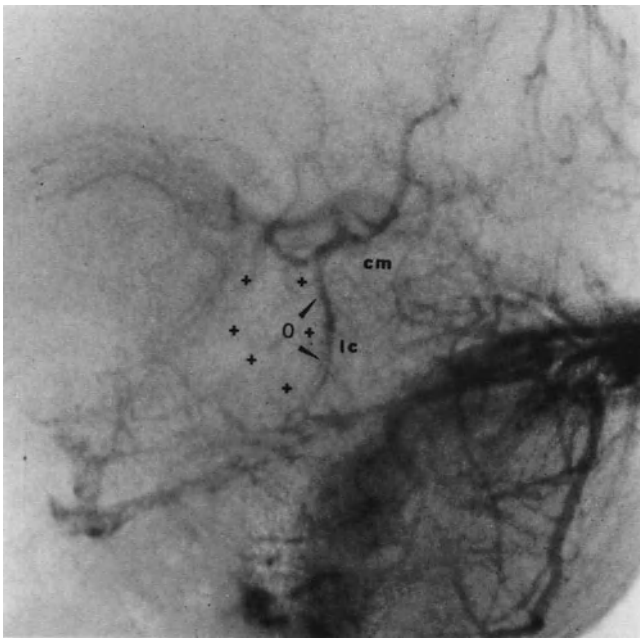
C



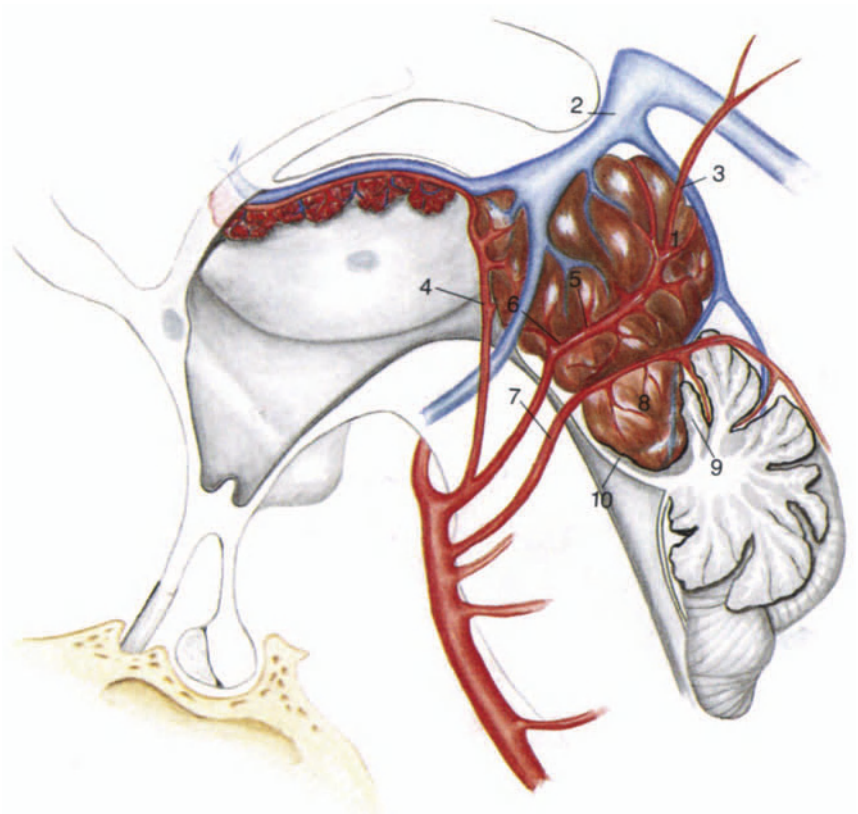
D



E



A



B

Figure 10.47. (A) Venographic study of the posterior fossa is of extreme value in that it allows one to identify, with precision, expansion of the tumor mass into the precentral and supraculminate veins (o) as they course superiorly in a vertical manner to enter the great vein of Galen. It is the tumor mass anterior to this vein which displaces the lobulus centralis (lc) and culmen monticuli (cm) posteriorly, producing this rather characteristic deformity, indicating that the tumor (+) is located anterior to this venous system. (B) 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*).

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, 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.*

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.

Medulloblastoma, brainstem 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 glioma are described in this section only to apprise the reader of its characteristics in the event it is encountered at surgery, not to recommend operating it for biopsy, "debulking," or "cyst" drainage. There is no reason to conclude that computed transmission tomography 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.

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-advised. One may, with some degree of certainty, determine whether the tumor is *primarily* in the vermis, the floor of the IV ventricle, the cerebellar peduncles. He may, almost invariably, identify tumors within the brainstem, and invariably localize those in the cerebellar hemispheres or the foramen magnum. Consequently, the decision whether to operate or to treat only with Roentgen therapy is predicated upon the anatomical location of the tumor: *Expanding masses within the brainstem are treated with Roentgen therapy only, those anywhere else in the posterior fossa, even if exophytic masses pedunculated from the brainstem, should be operated.* The simple fact that one may operate an intraparenchymal brainstem tumor and have the child survive is no justification for surgery. The only justifications for surgery here, as in all other instances of neurological pathology, are either to (1) cure the patient, (2) make the patient better, or (3) prevent the patient from worsening. Because, in brainstem tumors, there is no correleation between histological appearance of the space-occupying lesion, operative procedure performed, and quantitiy or quality of survival, it is not in the child's interests to undergo an operation.

Fourth ventricular tumors should be removed through an inferior cerebellar triangle craniotomy. It is not necessary to remove the arch of the atlas. 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 he 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 valeculla 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.

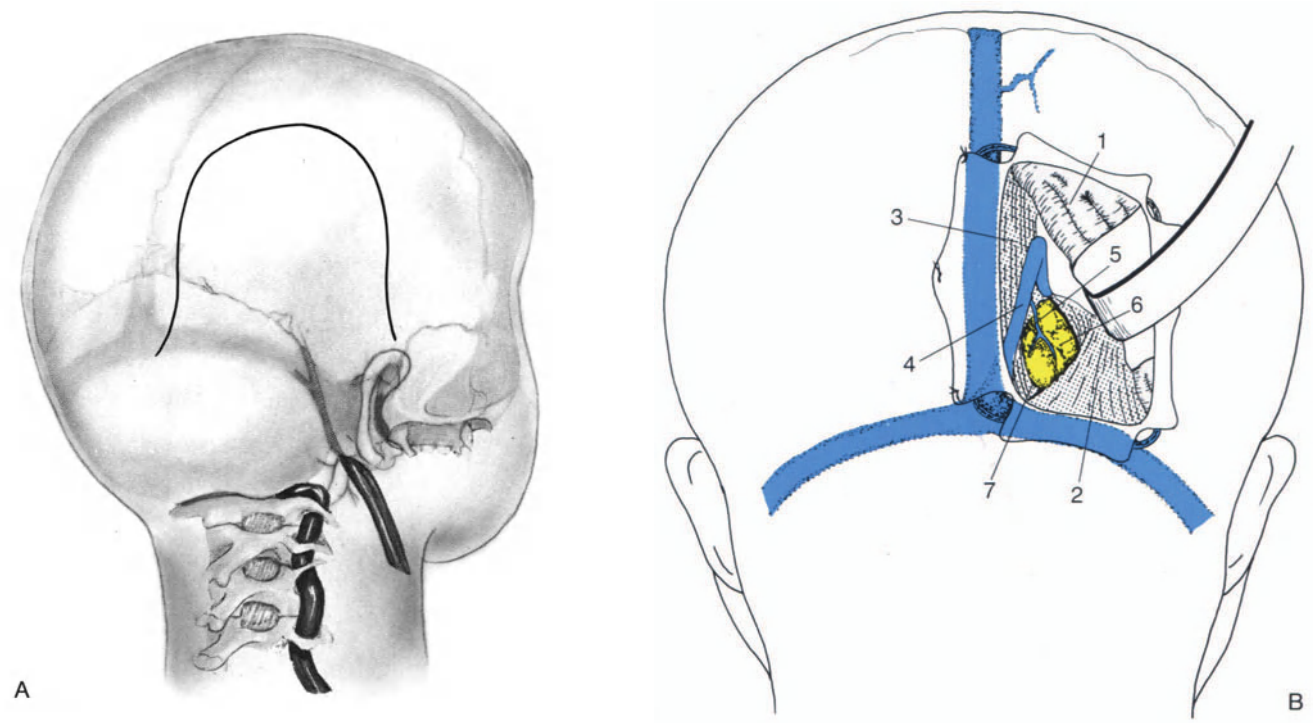
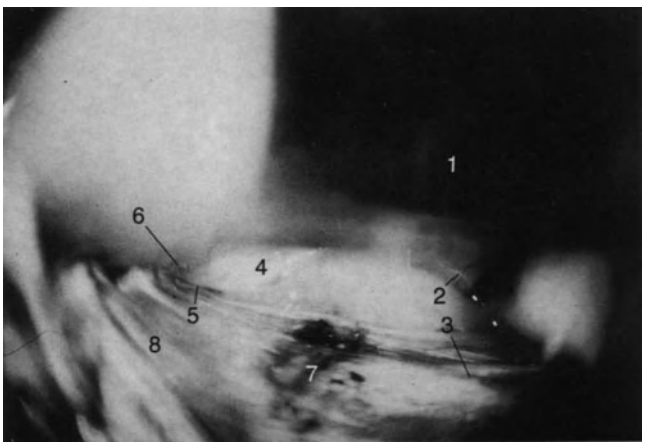
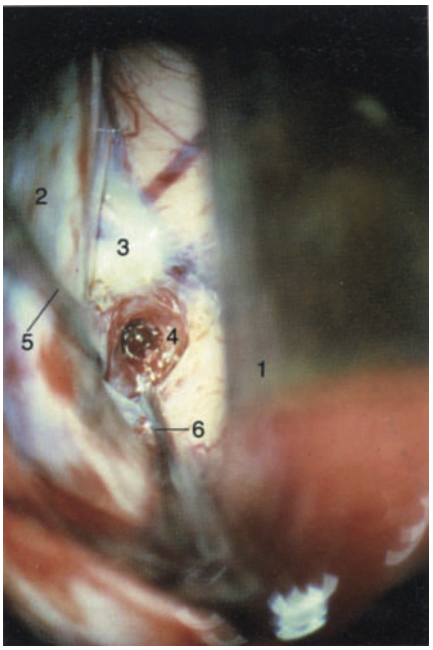


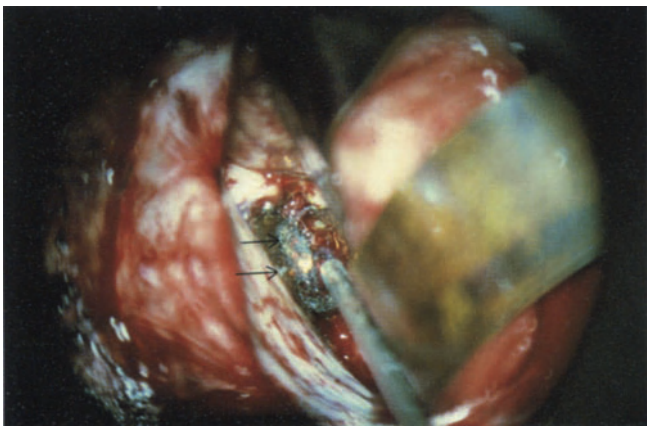
Figure 10.48. (A) The skin incision and bone flap for the occipital transtentorial approach are illustrated from the right posterior oblique perspective, illustrating both the torcular Herophili (*1*) and transverse sinus (*2*). The flap is planned so as to leave the occipital artery intact. (B) Diagrammatic representation of the occipital/transtentorial approach illustrating: (*1*) superolateral retraction of the occiptial lobe; (*2*) the tentorium; (*3*) the falx cerebri; (*4*) the straight sinus, and great vein of Galen; (*5*) the supraculminate vein, and (*6*) the tumor. The sectioning of the tentorium should extend along the line just lateral to the junction between tentorium and the falx (*7*). (C) 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*). (D) 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 ►



C



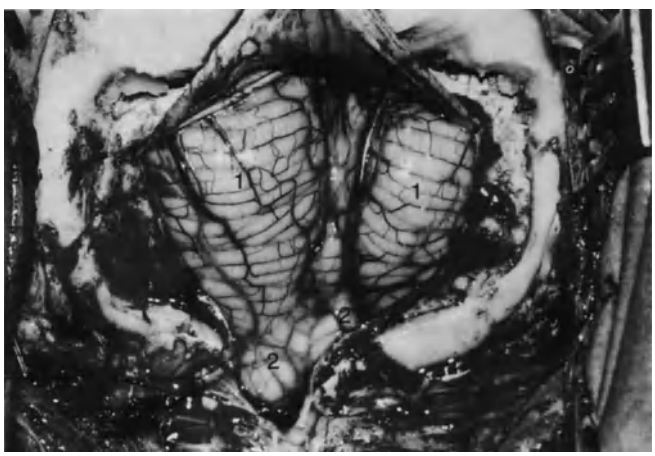
D

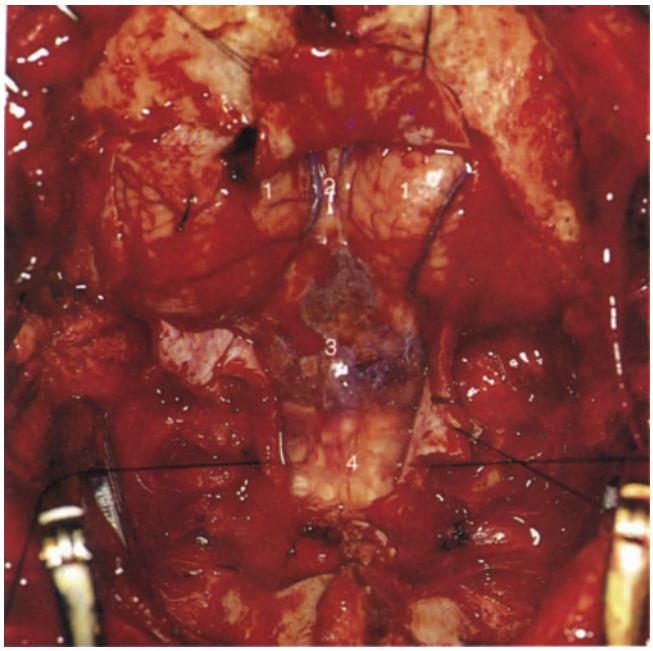


E

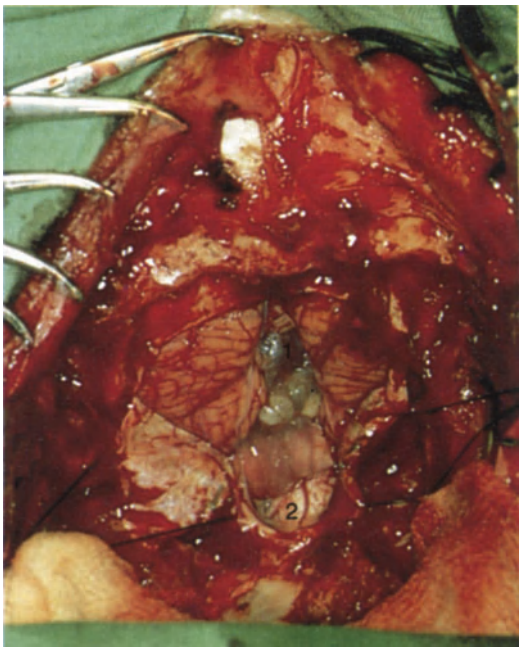
fibrosed arachnoid. (E) The laser is being used to vaporize the tumor *in situ*, minimizing retraction. One notes the neon laser (arrow) used as a visible tracer, since the CO_2 laser beam, well in the infrared wavelength, is invisible to the human eye. The carbonized tissue is lased blood.

Figure 10.49. Both cerebellar hemispheres (*1*) and tonsils (*2*) are remarkably deformed in this child with a vermian medulloblastoma which extended into the right cerebellar hemisphere and both tonsils.





A



B

Figure 10.50. (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 of 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*).

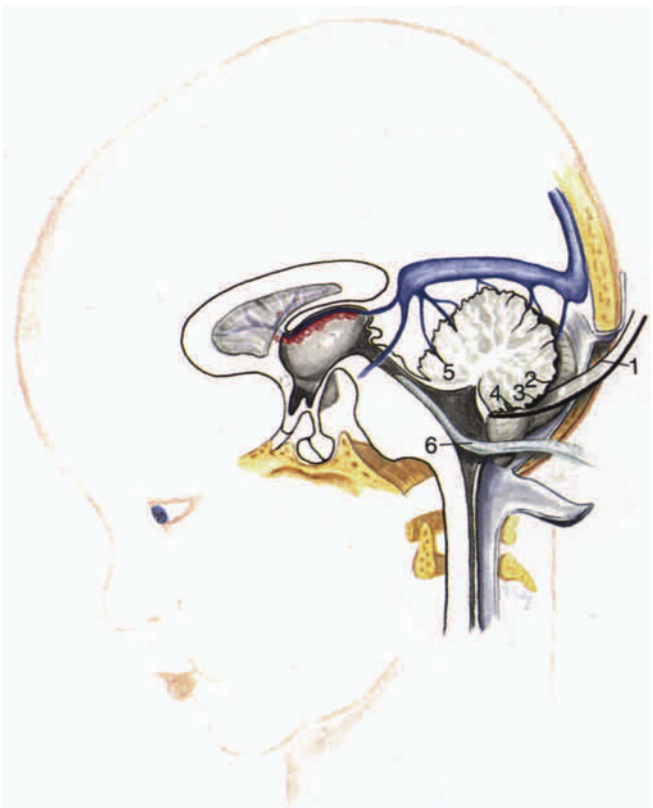


Figure 10.51. Exploration of the IV ventricle is best achieved by placing a narrow-blade spatula (*1*) beneath the pyramis (*2*) and uvula (*3*) of the cerebellar vermis, and then elevating the spatula so as to open the vallecula. The tip of the blade may then be inserted through the vallecula into the 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 one proceeds to inspect the anterior medullary velum.

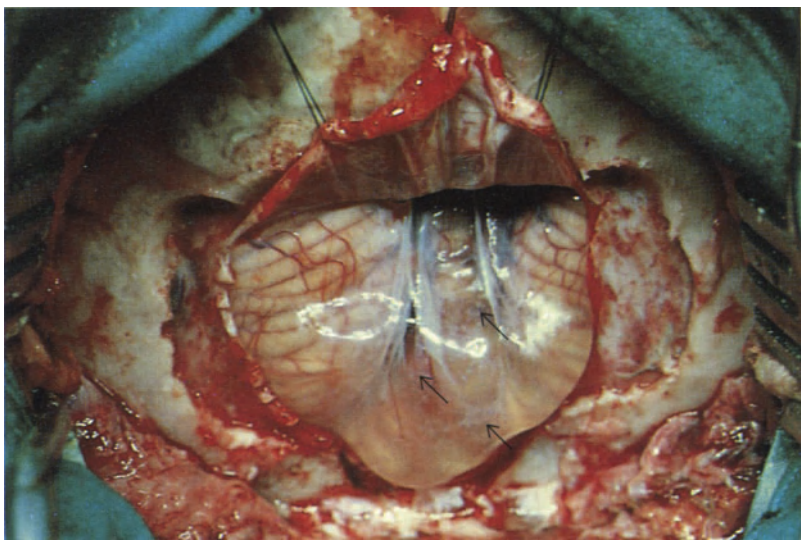


Figure 10.52. Meduloblastoma: The arachnoid of the cisterna magna is covered by a multitude of seedlike-appearing metastases (single arrows). This is common in medulloblastoma. It is not indicative of either a poor prognosis or subsequent central nervous system or peripheral metastases.

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 exludes ependymoma.

Medulloblastoma (Figures 10.52 to 10.56)
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, seedlike 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 rarely crystal clear, often being opalescent or having a 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 extend laterally into each cerebellar hemisphere. 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 generally are 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, it is much preferable to remove the tumor by vaporization: this avoids traction. A telfa must be inserted between the inferior surface of the

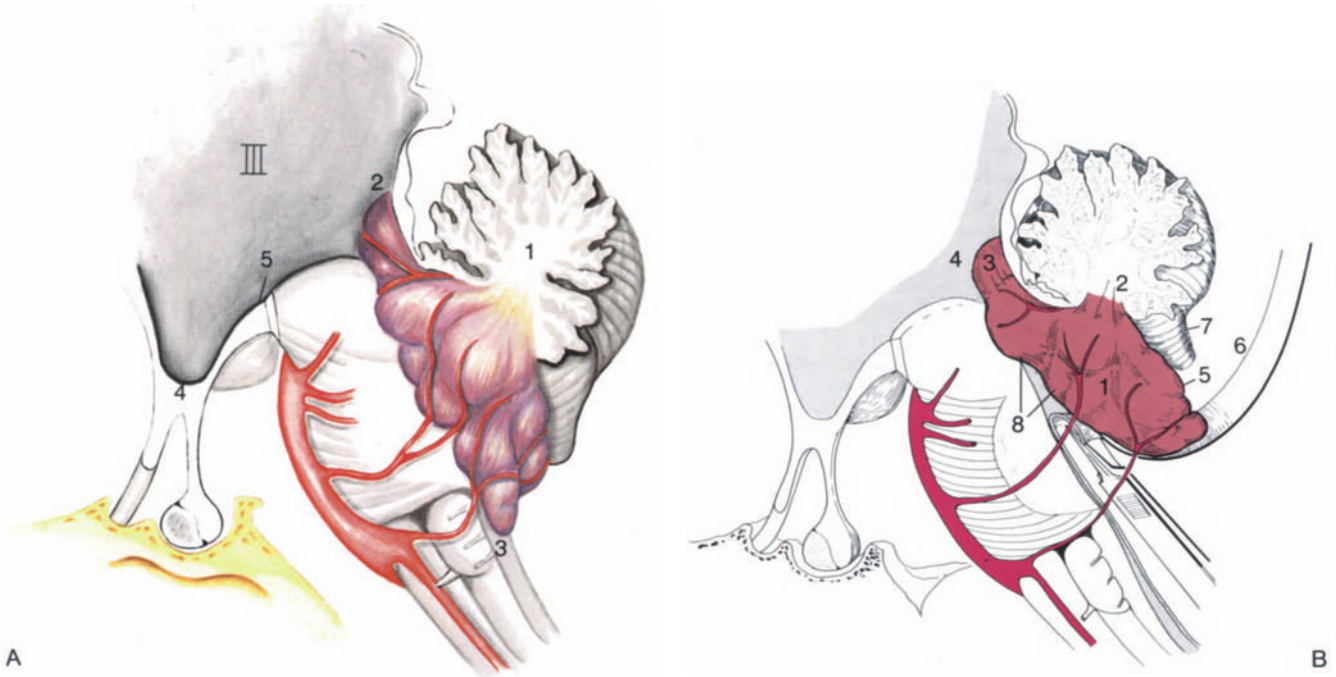


Figure 10.53. (A) Medulloblastoma growing from the region of the nodulus (*1*) fungates into the IV ventricle, often plugging both the aqueduct (*2*) and the foramen of Magendie (*3*). This results in dilation of the III, disappearance of the suprachiasmatic and infundibular recesses, and dilated III compressing both the optic chiasm (*4*) and stretching floor of the III ventricle (*5*). The gradual dilation 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*) of 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.

medulloblastoma and the floor of the IV ventricle before 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, 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 vaporized 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. At times, it is preferable to resect the pyramis and uvula. 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 should be placed over the cerebellar hemispheres 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 this structure is 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 surround-

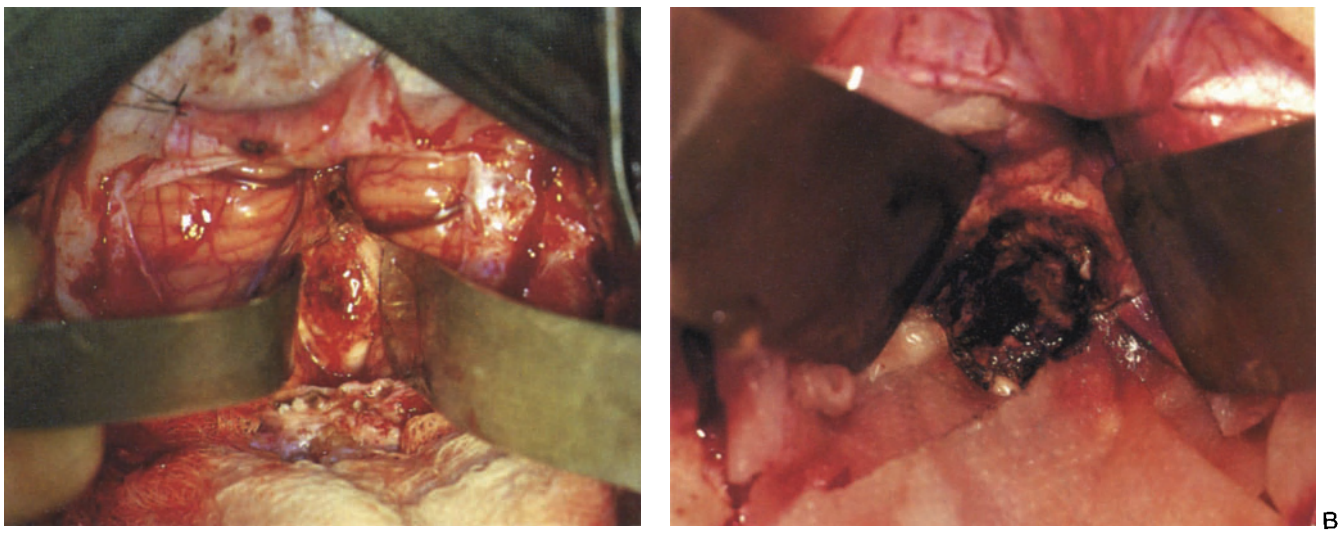


Figure 10.54. (A) The medulloblastoma was exposed after the vermian cortex over the folium, tuber, pyramis, and uvula were vaporized with CO_2 laser. Spatulae are positioned for photographic purposes. (B) After the obex and upper cervical cord, as well as both cerebellar hemispheres, were covered with telfa for protective purposes, the medulloblastoma was vaporized (with the laser).

ing neural structures upon which it borders or into which it extends. The tumor resection should be brought to the 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 hemispheres, tonils, cerebellar peduncles. *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.

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. 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 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 vaporized, the operating microscope should be used to scan the operative field, inspecting carefully the bed of the tumor for residual patches of neoplastic cells. These should be vaporized with a pulsed wave, defocused beam, at 5–7 W.

In the event a laser is 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, facilitates 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 completely freed, the sweeps are extended further laterally, freeing the tumor from the medial surface of each cerebellar hemisphere prior to

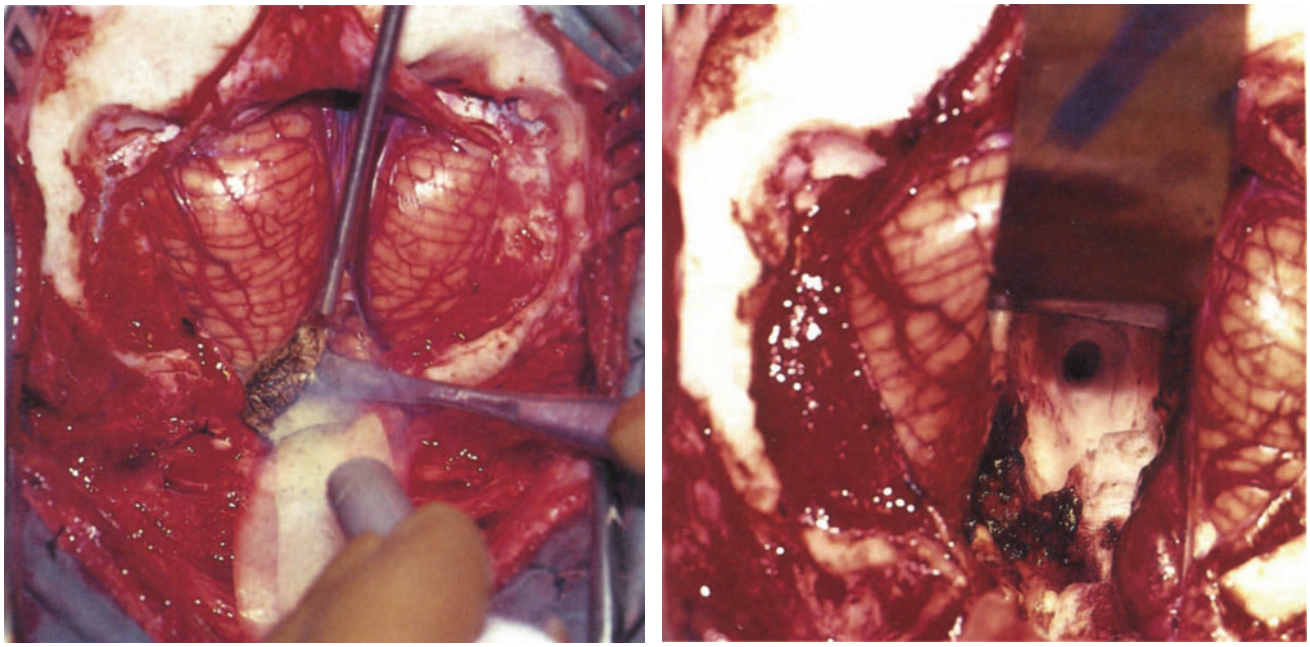
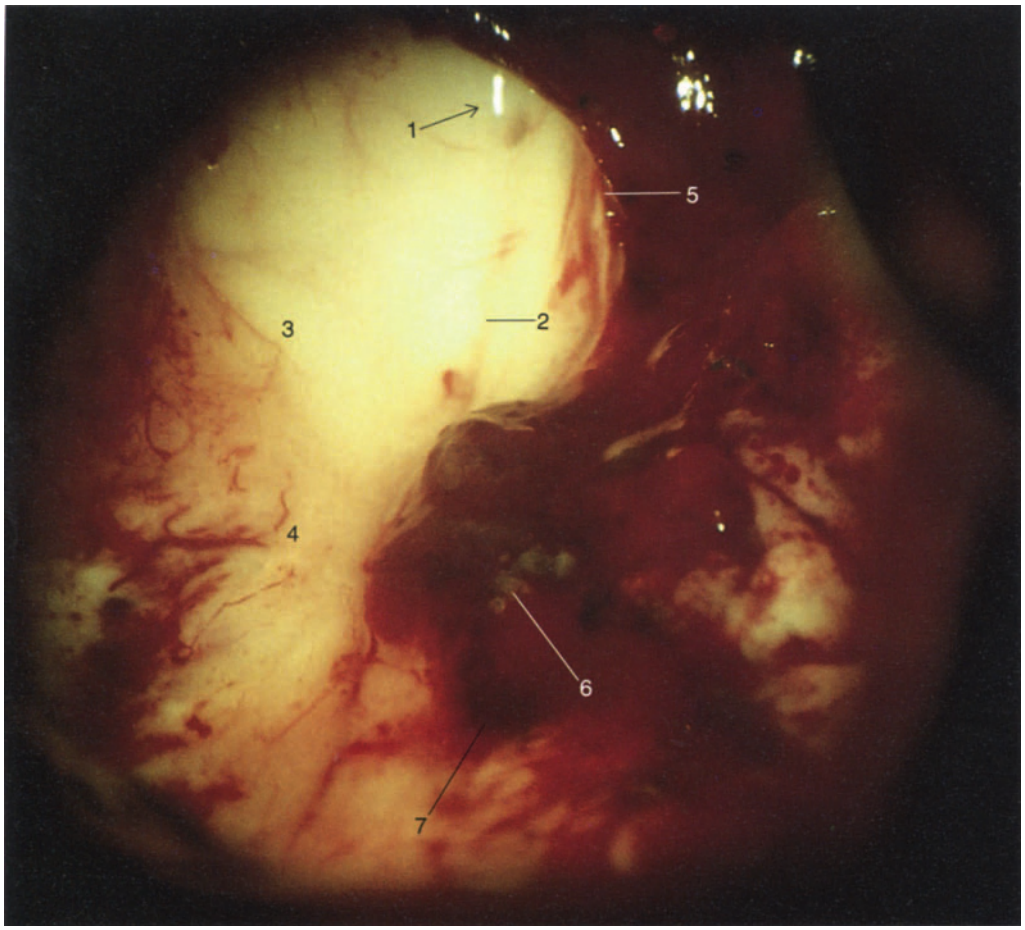


Figure 10.55. Varying stages in laser vaporization of a medulloblastoma which was extending primarily into the cerebellar hemisphere and tonsil. (A) The thin layer of normal cerebellar tissue overlying tumor extension into the left cerebellar tonsil is being vaporized. Note the characteristic appearance of cerebellar surface during vaporization. (B) 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 a Takahashi forceps, not with laser, to avoid inadvertent damage to the ependymal surface. (C) 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.

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 looking over its superior surface. He should now 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 connec-



C

tions between tumor and ependyma. This will free the tumor, and deliver it from the ventricular system and into the surgeon's hands.

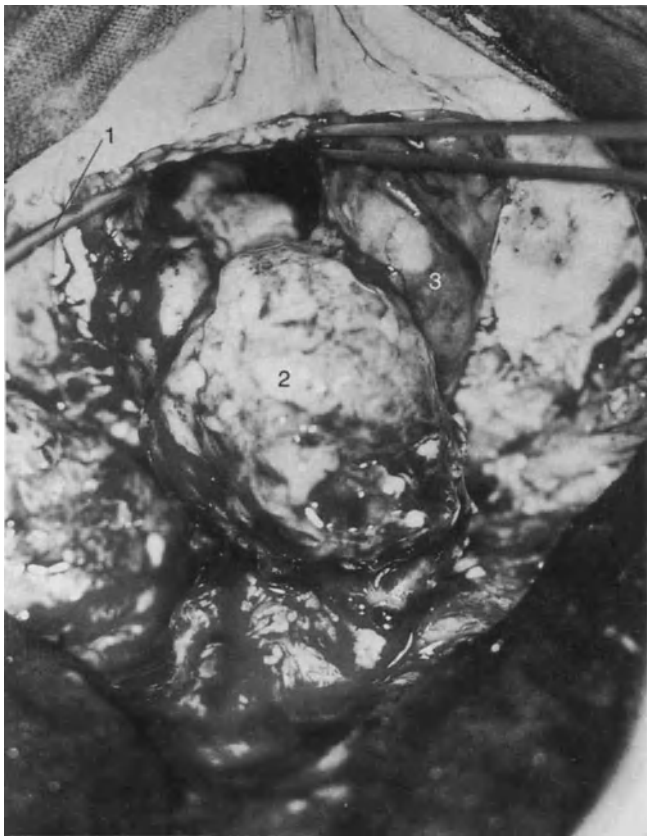
After the medulloblastoma has been completely removed, one should inspect the IV ventricle to determine whether there have been neoplastic extensions into its walls or floor. In the event they are identified, no attempt should be made to remove them from the surface of the IV ventricle unless one has a laser and is working with the operating microscope. 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 minutes to be certain that no bleeding persists before proceeding. Complete closure of the dura, in a water tight 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.

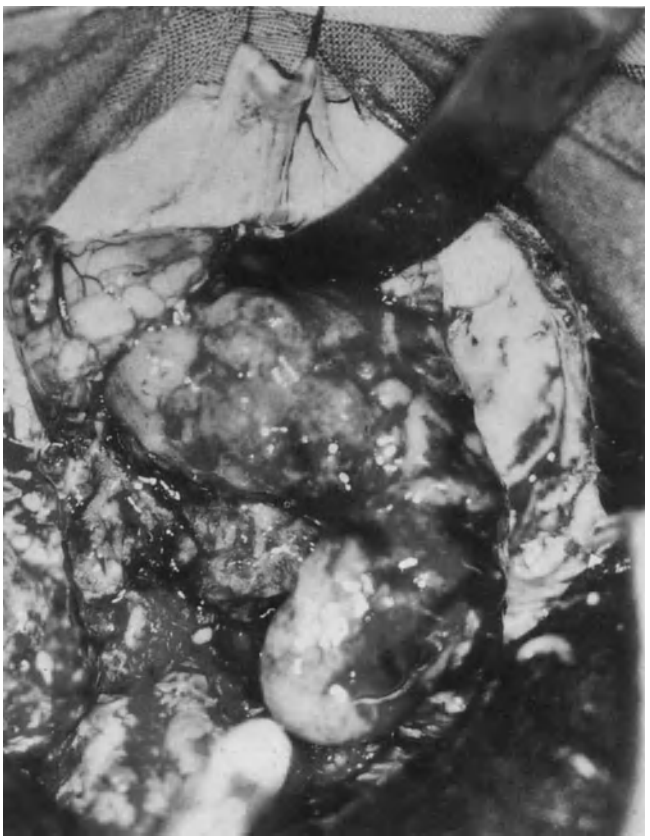
The great advantage of total resection, whether *en bloc* or by vaporization, of medulloblastoma is that it affords the child the greatest opportunity for cure: a 42% ten-year survival in our series. There is no greater morbidity associated with radical resection. Indeed, biopsy and partial resection are associated with a greater morbidity than "total" resection.

Ependymoma (Figure 10.57)

By and large the surface appearances of the cerebellum and cisterna magna in ependymoma are identical to

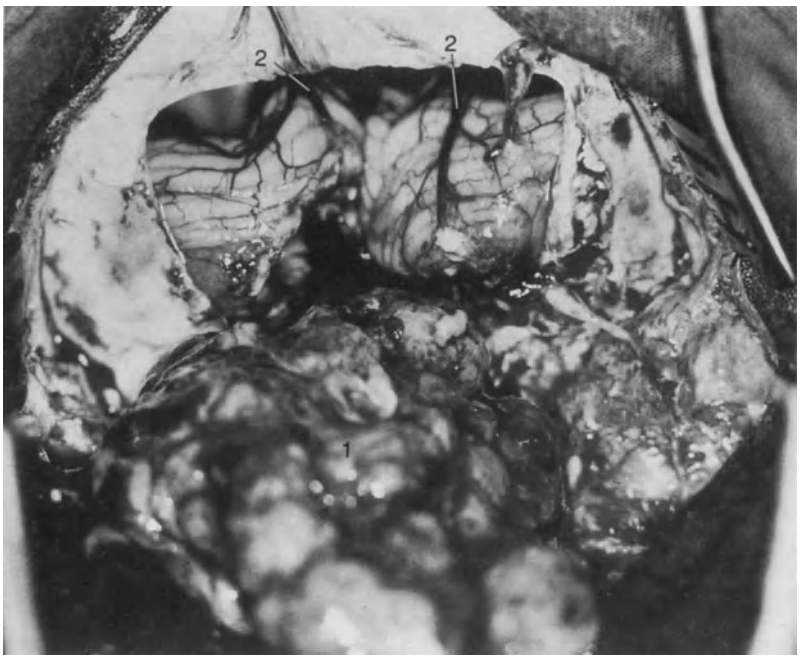


A

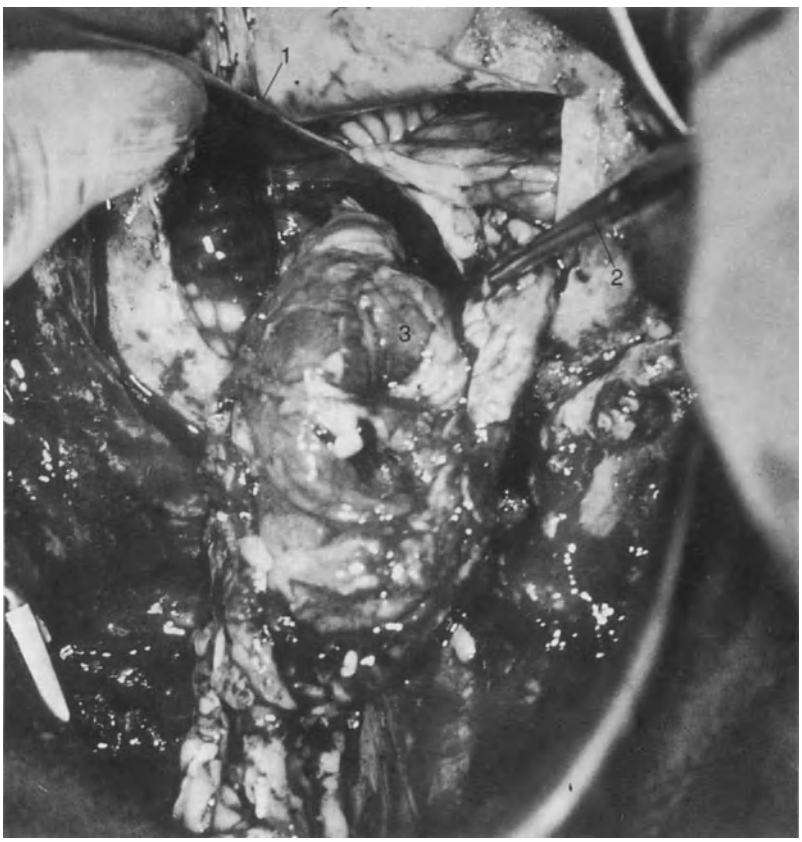


B

Figure 10.56. Varying stages of instrument resection of medulloblastoma, with the children in the lounging position. (A) The Penfield #4 dissector (*1*) is being used to separate the tumor (*2*) from the bordering surfaces of the left cerebellar hemisphere. The mass has already been removed from its lodging within the vermis and right cerebellar hemisphere, and is falling from the operative field. (B) A spatula is being used to elevate (hold in place) the cerebellum as the medulloblastoma delivers itself from its bed, leaving an enormous cavity in its wake. (C) As the tumor mass (*1*) leaves the posterior fossa, the cerebellar hemispheres hang from the superior cerebellar veins (*2*) bridging to the under surface of the tentorium. It is advisable to coagulate and transect these veins before closure, so as to minimize the risks of postoperative hematoma. (D) Here, in another child, the recommended technique for holding the cerebellar hemispheres in place with a spatula (*1*) (as a dissector (*2*) is guided along the surface of the tumor (*3*), separating it from cerebellum) is shown. ►



C



D

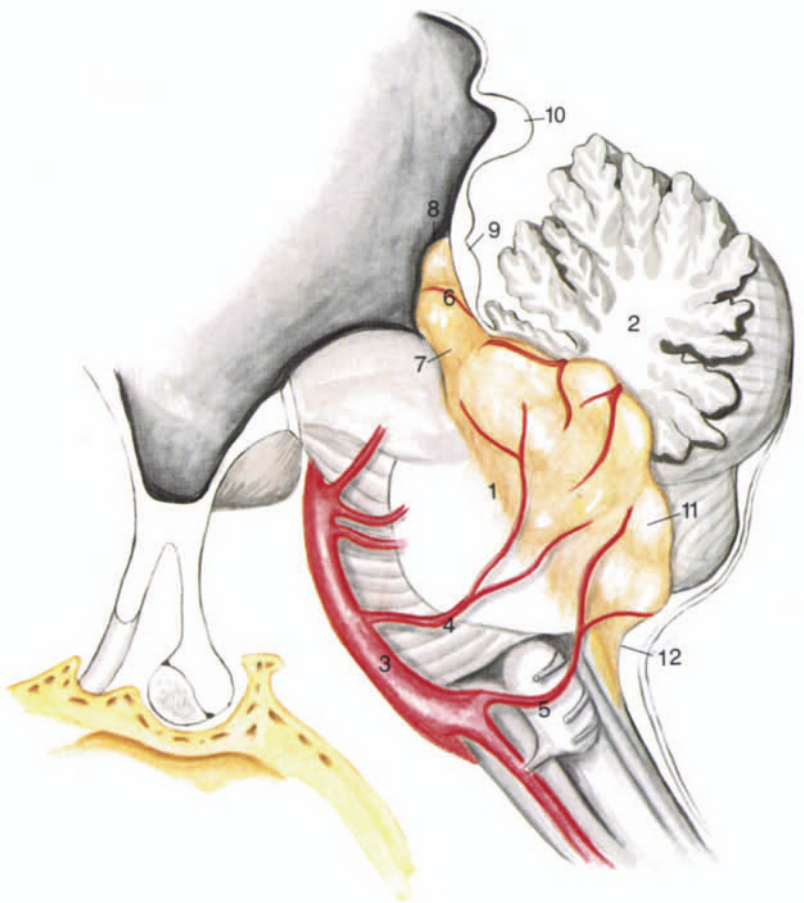


Figure 10.57. The ependymoma growing from the floor of the IV ventricle (*1*) displaces the cerebellar vermis (*2*) posteriorly. It causes the same changes in the IV ventricle and its bordering structures as the medulloblastoma. The basilar artery (*3*) is practically never deformed, though AICA (*4*) 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 magna (*12*), deforming the cerebellar vermis.

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 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. No attempt should be made to place the telfa between the tumor and the floor of the IV ventricle! When the telfa has been placed, a laser, if available, should be used to vaporize 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. The remainder of the laser technique is the same as for the medulloblastoma. 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.

If laser is not available the tumor should be aspirated gently, the aspiration should proceed slowly and, preferably, using the lowest pressure and the smallest size suction tip compatible with tumor removal. 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-minute wait preceed closure.

Choroid Plexus Papilloma (Figures 10.50 and 10.58)
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 ex-

tends into and occupies completely the cisterna magna, draping over the dorsal aspect of the medulla oblongata.

The primary surgical considerations for IV ventricle papillomas 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; and
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 entirety of 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 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 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 feeders from PICA may be identified along tumor surface, and coagulated 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, a hemostatic run, allows the surgeon to coagulate feeders bridging from parenchyma to tumor, layer at a time, with the tumor being gradually delivered into the field. This eliminates the risk of traction and compression on the brainstem. It also diminishes dramatically 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

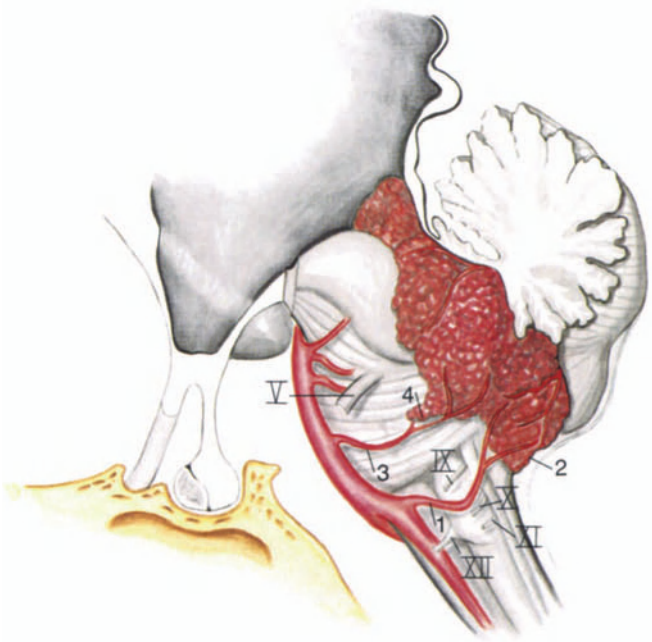


Figure 10.58. Choroid plexus papilloma. The tumor is lobulated and highly vascularized, being fed by branches of PICA (*1*) in the region of the vallecula (*2*) and AICA (*3*) at the lateral recesses (*4*). Cranial nerves are marked.

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 shrink the papilloma by coagulating portions of it with bipolar.

The dome of the tumor may gradually be shrunk 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 tumor is delivered into the field.

Wait approximately 10 minutes 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-minute wait, to assure hemostasis, precedes dural closure and bone flap reapproximation.

If laser is not available, one must 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 very 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.

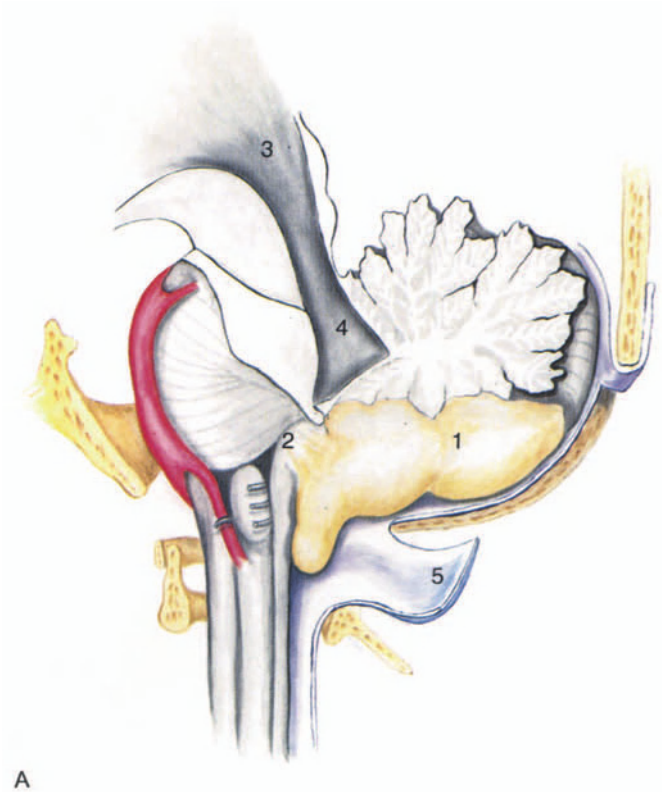
Brainstem Glioma (Figure 10.59)
If an intraparenchymal brainstem glioma, occluding the IV ventricle only because of fusiform tumefaction of the stem, is encountered, biopsy is not recommended because the complications could be severe. However, exophytic gliomas, fungating out of the restiform body or 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 invariably 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 cerebellum and lateral surfaces of the IV ventricle, as well as the intact 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 is the resection instrument of choice. Removal of the individual lobules, one at a time, exposes the floor of the IV ventricle 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.

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 or below 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.

Superior Triangle Tumors (Figures 10.60 and 10.61)
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 conjuncti-



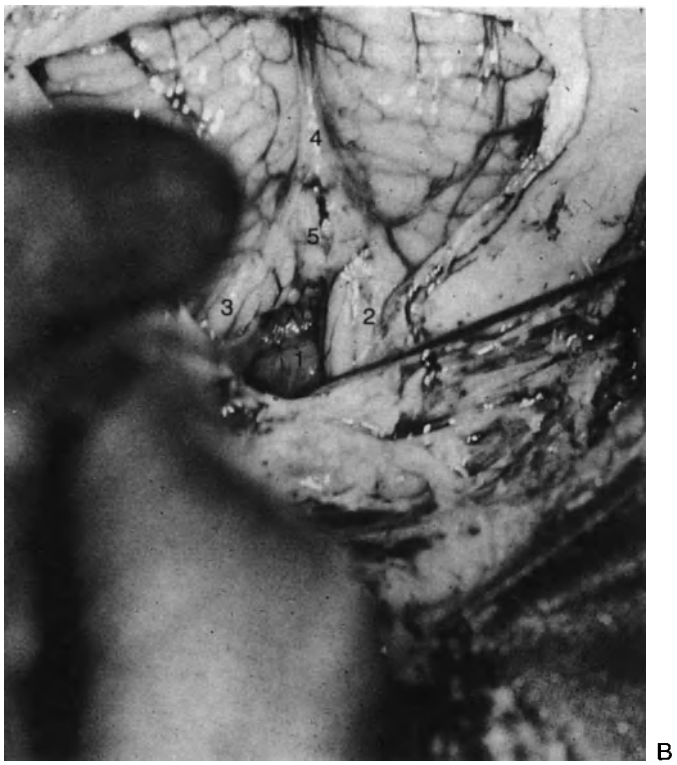


Figure 10.59. (A) Exophytic brainstem glioma. The tumor (*1*) has a pedicle (*2*), from which it extends, fungating into the cisterna magna. The aqueduct (*3*) and IV ventricle (*4*) are dilated, so one may expect a gush of cerebrospinal fluid at some time during the resection. Note the sling (*5*) fashioned from the dura. This should be sewn upward and outward to provide desired support. (B) The exophytic brainstem glioma (*1*) is nestled 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 entirety of the cisterna magna, elevate the vermis (*2*), and displace superolaterally the cerebellar hemispheres (*3*). The mass was attached to the clava by a 3-mm pedicle of neoplastic tissue, from which it was removed. The postoperative course was uneventful and the child a long-term survival.

vum, 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 hemispheral folia. It is unusual for superior vermian tumors to cause thinning of the squamous portion of the occipital bone, as commonly occurs in inferior vermian triangle tumor.

The cerebellotomy is performed 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.

Once the most posteroinferior portion of the vermian tumor is encountered, one makes the determination as

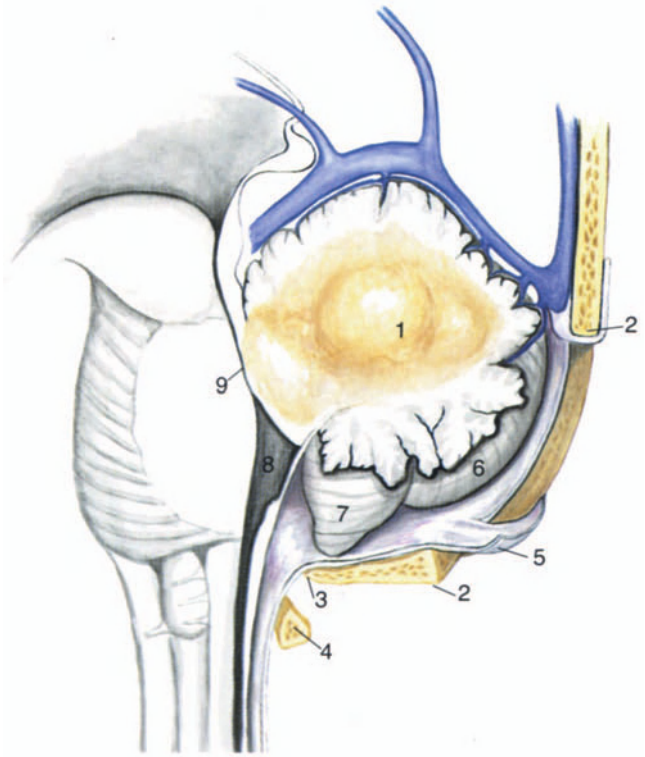


Figure 10.60. Solid, superior cerebellar triangle vermian tumor. The tumor (*1*) is primarily located above the line of the transverse sinus, so a quadrilateral bone flap (*2*) is reflected, neither opening the rim of the foramen magnum (*3*) nor taking away the arch of C1 (*4*). The dura is fashioned into a sling (*5*), which supports the cerebellar hemisphere (*6*). Horizontal squamous occipital bone supports the tonsil (*7*). The IV ventricle (*8*) may remain open, but the aqueduct (*9*) is almost always compressed.

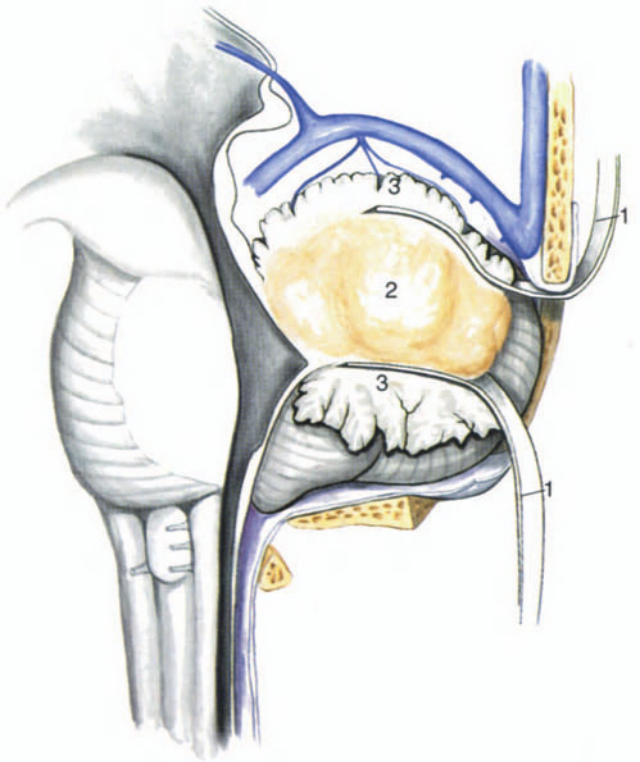


Figure 10.61. If one does not have access to the laser, then spatulae (*1*) are used for removal. They are insinuated between the tumor (*2*) and the cerebellar parenchyma (*3*), after having been bent to conform to what the surgeon estimates to be tumor contour, and then slid over the tumor so as to separate it from cerebellum. Delivering tumor through the cerebellotomy takes the compression from the anterior medullary velum and allows the IV ventricle to reexpand.

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 laser, vaporizing the tumor *in situ* without making any effort to remove it *en bloc*. Self-retaining retractors are inserted in order to hold the cystic cavity open and to prevent the superior portions of the cerebellar hemispheres and vermis from herniating into the operative field, collapsing the capsule and impairing the surgeon's vision. Once this is done, the laser is used to vaporize the tumor capsule, with a defocused, continuous wave, beam at 8 W. This is a bloddless procedure, one which allows the surgeon to determine immediately when the capsule is vaporized and cerebellar fibers are visualized.

In the event the tumor is solid, no attempt should be made to separate it from the surrounding, normal, cerebellar tissue, with spatulae, dissectors, cautery, suction. Rather, the cerebellum should be covered with telfa and then either fluffy cotton or telfa should be inserted along the border between tumor and surrounding cerebellar tissue, before beginning *laser* removal. This provides protection to the cerebellum. Small-bladed 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.

Laser vaporization, with continuous wave, of the tumor is then performed, using a defocused beam at 15 W. These bloodless tumors vaporize readily, without formation of coagulum along their surface. 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 with laser 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 a Cushing or, preferably, a micropituitary forceps. Suspending the tumor with one of these instruments allows the surgeon to use laser to vaporize its inferior pole, freeing small lobules one at a time. Attention is given to orientation, to avoid entering the region of the deep cerebellar nuclei at the center of each cerebellar hemisphere, immediately superior to the brachium pontis and the brachium conjunctivum. Care should be taken not to vaporize in the region of the culmen monticuli, with the beam pointed anterosuperiorly, without having placed telfa cotton 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 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 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 hemisphere 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 should be 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 apples indifferently whether the tumor is removed with *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 must take great care to maintain anatomical orientation, to avoid entering the brachium 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.

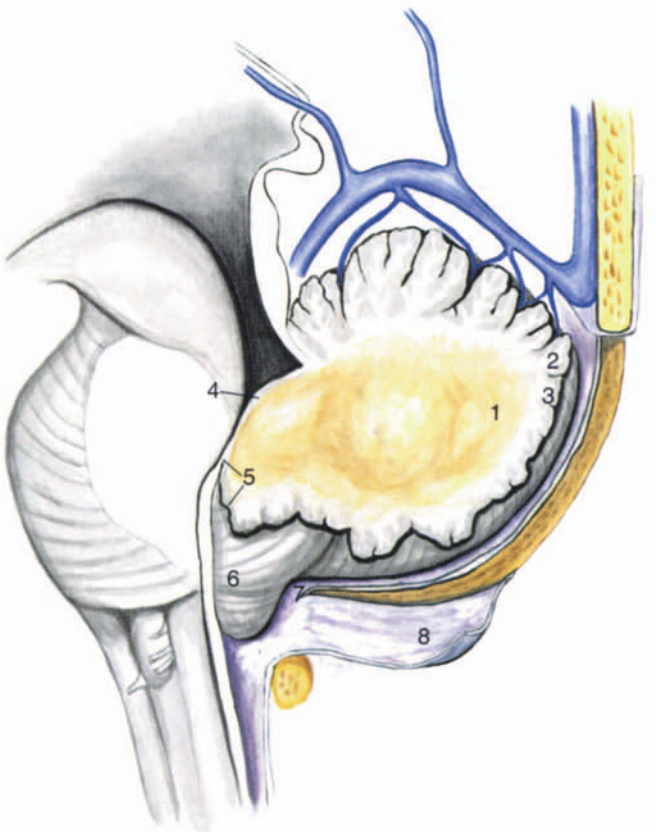


Figure 10.62. Suggested technique for nonlaser resection of inferior vermian astrocytoma. This astrocytoma (*1*) expands within the vermis beneath the level of junction of folium (*2*) and tuber (*3*), extending as far anteriorly as the posterior medullary velum (*4*). This occludes the IV ventricle 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.

Inferior Triangle Tumors (Figure 10.62 and 10.63)

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 choroid 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 hemispheres, posterior to the superior spinal cord and obex of the IV ventricle. The lateral and retrome-

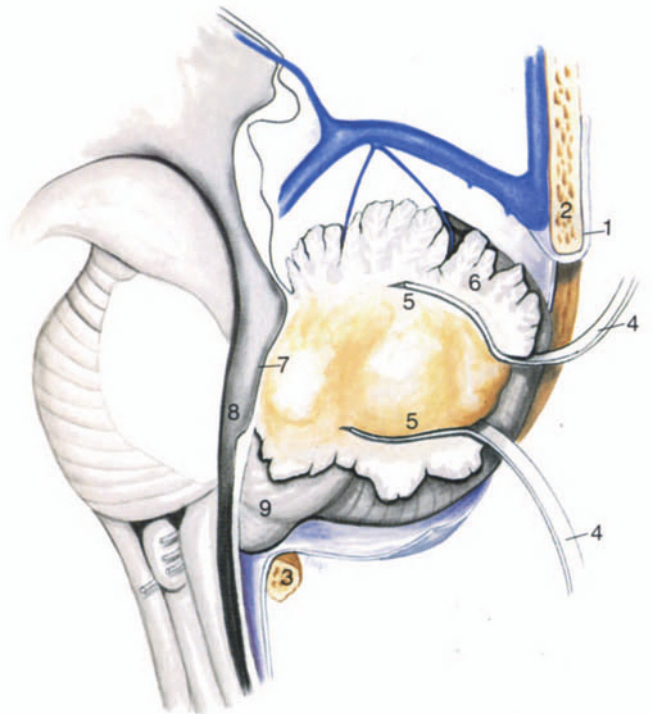


Figure 10.63. The tumor is exposed through an inferior cerebellar craniotomy, and the superior trapdoor of the dura (*1*) sewn over the squamous occipital bone (*2*) and then anchored into place. The opening extends into the cisterna magna, so that dura is exposed as far inferiorly as C1 (*3*). The spatulae (*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.

dullary 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 form the surface of the tumor with spatulae, until approximately a 2-cm surface area of tumor has been exposed. Telfa are then placed to protect the surfaces of the neural tissue and the vascular structures, before the laser is used. The same laser factors are used as for superior cerebellar vermian tumors. 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 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 superior spinal cord, the floor of the IV ventricle and its lateral recesses, the medial surfaces of the tonsils.

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, to plug the flow of cerebrospinal fluid, and to cause dilation of cisterns opposite to their pole 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 C1 has been osteotomized at its lateralmost portions, bilaterally, and dislocated posteriorly onto the spinous process of C2. This permits opening of the dura down to the C1/C2 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" its ventral portion 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, 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 a mesenchymal mass.

Dorsal Foramen Magnum Tumor (Figure 10.64)
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 form 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.

Once all tumor lobules have been vaporized, 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 laser is not available, bipolar/microscissor resection technique should be used. The resection should not extend to a level which is flush with the cervicomedullary structures: it should be stopped approximately 2 mm distal to this point!

Lateral and Ventral Foramen Magnum Tumors
Lateral Foramen Magnum Tumors (Figure 10.65)
Lateral foramen magnum tumors expand in the interval between the IX, X, and XI cranial nerves dorsally. The ventral, lateral, and retromedullary segments of 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

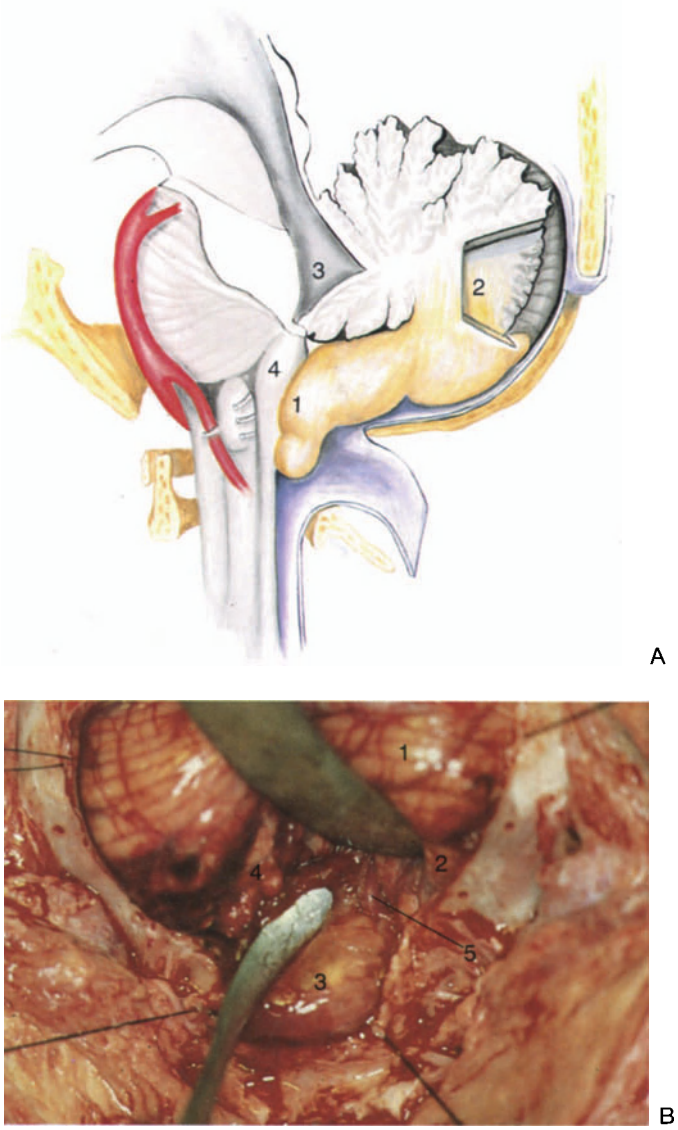
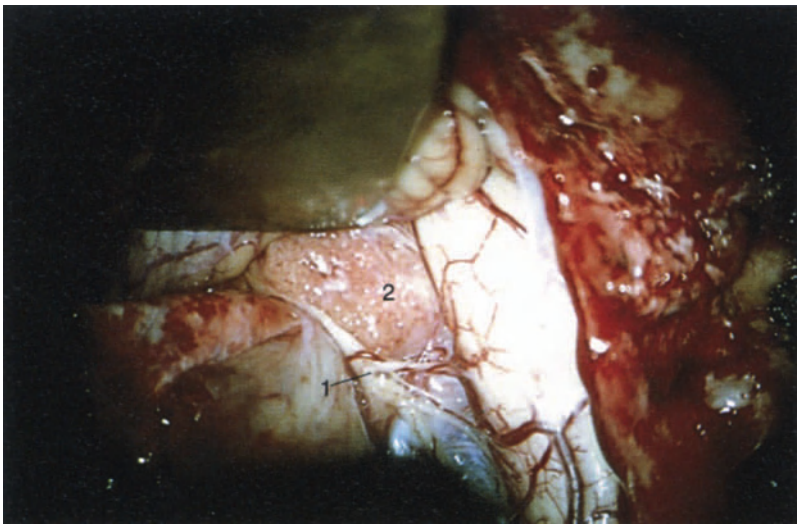
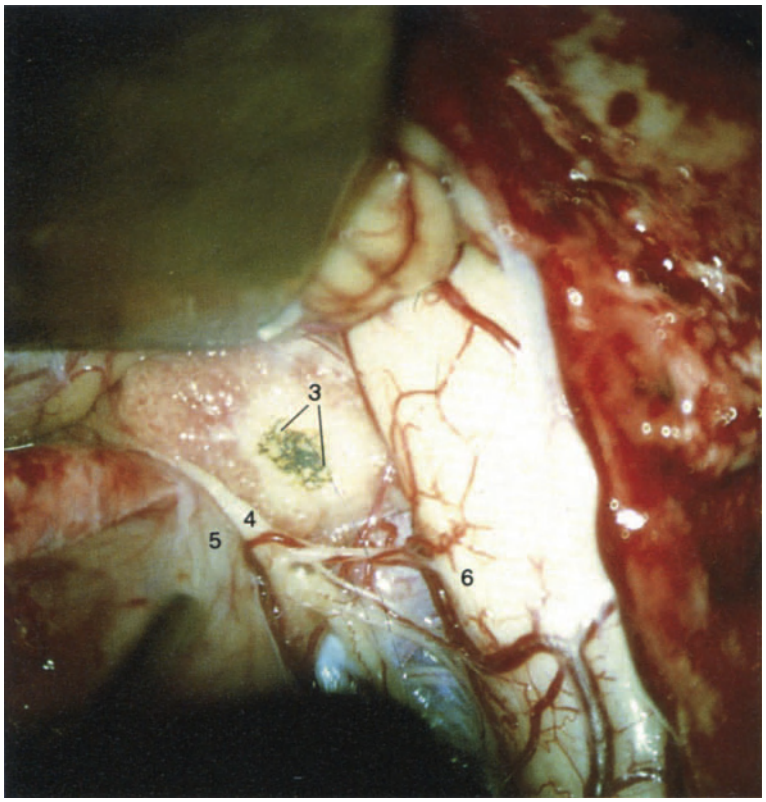


Figure 10.64. (A) Foramen mangum tumor (*1*) pedunculating from the inferior vermis (*2*). Gliomas have a tendency to become exophytic, so that one may expect to see signs of cerebellar dysfunction, obstructive hydrocephalus, spinal cord compression, in foramen magnum tumors. Here, the exophytic tongue 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.

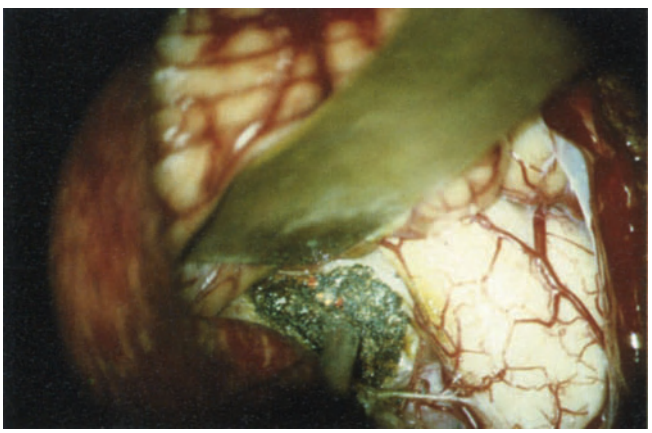


A

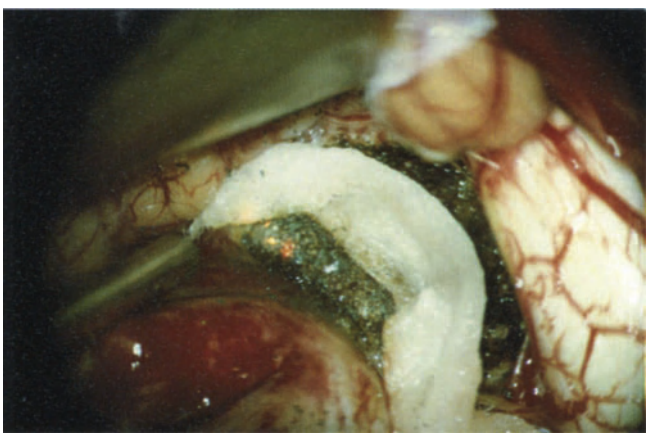
Figure 10.65. (A) Lateral foramen magnum tumor. This is a chordoma extending from the clivus, lateralward, to expand within the interval between IX, X, and the spinal accessory (*1*) cranial nerves dorsally, and the XII cranial nerve ventrally. The tumor (*2*) extends below the foramen magnum and superiorly 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) 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 terminology 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 slakening 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. ►



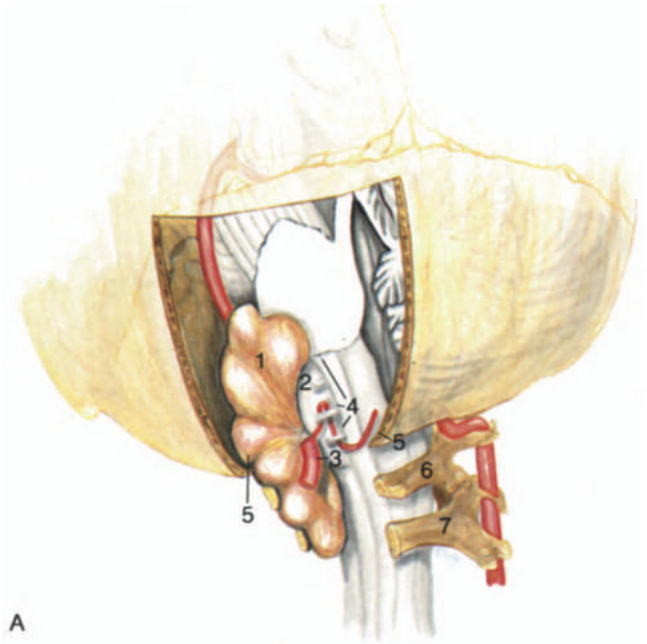
B

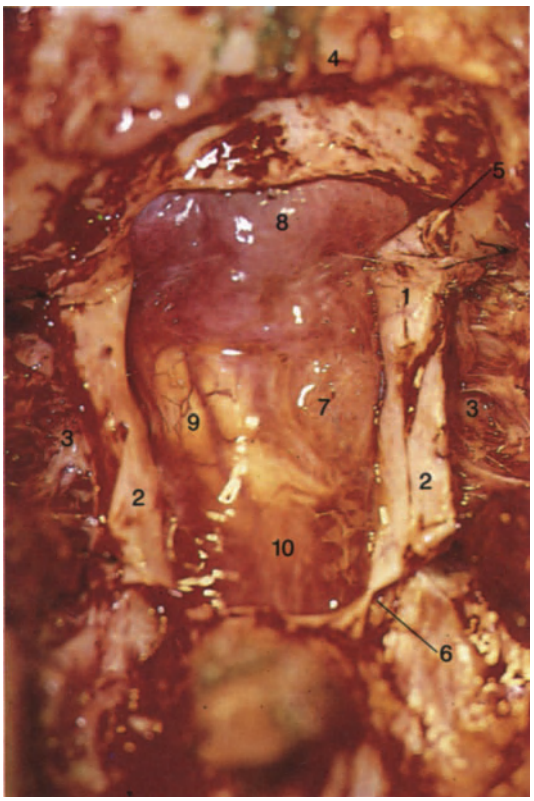


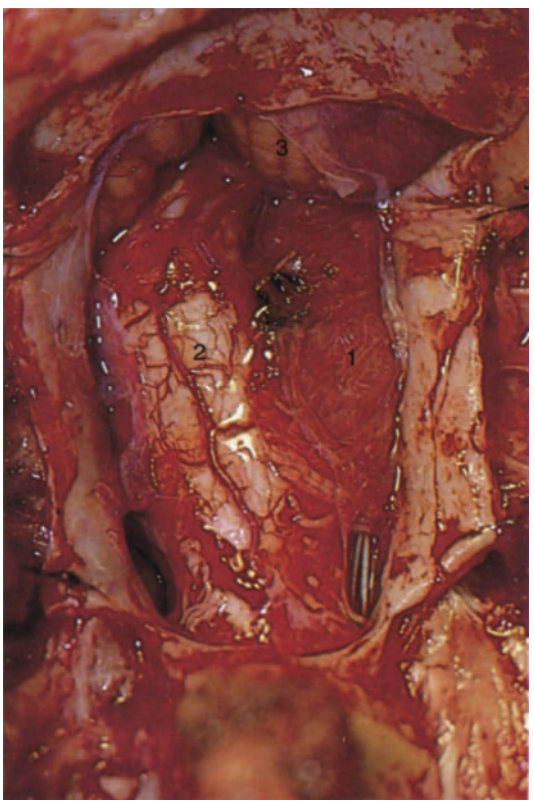
C



D







10-66C

Figure 10.66. (A) This is a posterooblique 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 diplaced. 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.

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, and the laser is set at a defocused beam, 3 W, pulse mode for vaporization of the minute portions of tumor within the intervals between individual cranial nerves and segments of PICA. 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.

Ventral Foramen Magnum Tumors (Figure 10.66)
Either purely ventral tumors or 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 a 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 he has protected it completely*. Different 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 hemisphere 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 hemispheral tumor more often extends into one or more cerebellar peduncles than does the vermian tumor.

Solid Cerebellar Hemisphere Tumors (Figure 10.67)
The removal of solid cerebellar hemisphere tumors is effected with the same laser technique 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, olive. Similar anatomical considerations apply to removal of cystic astrocytoma of the cerebellar hemisphere.

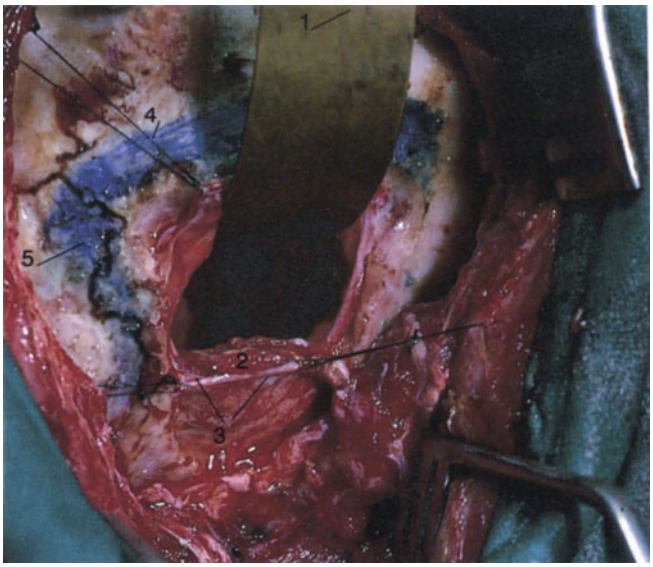


Figure 10.67. Nonlaser ("classical") technique for removal of solid cerebellar hemisphere astrocytoma. This procedure is used either when a laser is not available or when, as in this case, the solid tumor occupies the cerebellar hemisphere at the pontocerebellar angle, either adjacent or attached to 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 hemispheral tumors one is advised first to elevate the inferior cerebellar hemisphere, then to identify VII, VIII, IX, X, XI cranial nerves, and, finally, to place telfa cotton between hemisphere and cranial nerves. The same should be done for 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 (Figure 10.68)
Cystic cerebellar hemisphere tumors accumulate enormous volumes of fluid. After the cerebellotomy has

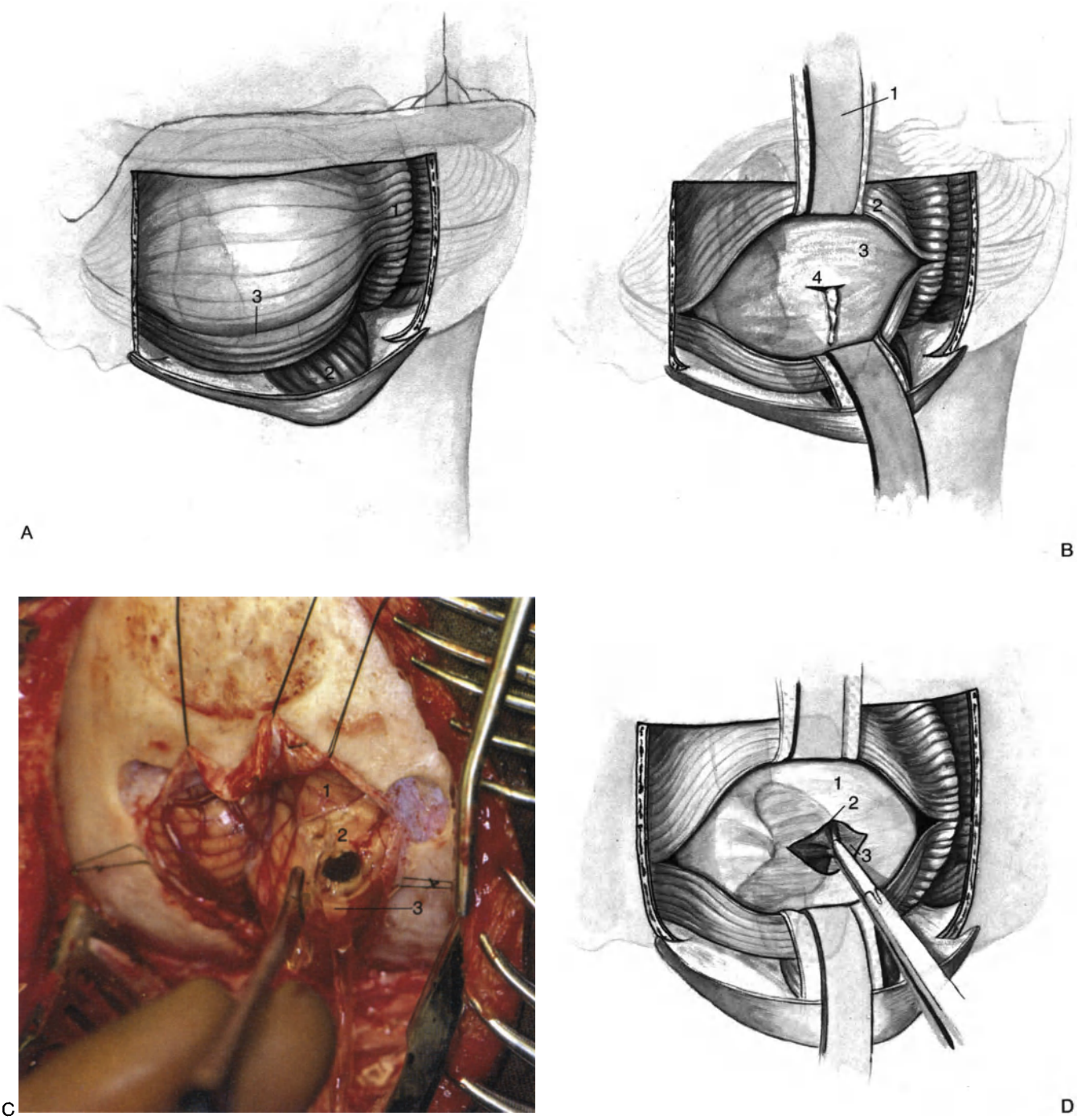


Figure 10.68. (A) Cystic tumor occupying the left cerebellar hemisphere, displacing the vermis (*1*) medially and the tonsil (*2*) inferomedially. The folia over the superior surface of the hemisphere are widened and flattened. The cerebellotomy (*3*) has been extended to the junction of hemisphere and vermis. (B) 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 cerebellar bed. Incision (*4*) is then made in cyst capsule and the fluid drained. (C) 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. (D) As the fluid is drained, the cyst wall (*1*) collapses, falling from its surrounding cerebellar bed. The tumor nodule (*2*) is grasped with a forceps and pulled from the inner capsule wall (*3*), facilitating delivery of the entire capsule from the cerebellum.

been performed 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 laser-vaporized along with the capsule of the cyst: the same technique is used in all cystic cerebral and cerebellar tumors. If a laser is not available, the nodule (if present) and cyst capsule should be removed, as already described for cerebral hemisphere cystic tumors and as herein illustrated.

Parasellar Tumors

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 nil.

Anatomy (Figure 10.69). 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, requires a great deal of time and effort to master. In the abnormal, especially for extraparenchymal tumors such as craniopharyngioma, the matter is compounded.

The parasellar area holds the following:

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, the anterior communicating artery (M-1 of the middle cerebral artery), the posterior communicating artery, the anterior choroidal artery
5. Hypothalamus: anterior-inferior portion of the III ventricle, infundibulum, tuber cinereum, mammillary bodies
6. Basal cisterns: suprachiasmatic, infundibular, mammillary, interpeduncular, pontine
7. Cranial nerves: olfactory, oculomotor, abducens, trochlear, trigeminal

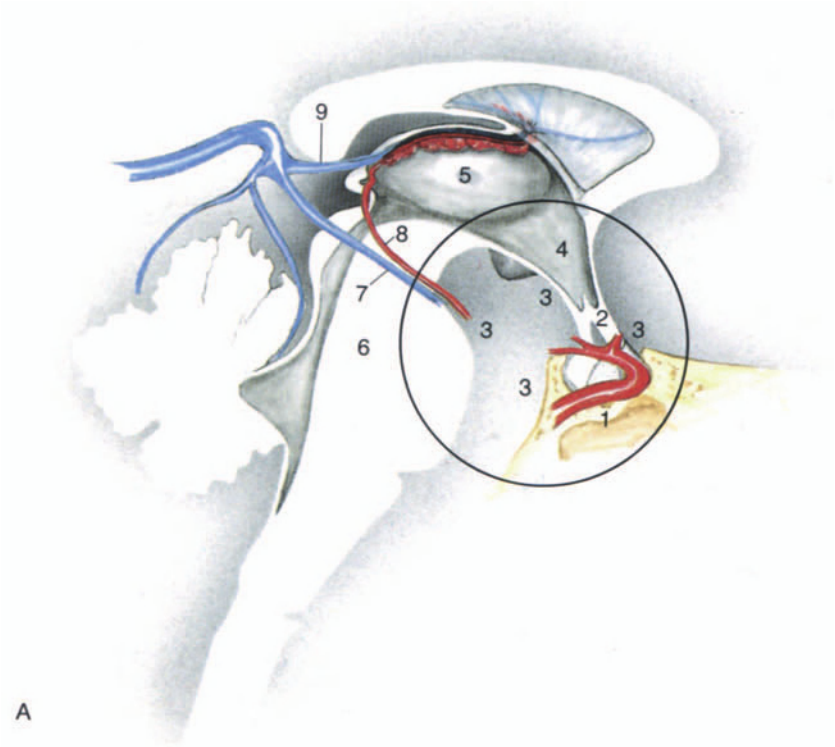
Intraparenchymal tumors of the parasellar area grow either from the optic pathways or the hypothalamus. Consequently, they expand within these structures, growing along the neural pathways and into the surrounding III ventricle and parasellar cisterns. Extraparenchymal tumors, on the other hand, deform, stretch, and compress all structures within the parasellar area, as they expand within the sella turcica, parasellar cisterns, 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 involved by craniopharyngioma are the optic pathways, hypothalmopituitary system, mammillary bodies, inferior and anterior III ventricle, vein of Rosenthal, posterior cerebral artery, and the anterior circle. The basilar artery may be deformed by craniopharyngioma, generally being displaced posteriorly, but, with the rarest of exceptions, it is neither engulfed by the tumor nor are neoplastic calcifications adherent to it. These exceptions occur in the "Formes Geants."[12]

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.

The hypothalamus, infundibular stem, and posterior lobe of the pituitary gland (pars nervosa) are derived embryologically from an outgrowth of the developing brain. They form one anatomical and functional unit (Fig. 10.70). Certain groups of hypothalamic neurons cluster to form nuclei with the supraoptic (vasopressin) 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 neurohypophysis and infundibular stem form the posterior portion. The infundibular stem and the associated pars tuberalis together 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 pars intermedia is adjacent to the neurohypophysis.

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.



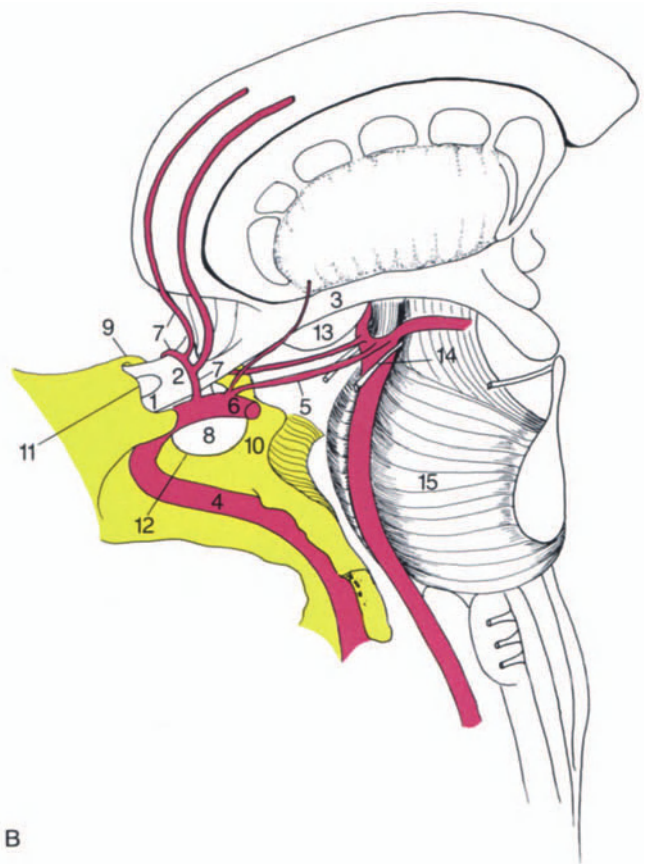


Figure 10.69. (A) This is a descriptive, not a spatially proportioned, illustration of the midline structures which may be involved by a parasellar tumor, depicting relative location of the sellar (*1*), chiasmatic (*2*), cisternal (*3*), hypothalamic (*4*), superior III ventricular (*5*), and brainstem (*6*) areas. Note the relative location of the basal vein (*7*), medial posterior choroidal artery (*8*), and internal cerebral vein (*9*). (B) The significant 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*).

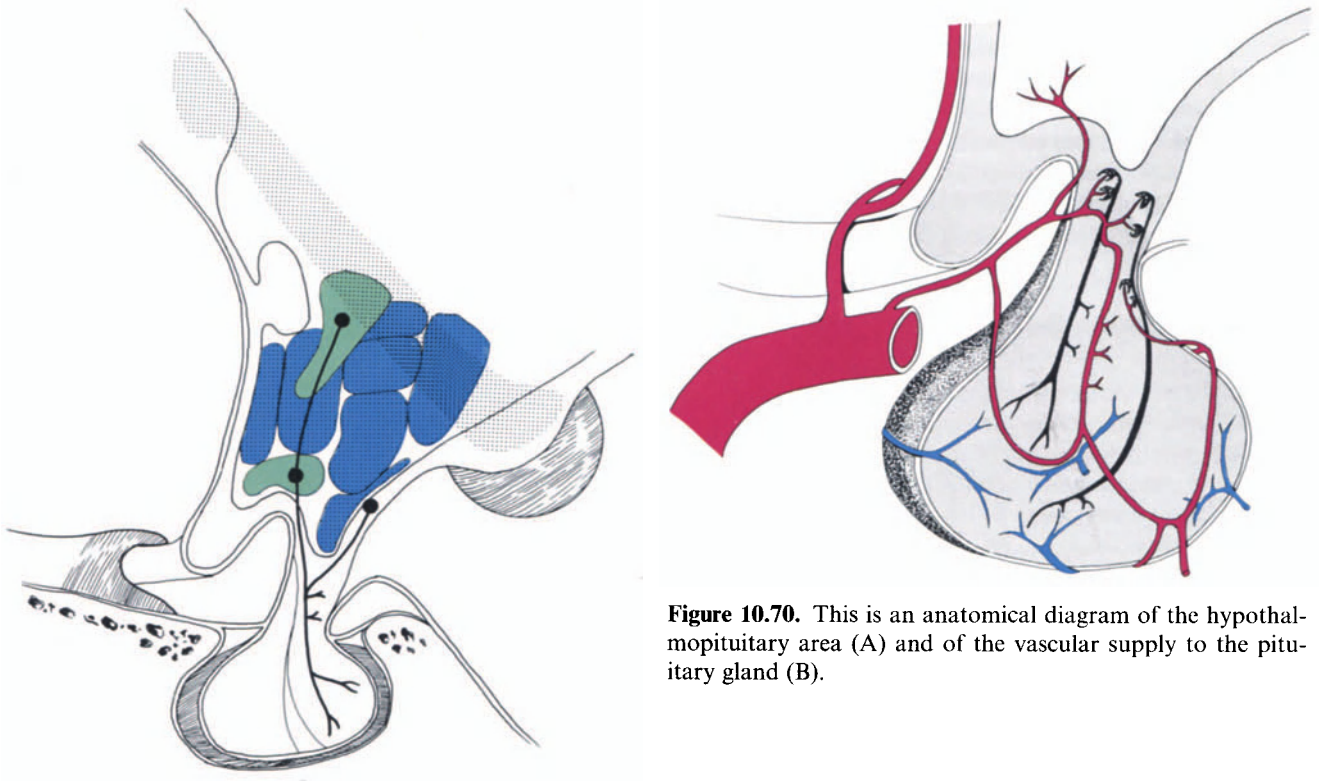


Figure 10.70. This is an anatomical diagram of the hypothalmopituitary area (A) and of the vascular supply to the pituitary gland (B).

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 when it reaches the hypophysis. In turn, these branches anastomose 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.

Craniopharyngioma. The craniopharyngioma most commonly encountered 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 entirety of 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 second 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 (Figure 10.71). The germinoma is a very distinct clinical entity, differing remarkably from the optic pathway and hypothalamic gliomas on one hand, and the craniopharyngioma on the other. Specifically, this tumor presents clinically with endocrine dysfunction, does not cause changes which may be identified with plain skull x-rays, 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.

The bifrontal craniotomy is recommended for resection of craniopharyngioma when the surgeon has evidence to suggest that he may attain a surgically "complete" resection. A unilateral, anterior frontal craniotomy is recommended for partial resection of a craniopharyngioma. There is no indication for surgery for

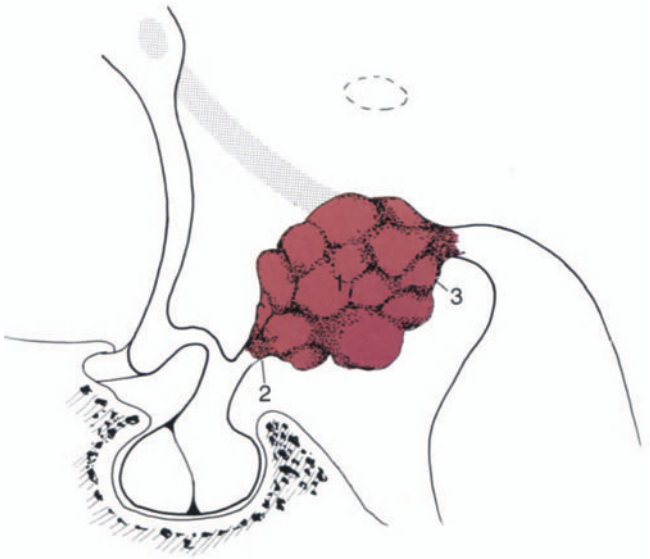


Figure 10.71. Hypothalamic *germinona* (*1*) grows from the region between the pituitary stalk (*2*) and the mammillary bodies (*3*). It may fungate into the III ventricle, but, generally, remains quite small.

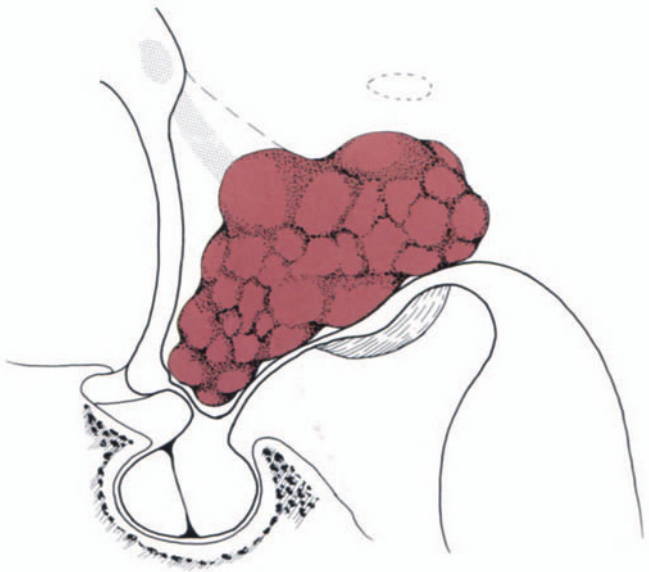


Figure 10.72. Hypothalamic *glioma*, different from the germinona, grows from the region of the optic chiasm, between the supraoptic and infundibular recessess of the III ventricle. The mass invariably fungates into the III ventricle, often becoming giant in size, and lobulates into the interpeduncular cistern.

germinoma, optic pathway or hypothalamic gliomas. Debulking of these tumors has not been demonstrated to provide benefits superior to those of Roentgen therapy alone. Indeed, the operative morbidity for children with tumors within the hypothalamus, children who are either already suffering from hypopituitarism or may develop a hypopituitary syndrome from the operative procedure, is much greater than any tumor, including IV ventricle lesions.

Parasellar Glioma (Figure 10.72 to 10.76)
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.

Parasellar glioma
1. is most common under 2 years of age;
2. causes roving nystagmus or spasmus mutans;
3. rarely causes diabetes insipidus, commonly causes precocious puberty;
4. invariably enlarges or deforms optic foramina and planum sphenoidale;
5. very seldom calcifies; and
6. enhances diffusely in CTT scan, showing lucent areas within its center.

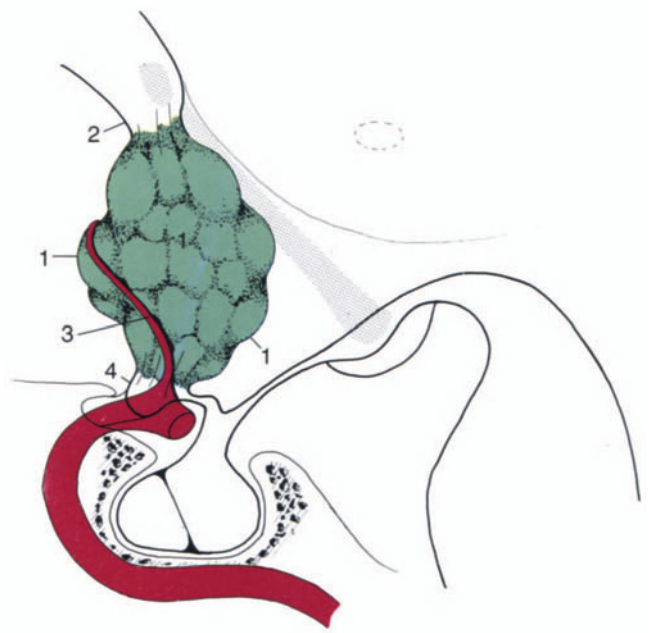
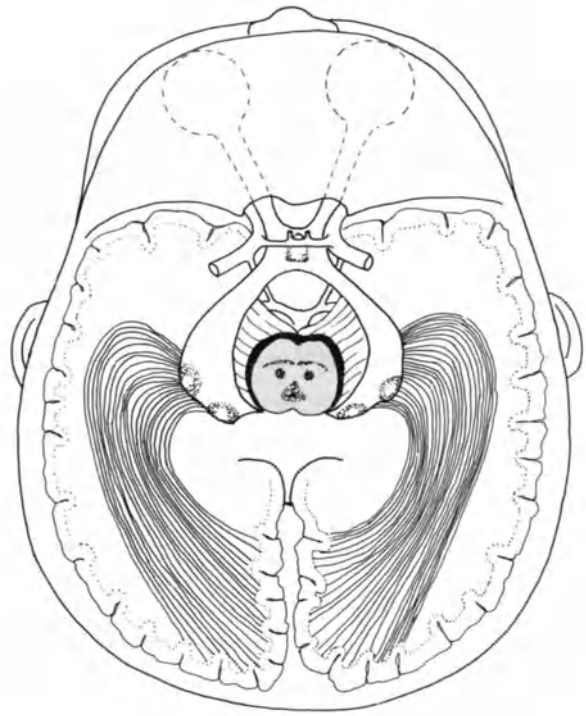


Figure 10.73. The glioma (*1*) expanding from lamina terminalis bulges posteriorly into the III ventricle and invades the rostrum of the corpus callosum (*2*). The A-1 segment of the anterior cerebral artery (*3*) is elevated and deformed, as the tumor extends inferiorly into the region of the optic chiasm (*4*).



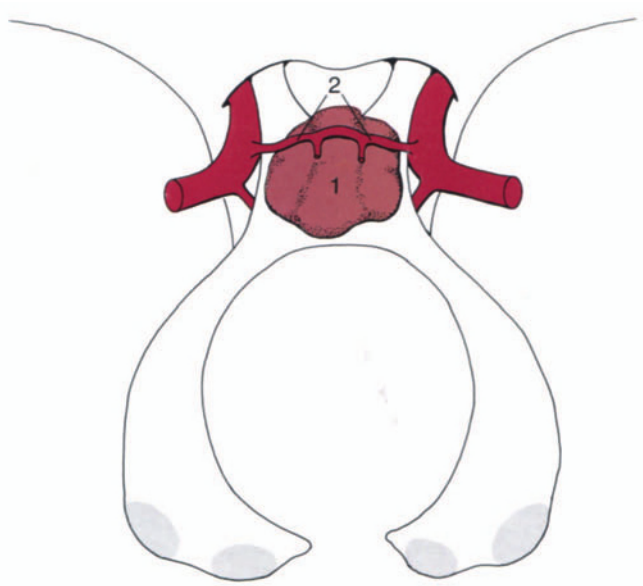


Figure 10.74. (A) The optic pathways, here reproduced, to permit a visual point of reference for discussion of gliomas of the optic pathways. (B) This is a coronal drawing of a glioma of the lamina terminalis (*1*) illustrating how its anterior and superior bulge deforms and stretches the A-1 segment of the anterior cerebral arteries (*2*).

Craniopharyngioma
1. with the *rarest* exception does not occur under 2 years of age;
2. causes visual field cuts;
3. commonly causes diabetes insipidus, never causes precocious puberty;
4. invariably calcifies;
5. very rarely enlarges optic canals; and
6. never deforms planum sphenoidale without enlarging sella turcica.

One may encounter a parasellar tumor which has grown to enormous size, with necrotic center, and which, consequently, may be causing difficulty because of its volume. These tumors should be debulked surgically: they are exophytic gliomas which are amenable to subtotal resection just as the exophytic glioma of the hindbrain.

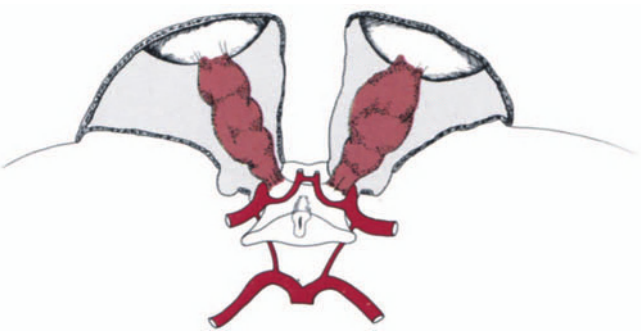


Figure 10.75. Bilateral optic pathway gliomas are not uncommon in children with Recklinghausen's disease. Here, tumor invasion of the intraorbital portions of the optic nerves is illustrated.

Biopsy of Parasellar Glioma
(Figures 10.77 and 10.78)
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 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 advised 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.

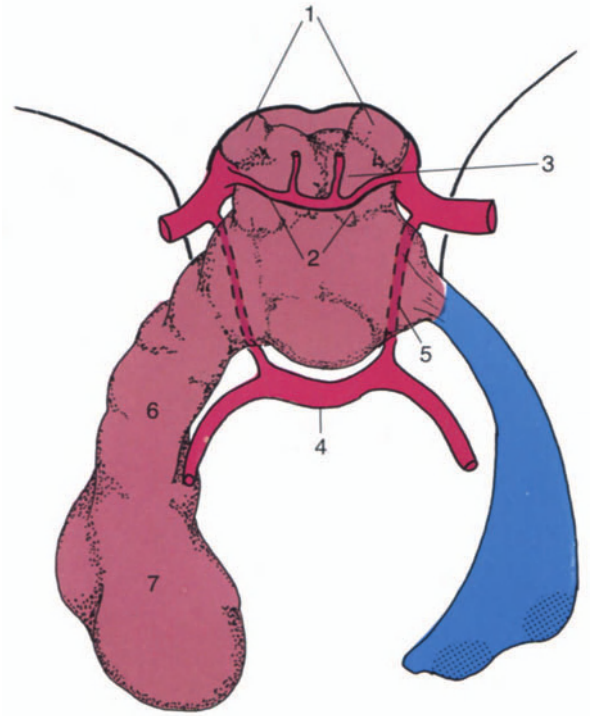


Figure 10.76. Optic pathway gliomas may involve the entirety of the intracranial optic nerve bilaterally (*1*), elevating and deforming the A-1 segments of the anterior cerebral arteries (*2*) at the transition point between optic nerve and optic chiasm (*3*). Tumefaction of the optic chiasm may fungate 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*).

Figure 10.77. Biopsy of hypothalamic glioma. Optic pathway gliomas may expand enormously and into either the frontal or, more commonly, temporal lobes. When this occurs, surgery is definitely indicated, so as to resect the intraparenchymal glioma and thereby provide maximal debulking and decompression. In this child, the optic pathway glioma (*1*) has expanded into both the superior temporal (*2*) and inferior frontal (*3*) areas.

A #15 blade is used to incise the tissue, and approximately 2 mm^3 of neoplasm are removed with a micropituitary biopsy forceps.

Hypothalamic Glioma

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.

Craniopharyngioma

In 1910 Dean Lewis[10] was the first surgeon to remove a craniopharyngioma. The longest recorded survival following craniopharyngioma surgery was in a patient operated by Harvey Cushing[11] 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 in 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 between the tumor capsule, hypothalamus, and optic chiasm. Consequently, one concludes that the most important factor in evaluating a child with a craniopharyngioma is anatomic location of the tumor. This determines the clinical

course and symptomatology, and causes the arterial and ventricular changes identified through neuroradiologic procedures.

The vascular changes permit an appraisal as to whether the tumor mass may 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 compose a significant, though generally minor, percentage of the total tumor. To a certain extent, CTT 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.

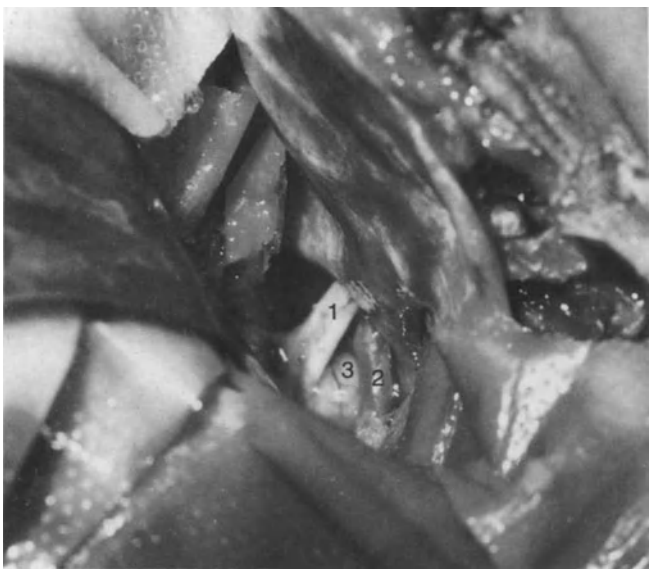


Figure 10.78. Hypothalamic glioma. Again, the hypothalamic glioma, as the optic pathway glioma, really should not be operated, since the diagnosis may be made preoperatively. However, in the event either of an inability to make this diagnosis (because of inadequate clinical information), or uncertainty 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 an hypothalamic glioma (*3*), which is expanding anteriorly beneath the optic chiasm and opening up the interopticocarotid space.

General Comments on Surgical Anatomy of Craniopharyngioma (Figures 10.79 to 10.81)

After the bifrontal craniotomy has been performed, the dura is opened over both frontal lobes and the superior sagittal sinus transected and cut. The falx cerebri is separated from the crista galli, and both olfactory nerves are coagulated and transected. 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. The frontal lobes fall posteriorly and medialward, the temporal lobes either remain in the anterior portion of the middle fossae or 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 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, 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 10.79, 10.80, and 10.81 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 optic nerve and internal carotid artery 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 cistern, 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

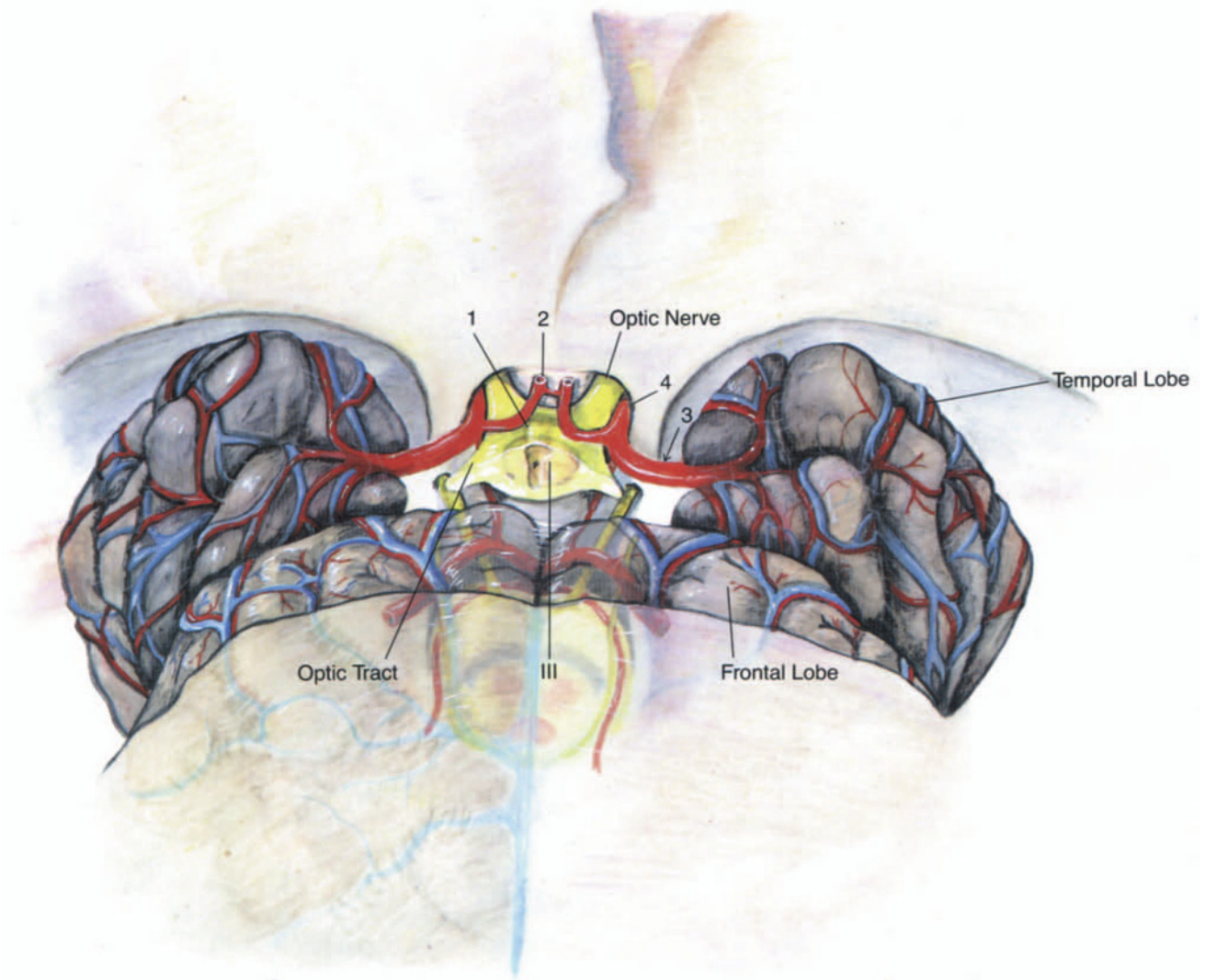
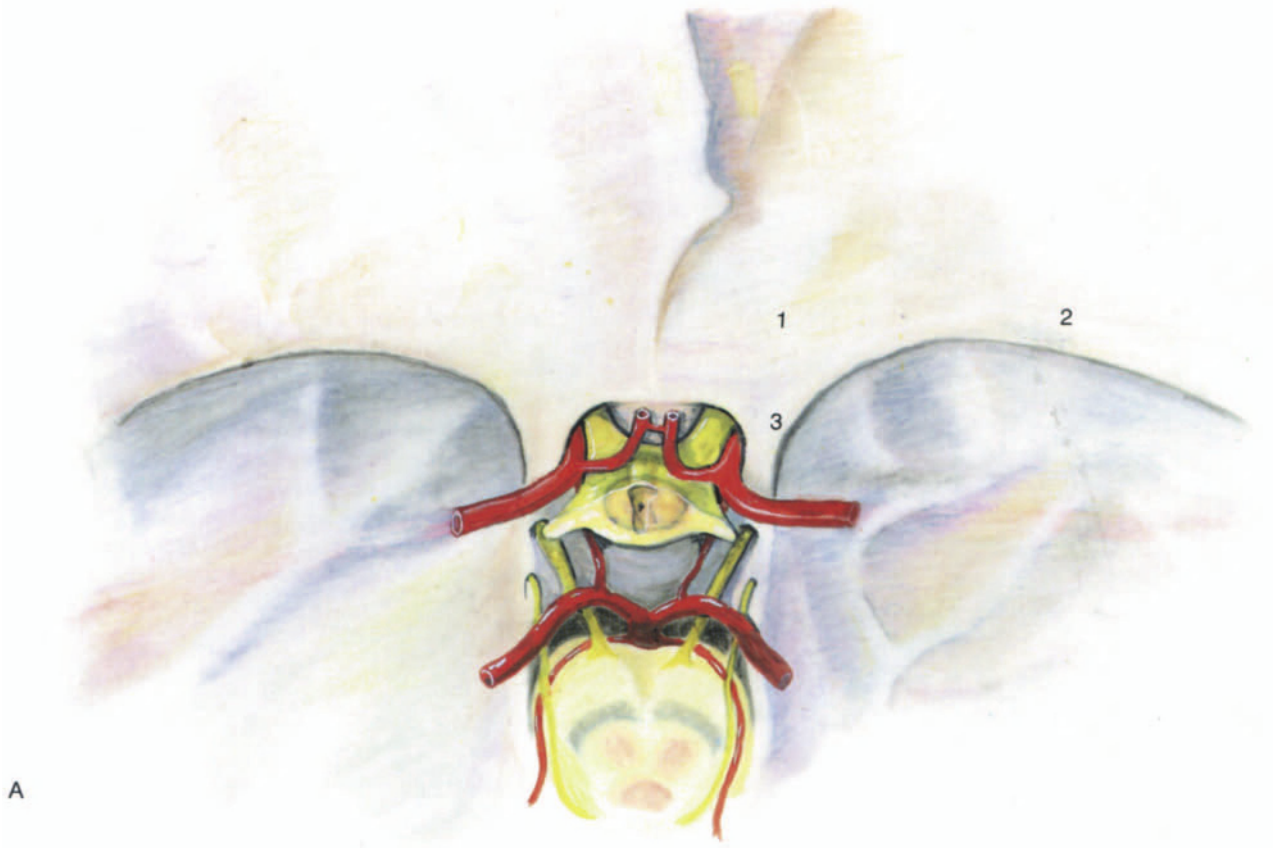
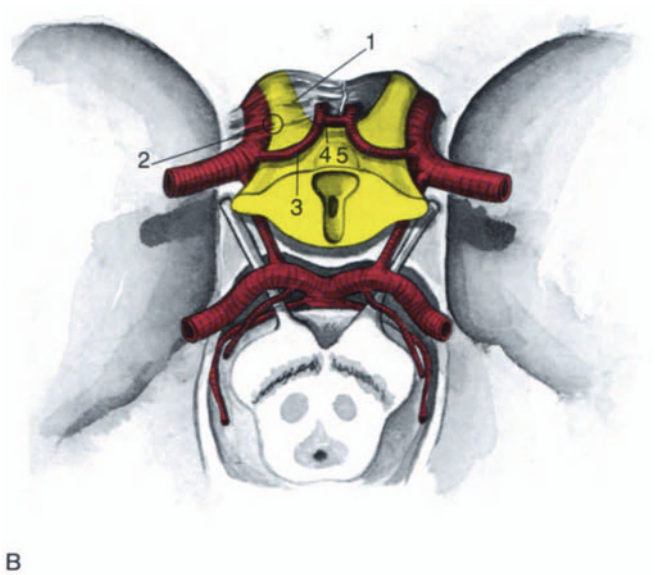


Figure 10.79. The surgeon's view of the parasellar area after the falx cerebri has been cut from the crista galli and both olfactory nerves transected. In this drawing the III ventricle above the hypothalamic sulcus and both optic tracts has been removed to allow the reader to conceptualize intraventricular 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.

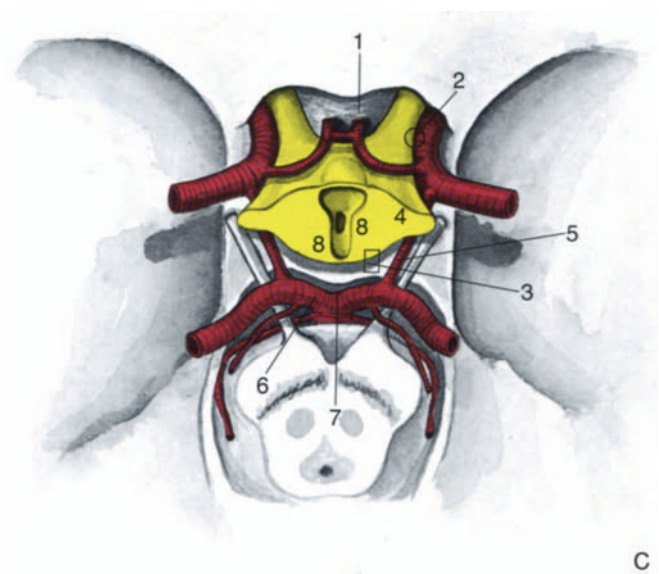
Figure 10.80. (A) The cerebrum and basal ganglia have been removed, allowing one to appreciate access to the parasellar area along the cribriform plate and planum sphenoidale (*1*), or the lesser wing of the sphenoid (*2*) and anterior clinoid (*3*). (B) The arachnoid membrane is intact over the prechiasmatic 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 three basic approaches to craniopharyngioma: prechiasmatic (*1*), interopticocarotid (*2*), and retrochiasmatic (*3*) between the optic tract superiorly (*4*) and the posterior communicating artery (*5*) inferiorly. One should note how very close the posterior cerebral (*6*) and basilar (*7*) arteries are to the hypothalamus (*8*). ►



A



B



C

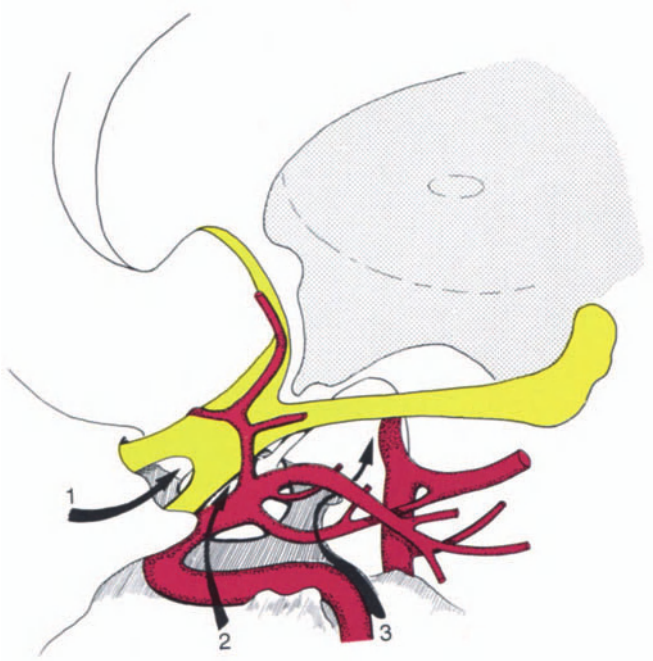


Figure 10.81. The three access (arrows) routes to infra- and retrochiasmatic craniopharyngioma: (*1*) The first route, between the two optic nerves, allows one to probe beneath the optic chiasm. It is unsatisfactory because one is obliged either to extend blindly his dissector along the optic chiasm 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, superior and inferior to the posterior communicating 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 telfa in order to protect them. Resection of the anterior 2 cm of the temporal lobe allows one to work his way superiorly along the internal carotid artery to its bifurcation without compressing the temporal lobe or internal capsule. This provides excellent visualization of the infundibular and interpeduncular cisterns, the basilar and posterior cerebral arteries, the peduncles.

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 facilitated by working through the three routes: prechiasmatic, interopticocarotid, and retrochiasmatic.

Classification of Craniopharyngioma

The recommended surgical classification of craniopharyngiomas, dividing them into five distinctly different anatomical forms (locations) is that proposed by Jacques Rougerie[12]:

Prechiasmatic Craniopharyngioma

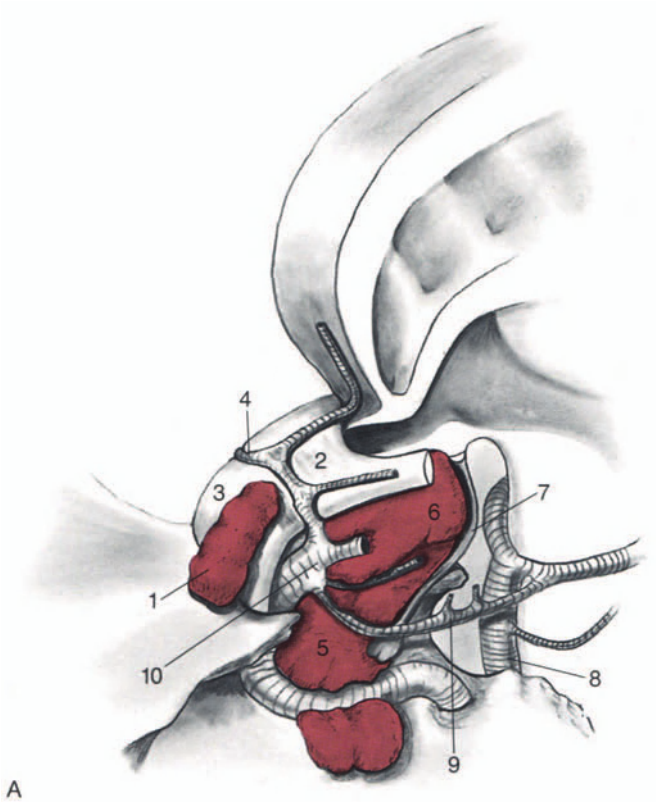
(Figures 10.82 and 10.83)

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.

Craniopharyngiomas 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, before extending anterior to the chiasm between the carotid arteries and optic nerves. The prechiasmatic cistern is obliterated as the tumor, invariably contained within its capsule, thus maintaining the globular form, grows anterior to the chiasm, and then superior to it, displacing it posterosuperiorly as it stretches and verticalizes 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 attenuation of branches from A-1 to the optic chiasm. For these reasons, funduscopic examination reveals optic atrophy, visual field examination reveals defects indicative of optic nerve and anterior chiasm involvement. The visual field defects are often "explosive" in onset and progression.

Cerebral 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 an attempt at complete resection is indicated by the venographic phases 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,



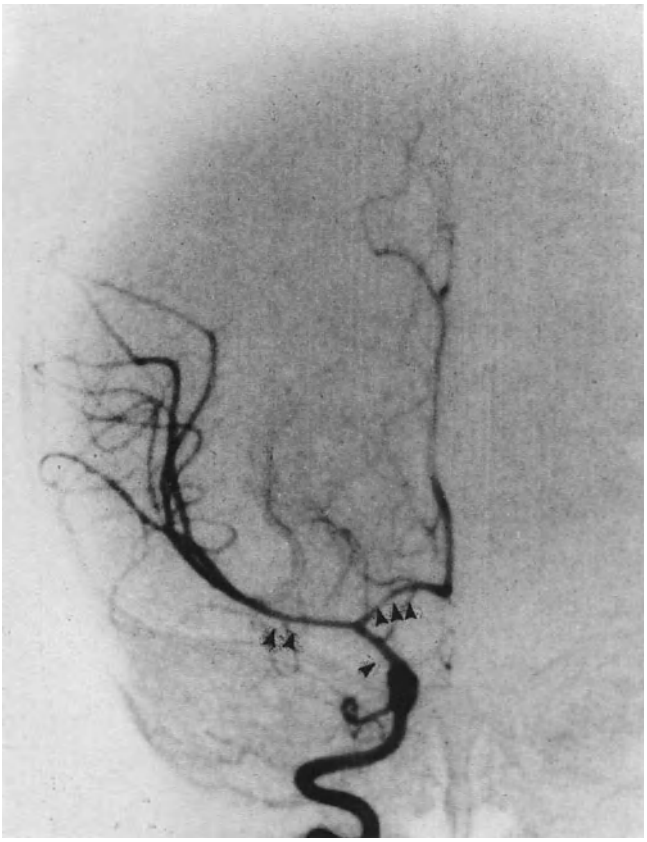


Figure 10.82. (A) The prechiasmatic extension of tumor (*1*) elevates the optic chiasm (*2*) and verticalizes the optic nerve (*3*). A-1 (*4*) is elevated with the chiasm, so that it may be used as an angiographic indicator of whether there is prechiasmatic extension of tumor. There is always infrachiasmatic (*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. The basilar artery (*8*) is not deformed, although the posterior communicator (*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) Prechiasmatic extension of the craniopharyngioma is indicated arteriographically in the half-axial projection by elevation of the A-1 segment of the anterior cerebral artery (triple arrows). The double arrows reveal elevation of the middle cerebral artery resulting from a dilated temporal horn, and the single arrow shows attenuation of the internal carotid artery resulting from stretching.

one must be prepared to remove retrochiasmatic lobules. The enlarged prechiasmatic area, made ample by partial tumor removal, facilitates this.

Intrasellar Craniopharyngioma

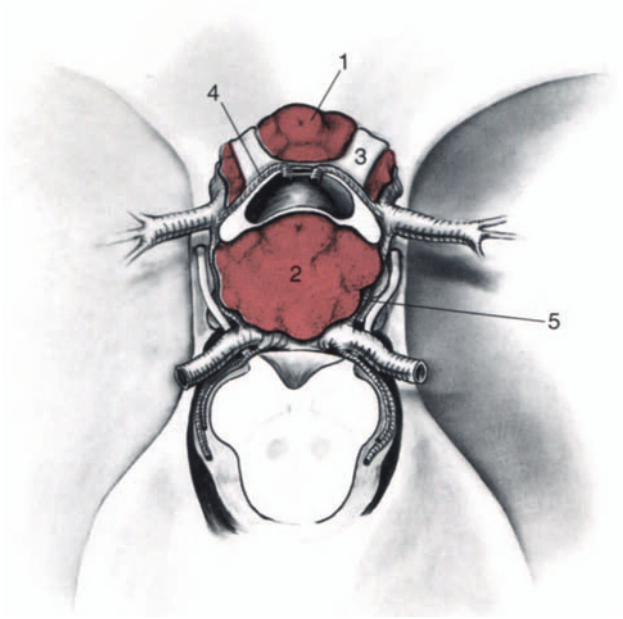
The intrasellar craniopharyngioma is located entirely within the sella turcica, beneath the *diaphragma sellae*, and causes enlargement of the sella turcica's volume, partial or total destruction of the clinoids, invasion of the sphenoid sinus, or compression and displacement of the cavernous sinuses. 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.

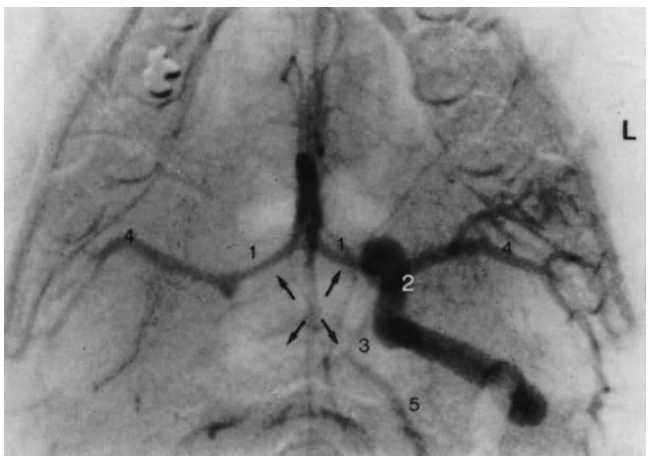
Previously, these tumors have been considered to present with signs indicative of pituitary dysfunction, giving cause to believe that some, or all, of the pituitary function could be preserved since the gland may not be totally destroyed. More recently, however, we have come to learn that intrasellar 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 (Figure 10.84)

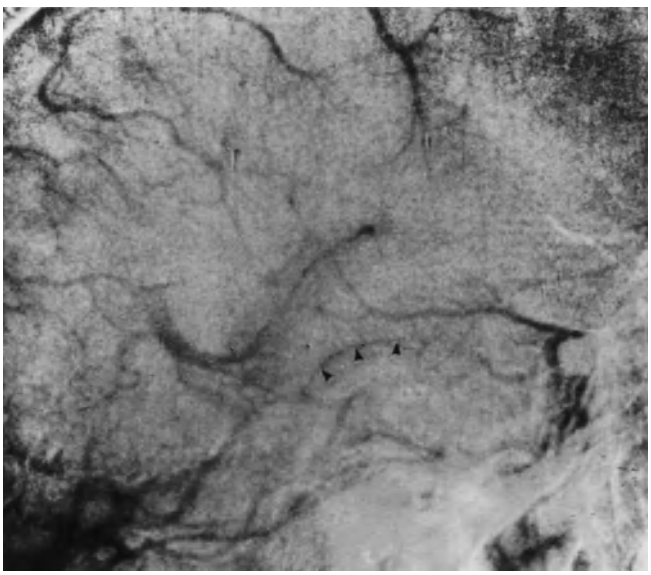
The tumor mass extends posterior to the optic chiasm, between it and the optic tracts, displacing and, at times, invading both the hypothalamus and III ventricles.



A



B



C

Figure 10.83. (A) 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. (B) The circle of Willis is invariably involved either partially or in its entirety, when a craniopharyngioma expands into the suprasellar region. The A-1 segments of the anterior cerebral artery (*1*), the bifurcation of the internal carotid artery (*2*), the posterior communicating artery (*3*), and the middle cerebral arteries (*4*), all may be studied to advantage in this child, as may the posterior cerebral artery on the left (*5*). The circle of Willis in this child is remarkably widened (arrows) by the symmetrical expansion of tumor mass within it. (C) Here one sees elevation of the vein of Rosenthal (arrowheads), indicating a "totally" resectable craniopharyngioma.

Approximately one third of the patients with retrochiasmatic craniopharyngiomas are under ten years of age. Less then 5% of these tumors may be "totally" removed. The mortality rate is high, ranging from 20 to 30%.

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. Angiography reveals anterior displacement of the A-1 segments of the anterior cerebral arteries and of the anterior communicating arteries, with posteroinferior displacement of the posterior communicating and basilar arteries, causing bowing of the perforating branches of these latter vessels. The lateral and posterior thalamoperforating vessels, generally numbering six on either side, are streteched and bowed with the concavity facing anteriorly. Because these tumors invariably either invade or nestle within the hypothalamus, displacing the anterior perforated substance downward, they cause an inferior displacement of the vein of Rosenthal. These vascular changes indicate to the surgeon that the tumor may not be totally resected.

The obstruction of the basal cisterns and the posterior portion of the III ventricle results in hydrocephalus, an increase in intracranial pressure, and 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.

The location of these tumors behind the optic chiasm is particularly grave in that it renders surgical removal more difficult, 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 thalamoperforating 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 capsule may engulf the posterior communicating arteries and their thalamoperforating branches. The combination of damaged hypothalamus plus severe increases in intracranial pressure is 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.

Removal of the tumor encumbers one to do so "piecemeal," through anatomical planes between the oculomotor nerves and caroticobasilar systems. *Total resection of these tumors is not advisable.* 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.

"Les Formes Geantes"

These giant forms "explode" from the sella turcica, expanding into the prechiasmatic region, directly superiorly, 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 the tumor extends into the anterior, middle, and posterior fossae. The apparently single globular mass, in fact, is generally polyglobular 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 of newborn. The dense capsule adheres to the pia archnoid 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, as well as limbic, lobe involvement accompanies destruction of the optic pathways, 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 a problem.

Atypical Craniopharyngioma

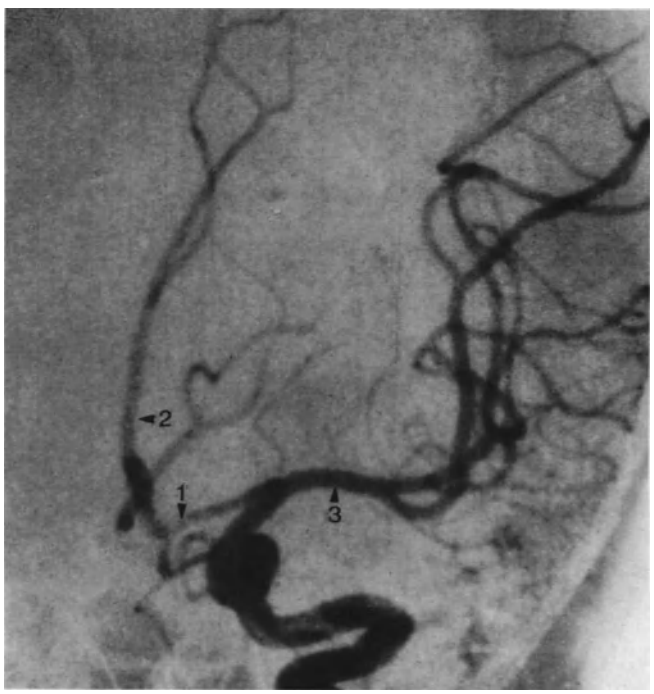
The atypical forms of craniopharyngioma may be located within the pharynx, the posterior fossa, limited entirely to the sphenoid sinus, even in the pineal region. When craniopharyngioma is not located in the parasellar area, but either within the sphenoid sinus, clivus, pharynx, posterior fossa, or pineal region, it does not behave clinically as a parasellar tumor. Accordingly, it is of embryologic and histologic 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.

Supplemental Surgical Management of Craniopharyngioma

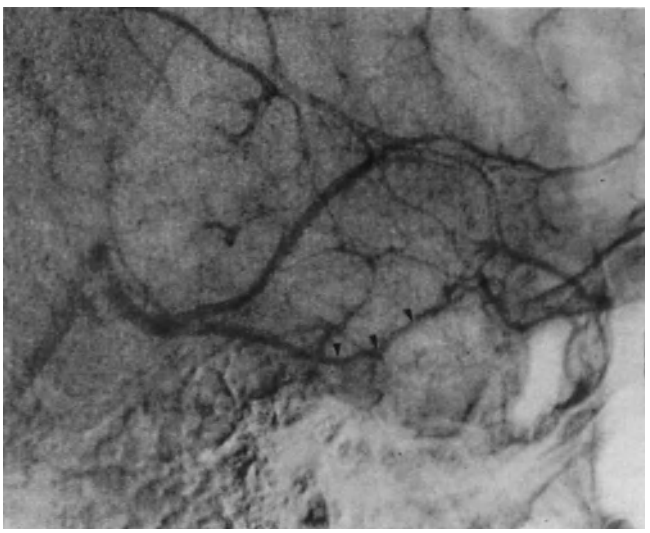
Hydrocephalus complicating craniopharyngioma may result from tumor obstructing the basal cisterns, invasion of the III ventricle and occlusion of the foramina of Monro, and, very rarely, posterior displacement of the brainstem with occlusion of the aqueduct of Sylvius. A unilateral ventriculoperitoneal 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 diminishes immediately 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 diminish intracranial fluid volume.

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 into the cystic cavity, so as either to diminish the secretion of cystic fluid or destroy the secretory cells lining the cavity. This procedure is theoretically attractive but has not been widely accepted, very likely because it is not as easy and safe as direct surgical exposure of the tumor mass. It is not possible 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

A

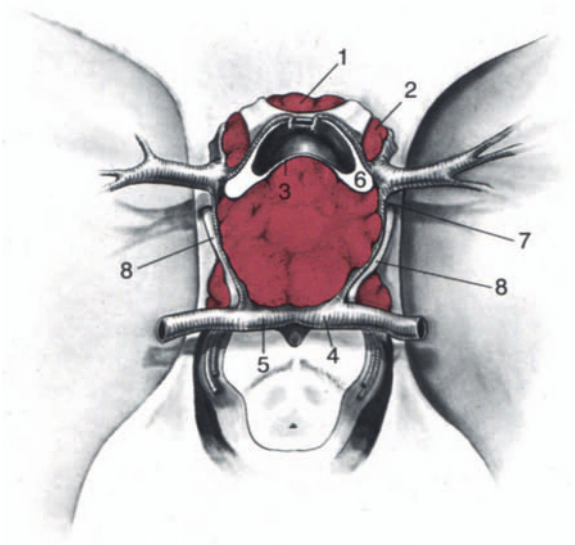


B

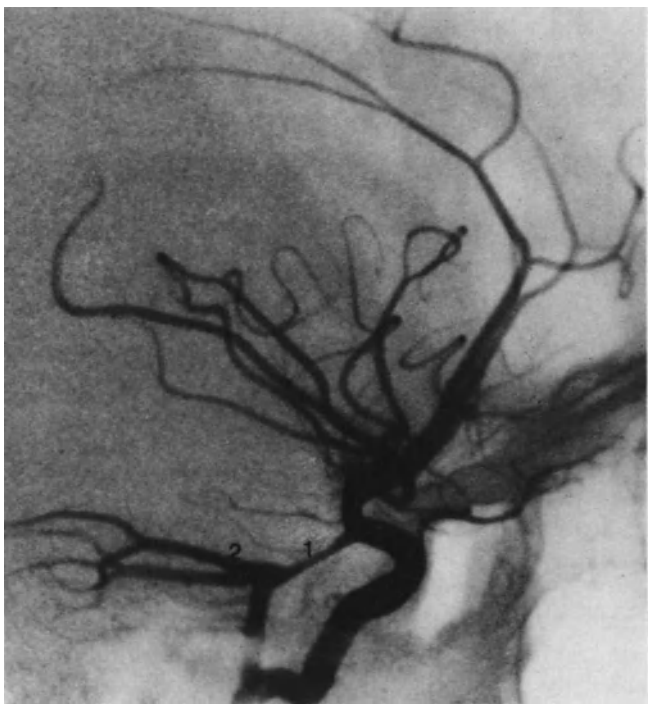


C

Figure 10.84. (A) Retrochiasmatic tumor extension (*1*) results in forward displacement of the optic chiasm (*2*), an anatomical change illustrated angiographically by forward and, at times, downward displacement of A-1 (*3*). The tumor elevates remarkably the floor of the III, at times fungating into it (*4*), and engulfs the pituitary stalk so that 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) The depression of A-1 (*1*) and the contralateral bowing of A-2 (*2*) permit one to identify the tumor mass as being located behind the prefixed optic chiasm and thus within the III ventricle. Depression of A-1 signifies anterior and inferior displacement of the chiasm. Bowing and shifting of A-2 signifies invasion of the III ventricle and forward displacement of the lamina terminalis. The elevation of M-1 (*3*) indicates cystic transformation of the Sylvian cistern and extension of tumor into the middle fossa. (C) Depression of the vein of Rosenthal (arrowheads), pushing it downward and upon the posterior clinoid processes, confirms the impression gained that this craniopharyngioma is not totally resectable because it is invading the hypothalamus. (D) 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, debulked, by working through the interopticocarotid space and the retrochiasmatic region between optic tract (*6*) and posterior communicating artery (*7*). The III cranial (*8*) nerve is displaced laterally. (E) The downward displacement of the posterior communicating (*1*) and posterior cerebral (*2*) arteries indicates expansion of tumor mass within the III ventricle, hypothalamus and thalamus: this tumor is not totally resectable. ►



D



E

vision radioactive substances of his choosing. The most significant advantage of direct exposure of the tumor is that it allows the surgeon to evaluate the resectability of the tumor mass, and remove it if feasible.

Surgical Approach to Craniopharyngioma: General Comments

Irrespective of the surgical approach to the craniopharyngioma and its intrasellar, prechiasmatic, retrochiasmatic, or other location, the technique for removing the tumor is always the same, *but not the anatomical approach.* This technique is predicated upon the fact that craniopharyngioma may be solid, cystic, solid/cystic, well-encapsulated, 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 system, 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 then the capsule opened by the blades of the bipolar forceps. Cerebral 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 the 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 should then be used to separate the capsule from surrounding tissue by pressing on capsule and pushing it toward the sella turcica, 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, bit at a time. *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 either 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 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 of entry to the neoplasm. Thus, one may be working through the interopticocarotid space as his 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 piecemal removal.

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 he is able to get around the capsule with a dissector before delivering it through his 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 adherent 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 be to perform a complete resection of the tumor. Surgical removal of the intrasellar and prechiasmatic craniopharyngiomas is benign, attempts at total removal of retrochiasmatic and giant forms subjects the patient to very highy morbidity and mortality risks.

Surgical Technique: Specific Procedures

Rhinoseptal, Transphenoidal 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. Particular attention should be 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 particular 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 possible. If not, it should be clipped or coagulated and sectioned.

The high incidence of recurrences of intrasellar craniopharyngioma speaks eloquently of the difficulty to be 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. Recent use of the operating microscope has brought us to extend our procedures slightly more lateralward (*the bifrontopterional 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: *Prechiasmatic Approach* (Figure 10.85). 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 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. The dissection of the optic nerves is thereby facilitated. At this time one should attempt to identify the pituitary stalk. If it is found intact, it should be sectioned. 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 performed behind the tumor, *with the intention to deliver too soon the posterior portion of the tumor through the prechias-*

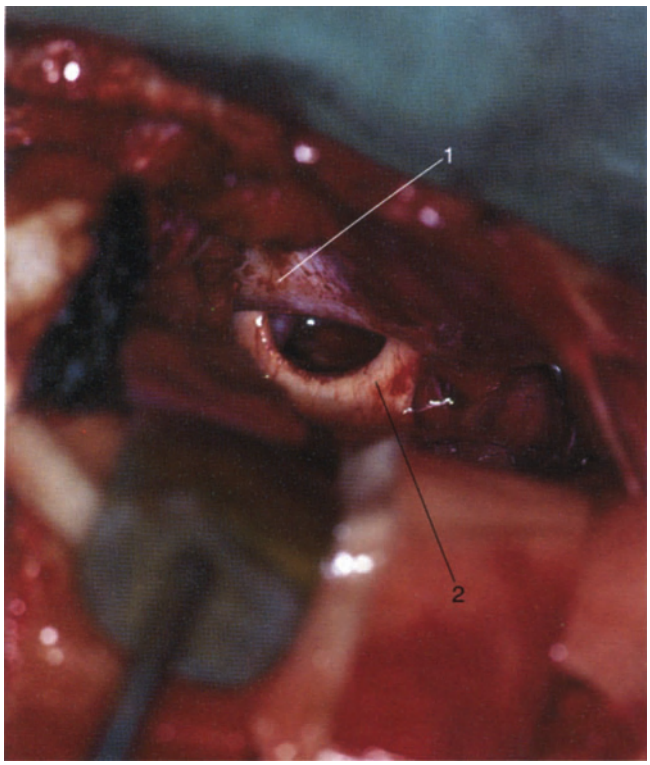


Figure 10.85. Prechiasmatic extension of craniopharyngioma presents no particular surgical problems. There is an adequate amount of tumor protruding between the planum sphenoidale (*1*) and the optic chiasm (*2*) that the surgeon may immediately enter the tumor, drain the fluid within it so as to attain decompression, and then proceed to separate 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 planum sphenoidale and optic chiasm.

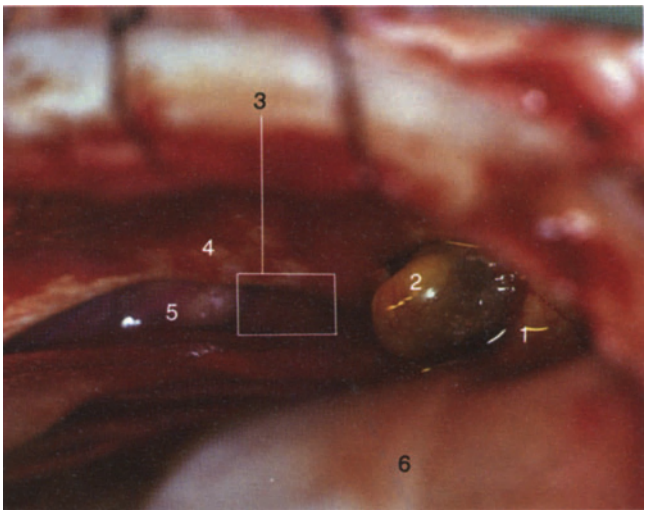


Figure 10.86. The interopticocarotid space provides the surgeon a second portal for access to the craniopharyngioma expanding beneath the optic chiasm (*1*). This photograph illustrates a craniopharyngioma (*2*) which dissected its way through the interopticocarotid space, elevating the optic nerve and 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.

matic space. The next step is dissection of the tumor and removal of its retrochiasmatic extensions, delivering them through the prechiasmatic space. In this portion of the procedure, the surgeon stays within the subarachnoid space. At times, it is possible to separate more or less completely the tumor capsule from the arachnoidal membranes composing the basal cisterns in and around the sella turcica.

The difficulties encountered in this approach are 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 greatly the anatomic exposure of the area.

Problems in the intrasellar area include precise separation of tumor tissue from residual pituitary gland, which is difficult even when the operating microscope is used, 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, a view into the retrochiasmatic area and superior portion of the interpeduncular and pontine cisterns.

Interopticocarotid Approach (Figures 10.86 and 10.87). 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,[13] and has been used by various authors since that time. The only additional contribution herein suggested 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 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.

Retrochiasmatic Approach (Figure 10.88). One may choose to dissect completely the optic nerves, passing along their superior surfaces to the optic chiasm, and then continuing the dissection over the optic chiasm posteriorly to the infundibulum, sectioning this latter after coagulation or clipping, entering the retrochiasmatic space by separating completely the optic path-

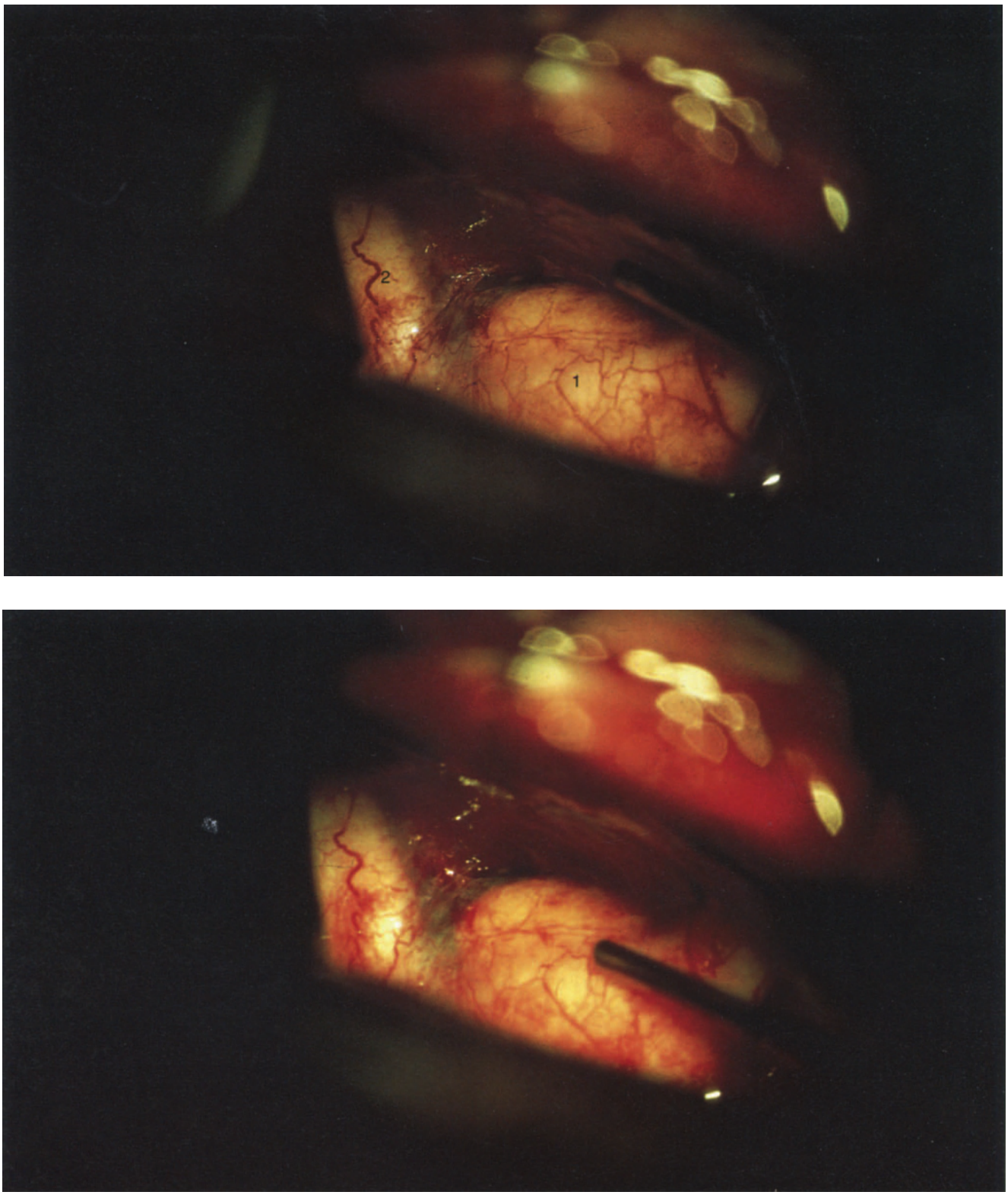
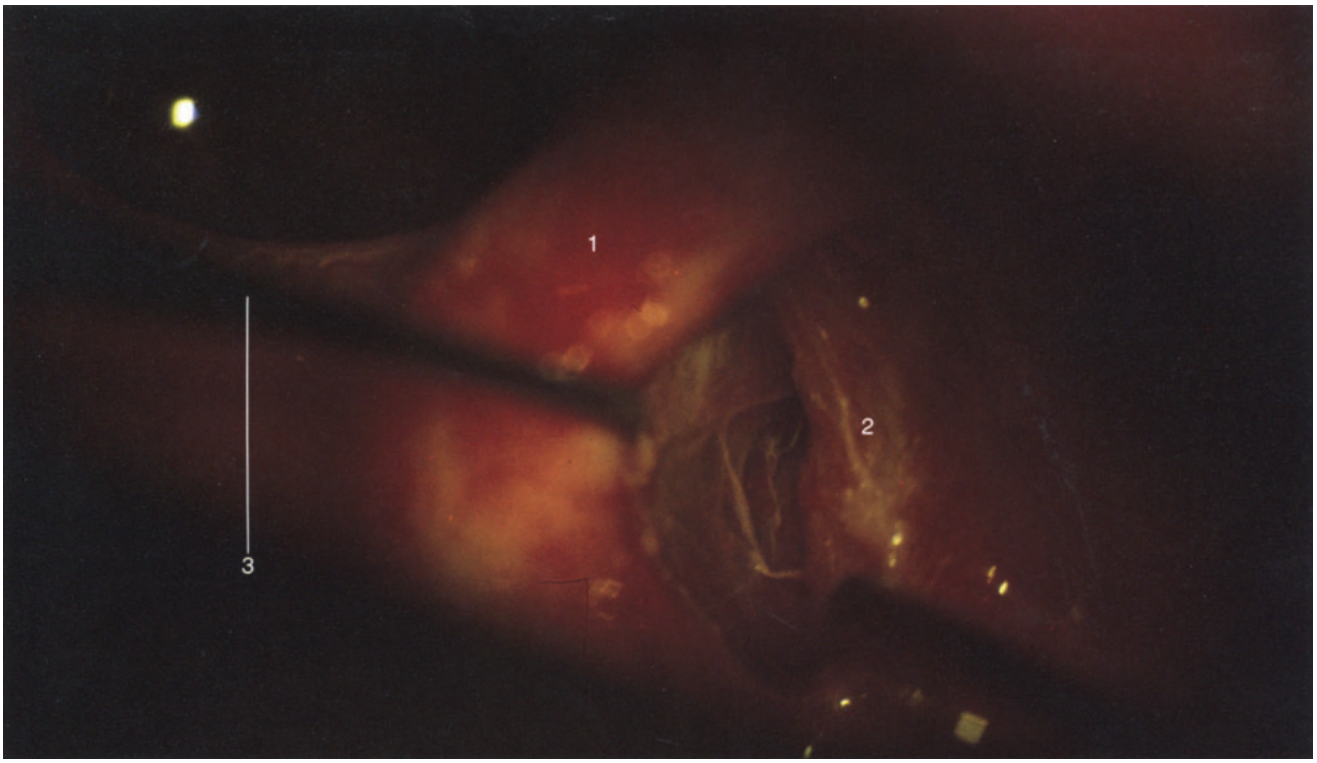
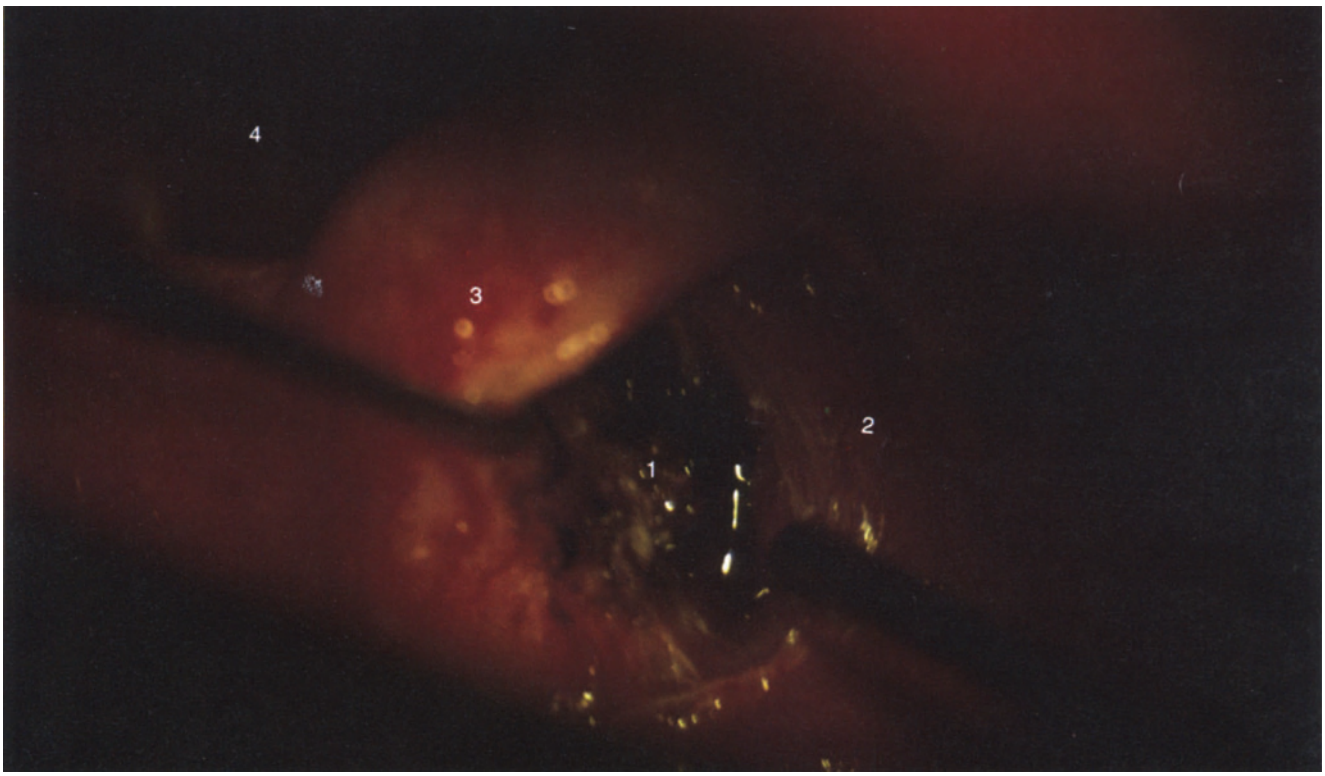


Figure 10.87. (A) This is an example of a child whose craniopharyngioma expanded beneath the optic chiasm (*1*), elevating it and displacing its most anterior surface onto the planum sphenoidale, elevating and flattening the left optic nerve (*2*). One sees a small amount of craniopharyngioma protruding 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) Consequently, one may open the interopticocarotid space. The ►

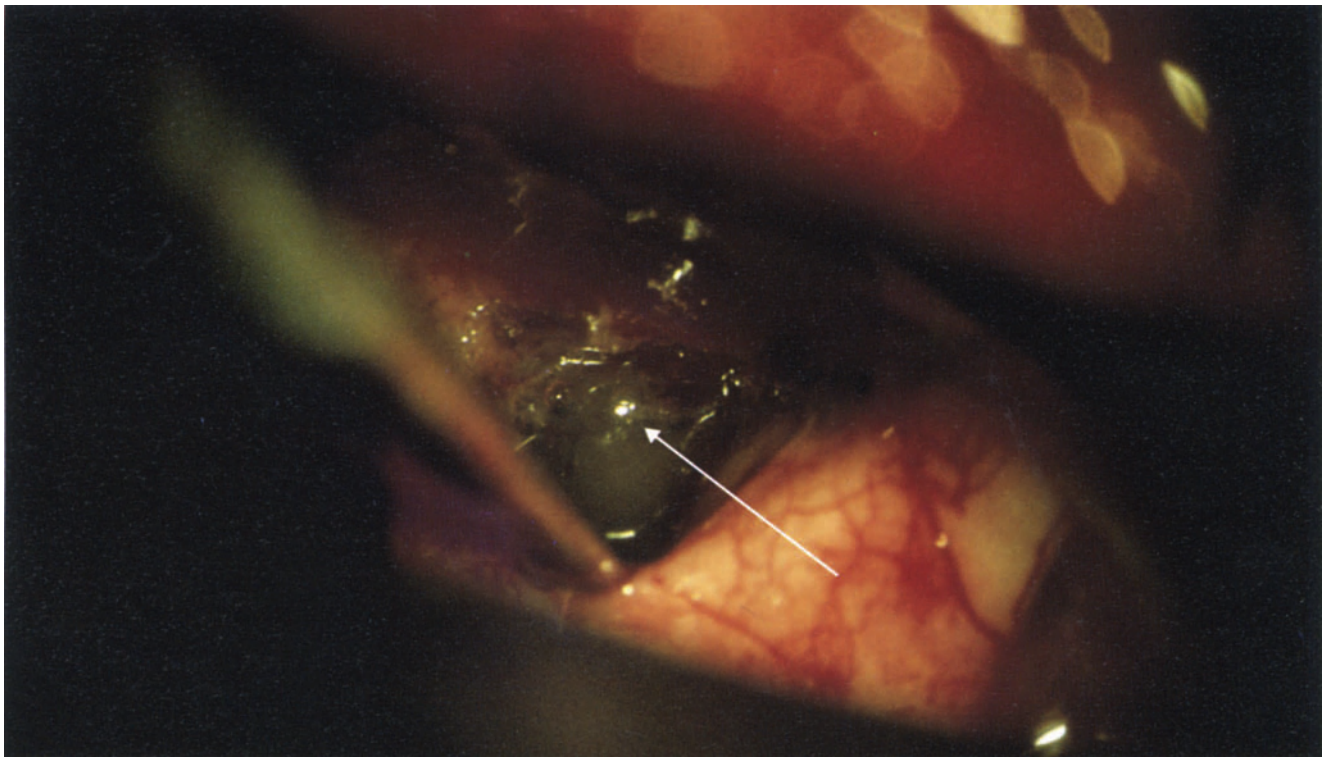


C

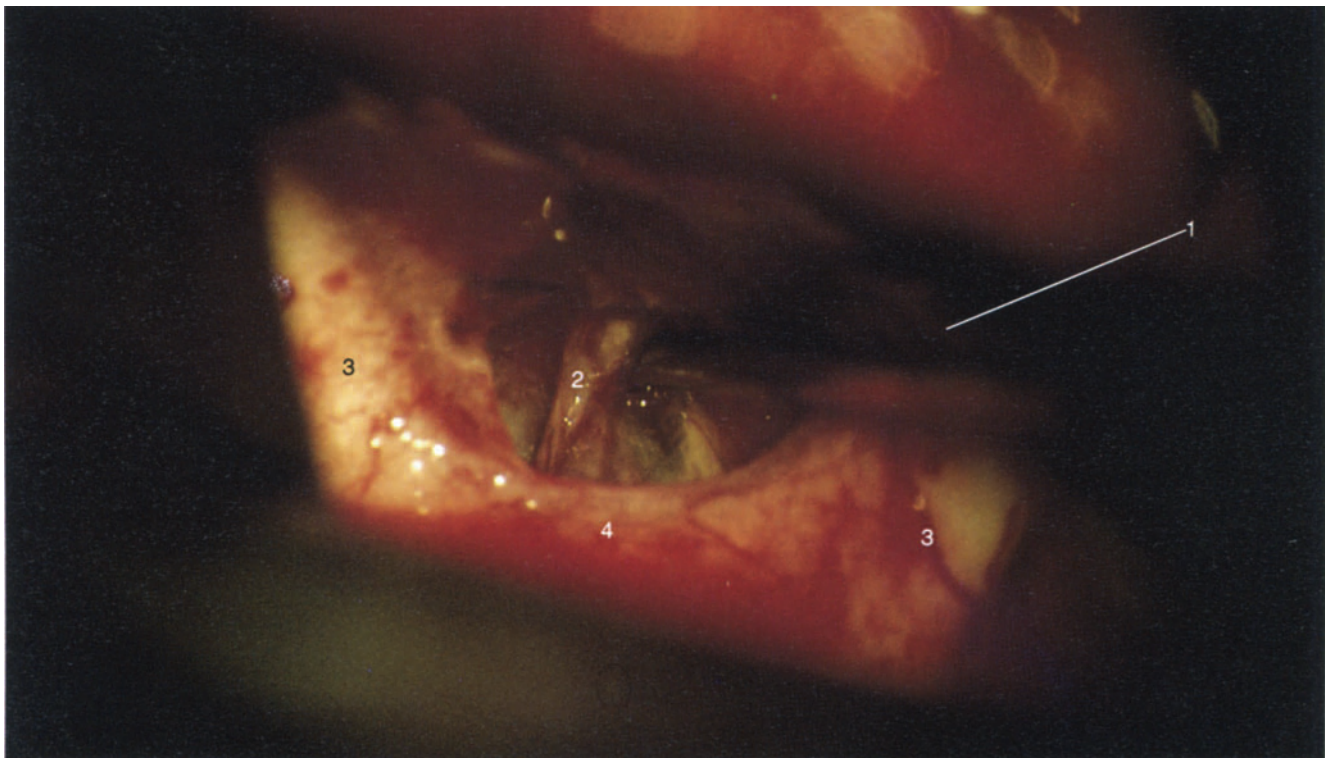


D

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. (D) 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. Then, either a dissector or the suction is used to "feed" tumor from the interopticocarotid opening beneath the optic ►



E



F

nerve (*3*) anteromedially to the prechiasmatic opening (*4*). (E) The tumor in the prechiasmatic space (arrow), pushed there through the interopticocarotid opening, may now be lifted from the field. (F) 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*).

ways 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 Subtemporal Approach

One may perform the subtemporal approach (called "pterional" by some) in either one or two stages. Cerebral spinal 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 to dissect 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, sectioning one column of the fornix, 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 an extremely difficult undertaking, one which presents an unacceptable risk of damage to the hypothalamus. It may be undertaken only as a supplementary approach, for multilobular 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. And then, maybe it is best to avoid it!

Corpus Callosum and Septum Pellucidum Tumors

Corpus Callosum: Surgical Anatomy

(Figures 10.89 and 10.90)

The corpus callosum is the largest commissure in the central nervous system, connecting the two cerebral hemispheres at the frontal lobes anteriorly, the parietal lobes in the center, and the occipital lobes posteriorly. When viewed in coronal sections, it is seen to extend from hemisphere to hemisphere, superior to the lateral ventricles, radiating into frontal lobes as the forceps minor and into the occipital lobes as the forceps major. In sagittal section its anatomical subdivisions are the rostrum, genu, body, and splenium. The rostrum extends directly to the lamina terminalis and the anterior commissure, at the point where the columns of the fornix turn posteriorly into the body of the fornix. As the rostrum extends anterosuperiorly it widens, gently expanding into the anatomical subdivision known as the genu. This is the most anterior extension of this commissure in the sagittal plane. Its curvilinear course brings it posteriorly into the body, which, in turn, extends posteriorly into the splenium which hangs over the superior and posterior aspects of the III ventricle and the pineal gland. It is immediately posterior to the quadrigeminal plate and anterosuperior to the great vein of Galen.

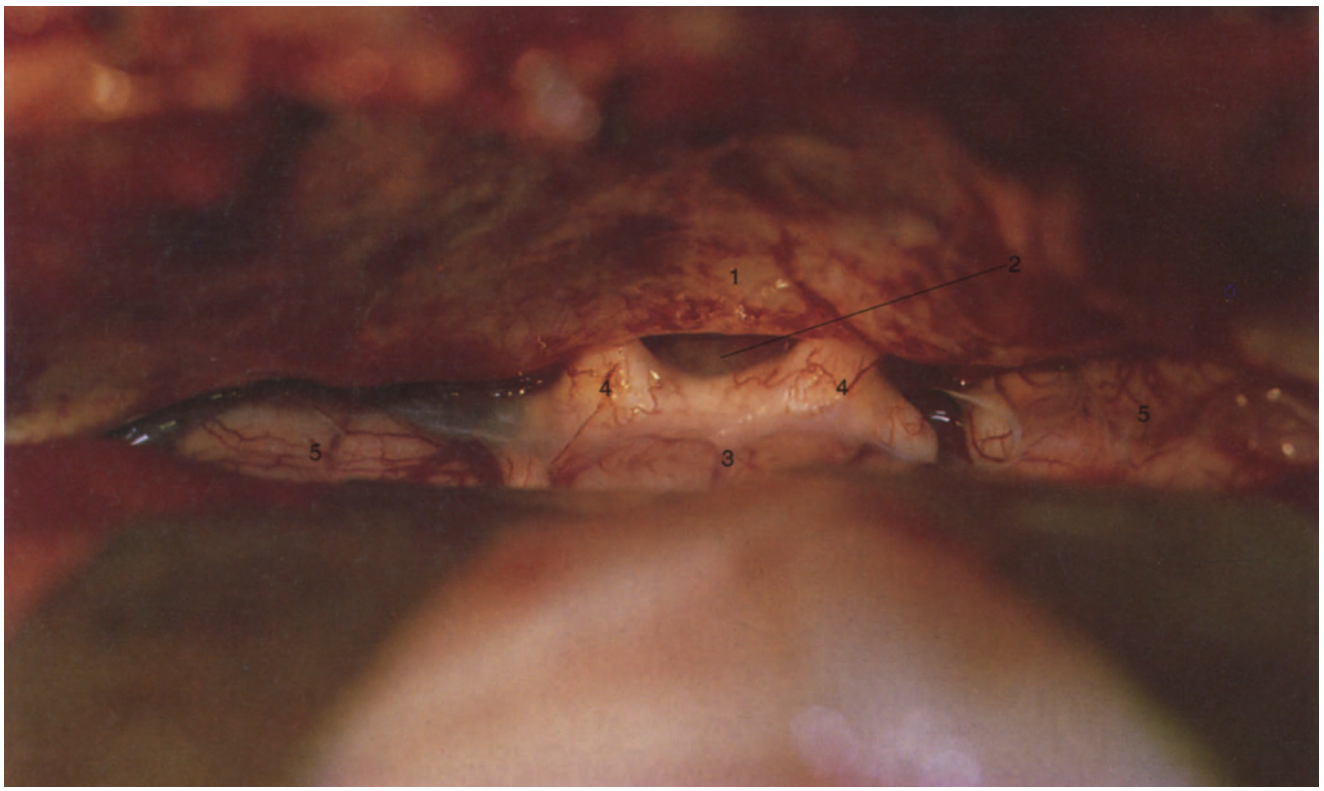
Corpus callosum tumors should not be operated since they are either highly malignant or completely benign. The former are represented by astrocytoma diffusum, primitive neuroectodermal tumor, diffuse intraprenchymal ependymoma, glioblastoma multiforme, lymphoma, atypical teratoma. All of these extend into the corpus callosum from surrounding parenchymal structures. Though partial or "total" resection is technically possible, the invasion of the parietal lobes and fornices, invariably 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.

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.

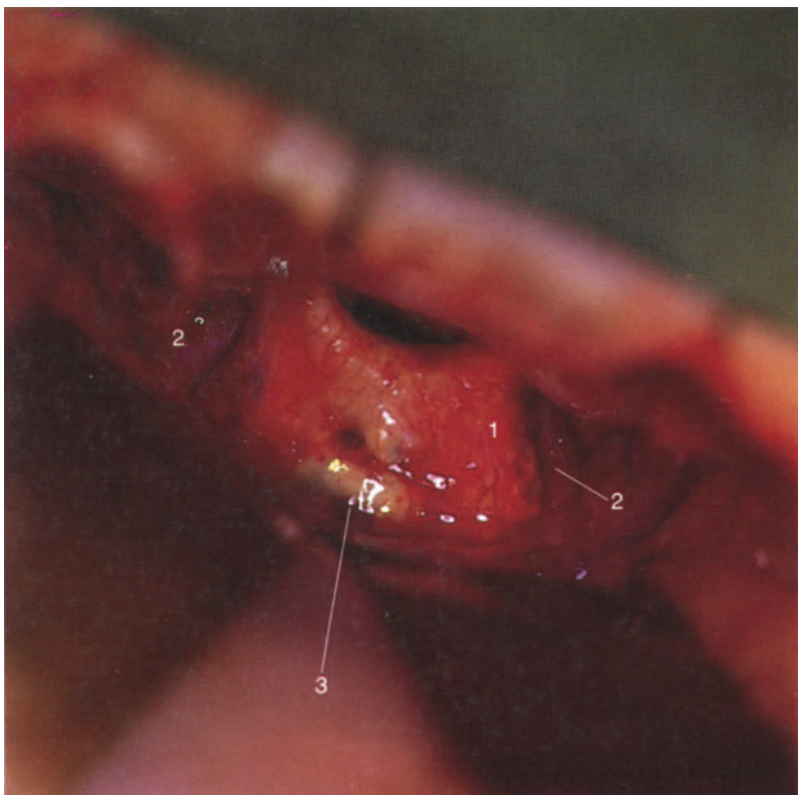
Septum Pellucidum: Surgical Anatomy

(Figures 10.91 to 10.94)

The septum pellucidum is a double membrane strung tautly between the genu and body of the corpus callosum anteriorly and superiorly, and the body of the fornix and transverse fissure anteriorly 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 between the two surfaces of the septum pellucidum 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

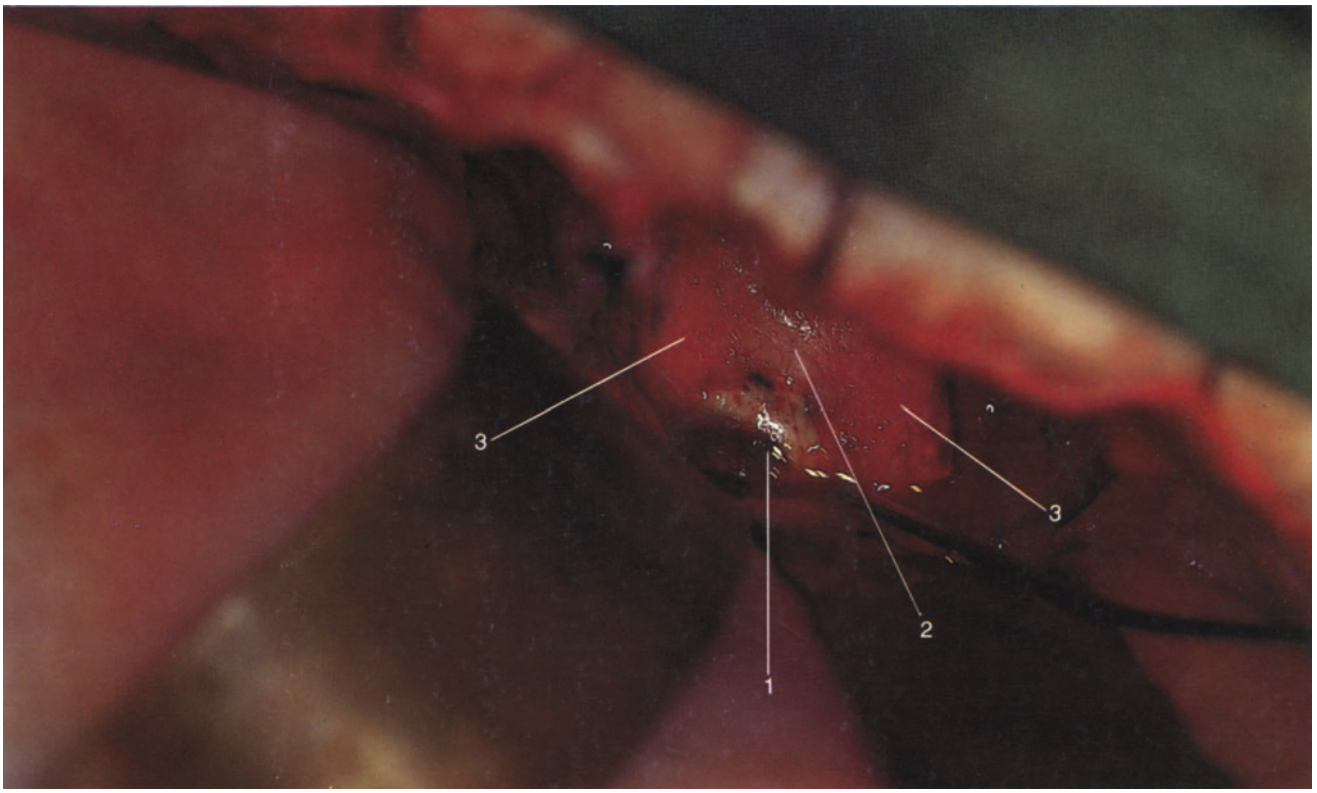


A



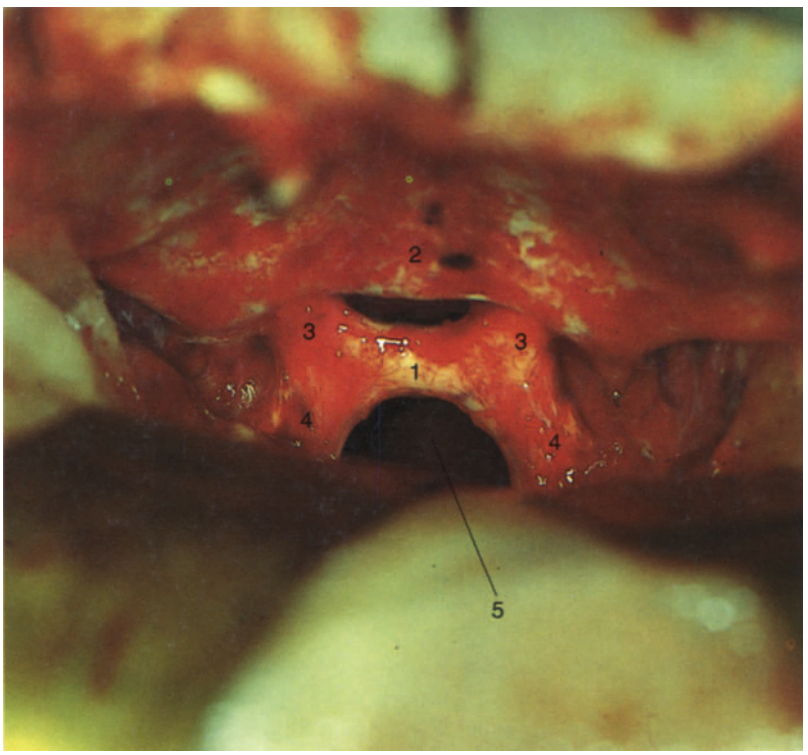
B

Figure 10.88. Retrochiasmatic tumors expand almost entirely behind the chiasm, though certainly there are both infrachiasmatic and, at times, small amounts of prechiasmatic neoplasm. (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 left temporal lobe are well visualized. The arachnoid bridging from the right 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 these latter, in turn, freed from the internal carotid arteries, (*2*) one proceeds to identify the A-1 segments of the anterior ►



C

cerebral artery so as to dissect the lamina terminalis (*3*) from the chiasm. (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. (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*).



D

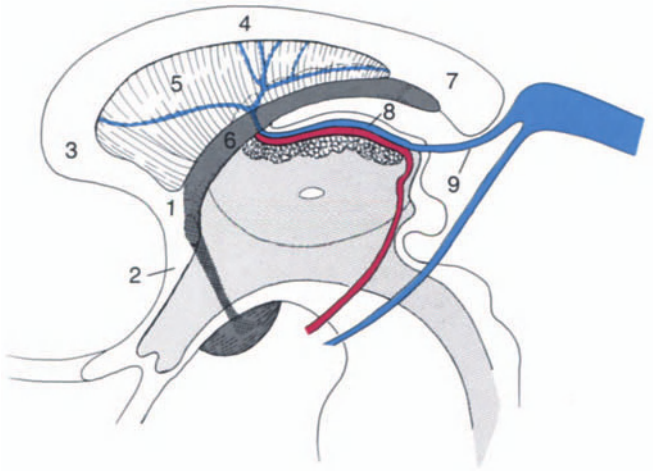


Figure 10.89. The rostrum (*1*) of the corpus callosum is in continuity inferiorly with the lamina terminalis (*2*) and superiorly with the genu (*3*). The genu and body (*4*) give purchase to the septum pellucidum (*5*), which also attaches inferiorly and posteriorly to the body of the fornix (*6*). The splenium (*7*) overhangs the internal cerebral (*8*) and Galenic (*9*) veins.

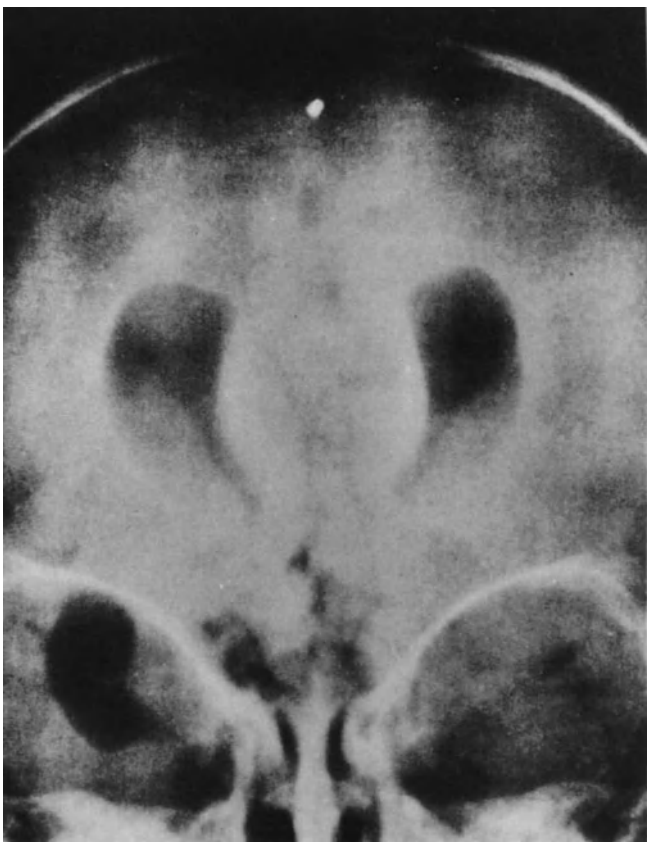


Figure 10.90. Lipoma of the corpus callosum revealed in a pneumogram.

ambient and quadrigeminal cisterns. Consequently, the distinction between 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 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.

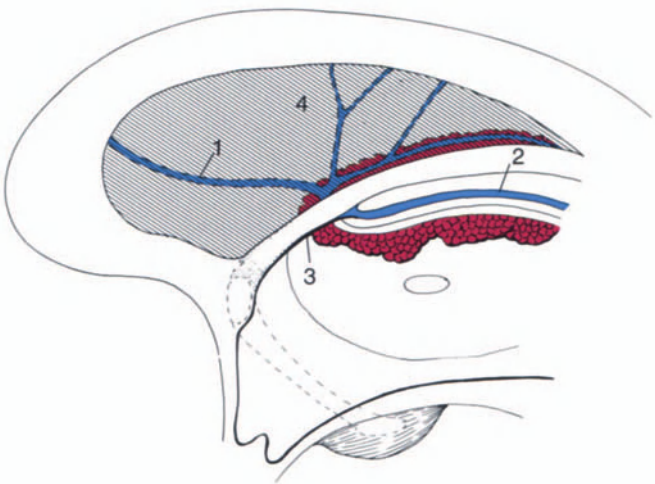


Figure 10.91. The anterior septal vein (*1*), a tributary to the internal cerebral vein (*2*), generally at the foramen of Monro (*3*), is located within the septum pellucidum (*4*).

Spinal Tumors

Such vertebral column tumors as the osteoma, hemiangioma, 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, suffice it to state that the aneurysmal 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 hematagenous, renal, or neuroblast in nature. Almost never seen by a pediatric neurosurgeon, they are treated with Roentgen therapy and chemotherapy.

Metastatic Spinal Tumors

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. The author recommends radiation therapy rather than decompressive laminectomy.

Intradural-Extramedullary Tumors

Only the arachnoidal cyst and neurofibroma occur in this location in childhood.

Arachnoidal Cyst

The arachnoidal cyst is seldom diagnosed preoperatively. Surgery consists of resecting as much of the cyst wall as possible.

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 foramen, neither the spinal nor paravertebral routes performed independently permit this. Consequently, these hour glass neurofibromas should be removed in a single sitting,

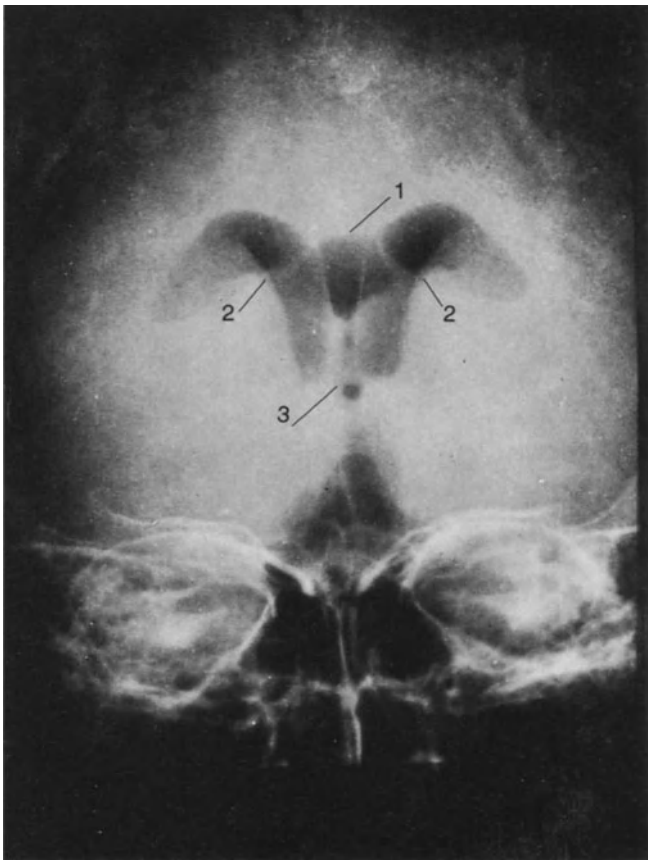


Figure 10.92. Gas has filled the ventricular system directly, not the subarachnoid spaces. There is a cavum septi pellucidi (*1*) between the lateral (*2*) and III ventricles (*3*).

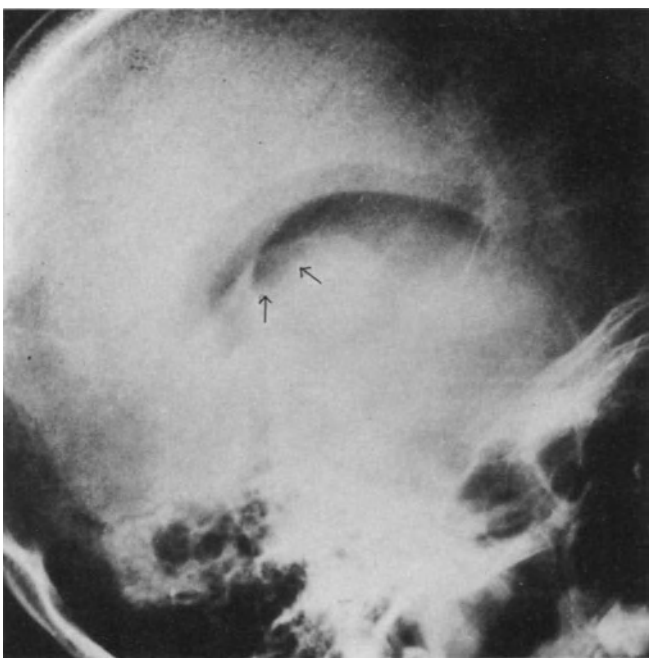


Figure 10.93. Gas has passed from the cavum septi pellucidi into the cavum vergae (arrows).

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 spinal surgical procedures, removal of an hour glass neurofibroma is best 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 neurosurgeon and general surgeon 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 intervertebral 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 intraabdominal tumor. Repositioning the laminotomy flap may be performed before the general surgeons proceed, or after they have completed their work.

Intramedullary Tumors

Cystic Intramedullary Tumors

Since it is impssible to distinguish an intramedullary astrocytoma from syringomyelia, the treatment is the same.

Solid astrocytoma

These tumors, whether benign or malignant, invade diffusely the entire spinal cord. There is often a plane of demarcation between pure tumor and normal spinal cord. Resection, consequently, is possible, through a myelotomy.

Ependymoma

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.

Cauda Equina Ependymoma

When ependymoma involves the cauda equina, it layers all nerve roots as icing on a cake, rendering it impossible to remove tumor and preserve neural elements. Surgery should be limited to decompression.

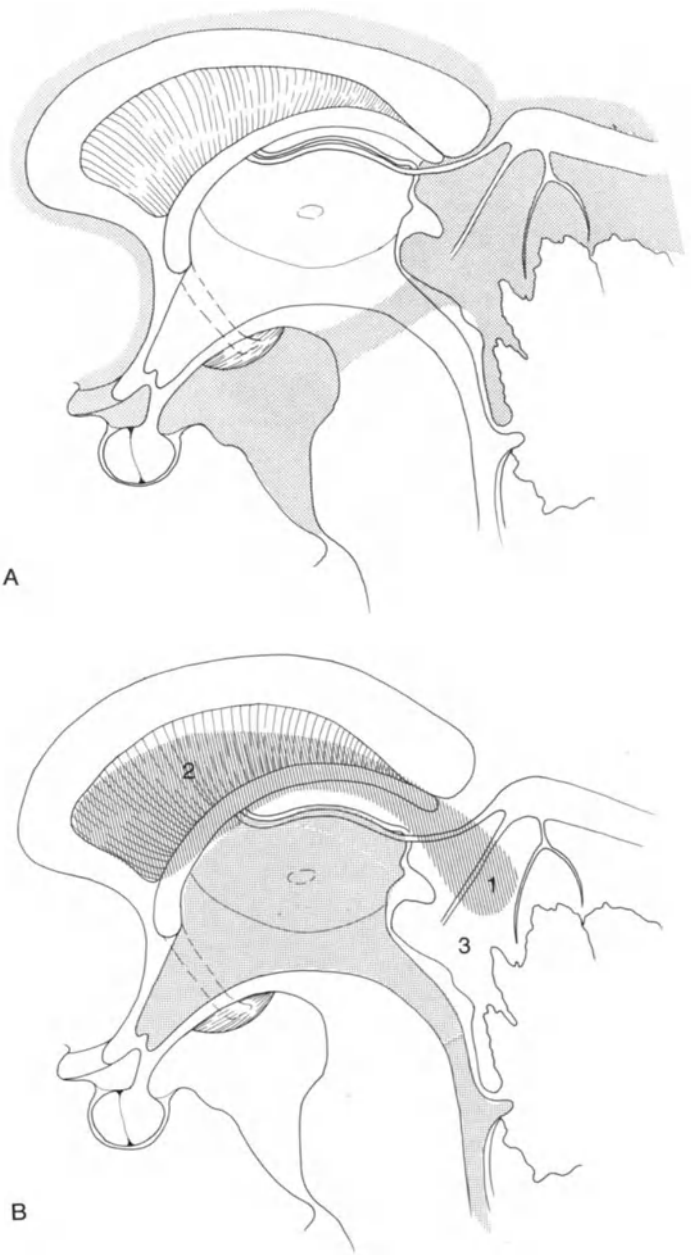
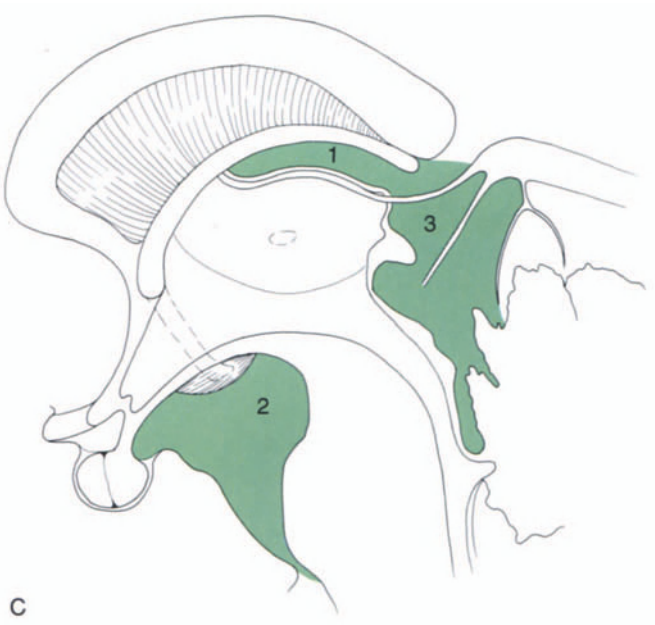
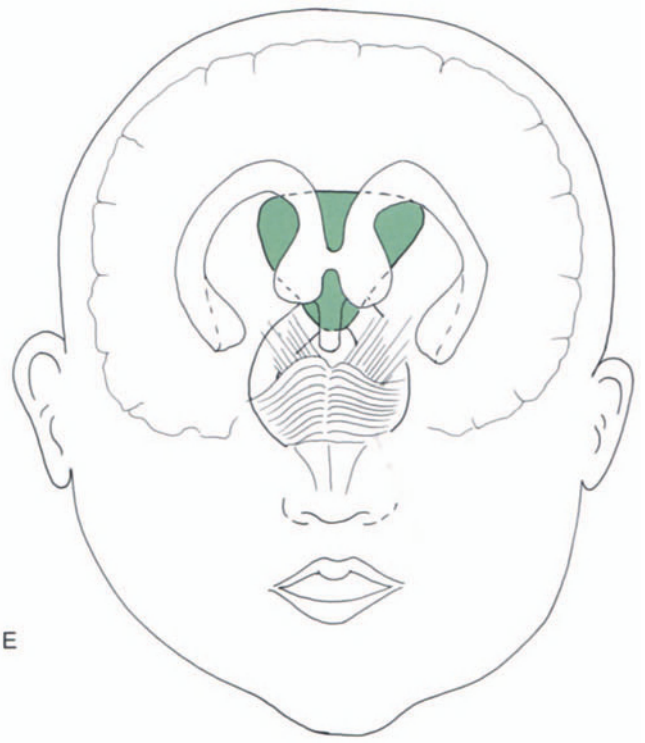


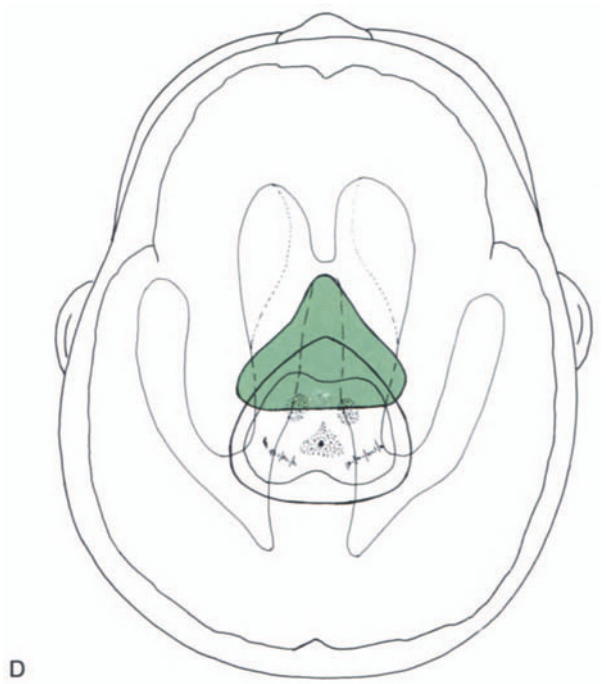
Figure 10.94. The normal cisterns and ventricles, as well as the anatomy and flow of cerebrospinal fluid into a cavum veli interpositi and cavum vergae are illustrated. (A) The normal cisterns. (B) The normal midline ventricles. Splenium (*1*) and genu (*2*) of corpus callosum, as well as quadrigeminal cistern (*3*) are missing. (C) The cavum veli interpositi (*1*) is filling from the basal cistern (*2*) through the quadrigeminal cistern (*3*). (D) In the axial projection, the cavum veli inter- ►

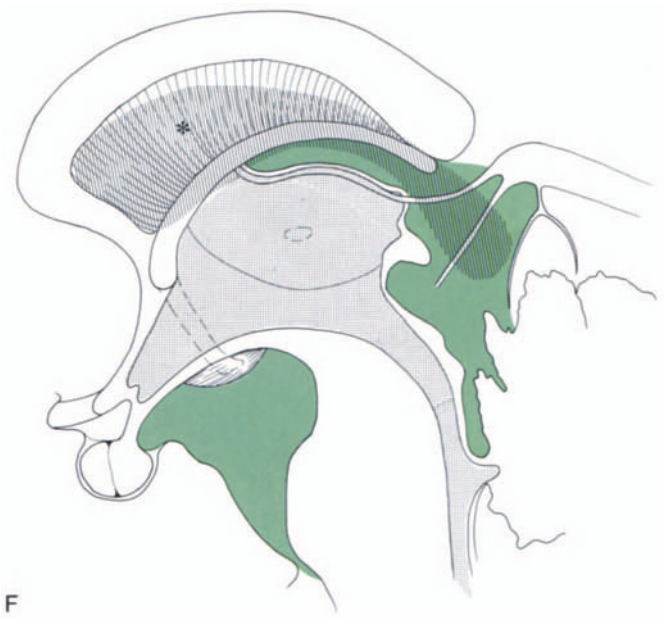
Arteriovenous Malformations of the Spinal Cord

These are so rare in childhood that no attention will be given the technique for their removal in this text.

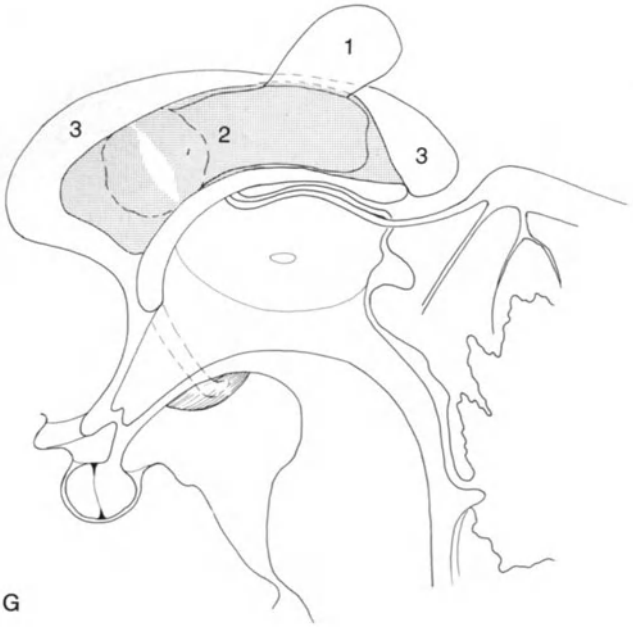




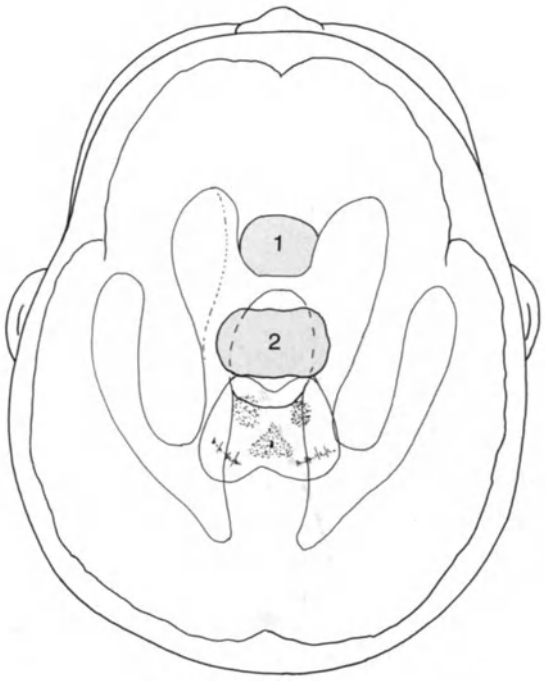




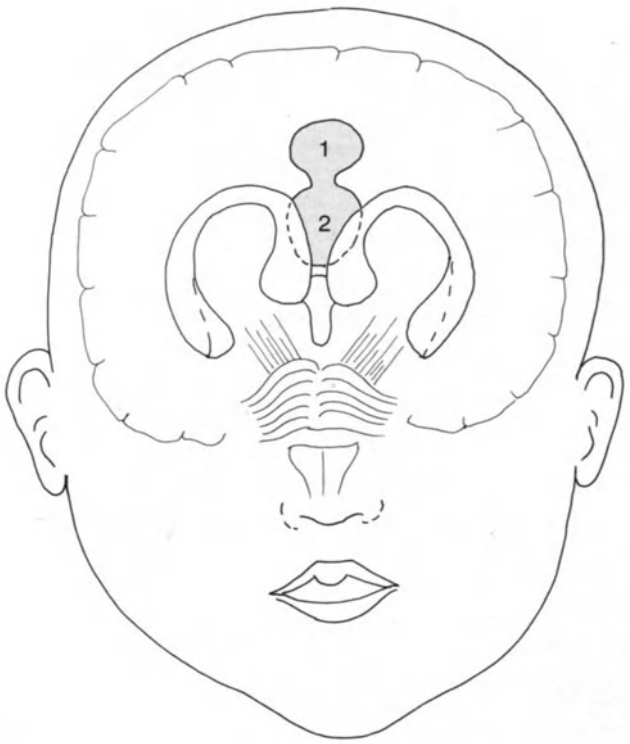
positi splays out beneath the lateral ventricles, extruding laterally at each choroidal fissure. Since this drawing was taken from an air study, the gas is projected over the lateral ventricles. (E) In the half-axial projection, we see that the cavum veli interpositi insinuated beneath the lateral and superior to the III ventricles. (F) The cavum vergae fills from the ventricular system, through a cavum septi pellucidi (*). It may hang into the quadrigeminal cistern, as here illustrated, or (G) ex- ►



G



H



I

tend (*1*) superior to both the cavum septi pellucidi (*2*) and the corpus callosum (*3*). (H) The axial projection illustrates how the cavum septi pellucidi (*1*) and cavum vergae (*2*) displace the lateral ventricles. (I) The half-axial projection illustrates superior (*1*) and inferior (*2*) extension of the cavum vergae.

References

1. Raimondi AJ, Yashon D, Matsumoto S, Reyes C: Increased intracranial pressure without lateralizing signs: The midline syndrome. Neurochirurgia 10:197–208, 1967.
2. Krabbé KH: Histologische und embryologische Untersuchungen über die Zirbeldrüse des Menschen. Ana Hft 54:187–319, 1916.
3. Cushing H: The intracranial tumors of preadolescence. Am J Dis Child 33:551–84, 1927.
4. Bailey P: Intracranial tumors. Springfield, IL, CC Thomas, 1948.
5. Rubinstein LJ: Tumors of the central nervous system. Atlas of Tumor Pathology 2nd series, phase 6, pp 269–84, Washington, DC, Armed Forces Institute of Pathology, 1972.
6. Dandy WE: An operation for the removal of pineal tumors. Surg Gynecol Obstet 33:113–19, 1921.
7. Poppen JL: The right occipital approach to a pinealoma. J Neurosurg 25:706–11, 1966.
8. Jamieson KG: Excision of pineal tumors. J Neurosurg 35:550–3, 1971.
9. Krause F: Operative Freilegung der Vierhügel nebst Beobachtungen über Hirndruck und Decompression. Zentralbl Chir 53:2812–20, 1926.
10. Lewis DD: A contribution to the subject of tumors of the hypophysis. JAMA 55:1002–8, 1910.
11. Streja D, Teichner F, Marliss E: Fifty-year survival after surgery for craniopharyngioma. JAMA 234(5):510–11, 1975.
12. Rougerie J, Raimondi AJ: Craniopharyngiomas. In Brain Tumors in the Young. Luis V. Amador (Ed.) Springfield IL, CC Thomas, 1983, pp 599–618
13. Dandy W: The Brain. New York, Harper & Row, 1969, pp 554–88.

Uncited References

Kuwamura K, Matsumoto S, Raimondi AJ, Gutierrez FA: Tumors of the central nervous system in children under 2 years of age. Neurol Med Chir (Tokyo) 18:109–15, 1978.

Oi S, Raimondi AJ: Ependymoma. In Pediatric Neurosurgery, New York, Grune & Stratton, 1982, p 419–28.

Oi S, Raimondi AJ: Hydrocephalus associated with intraspinal neoplasms in childhood. Am J Dis Child 135:1122–4, 1981.

Raimondi AJ, Gutierrez FA: Diagnosis and surgical treatment of choroid plexus papillomas. Child's Brain 1:81–115, 1975.

Raimondi AJ, Rougerie J: A critical review of personal experiences with craniopharyngioma: clinical history, surgical techniques, and operative results. In Concepts in Pediatric Neurosurgery 3, Basel, S. Karger, 1983, pp 1–34.

Raimondi AJ, Shimoji T, Gutierrez FA: Suprasellar cysts: Surgical treatment and results. Child's Brain 7:57–72, 1980.

Raimondi AJ, Tomita T: Brain Tumors in children. Advantages and disadvantages of individual treatment. In Raimondi AJ, Hoffman H, Epstein F (eds): Concepts in Pediatric Neurosurgery I, Basel, S. Karger, 1981.

Raimondi AJ, Tomita T: Hydrocephalus and infratentorial tumors incidence, clinical picture, and treatment. J Neurosurg 55:174–82, 1981.

Raimondi AJ, Tomita T: Medulloblastoma in childhood. Acta Neurochir (Wien) 50:127–38, 1979.

Raimondi AJ, Tomita T: Medulloblastoma in childhood; comparative results of partial and total resection. Child's Brain 7:310–28, 1979.

Raimondi AJ, Tomita T: Pineal tumors in childhood: Epidemiology, pathophysiology and surgical approaches. Child's Brain 9:239–66, 1982.

Raimondi AJ, Tomita T: Tumors during the first year of life. Child's Brain 10:193–207, 1983.

Shkolnik A, Tomita T, Raimondi AJ, Hahn Y, McLone DG: Work in progress: Intraoperative neurosurgical ultrasound localization of brain tumors in infants and children. Radiology 148:525–7, 1983.

Tomita T, Raimondi AJ: IV ventricle tumors. In Pediatric Neurosurgery. New York, Grune & Stratton, 1982, pp 383–93.

Velasco JM, Raimondi AJ: Choroid plexus papilloma. In Pediatric Neurosurgery. New York, Grune & Stratton, 1982, pp 451–60.

"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

Chapter 11

Vascular Disorders: Surgical Approaches and Operative Technique

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.

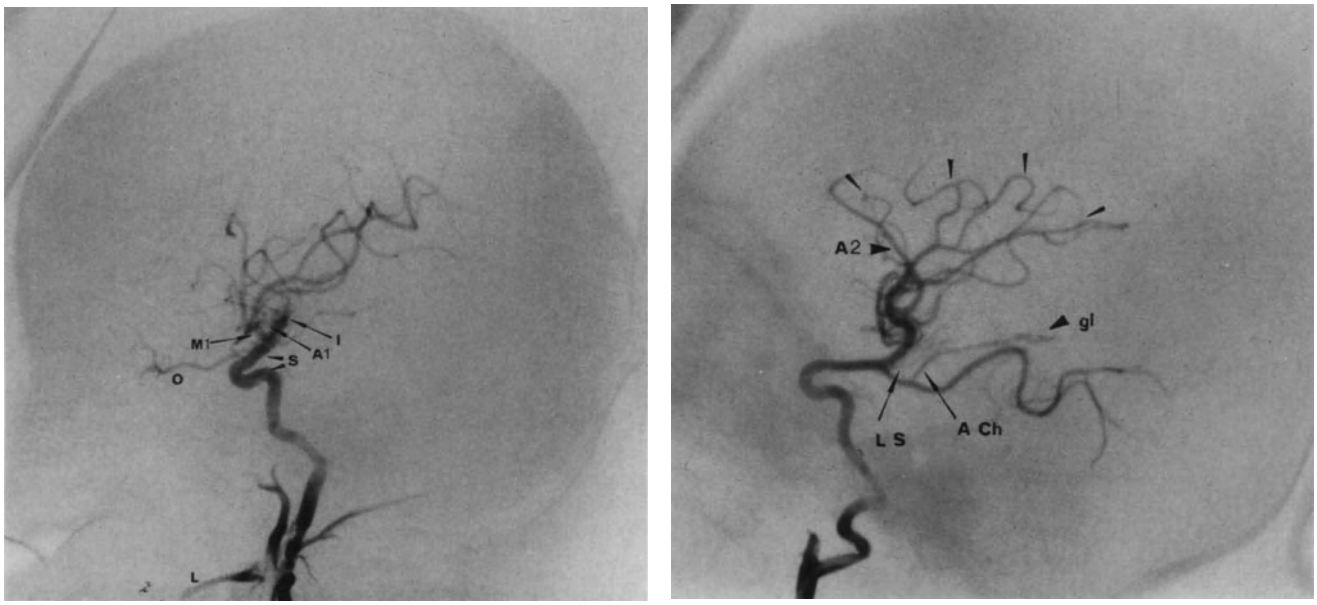
Specific vascular anatomical characteristics of newborn and infants are as follows: The anterior communicating artery does not exist as a radiologically identifiable anatomical structure, the internal carotid artery is large and continues directly into the middle cerebral artery, and PICA is located 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. 11.1 A). Similarly, the branches of the middle cerebral artery are linear extensions of this vessel, at the limen of the insula (Fig. 11.1 B). Hence, one may understand why aneurysms are more common at the bifurcation of the internal carotid artery and trifurcation of the middle cerebral artery.

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 atteries 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 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.



A B

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-1 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 (*O*) takes 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).

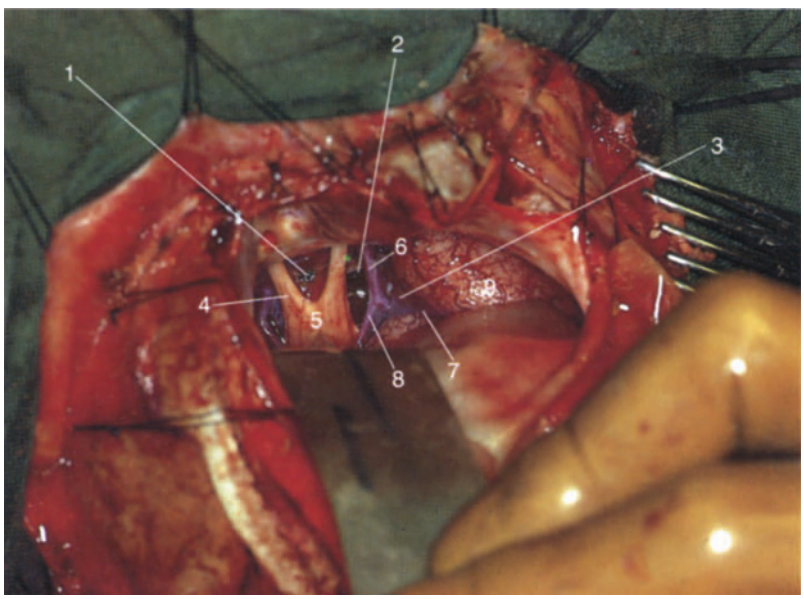
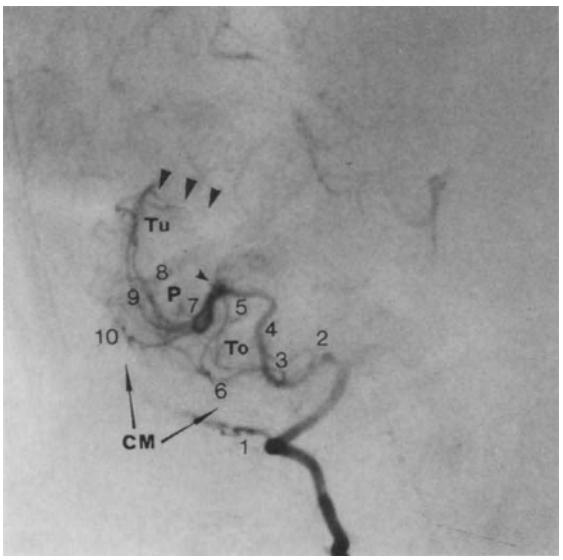
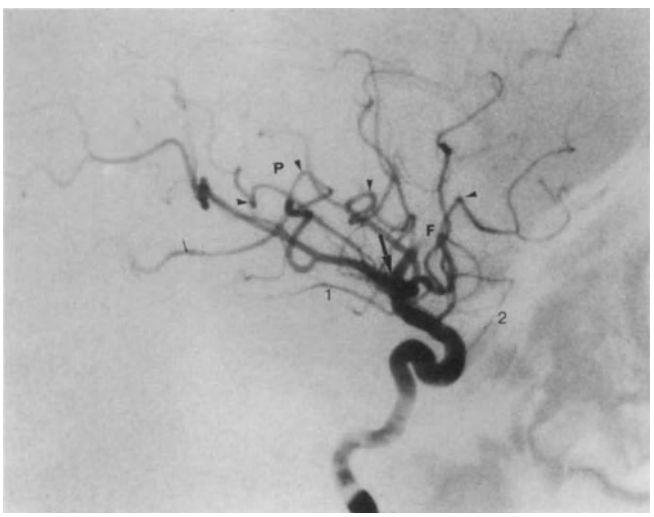


Figure 11.2. This is a photograph of a right medial frontal craniotomy in an infant, illustrating the enormity of basal cisterns and relative anatomy of optic pathways and carotid arteries. The prechiasmatic (*1*), interopticocarotid (*2*), and Sylvian (*3*) cisterns have been opened, permitting complete 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.



A



B

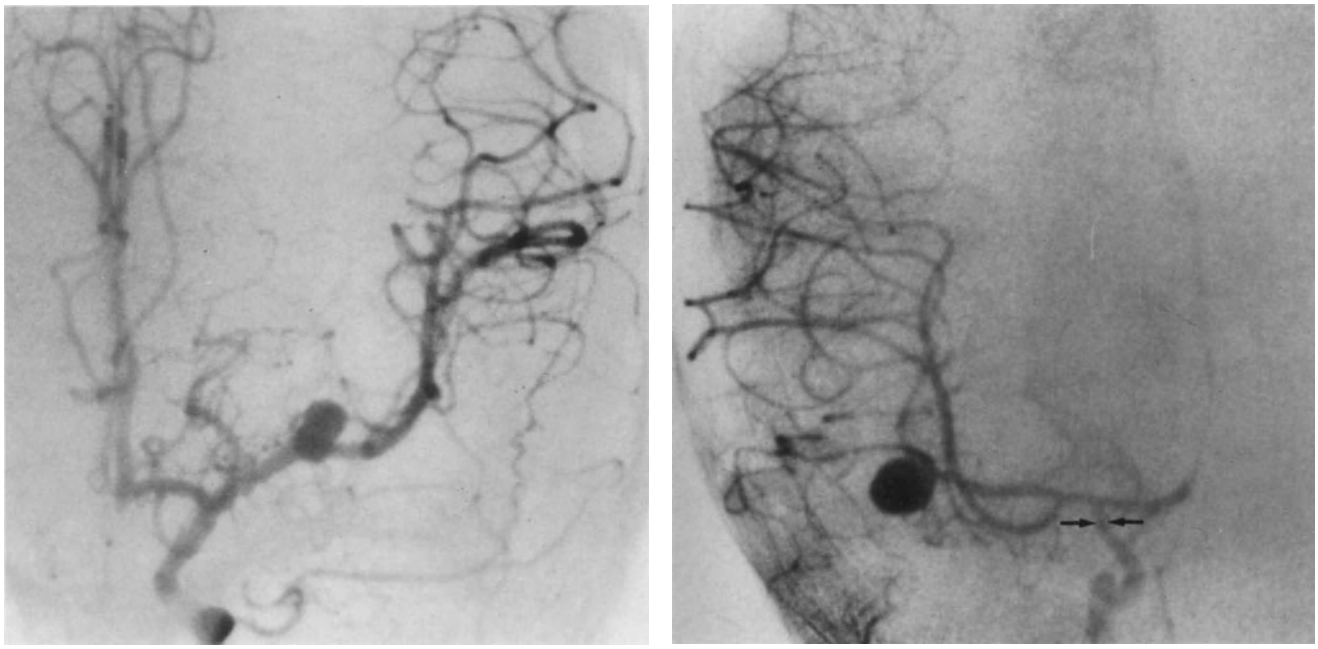
Figure 11.3. (A) Muscular branches (*1*) are quite prominent and establish anastomotic routes between the vertebral and occipital arteries. The posterior inferior cerebellar artery (PICA) most often originates from the vertebral and courses around the medulla oblongata, where it may be divided into ventral (*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 hemispheral (*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.

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 (Figures 11.4 and 11.5)

These aneurysms are approached in identically 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 ar-



A B

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).

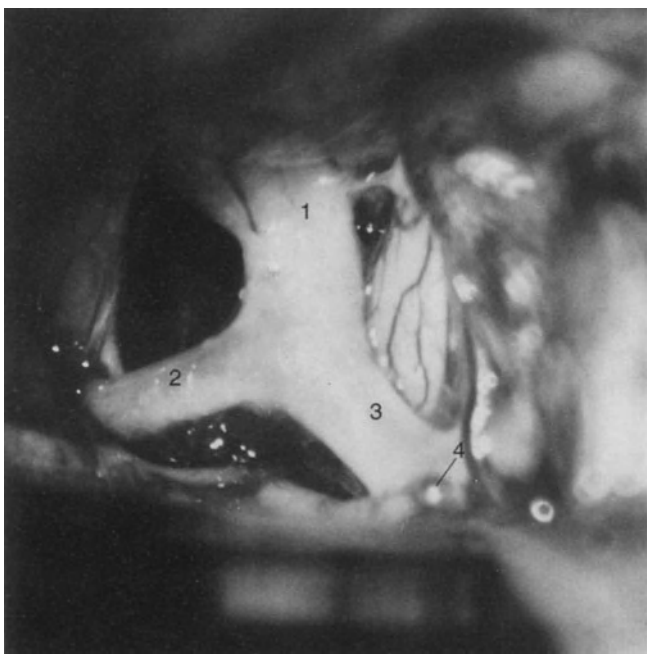
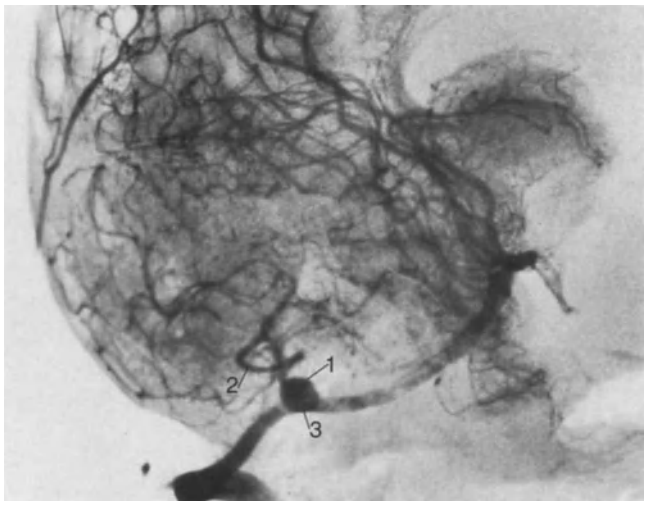


Figure 11.5. The internal carotid artery (*1*), anterior cerebral artery (*2*), middle cerebral artery (*3*), and aneurysm (*4*) at the trifurcation of the middle cerebral artery within the Sylvian fissure.

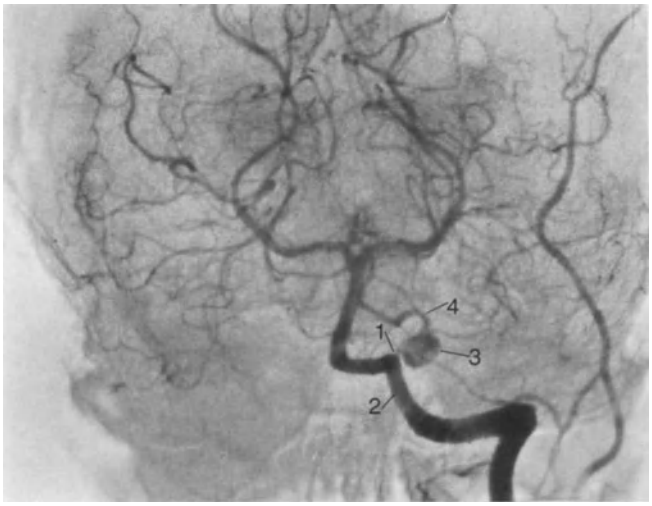
tery. The Sylvian fissure is opened from medial to lateral, quite an easy undertaking because of its enormous size and the thinness of the arachnoid. This separates the temporal and frontal lobes from one another, exposing the limen of the insula and the triangular, olfactory, and temporal operculae. 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.

Posterior Inferior Cerebellar Artery (PICA) Aneurysms (Figures 11.6 to 11.8)

With the child in the sitting position an inferior cerebellar triangle craniotomy is performed, and the dura is opened, as illustrated in Figure 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 C1 should be removed as far laterally as the posterior strut of the foramen transversarium. Because of the smallness of size and incomplete ossification of C1, it is best to use a very delicate, sharp–tipped, rongeur to take away the bony spicules, taking care to remain within the perios-



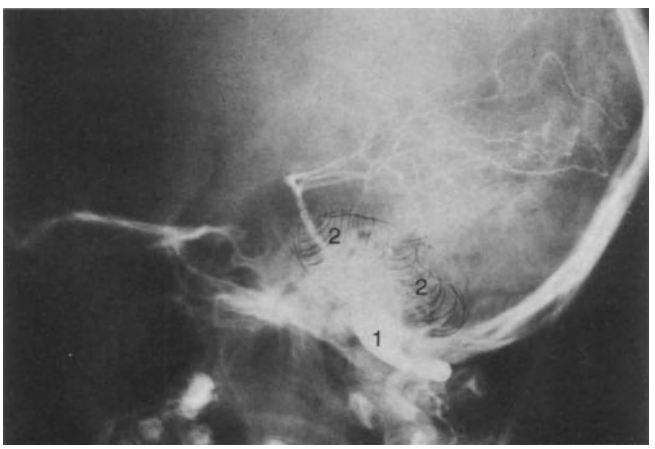
A



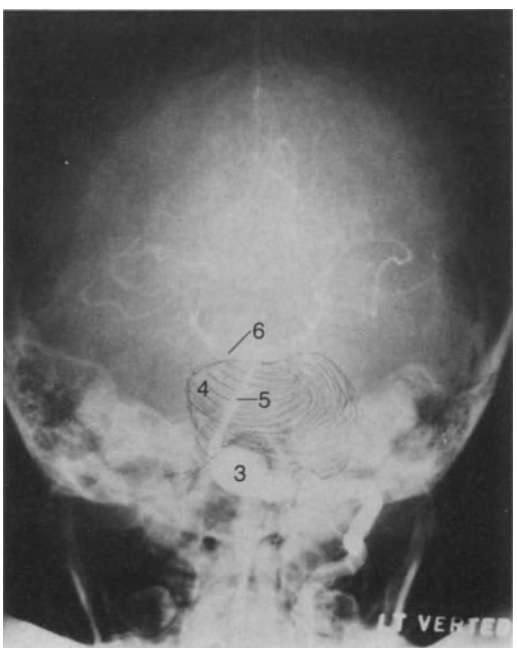
B

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*).

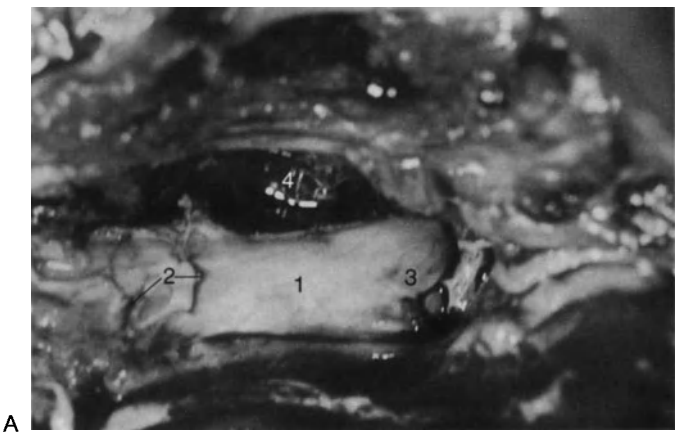
Figure 11.7. Posterior inferior cerebellar artery (PICA) aneurysms are well known to develop into giant aneuryms in infancy. Why this is so remains doubtful. Because of the giant size, and the presence of clot laminated in various stages of aging, one may invariably consider PICA aneuryms to be posttraumatic 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.



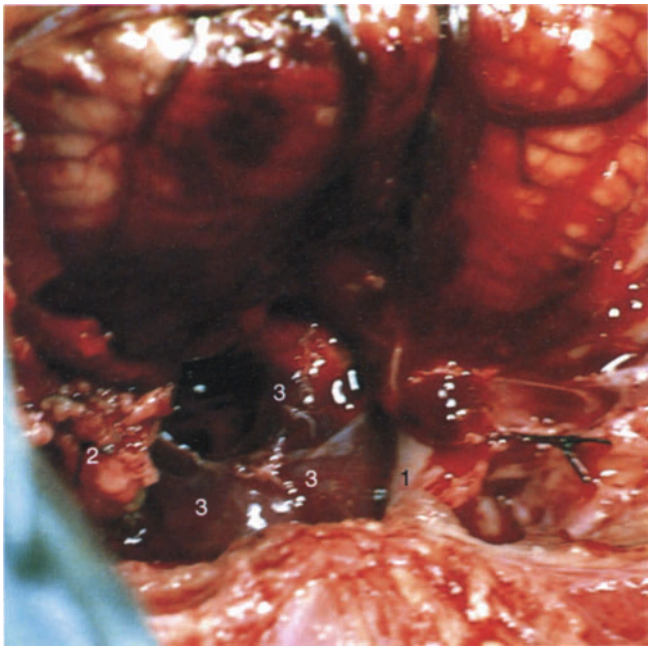
A



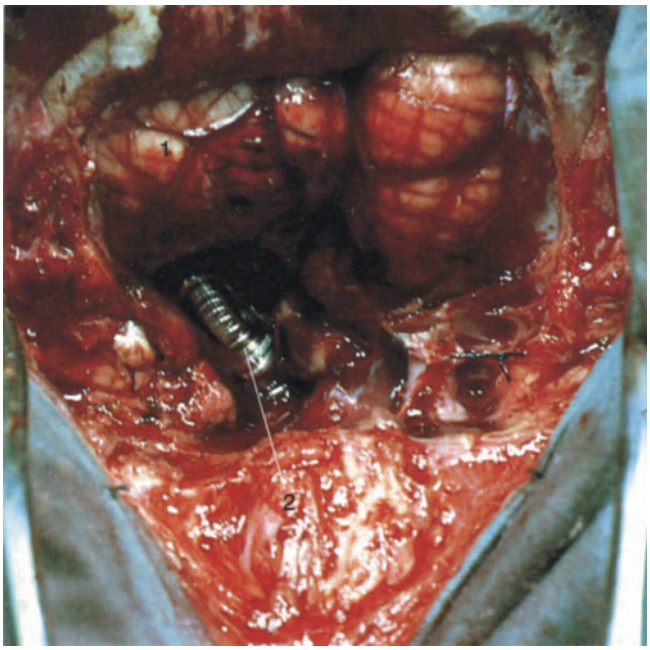
B



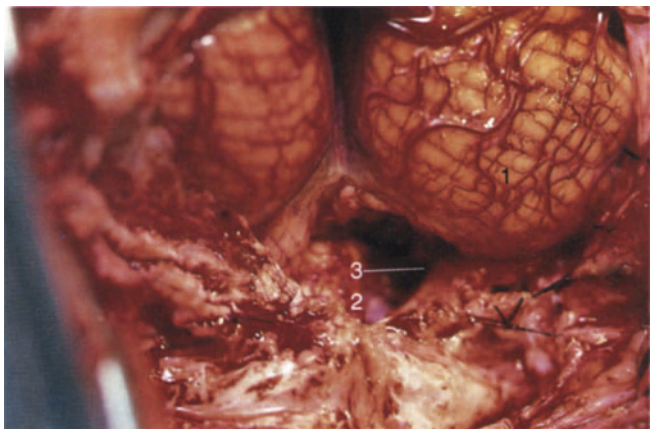
A



B

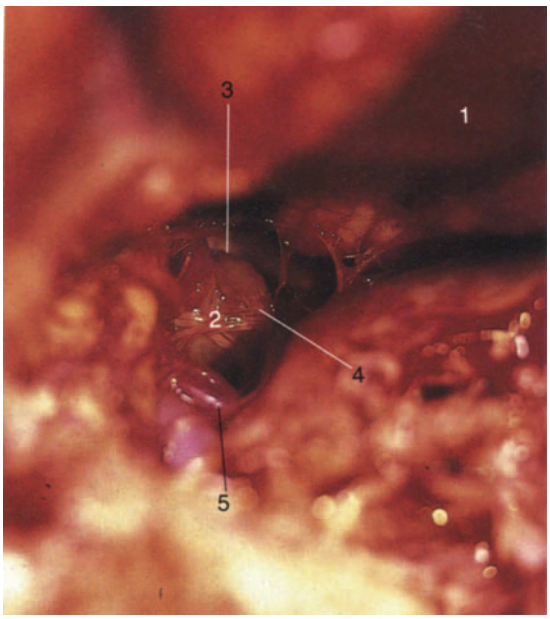


C

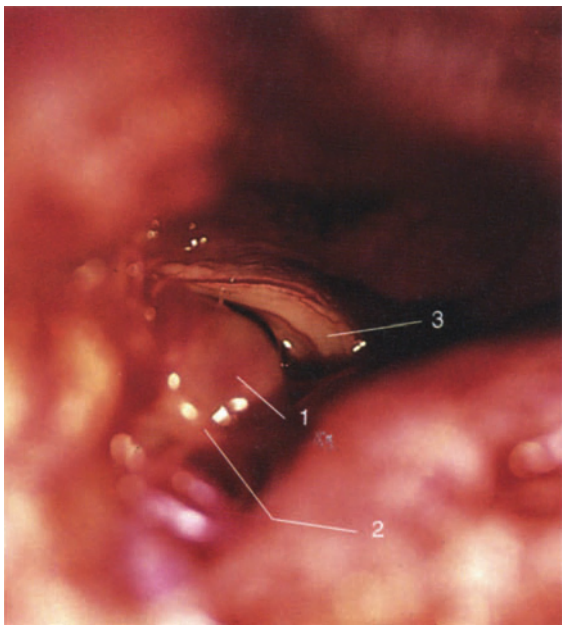


D

Figure 11.8. These photographs were taken during the repair of the aneurysm illustrated in Figure 11.7. (A) The arch of C1 has been exposed (*1*), allowing one to identify fracture line (*2*) at the region of the foramen transversarium and (*3*) just lateral to the spine C1. The aneurysm (*4*) bulges just above the arch of C1. (B) The arch of C1 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 C1, 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*). ►



E



F

teum. 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 whether the exposure is adequate. The enormous size of the cisterna mangna 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 $3 \times 2 \times 3$ 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.

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. *At no time should the technique of dissection be such as to push the aneurysm in the direction of the medulla oblongata.* Also, care must be exercised in separating the wall of the aneurysm from 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

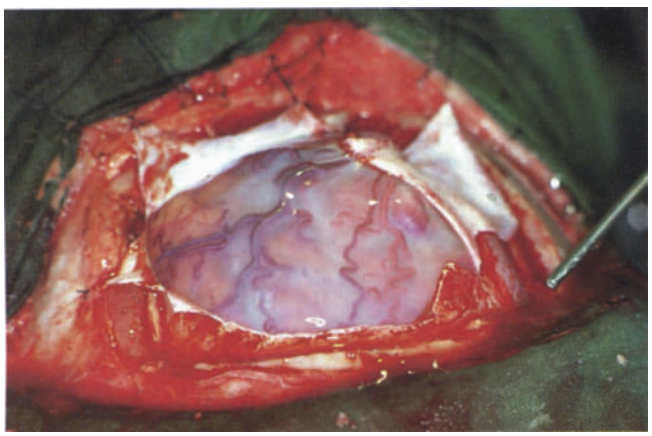


Figure 11.9. Opalescence of the arachnoid, which one almost invariably observes in children with arteriovenous malformations. Whether this is secondary to the shunting of arterial blood into the venous system, the increase in intracranial pressure, repeated microscopic subarachnoid bleeds, is purely speculative.

Figure 11.10. Here, the cortical surface of the hemisphere of a child with a deep (centrencephalic) arteriovenous malformation is illustrated. One notes the absence of opalescence of the arachnoid membrane, shunting of arterial blood into the venous system (arrows).

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.

Vascular Malformations

(Figures 11.9 to 11.14)

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 arteriovenous, *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 anatomic basis for air emboli, at most a very theoretic chain of events. They are, however, unsightly, and often frighten the parents.

Dural arteriovenous fistuale 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.

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.

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. The author has never seen an associ-

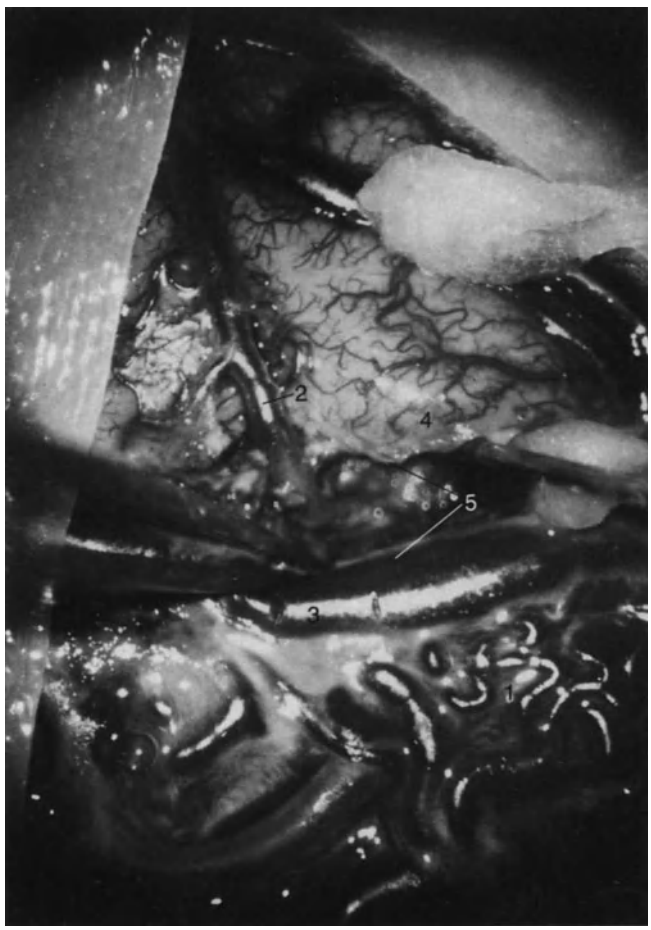


Figure 11.11. The ideal technique for resecting arteriovenous malformation consists of separating the malformation—racemose (*1*); efferent (*2*); afferent (*3*)—components from the surrounding brain (*4*) by performing a cerebrotomy along the border between normal brain and vascular anomaly (*5*).

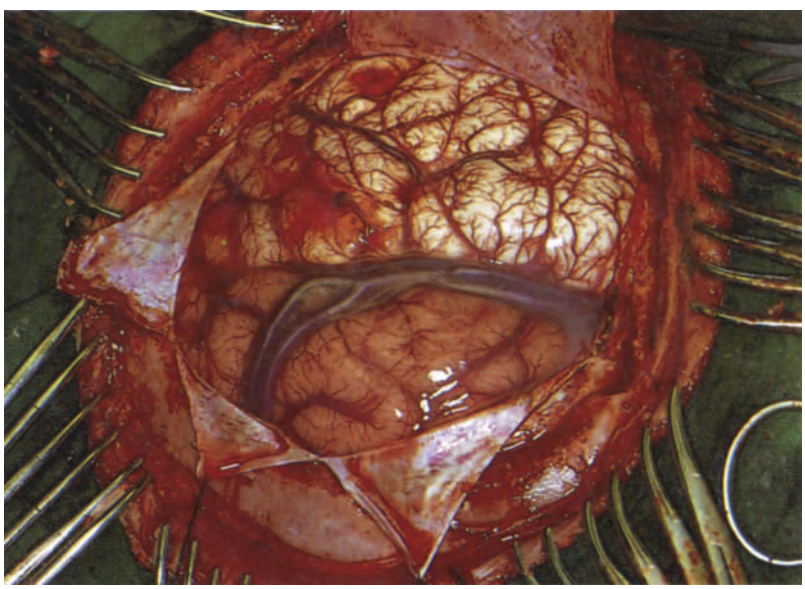
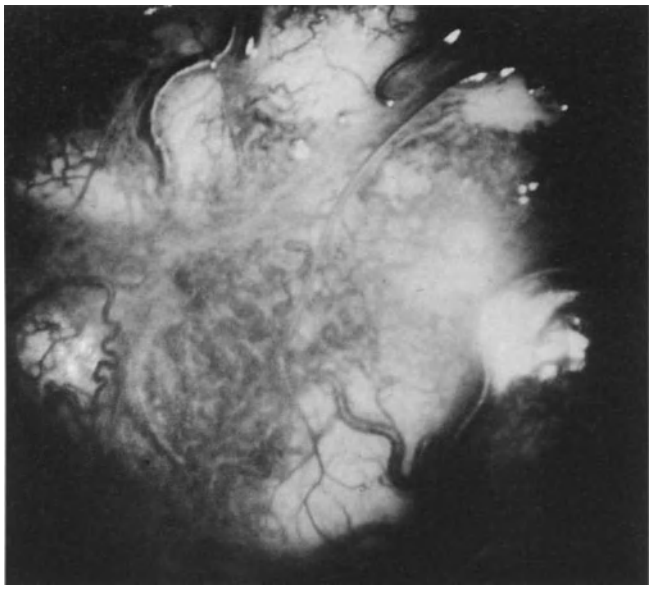
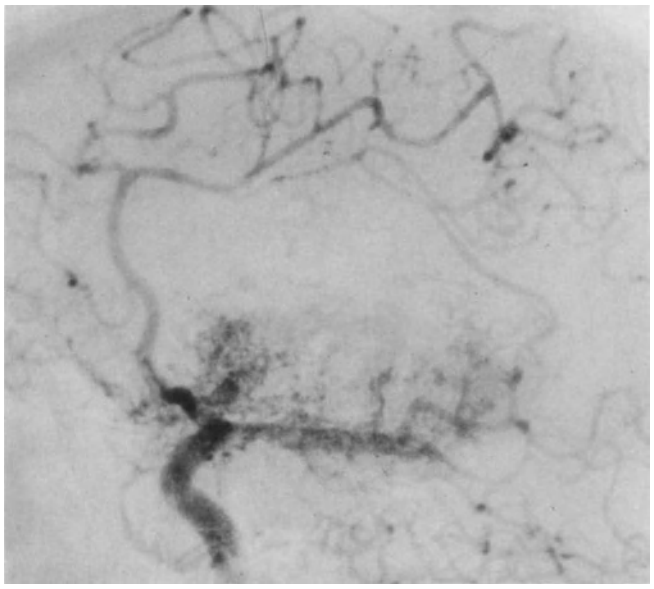


Figure 11.12. This is the appearance of the cerebral cortex in a child with an arteriovenous malformation of the Galenic system. Note the granular appearance of the arachnoid, the "bag of worms" appearance of the cortical draining veins, and arterialization of some cortical vessels.

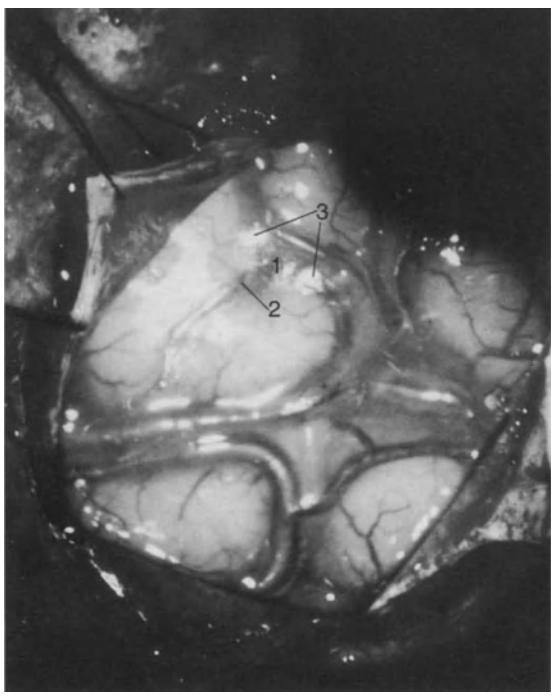


A

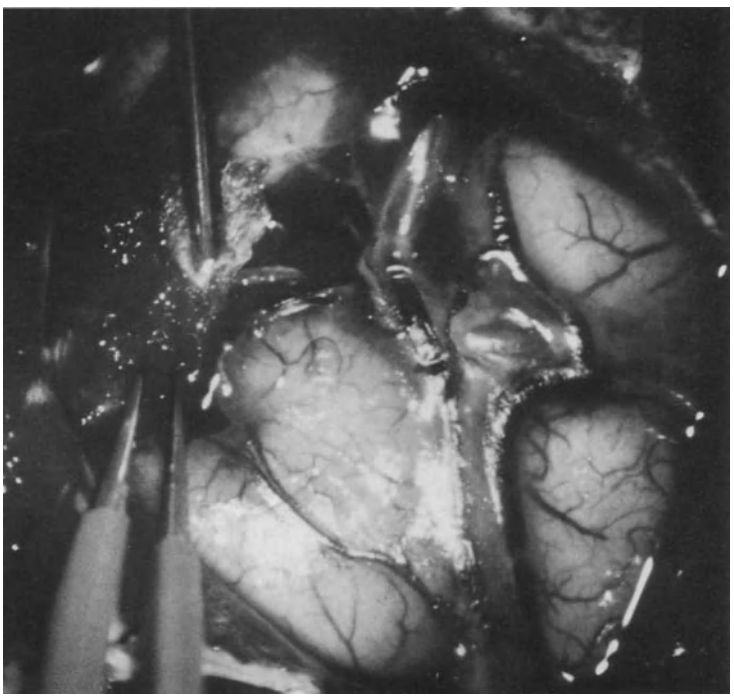


B

Figure 11.13. (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 uncommon for arteriovenous malformations of the Galenic system to have a predominantly racemose component, nourished by perforating branches of the anterior, middle, and posterior cerebral systems.



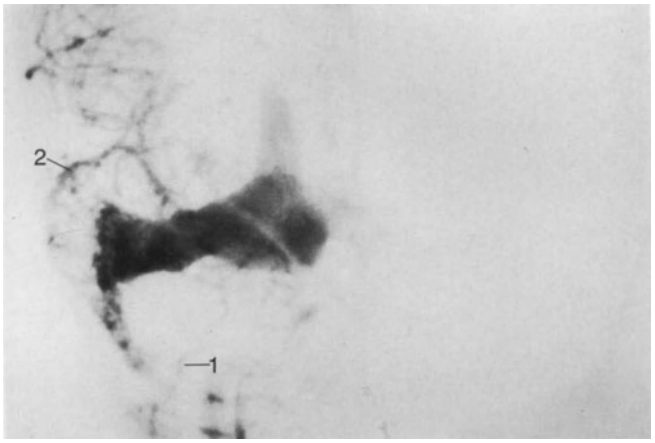
A



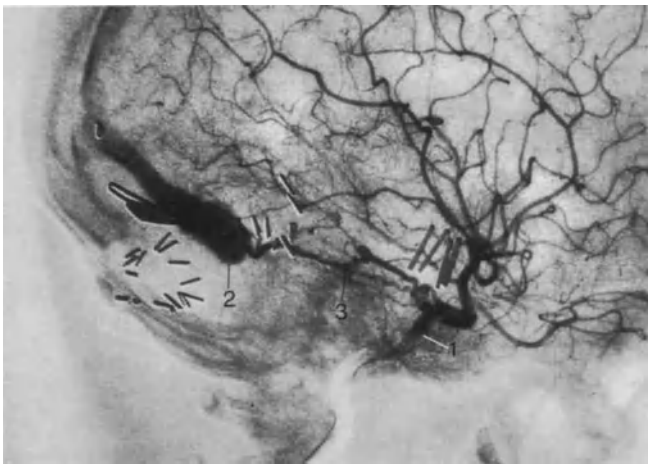
B

Figure 11.14. Until the present, it is not advisable to use laser for resection of arteriovenous malformations, whether one is speaking of carbon dioxide, argone, or neodymium YAG. This does not mean that some pathological vessels may not be successfully occluded with one of the three lasers. It does mean that the resection of the malformation cannot be completed with a laser. This is an illustration of such an attempt. (A) A small area of laser destruction of the arachnoid and cortex (*1*) in an attempt to coagulate a minuscule anomalous vessel (*2*). The neon laser tracer (*3*) is aimed at another anomalous vessel. (B) Extensive hemorrhage from both sites.

A



B



C

Figure 11.15. (A) Hypertrophied meningeal arteries (arrows) feeding into the transverse sinus. (B) Preoperative arteriogram illustrating middle meningeal (*1*) and occipital (*2*) feeders to the transverse 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 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.

ation between either transcranial or dural arteriovenous fistulae and leptomeningeal or choroidal malformations. The arteries afferent to the fistuale 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 (Figure 11.15)
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, the cavernous sinus is involved only after a sphenoid sinusitis or an injury.

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

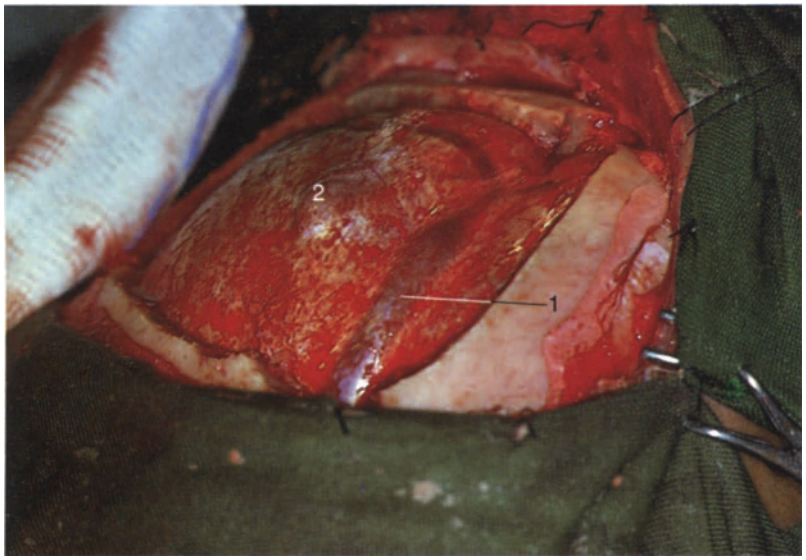


Figure 11.16. For hemispheral arteriovenous malformations feeding extensively into the superior sagittal sinus (SSS) (*1*), with aneurysmal dilation of some elements of the malformation (*2*), it is best to reflect a craniotomy that extends on both sides of the SSS.

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 burr 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.

Parenchymal Arteriovenous Malformations

Hemispherical Arteriovenous Malformations

(Figures 11.16 to 11.18)

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 he 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 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 malformation 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 an-

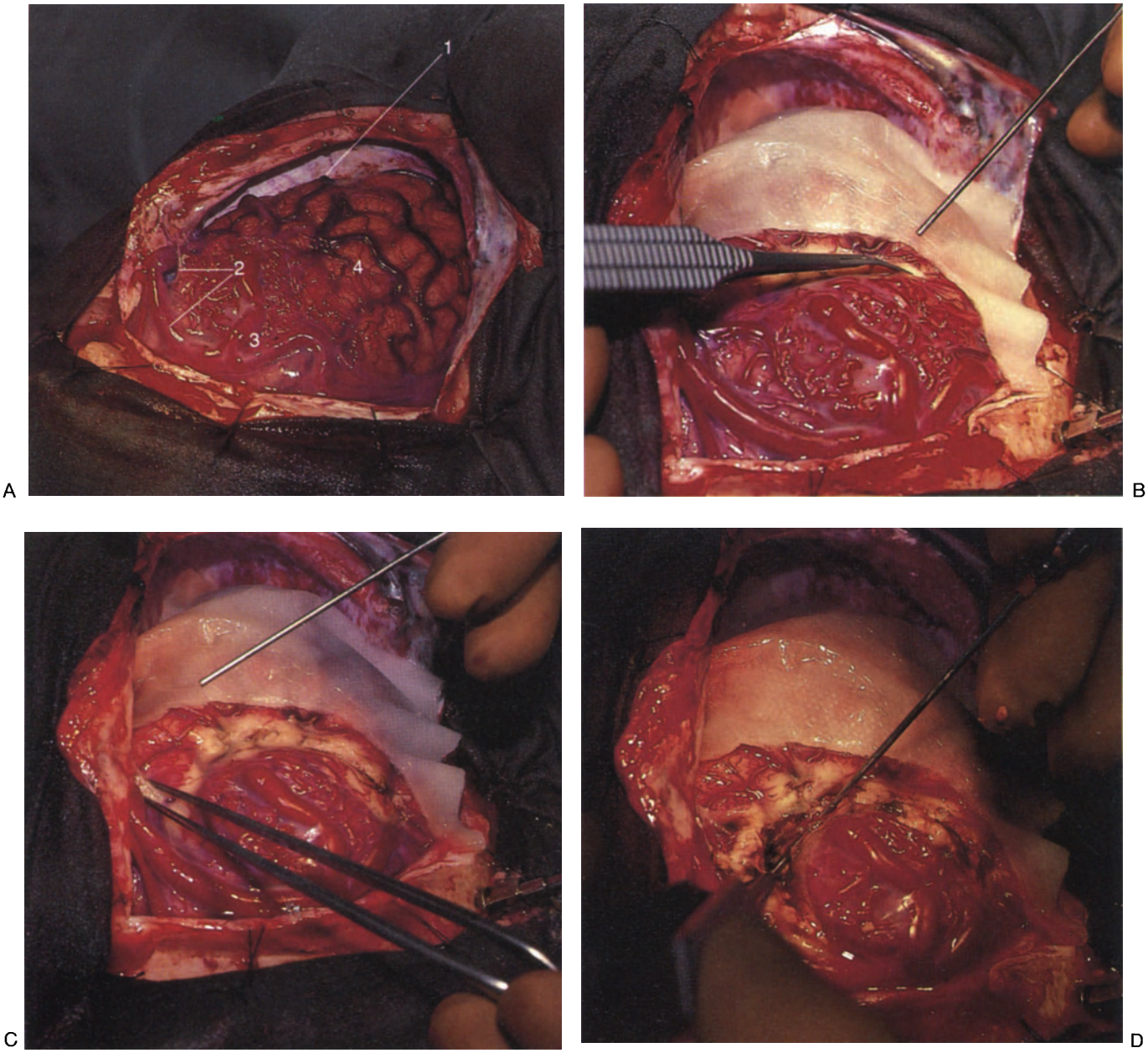


Figure 11.17. This large arteriovenous malformation of the right frontal lobe drained into the anterior two thirds of the SSS. (A) Three of these draining veins were coagulated and transected, allowing the frontal lobe to fall from the SSS, exposing the falx cerebri (*1*). The two largest draining veins (*2*) were left intact because of the sharp border between 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.

eurysmal 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.

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 arteriovenous 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 circumferential 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).

Brainstem Arteriovenous Malformations

(Figures 11.19 and 11.20)

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 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 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

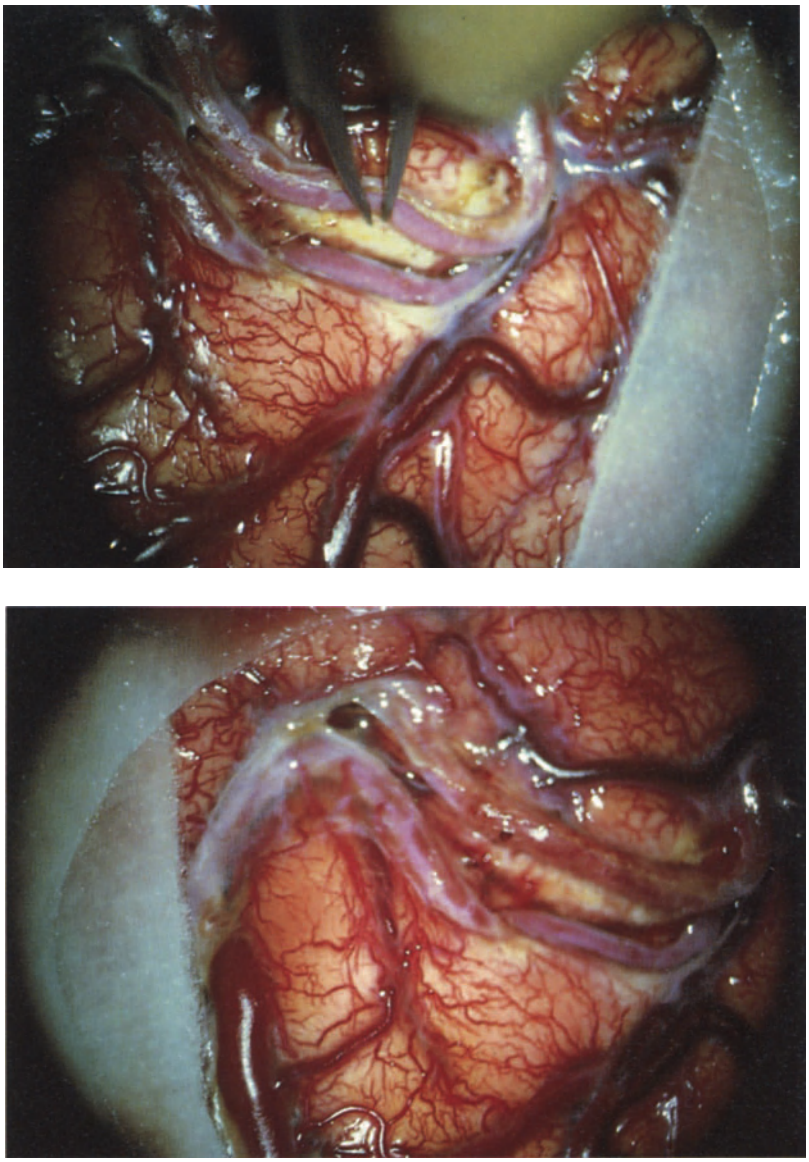
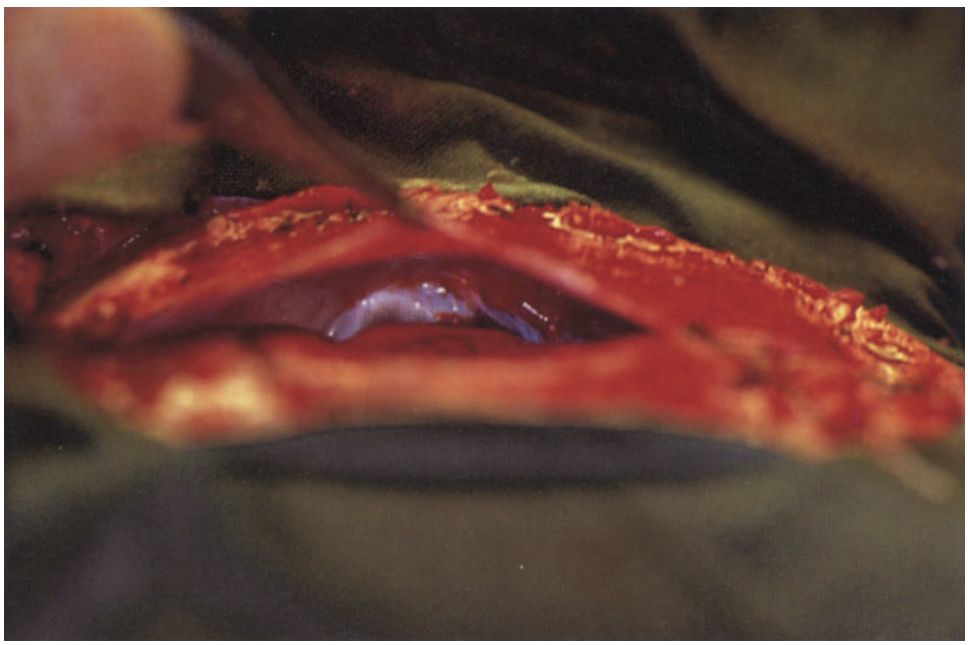


Figure 11.18. Smaller arteriovenous malformations, such as the microarteriovenous fistula (which would be called cryptic if it had bled, since surely it would not be identifiable), are resected by separating the anomalous vessels from the surrounding brain in stages, as shown in A and B.

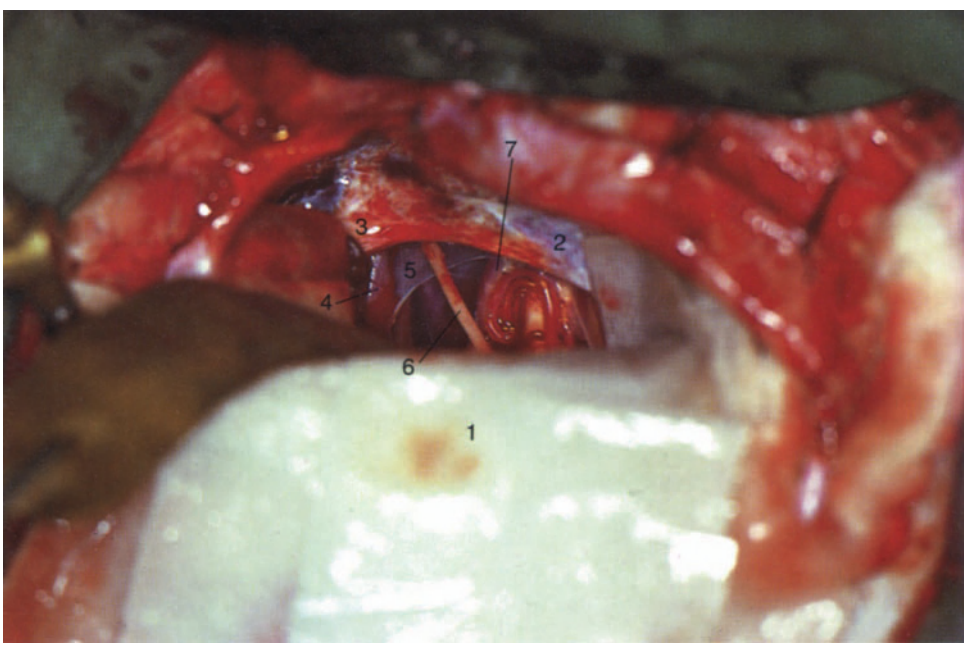
afferent vessels, and then coagulate the efferent vessels over a 3- or 4-mm distance before their entrance into the racemous or aneurysmal components. It is best to avoid clipping and transecting these vessels as long as possible, so as to minimize risk of dislodging the clip from a transected vessel during subsequent dissection. As a group of afferent vessels are dissected, coagulated, clipped, and transected, they should be covered with moistened telfa strips. 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 tend 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

Figure 11.19. 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 (arrows) along the surface of the pons. (E) Two of the anomalous vessels have been ligated and two remain to be ligated.

A

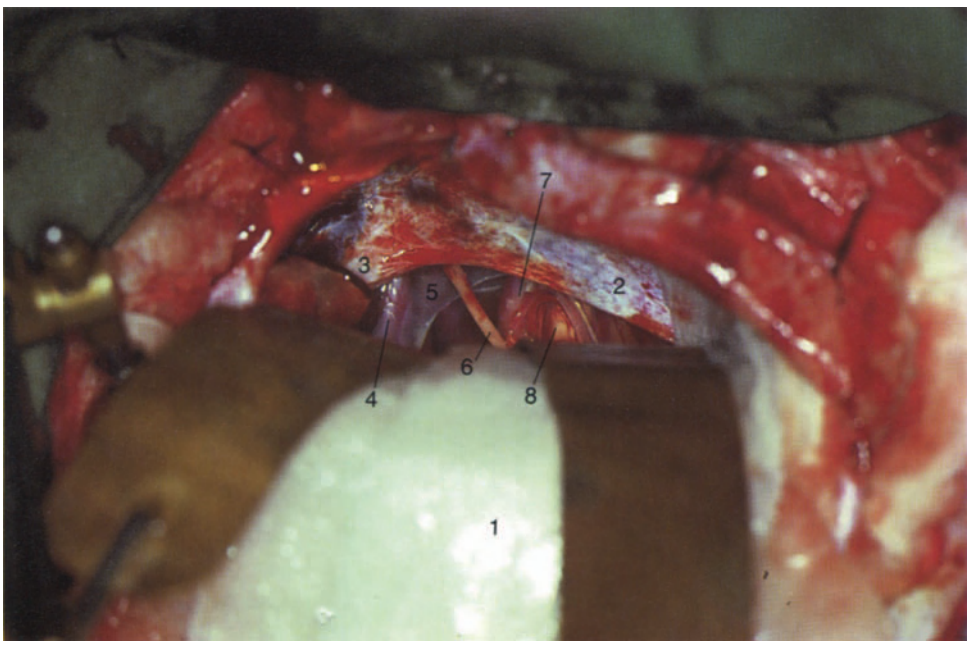


B

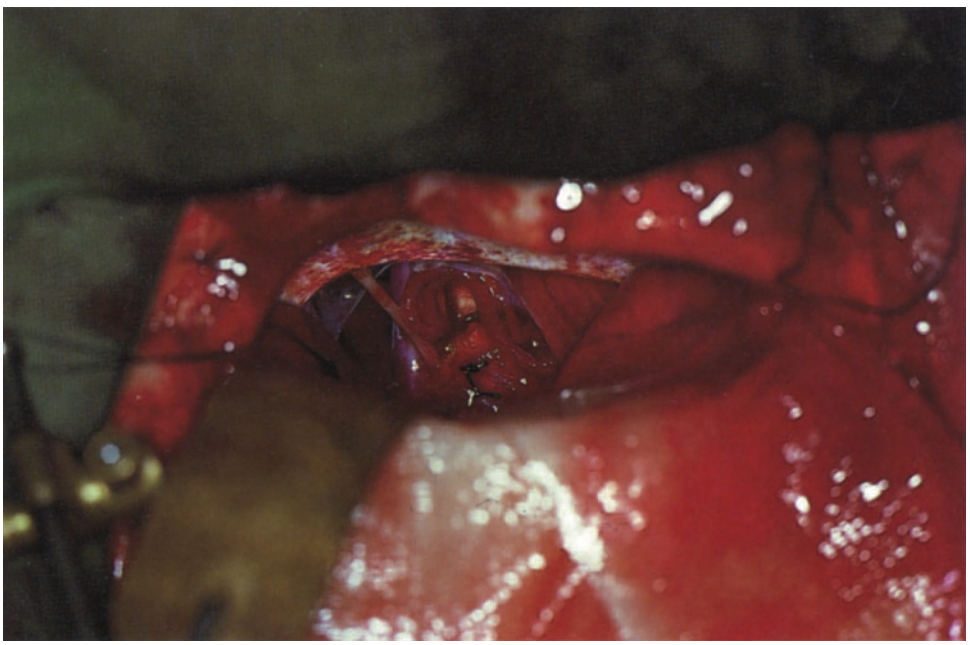


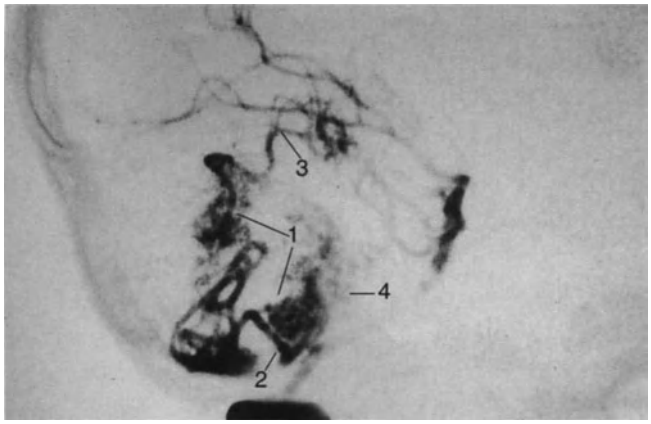
D

C

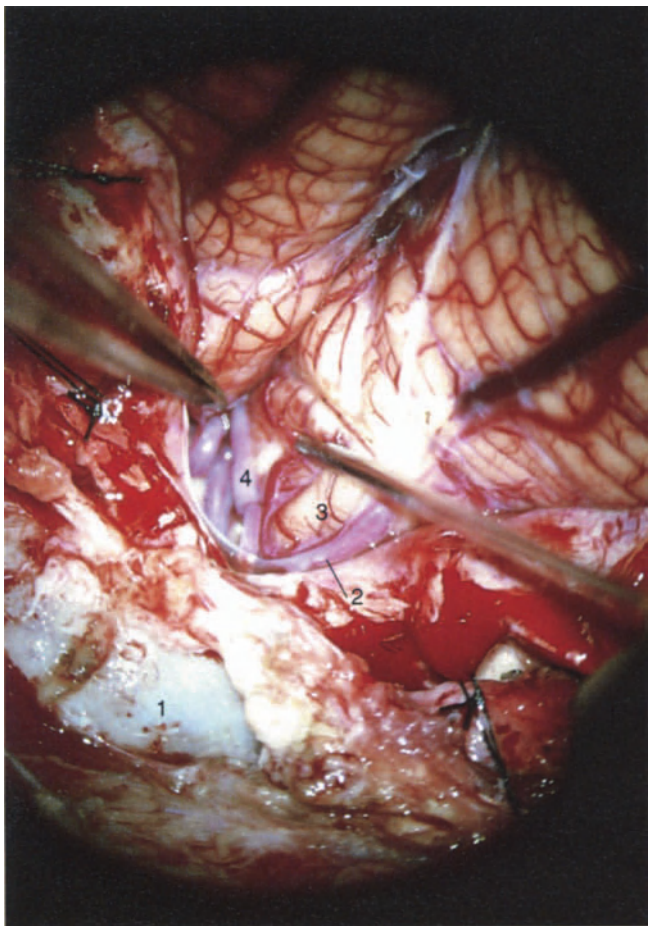


E

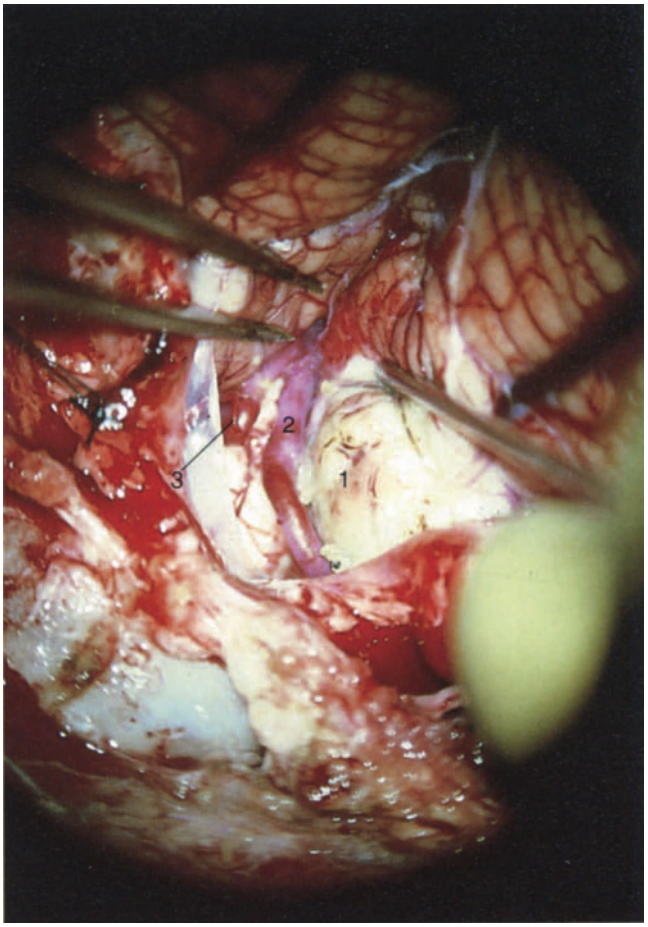




A



B

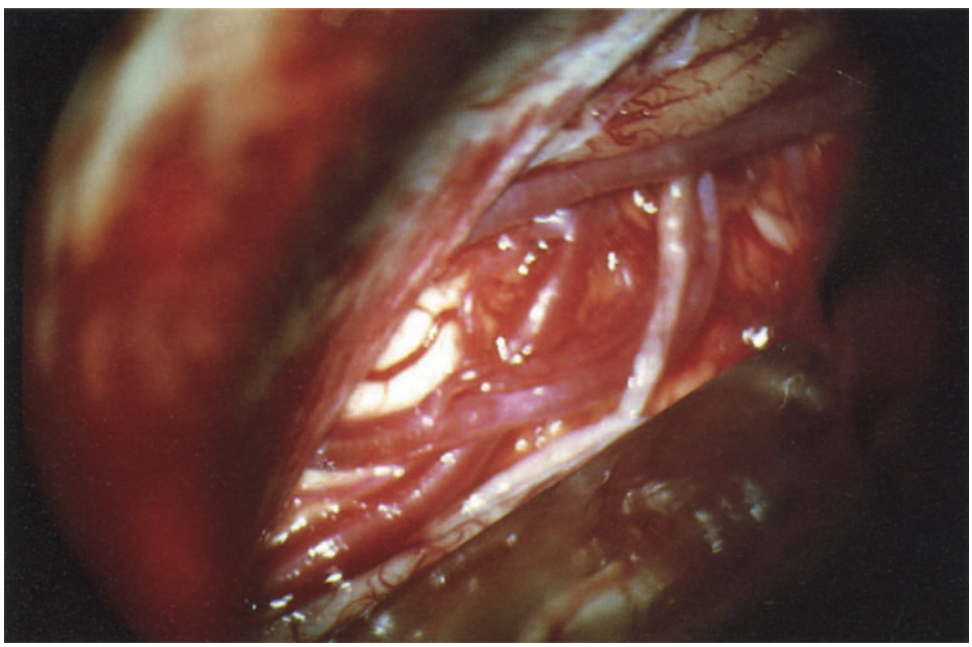


C

Figure 11.20. (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) A sub occipital 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) The vallecula (*1*) has been opened, exposing the floor of the IV ventricle (*2*), in order to permit access to the entry of the PICA into the arteriovenous malformation involving the choroid plexus and floor of the IV ventricle (*3*). (E) The arteriovenous malformation of the choroid plexus of the IV ventricle (*1*) has been coagulated, as have the feeding vessels to it (*2*) and the malformation within the floor of the IV ventricle (*3*). A hemoclip is being applied to the coagulated feeder (*4*). (F) 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 superior cerebellar and anterior inferior cerebellar arteries are exposed, coagulated and transected. ►

D

E



F

the IV ventricle, should not be coagulated or dissected. One may hope that they will subsequently coagulate spontaneously. He should not risk opening into them.

Intraventricular Arteriovenous Malformations

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 technical 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 (Figure 11.21)

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!*

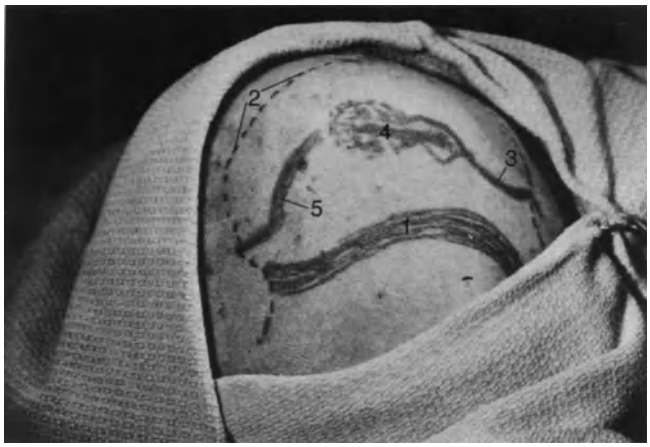
For the nondominant hemisphere, an osteoplastic temporal flap is reflected and the dura is opened in a double trapdoor fashion, prior to performing a cerebrotomy and entering the right lateral ventricle at the trigone. Self-retaining retractors are then placed into the temporal horn and body of the right 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 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.

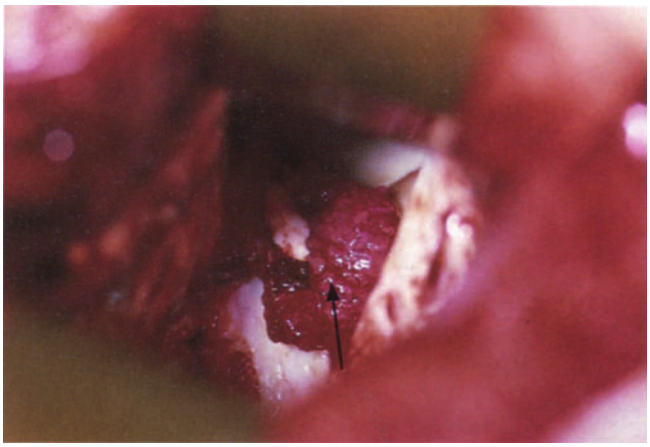
The malformation may now be lifted from its bed if it had been limited to the glomus, or the dissection may be continued to the foramen of Monro if it extends that far anteriorly. There is no need to resect normal choroid plexus from the terminal sulcus or the foramen of Monro. Those portions of malformation 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 (Caution. Oh, be cautious!), a frontoparietotemporal bone flap should be reflected and the dura opened.

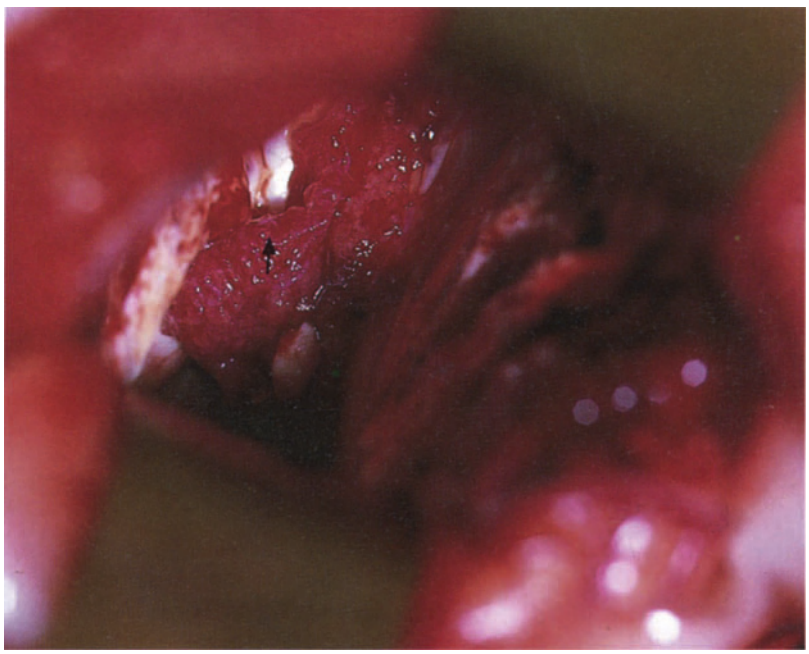
Access to the glomus is attained through a cerebrotomy placed posterior and superior to the supramarginal and angular gyri, entering behind Wernicke's area in the cortex and superior to the radiations of Gratiolet along the lateral surface of the trigone. This minimizes risks of speech impairment or 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, then 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 surgeon the same access to the most anterior portion of the lateral ventricle choroid plexus, without undue risk of damage from retraction.



A



B



C

Figure 11.21. 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.

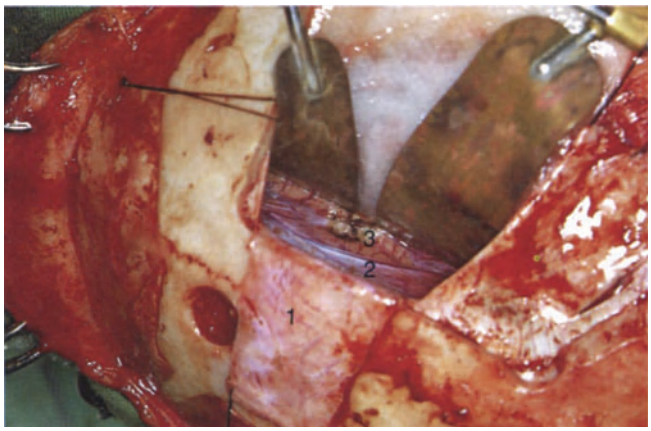
Third Ventricle Arteriovenous Malformations

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.

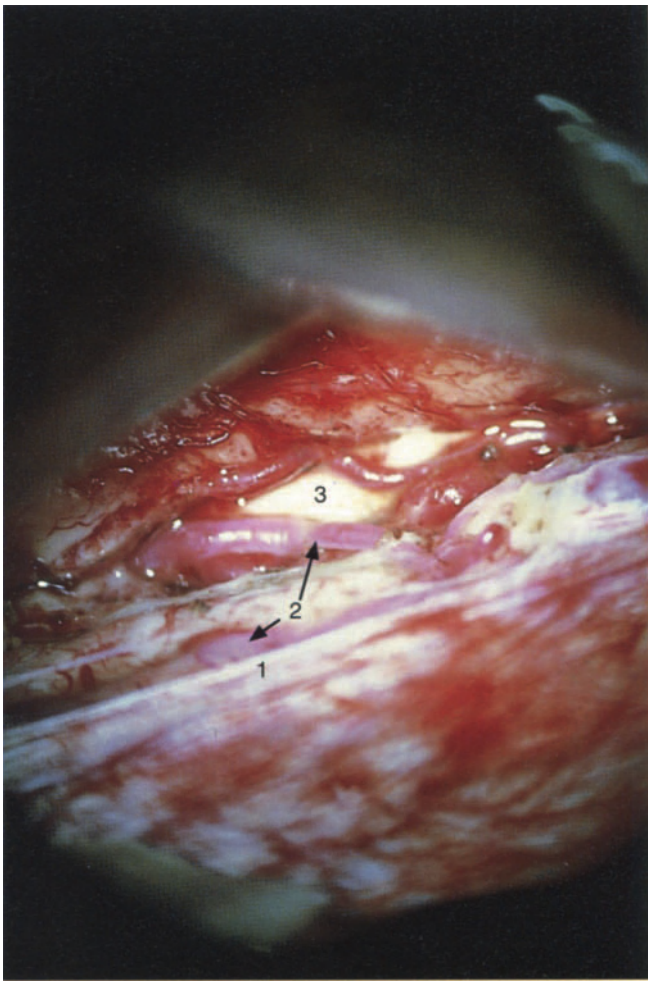
The surgical approach and technique for obliteration of arteriovenous malformations within the roof of the III ventricle is the same as for resection of papillomas of the roof of the III ventricle. The arterial feeders and the venous drainage are the same. The fundamental difference between the two is 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, undertaken, since generally it is not indicated, be limited to coagulating the choroid plexus within the roff of the III ventricle and that no attempt be made to identify and obliterate afferent and efferent vessels.

Arteriovenous Fistulae of the Galenic System

In infants and children, arteriovenous malformations of the Galenic system may be subdivided *anatomically* into 3 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 Galenic system; "inferior" when they enter the ventral surface of the Galenic system; and "posterior" when they course around the aneurysmal dilation



A



B

Figure 11.22. 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*).

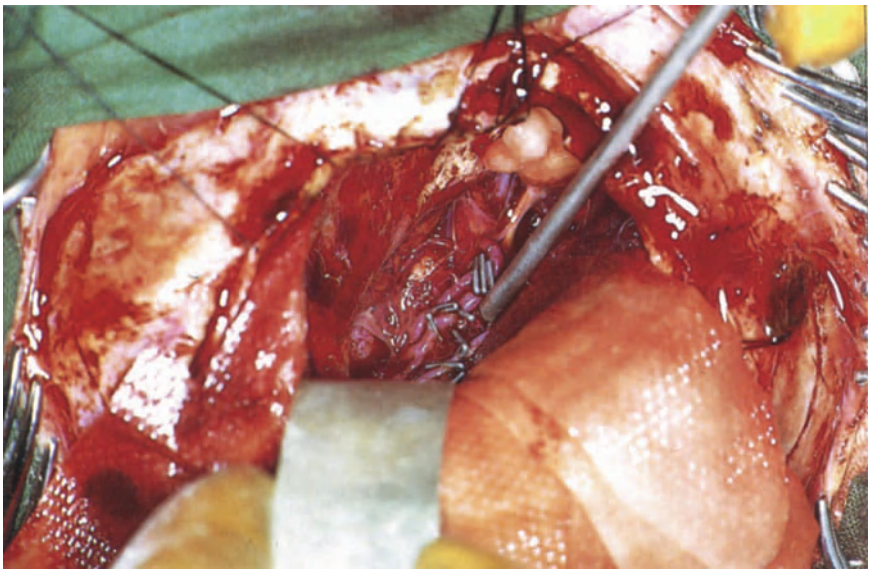
of the great vein of Galen to enter the posterolateral, tentorial surface of the Galenic system. This classification permits the surgeon to plan a biparietal craniotomy approach along the falx cerebri for the "superior" group; a two-stage, bilateral temporal craniotomy and supratentorial approach to 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 Galenic system is left as is, without any attempt to diminish its size either by inserting imbricating sutures or opening and reconstructing it.

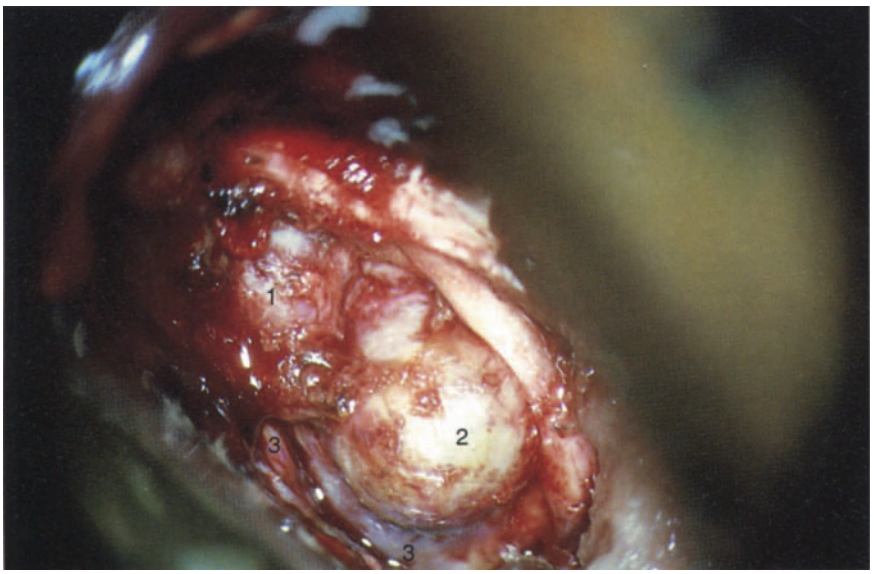
Superior Category (Figures 11.22 and 11.23)

In the Superior category, the pericallosal and posterior cerebral arteries enter the aneurysmally dilated 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 carotid artery are coursing inferosuperiorly, nestled within the wall of the aneurysmal dilation of the great vein of Galen. One cannot expose an adequate length by working over the dome of the aneurysm, so that he is obliged to be in a position to expose the lateral surface without compressing or excessively retracting

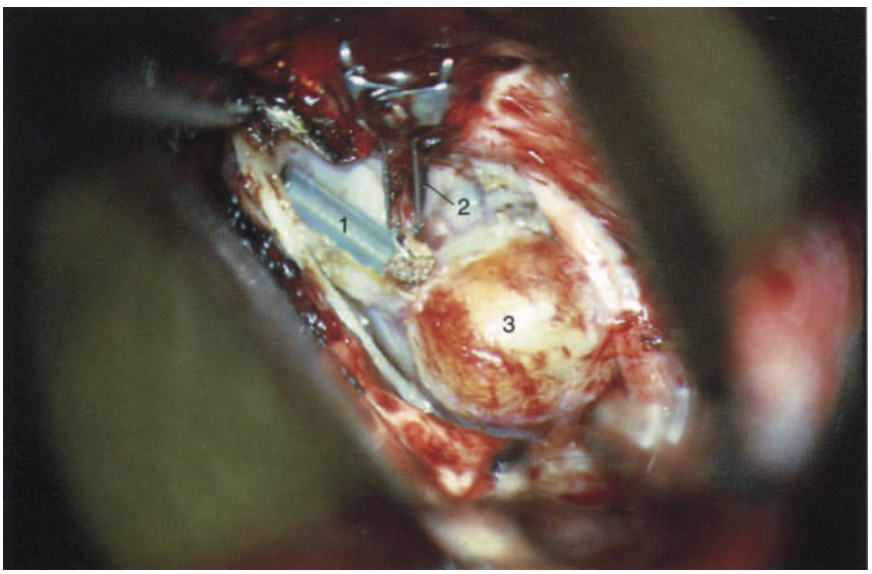
C

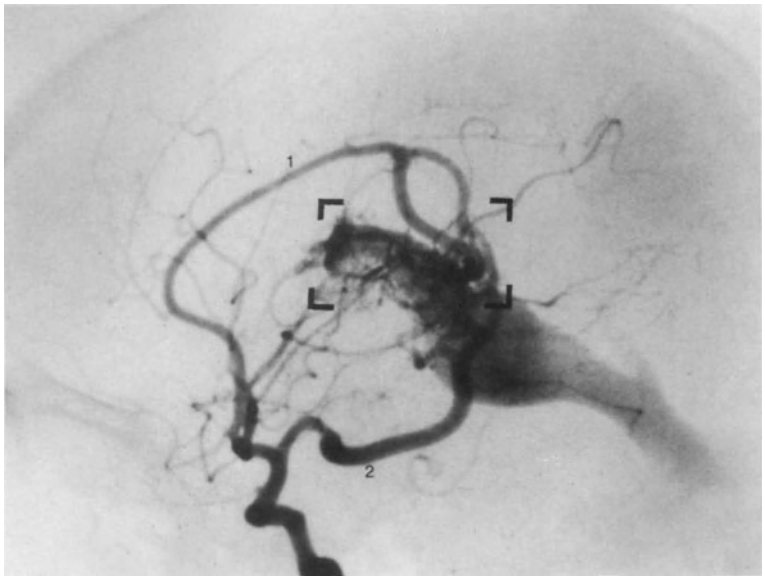


D

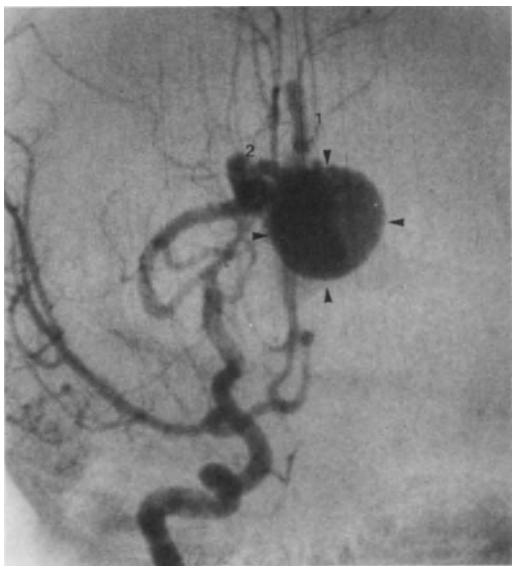


E





A



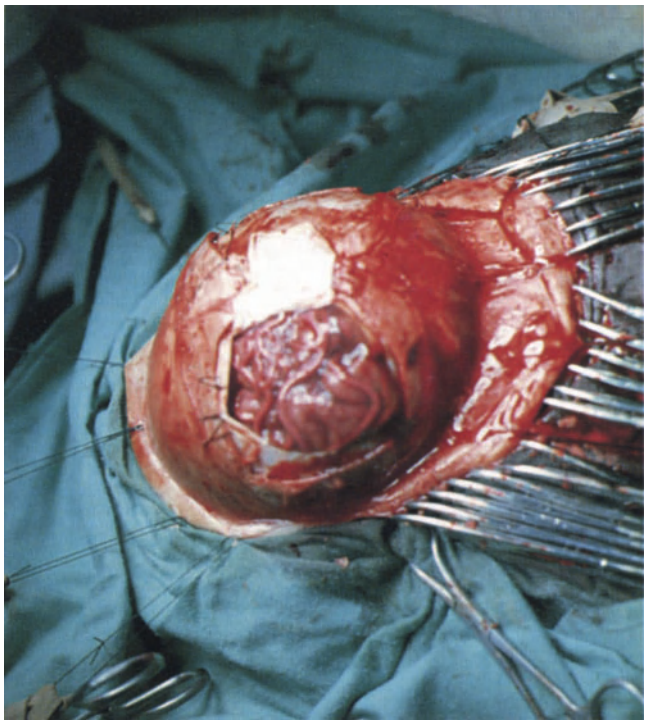
B

Figure 11.23. (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) 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 newborn in progressive, high output failure. ►

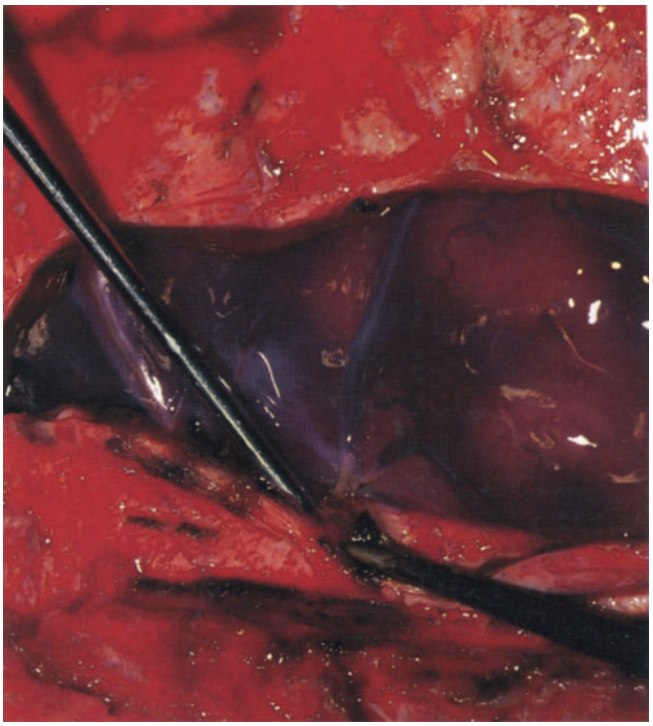
the dome. If a biparietal flap is indicated, then it is best to do this in one stage, rather than to perform a two-stage operation. Access to the corpus callosum and Galenic system is facilitated somewhat by the fact that children with arteriovenous fistulae involving the Galenic system generally have 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, the biparietal flap should be such as to leave a strip of bone over the superior sagittal sinus.

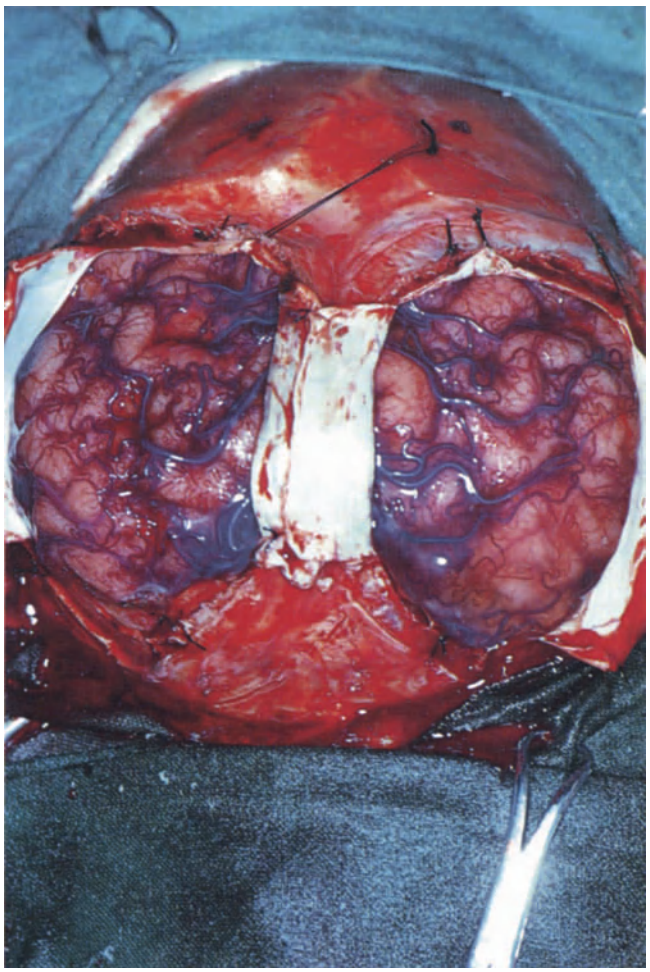
After reflection of the flap(s) and opening of the dura, one may observe a cerebrum which has the appearance of a ball of vessels containing oxygenated blood (in the newborn who is in high output failure, as illustrated in Figures 11.12 and 11.23C), or a normal-appearing cerebrum with few cortical veins 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 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 ex-



C



D



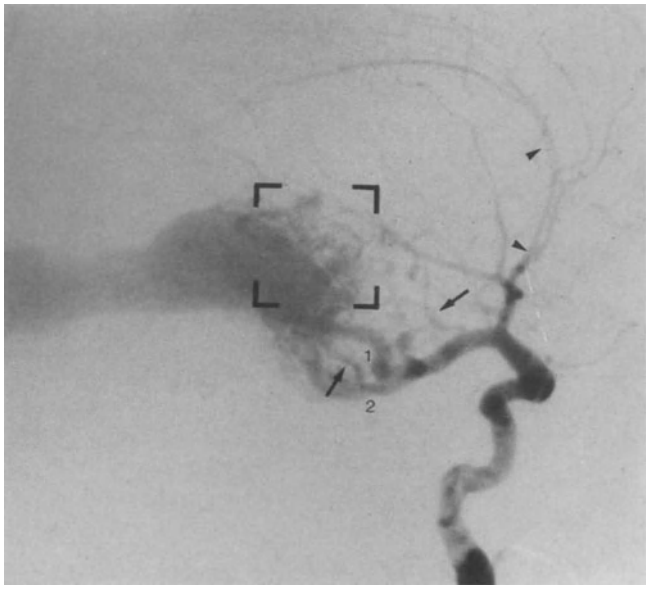
E

ample, 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, clipping, and transection. The entire superior lateral surface must be inspected for tributaries.

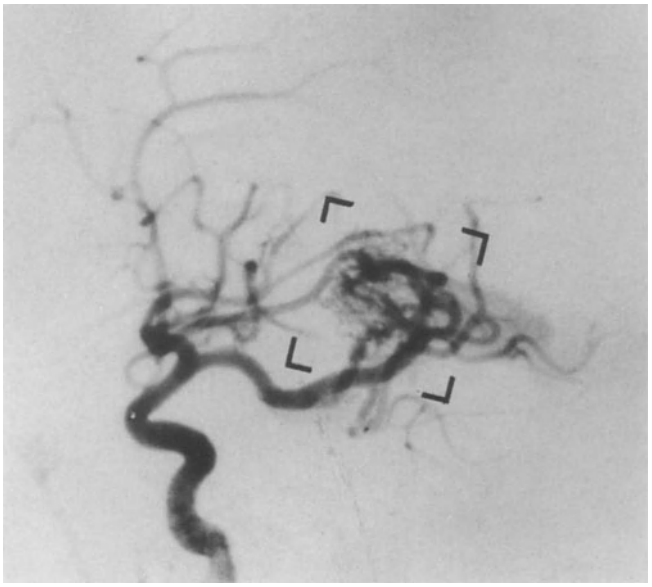
After all tributaries, on both sides (median and paramedian along the homolateral side, paramedian on the contralateral side), have been isolated, one proceeds to clip and transect. Depending upon the size of the tributaries, either small or medium hemoclips are applied. The recommended technique for 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 applicator 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 (Figure 11.24)

The "inferior" category is characterized by tributary branches from the posterior cerebral and posterior chorodial arteries entering the Galenic system along its



A



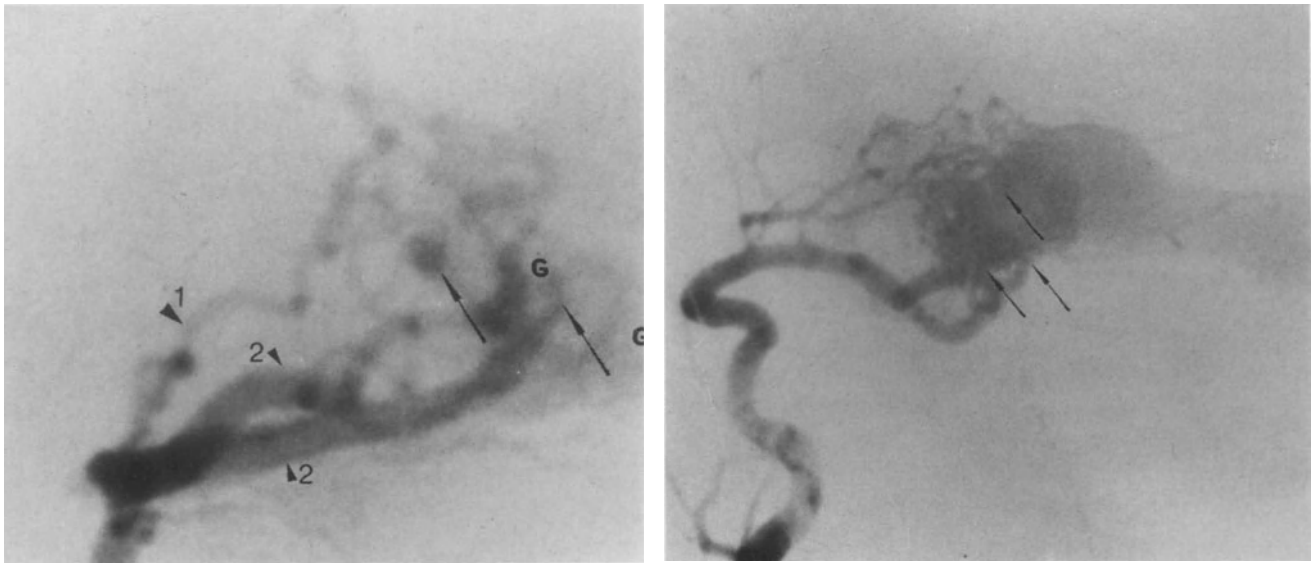
B

Figure 11.24. (A) The medial (*1*) and lateral (*2*) posterior choroidal 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."

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 to the Galenic aneurysm within 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 he 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 Galenic aneurysm being located deep to this cranial nerve. These tributaries are branches of the posterior cerebral system.

Though the inferior supply to Galenic aneurysms is almost invariably bilateral, the degree of dilation and number of tributary vessels varies 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 to 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. 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 Galenic aneurysm. 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, double clipped on either end, and transected.



A B

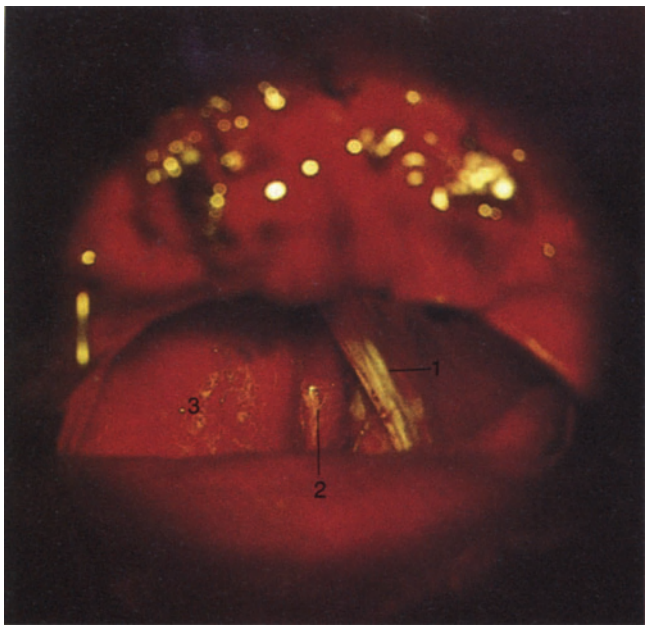
Figure 11.25. (A) The hypertrophied interpeduncular perforating branch (*1*) of the basilar fundus is passing through the brainstem to gain access to the quadrigeminal cistern. The remainder of the afferent vessels (*2*) to the aneurysmal dilation of the great vein of Galen (*G*) course almost horizontally within the ambient cistern, around the brainstem, and over the quadrigeminal plate. They are entering the great vein of Galen along its 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.

Posterior Category (Figures 11.25 and 11.26)
When the tributary vessels to the arteriovenous fistulae 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 and inferior lateral surfaces of the Galenic aneurysm at the point of entry of the afferent vessels.

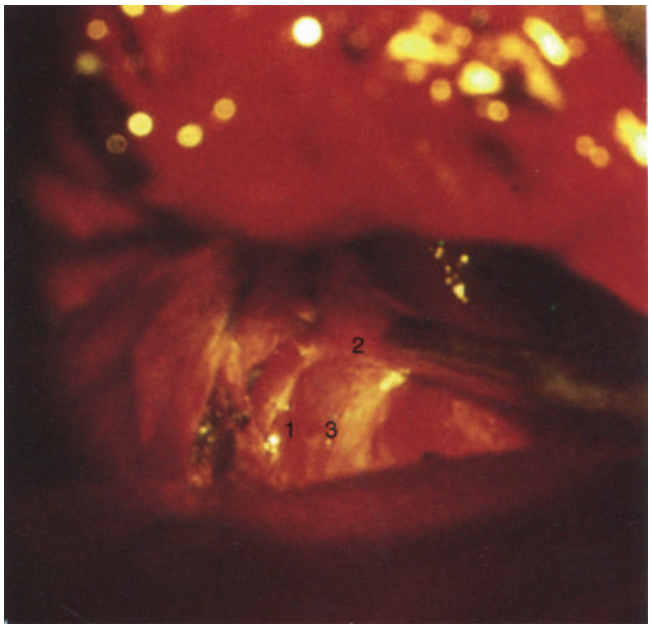
The significant anatomical characteristics are an almost complete absence of bridging tentorial veins, running from the superior surfaces of the cerebellar hemisphere and vermis to the transverse sinus and tentorium. The superior cerebellar cistern is often surrounded by thickened arachnoid, as is the quadrigeminal cistern. Both of these are densely adherent to the Galenic aneurysm superiorly, the culmen monticuli of the cerebellar vermis posteroinferiorly, and the dorsal pontomesencephalic surface anteroinferiorly.

The great vein of Galen, 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 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.

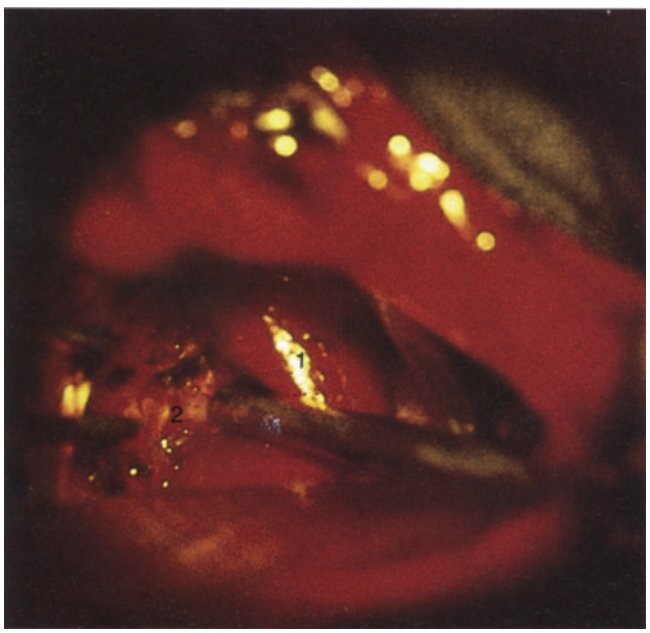
After a superior cerebellar triangle free bone flap has been reflected, the dura is exposed and a single trapdoor dural opening is made. Neither the transverse sinus nor the torcular Herophili should be exposed. The superior bridging veins are identified, coagulated, and transected. This allows the superior 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 of the great vein of Galen falls immediately into the operative field. The tentorial edges are then identified and, subsequent to this, the ambient cisterns are opened, exposing the major trunks of the posterior cerebral arteries and their tributaries to the aneurysm. These tributaries nestle within the Galenic



A



B



C

Figure 11.26. 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 cerebral feeder (*2*), so that the point of entry of this latter vessel into the former may be identified, freed, and occluded.

aneurysm, so that it is necessary to dissect each from the aneurysm, using blunt dissection to separate one from the other, and taking great 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 Galenic aneurysm as each set of tributaries are occluded. The aneurysm is not opened or imbricated.

Uncited Reference

Raimondi AJ: The surgical management of vascular disease in childhood. Amsterdam, Excerpta Medica International Congress Series, No. 310, 1973, p 44.

"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 over-run and oppressed by those of a stronger nature."

ALEXANDER POPE
Essay

Chapter 12

Infections

In few places 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 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.

Osteomyelitis (Figure 12.1)

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 currette these edges until diploic bleeding is encountered.

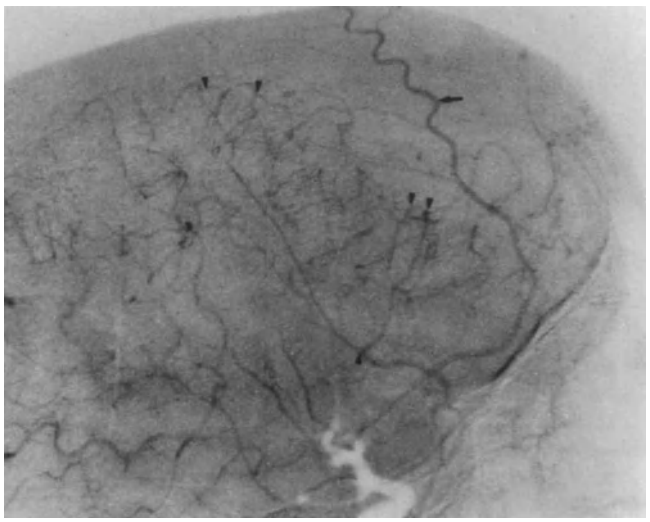
One should avoid opening the dura when operating on a child for osteomyelitis, whether it be secondary to a systemic infection, an infection of the nasal passages, or craniotomy, lest a pathway be opened for extension of the infection into the subdural or intraparenchymal tissue. Computed transmission tomography (CTT) scan and/or cerebral angiography should be used preoperatively to learn whether the infection is limited to the skull. Even when the osteomyelitis is posttraumatic, 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. Intracerebral 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).

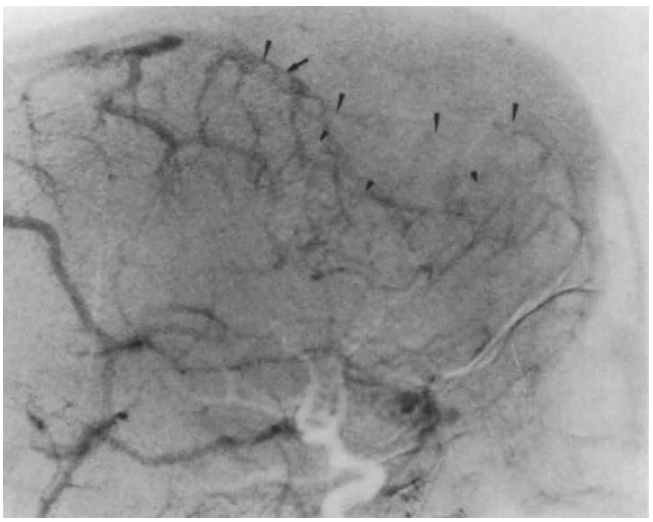
Epidural Empyema (Figure 12.2)

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 dis-

Figure 12.1. This child suffered a head injury, with a skull fracture, and subsequent osteomyelitis. Thereafter an enormous accumulation of liquid epidural pus developed, depressing the superior sagittal sinus (*1*). Surgery consisted of resection of the osteomyelitic tissue, resulting in an irregular skull defect (*2*) and evacuation of the epidural pus. Approximately 9 months later, autogenous rib grafts were fashioned into a cranioplasty, which gave him complete healing of his skull.



A



B

Figure 12.2. (A) The extradural space filling defect over the convexity of the right frontal lobe is expressed angiographically by the displacement and depression of the terminal branches of the anterior meningeal artery (arrowheads). One should note the terminal dilation of these vessels, the stain around them, and the relatively great vascularization of the frontal artery (arrow), all expressive of increased flow around and to an inflammatory process. The relative staining within the right frontal lobe results very likely from compacted brain secondary to displacement and cortical vasodilation. This superimposition study reveals carotid, posterior communicating, and middle cerebral arteries in white—an early arterial phase. The late arterial phases, showing the vessels in black, reveals the stain. (B) Note the irregular contour of the epidural space filling defect (small arrowheads for lateral defect and bold arrowheads for medial defect). It is of particular interest to observe that the venous system draining into the superior sagittal sinus (SSS) is somewhat depressed as is the SSS.

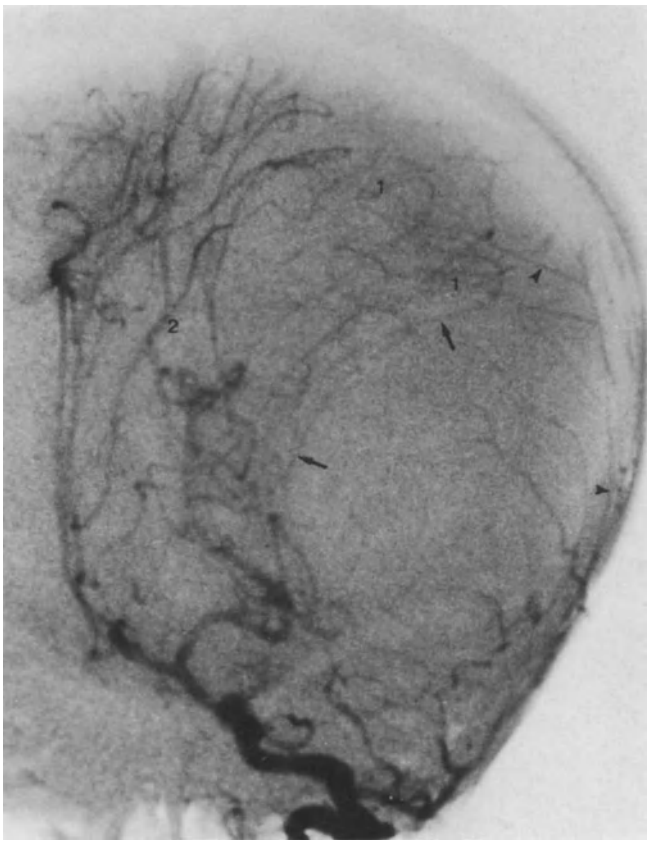


Figure 12.3. Subdural abscess. The accumulation of a discrete pocket of liquid pus within the subdural space is outlined laterally by the middle meningeal artery and its terminal branches (arrowhead) and medially by the displaced Sylvian complex (arrows). The staining of the inferior temporal gyri (*1*) indicates cerebritis, and the "halo" formation surrounding the displaced frontopolar artery (*2*) indicates meningitis. There is no evidence of capsular stain.

tinguish it from an intraparenchymal abscess. A CTT 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 burr hole over the suspected area of loculated pus within the epidural space.

Treatment of the epidural empyema consists of placement of a single burr 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 to 10 days, depending upon reexpansion of the brain and obliteration of the cavity. CTT scan suffices to monitor the healing process.

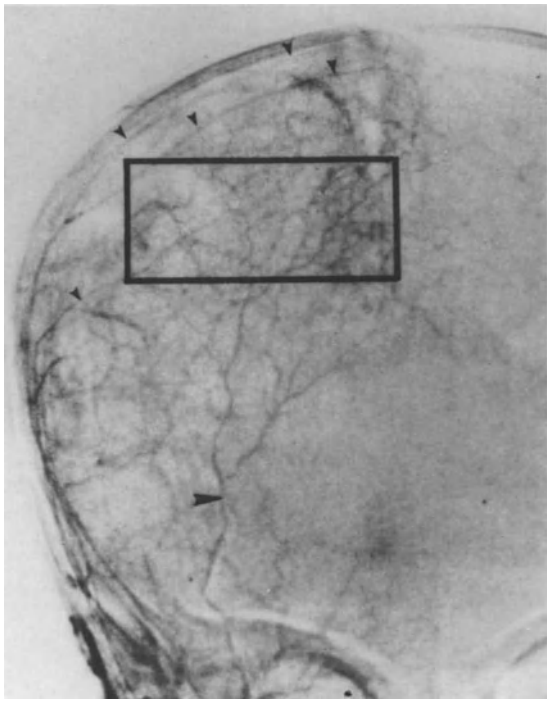
Subdural Abscess and Subdural Empyema

(Figure 12.3 and 12.4)

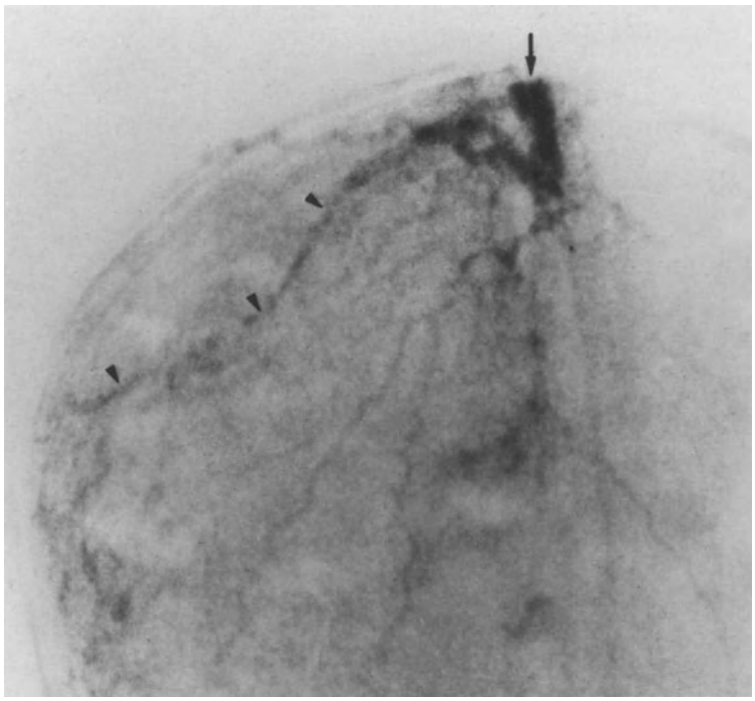
The difference between subdural abscess and subdural empyema is that the former is a well-encapsulated collection of liquefied exudate (pus), and the latter is a diffuse, fibrinous, pyogenic exudate adherent to adjacent pachy and leptomeninges, and subjacent cortex.

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!

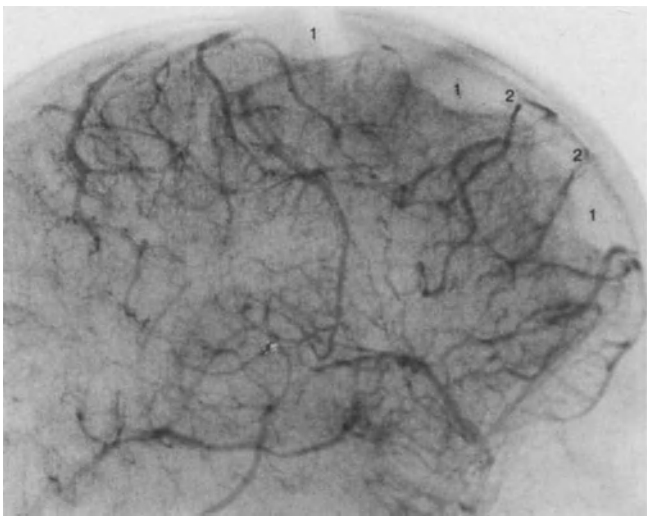
Anatomopathologically speaking the subdural empyema consists of a collection of fibrinous exudate within the subdural space, varying in thickness from 0.5 to 2 mm, intimately adherent to the inner surface of the dura externally and the arachnoid membrane internally. At times, it may be so thick and globular 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 burr 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 to 6 weeks of intravenous antibiotic therapy. The subdural *abscess*, however, is treated surgically with craniotomy, or burr holes, and evacuation of the pus, followed by antibiotic therapy. Anticonvulsant medication is essential because of the high incidence of seizures in these patients.



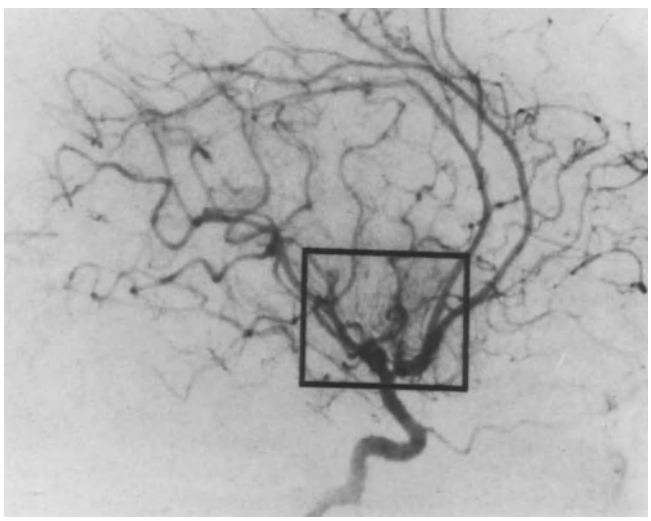
A



B

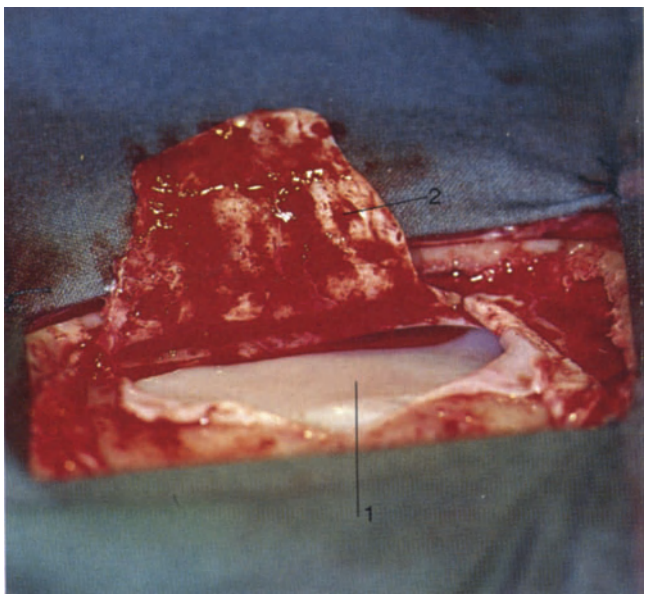


C

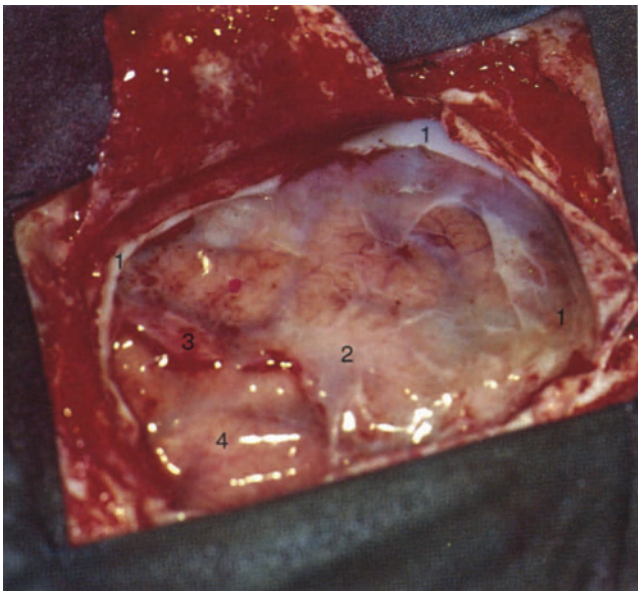


D

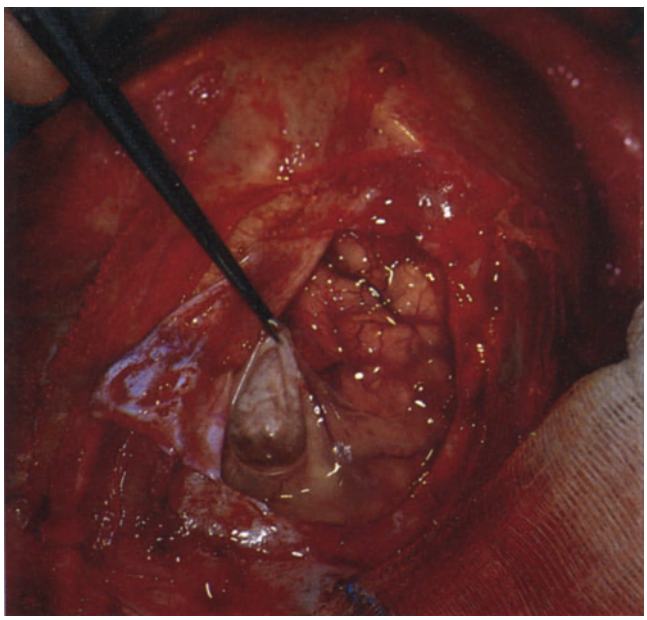
Figure 12.4. (A) This is an earlier phase of the same angiographic study as illustrated in Figure 12.5. Here, one may appreciate the normal course and location of the terminal branches of the middle meningeal artery (arrowheads), as well as the increased flow through the occipital artery (bold arrowheads) and the dilation of the terminal branches of the latter vessel (square). These dilated vessels nourish the "rete mirabile." (B) The cortical vein (arrowheads) is depressed, outlining an irregular space filling defect within the subdural compartment. The superior sagittal sinus (SSS) (arrow) is not depressed. (C) The loculation of purulent exudate in the subdural space is irregular (*1*), with "tenting" occurring at the point of entry of the bridging veins into the SSS (*2*). (D) One of the characteristics of subdural empyema is that the phlogistic process involves not only the visceral layer of the dura mater but, more importantly, the cerebral cortex. This is characterized angiographically by well-circumscribed dilation of cortical vessels, generally on a gyral pattern, with staining (square). These changes put into relief the futility of surgical treatment. (E) Here one notes the pearl white subdural empyema mem- ►



E



F



G

brane (*1*) beneath the dural (*2*) opening. (F) 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*). (G) The subdural empyema is adherent to a necrotic portion of brain.

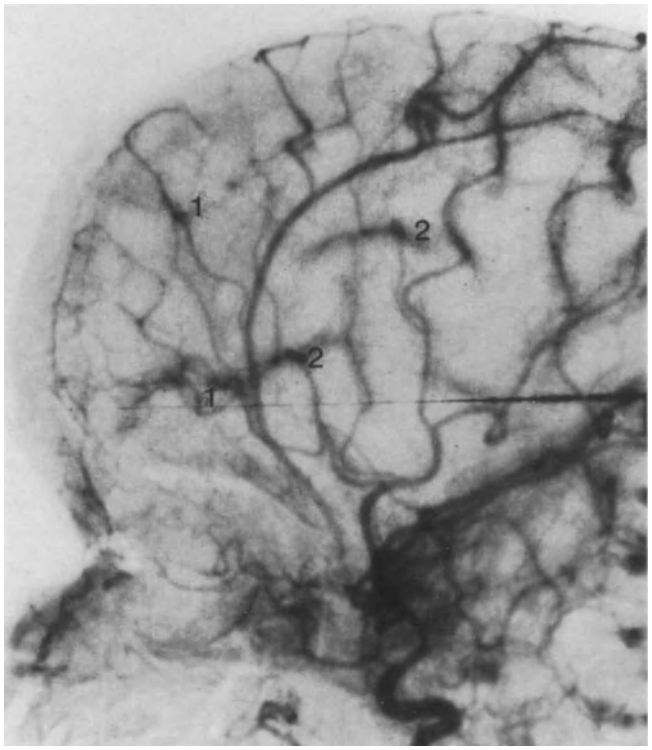


Figure 12.5. The arterial phases are characterized by "halo" formation around the sulcal branches of the anterior (*1*) and middle (*2*) cerebral systems.

Acute Meningitis with Hydrocephalus

(Figure 12.5 and 12.6)

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 Chapter 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 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 burr hole, abscess puncture, and drainage of pus; (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."

Burr Hole and Cannula Drainage

A single burr 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, 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-cc syringe to the cannula so as to aspirate

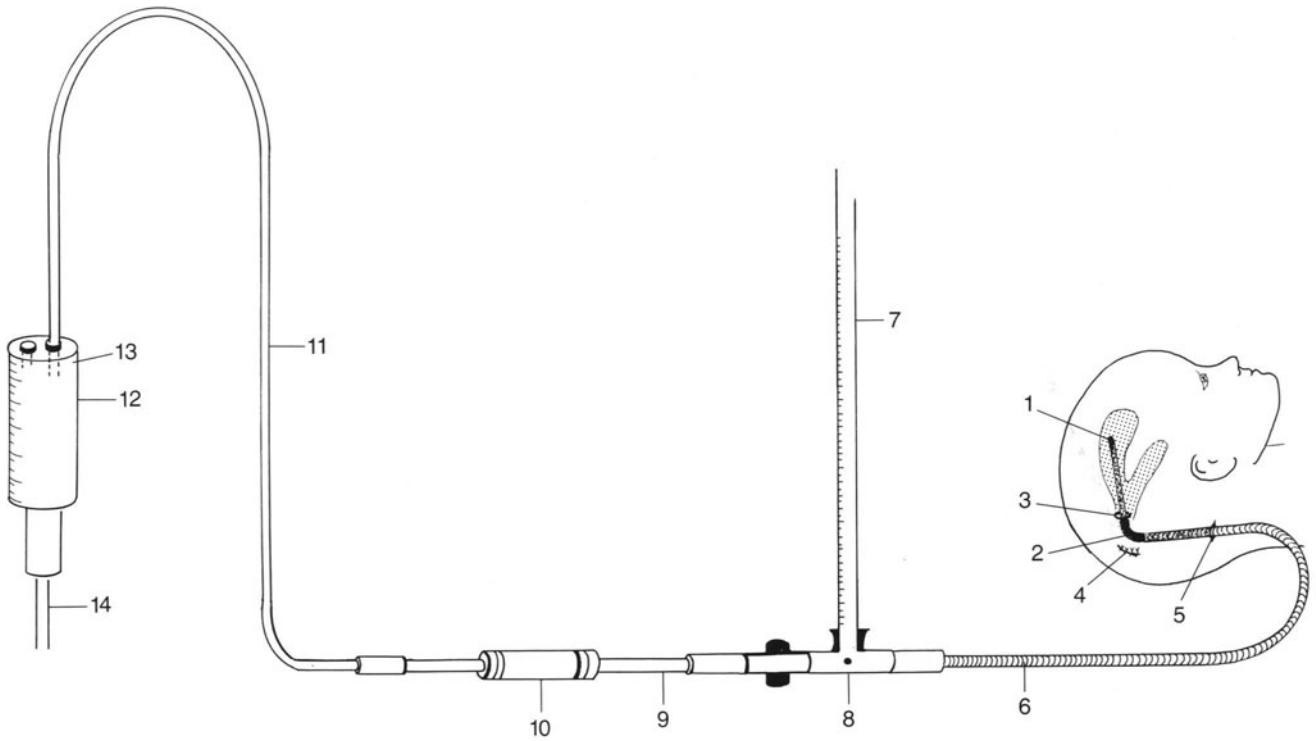


Figure 12.6. External ventricular drainage. *1* = Ventricular catheter; *2* = right-angle connector, optional, since the ventricular catheter may be run under the skin and then thru the stab wound; *3* = drill hole; *4* = skin incision; *5* = stab incision; *6* = spring tubing; *7* = manometer; *8* = 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 same level as head and heart; *14* = cerebrospinal fluid collection tubing.

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 CTT scans should be performed the evening of surgery, and then at three day intervals until the abscess cavity has collapsed completely. If there is CTT 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, of 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 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 flattend 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 cavity, 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 a 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 a 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 cavity, 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, 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 CTT 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 two 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 to remove the external ventricular drains after the infection has cleared. If, subsequently, hydrocephalus becomes apparent, a shunt may be inserted.

Ventriculitis (Figure 12.6)

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); (3) *external ventricular drainage, with appropriate antibiotic treatment, is, in fact, the treatment of choice.*

The external ventricular drain consists of a single Spring Catheter* shunting system inserted into the lateral ventricle, through a split trochar (see the section on shunt Revisions in Chapter 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%.

Subdural Effusions (Figure 12.7)

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 2 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 fontanel. Other

*Codman, Randolph, Massachusetts.

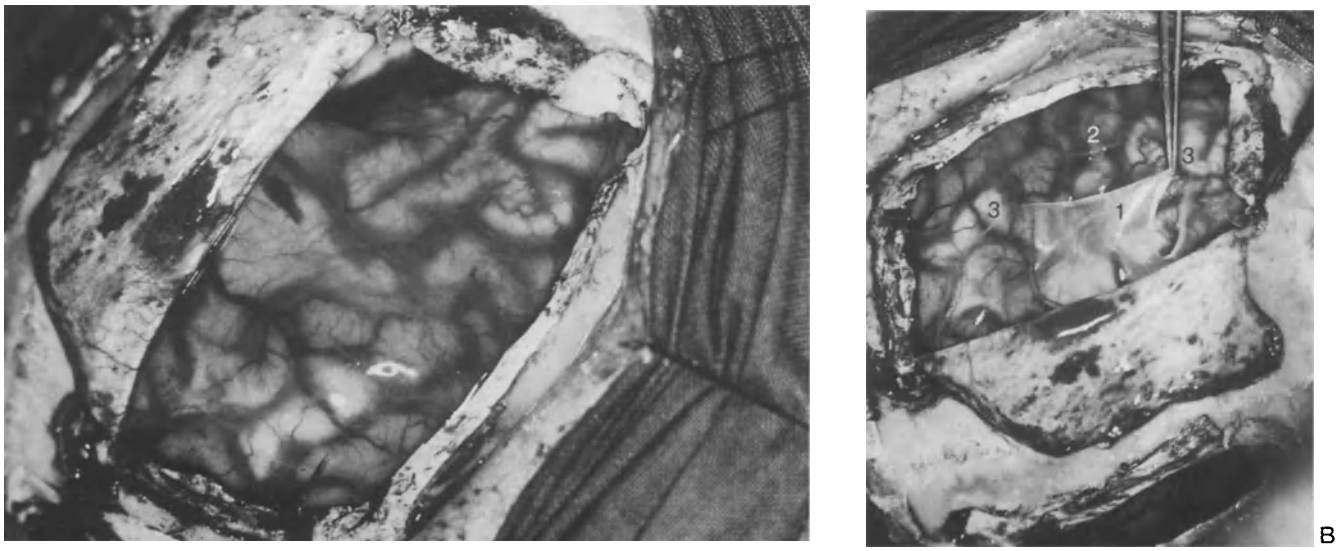


Figure 12.7. (A) The remarkable cerebral atrophy that occurs in association with long-standing subdural effusion, in a child who had suffered meningitis. One notes the opalescent membranes over the surface of the hemisphere, the enormous sulci, the shrunken gyri. (B) The outer leaf (*1*) of the subdural effusion membrane is elevated, exposing underlying effusion, inner leaf (*2*), and extensive areas of devascularization of cerebral gyri (*3*).

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, shunting procedures, have all failed. The treatment of subdural effusions consists of burr 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 Chapter 13) when there is progressive diminution in cerebral volume and "hanging veins."

Cerebritis and Cerebellitis (Figure 12.8)

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 processess 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 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

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.

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.

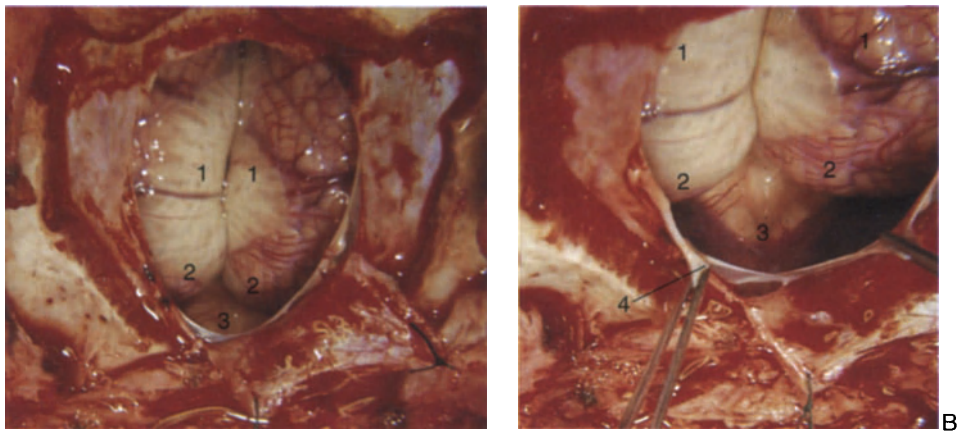


Figure 12.8. Cerebellitis. (A) The degenerative and necrotic changes within the cortex of both cerebellar hemispheres (*1*) has resulted in adhesions of the hemispheres to one another. The membrane of the cisterna magna has been removed, exposing the tonsils (*2*) and the dorsal aspect of the medulla 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 a 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.

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; 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, 3 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.

Uncited References

McLone DG, Czyzewsky D, Raimondi AJ, Sommers R: Central nervous system infections limiting intelligence. Pediatrics 70:338–42, 1982.

Raimondi AJ, DiRocco C: The physipathologic basis for the angiographic diagnosis of bacterial infections of the brain and its coverings in children: I. Leptomeningitis. Child's Brain 5:398–407, 1979.

Raimondi AJ, DiRocco C: The role of cerebral angiography in meningocerebral inflammatory disease in infancy and childhood. Neurosurgery 3:37–44, 1978.

Raimondi AJ, Matsumoto S, Miller RA: Brain abscess in children with congenital heart disease. J Neurosurg 23:588–95, 1965.

"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

Chapter 13

Trauma

The management of craniocerebral trauma in the child is, in contrast to the adult, if anything, more dependent upon critical neurologic 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 his 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 and the time he reaches a well-staffed and fully equipped nursing station.

The proper time to request skull x-rays, or computed transmission tomography (CTT) scans, for that matter, is after the child has been examined by a qualified pediatric neurosurgeon. He will then determine, on the basis of 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 CTT 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 on subdural), which may result from the same sequence of traumatic events that would cause a linear skull fracture: diploic hemorrhage from a diastatic fracture, epidural hemorrhage over the superior sagittal sinus from a linear or depressed fracture across the sinus.

The current state of the art of CTT scanning is such that one may obtain much more information from a carefully performed CTT scan than a combination of plain skull x-rays, cerebral angiography, and a hastily performed CTT 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 nuclear magnetic resonance in the management of acute, or chronic, head injuries.

Younger children suffer much greater damages from such deceleration impact injuries as falls, and acceleration burst injuries such as blows to the head. Approximately 50% of children whose fontanels have not closed suffer skull fractures from either acceleration or deceleration injuries, whereas only 29% of children with closed fontanels and sutures suffer fractures. Similarly, children less than six months of age suffer the highest death rate among motor vehicle occupants (9/100,000), one-year-old children suffer the second highest rate (4–5/100,000), and children between the ages of six and twelve 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 fontanel closure. If one studies age distribution histograms of children with open and closed fontanels, it becomes apparent that fontanel closure occurs at approximately one year of age, permitting the clinician to superimpose the closed fontanel population upon children older than one year of age. These observations (a higher incidence of skull fracture in children with open sutures and fontanels, 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, are the causative factors is not possible to ascertain at this time. The facts are that infants have less chances of a good recovery after blunt head injury than do children older than one year of age, 13.4% poor recovery in children less than one year of age and 4.9% in those between one and three years of age. Similarly, the highest incidence of admission to hospital for head injuries in childhood occurs during the first three 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 off at a plateau of (7%) during each trimester between the 21st and 36th months of life.

One may not, with certainty, explain 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 fontanel and sutures predispose a child to a higher incidence of subdural hematoma. It has been observed that 32% of the open fontanel population suffer subdural hematoma as a consequence of blunt head injury, whereas only 5% of the closed fontanel population suffer this complication. In fact, in a review of my own work, I observed that 79% of all posttraumatic intracranial mass lesions were in infants and that 93% of these were subdural hematomas! 7.5% (4 of 53) of these patients were toddlers, most suffering a CCS admission of 11 and none suffering a CCS below 8.

The newborn and infants suffering subdural hematomas are generally admitted late, at a time when they are symptomatic (failure to thrive, seizures, vomiting, lethargic) with clinical evidence of increasing intracranial pressure (split sutures, bulging fontanel, 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 fontanels 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 fontanel populations. This is expressed by the fact that excluding all subdural hematoma patients (83 of 462 in those studied by the author) results in a drop of poor outcome from 4.9 to 2.8% in the closed fontanel population and from 13.9 to 4.6% in the open suture population. Still, however, children under one 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 different 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 he ages. During the first three years of life, the central nervous system continues to mature at almost the same rate as during the intrauterine period. Progression from the newborn existence (approximately 22 hours of sleep, with the waking intervals being limited almost entirely to feeding) to the 30-month-old child (who speaks, obeys command, 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 two years of life, whereas myelinization of the central nervous system begins within the first year of life, but then continues progressively through the 10th year. By four 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 two 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, large basal cisterns.

On the other hand, older infants suffer tears at the pontomedullary junction, diffuse petecchial hemorrhages, 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 first approach and then join one another, the fontanels become obliterated as membranous bone forms between the two layers of the periosteum, the sutures lose their anatomical definition as a mesenchymal bridge between the outer layer of the dura (endosteum) and the periosteum, the tables of the skull thicken and the diploë convert from sinusoidal chambers to an irregular mass of channels running perpendicular to the surface of the skull. Thus, by one 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.

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, adolescents (and, alas, young adults) have invariably been considered a homogeneous group. Of especial significance is the fact that, with only one exception, this subject has never been studied—reported—chronologically, analyzing clinical observations in monthly, or yearly age-time frames in the critical first three years of life.

The efficacy of surgical or medical (e.g., the use of Decadron), as well as intensive care, management with the institution of bartiturate coma, has been measured using the GCS and 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 demand (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, meningitis, porencephaly, vasospasm, 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 two 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 resistent to cerebral damage. Approximately 20% of head-injured children suffer hyperkinesis, difficulty in anger control, impaired attention, and headache: elements of the "posttraumatic 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, the author 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. 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 to 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 to the CCS for higher integrative functions reveals that in the GCS a score of 12 to 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 fontanel, 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 the children with an ocular examination of 4 do well, whereas no child with an ocular examination of 1 sur-

vives. 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 category 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 two 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 two months of age. On the other hand, only the most basic motor functions—sucking, reflex grasp, startle reflex—are present in a two-month-old, whereas a two-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 fontanels and 94% of those with open fontanels 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 fontanel and 94% of the open fontanel 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 fontanel and 92% of the open fontanel 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 posttraumatic splitting of the sutures is indicative both of a "poor" outcome and a higher incidence of posttraumatic seizures. Palpation of the open fontanel provides good evidence regarding outcome, since only 5% of the children with a soft fontanel suffer a "poor" outcome, 16% of those with a full fontanel suffer a "poor" outcome, and 50% of those with a tense fontanel suffer a "poor" outcome. Only 12% of the children with a soft fontanel have subdural hematomas, whereas the percentages rise to 71 and 83, respectively, for those with full and tense fontanels. 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.

Posttraumatic seizures occur in 10% of the children with depressed fractures and 7% of those without fracture. Irrespective of whether a child's sutures and fontanel 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 three 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 posttraumatic 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 posttraumatic 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 posttraumatic seizure patients being less than one 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 posttraumatic seizures is higher in infants than in toddlers (26% vs 10%), with evidence that the toddler has the same incidence of posttraumatic 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 one year of age, one realizes that the seizure rate drops after the first month of life, remaining high during the first 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 indi-

cate 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 extraaxial 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 fontanel, since an expanding extraaxial hematoma does not invariably cause a tense or bulging fontanel.

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 burr 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 fontanel and 62% "good" results in closed fontanel 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 fontanel 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 Chapter 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 superior sagittal sinus (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

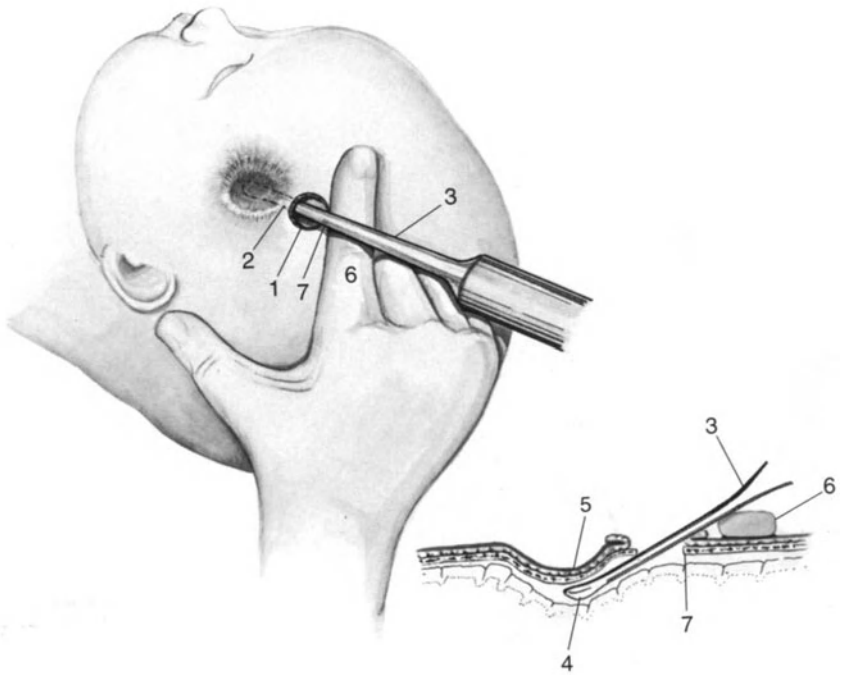


Figure 13.1. The "ping-pong" fracture is elevated by placing a burr hole opening (*1*) immediately peripheral to the point where normal skull ends and the depression begins (*2*). A periosteal elevator (*3*) is then inserted through the burr hole, bringing its tip (*4*) to the very nadir of the "ping-pong" 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 burr hole opening (*7*), lest he inadvertently cause another depressed fracture.

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, that the child be followed closely, 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 he finds either hemotympanum in the absence of a ruptured drum, battle sign (or raccoon eyes) in the absence of direct injury to the temporal or nasoorbital 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 (Figure 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, and that it affords the patient the added protec-

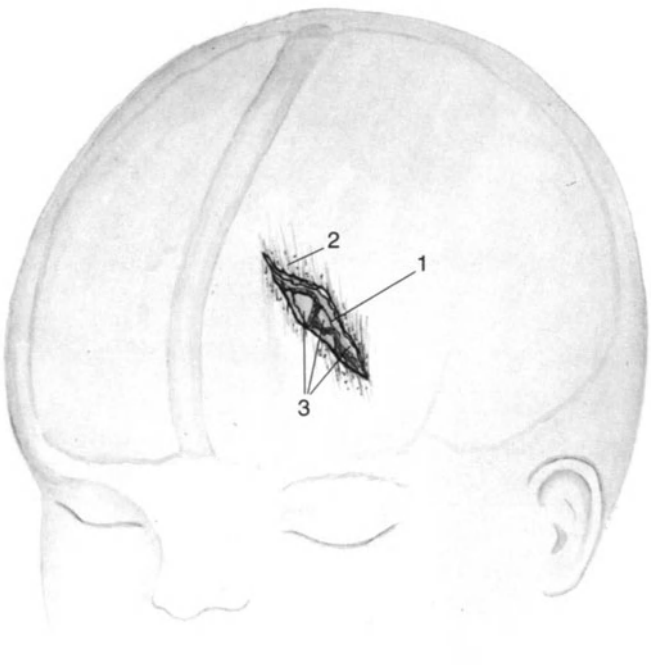


Figure 13.2. A compound depressed fracture is characterized by a full-thickness scalp laceration almost invariably accompanied by extensive tissue destruction from the compressive force, and single or multiple bony spicules, fragmented and, at times, impacted into the underlying dura and brain. The laceration (*1*) and surrounding areas of tissue damage (*2*) may overlay the bony spicules (*3*). This does not invariably occur.

tion 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 concavity of the skull into a 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.

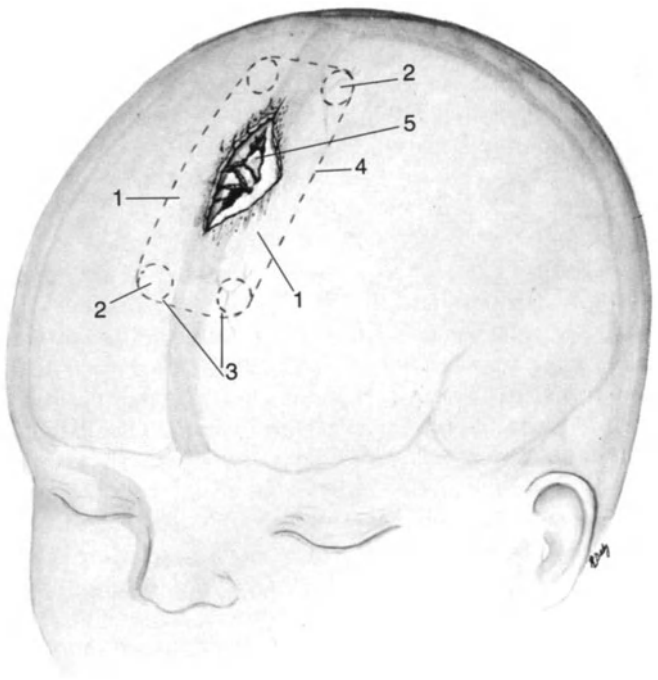
The ping-pong fracture is treated by making a perforator opening in the skull just peripheral to the limbus of the depressed area, and then slipping a Penfield #4, or #1, dissector, whichever is most appropriate, between the skull and the dura. The depression is then elevated, with the surgeon taking 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 burr 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.

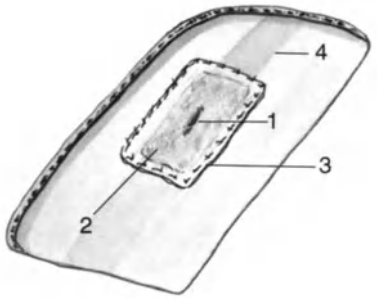
Compound Skull Fractures (Figures 13.2 to 13.4)
The compound depressed skull fracture represents an acute neurosurgical emergency, since one is obliged to debride the wound, inspect the fracture area for persistent bleeders, and remove bone spicules and foreign substances from the epidural space and surface of the brain. One should not explore the subcortical area of the brain for foreign substances nor should he enter this area in search of spicules identified either on CTT 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 communited 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.

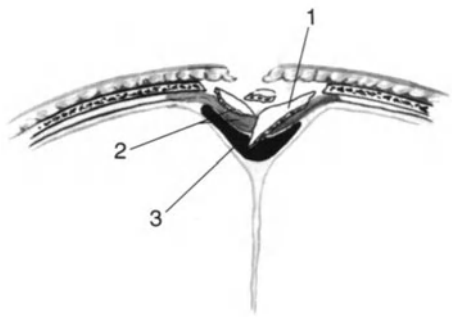
The compound depressed skull fracture over the superior sagittal or transverse sinuses presents entirely different surgical implications, operative risks, technique. Once one diagnoses a compound depressed fracture over one of these sinuses, either clinically or with skull x-rays, he should proceed immediately to CTT 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 cerebral angiography, taking care to obtain excellent venous phases 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 himself 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.*



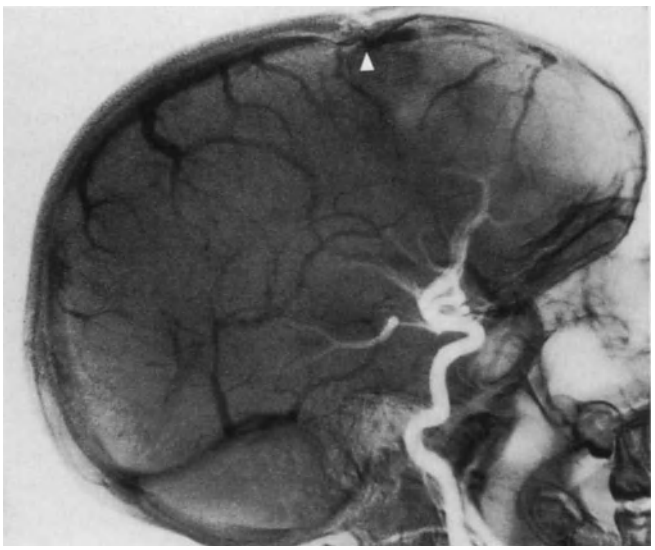
A



B



C



D

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 burr holes (*2*) so that their medial convexity borders upon the lateral surface of the SSS (*3*). Once the burr 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) In the so-called coconut fracture, the vertex of the skull is depressed to such a degree that it impinges upon the SSS with resultant constriction, occlusion, or tearing of this major dural venous channel. The arrowhead indicates the point at which the depressed fracture indents the SSS, apparently occluding it completely. This study illustrates the arterial system in white, the venous system and sinuses in black.

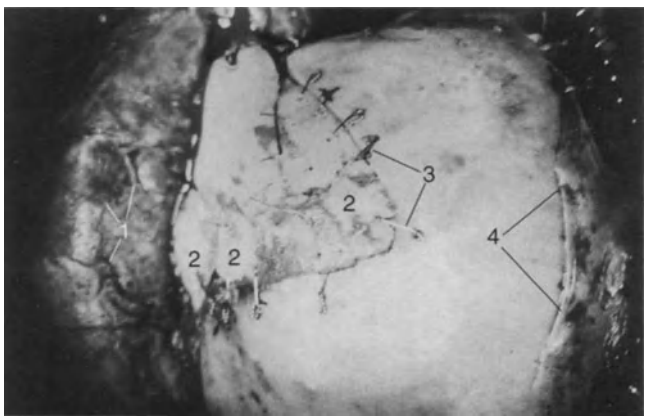


Figure 13.4. Improper technique for management of compound depressed skull fractures. One notes the scalp laceration (*1*), multiple bone fragments (*2*), and wiring of these back into position at the time of closure (*3*). The compound fracture, by definition, *contaminates* the wound. The bone fragments 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 risk of osteomyelitis.

In the event one is obliged to remove the depressed fracture fragments from the superior sagittal sinus, he 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. Burr 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 burr 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, 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 unfortunate 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 he 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 to 15 minutes. 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, 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, 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 (Figure 13.5 and 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

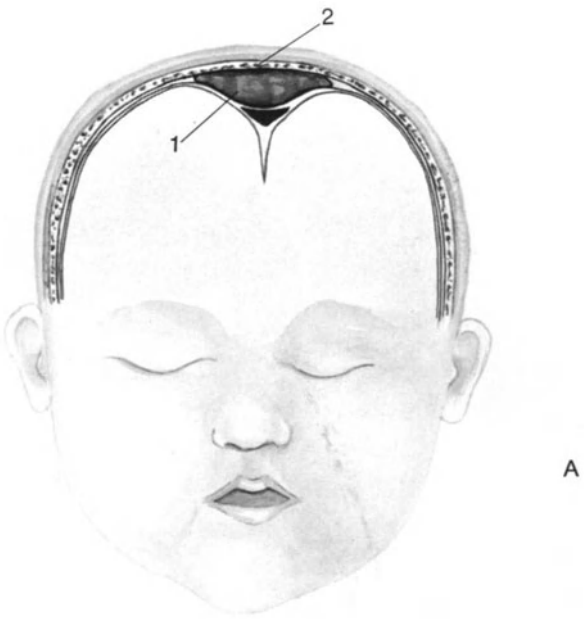


Figure 13.5. These drawings represent the two types of venous epidural hematoma, one over the SSS and the other over the convexity. (A) The SSS may be depressed either by a pure clot (*1*), secondary diastatic splitting of the sagittal suture (*2*), or as illustrated in Figures 13.B and 13.C. (B) Convexity (*1*) epidural hematomas are, on the other hand, almost invariably arterial, with the exception of bleeding from the transverse sinus. They may expand over both the supra- and 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 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.

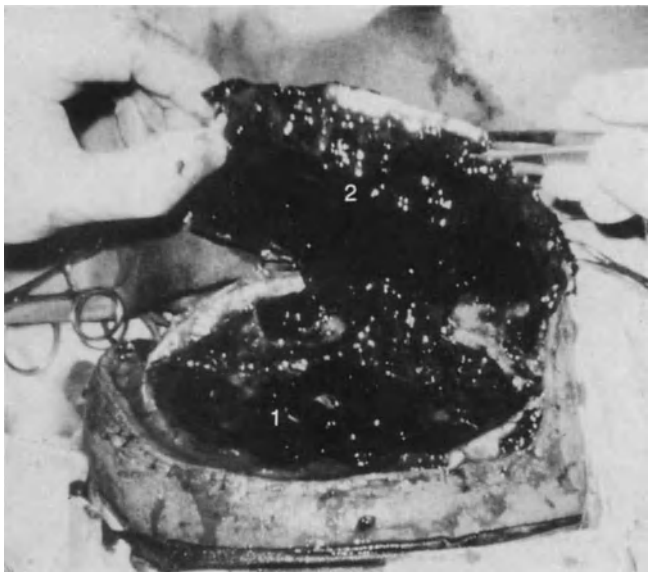


Figure 13.6. Epidural hematoma, illustrating a clot over the dura (*1*) and along the inner table of the skull (*2*), so as to demonstrate that the craniotomy line may not always border upon the most peripheral portion of the blood clot. Consequently, epidural hematoma extends beyond the line of craniotomy, separating 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.

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.

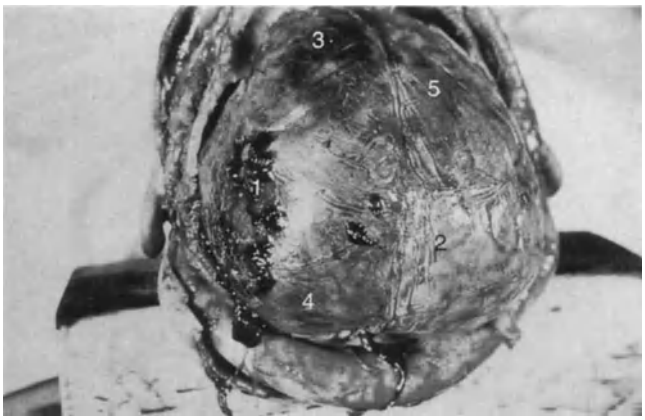
One must not expect invariably to find a torn meningeal vessel when lateral epidural hematoma is surgically evacuated. Rather, in the newborn and infant this is an exception. In the toddler and juvenile, it is a rarity. In the adolescent, it is exactly the same as in the adult. Following evacuation of 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 which is bleeding from the inner table, 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, not from rebleeding meningeal arteries.

Posterior Fossa Epidural Hematoma

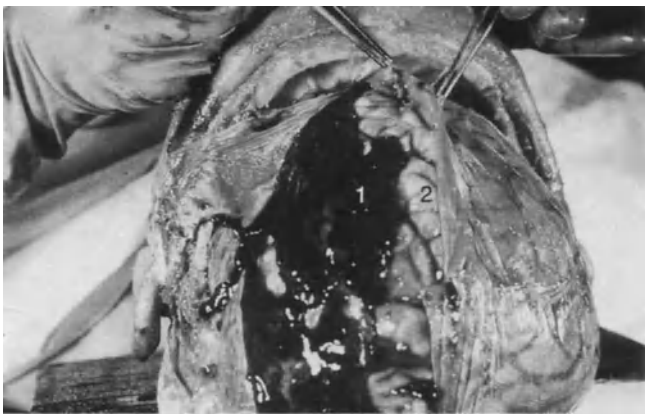
The posterior fossa epidural hematoma results either from a diastatic fracture of the squamous portion of the occipital bone or a tear in the transverse sinus. They expand rapidly, result in a massive collection of blood in the epidural space and obliteration of the cisterna magna, and cause (1) compression 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.

A



B



C

Figure 13.7. Acute subdural hematoma. The acute subdural hematoma is characterized by acute cerebral edema, diffuse cerebral contusion, subarachnoid bleeding, subdural collections of blood varying in thickness from one to approximately four mm. This compound cranial cerebral injury is often associated with dural lacerations; linear, depressed, compound skull fractures; scalp lacerations or abrasions. (A) This child suffered an acute subdural hematoma. One notes the freshly formed subdural clot (*1*), contusion of some of the gyri (*2*), flattening of all of the gyri. (B) This is an autopsy specimen of a young child with compound cranial cerebral injury resulting in acute subdural hematomas. One notes the epidural clots (*1*) which were underlying skull fractures, almost transparent dura over the right hemisphere (*2*) which is expressive of a severe increase in intracranial pressure and cerebral edema, accumulations of subdural blood over the left frontal (*3*) and occipital (*4*) lobes, smaller collections over the right frontal (*5*) lobe. (C) The dura on the left has been opened, exposing the flattening clot (*1*) and contused gyri (*2*).

Subdural Hematoma

The subdural hematoma is really 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 (Figure 13.7)
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.

Subacute Subdural Hematoma (Figure 13.8)
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. CTT studies suffice to make the diagnosis, so that cerebral angiography is not indicated. 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

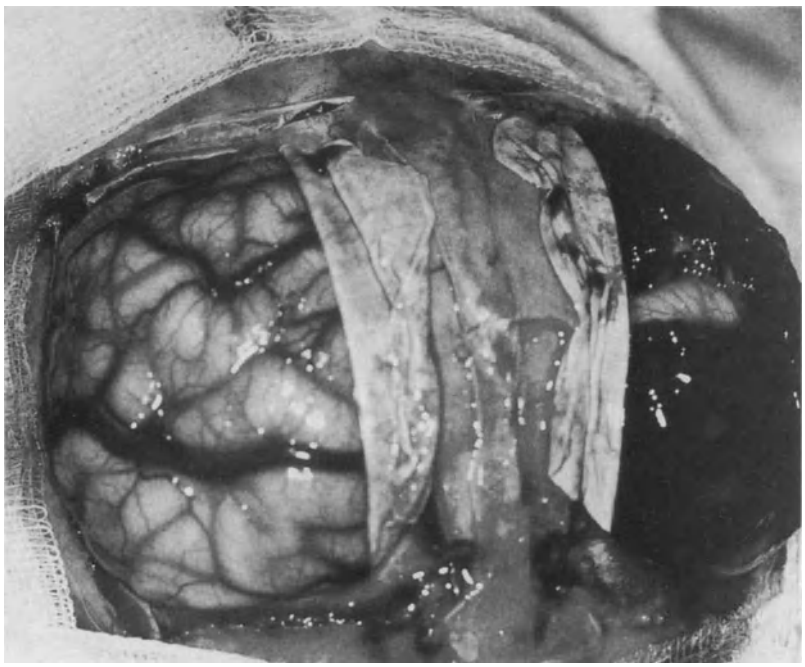


Figure 13.8. This is an operative photograph of a child with bilateral subacute subdural hematomas. The hematoma from the left has been removed and the one on the right exposed. In this child, the biparietal craniotomy was followed by simultaneous openings of the dura, so as to minimize chances of transfalcical 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.

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 structure. No drain is left in the subdural space, the dura is closed, and the bone flap is anchored into position.

Chronic Subdural Hematoma (Figures 13.9 to 13.12)
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 posttraumatic, 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 to be the result of its oncotic pressure. 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 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.

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 CTT scan or, now less commonly, cerebral arteriography. The CTT scan or cerebral arteriogram permit

1. the diagnosis of a chronic subdural hematoma;
2. an evaluation of the location and extent of the subdural hematoma; and
3. information concerning the degree of secondary cerebral atrophy and/or ventricular enlargement.

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[1] and then Gilles[2], independently, recommended subdural taps as a useful method of diagnosis and treatment. In 1933 Peet and Kahn[3] performed

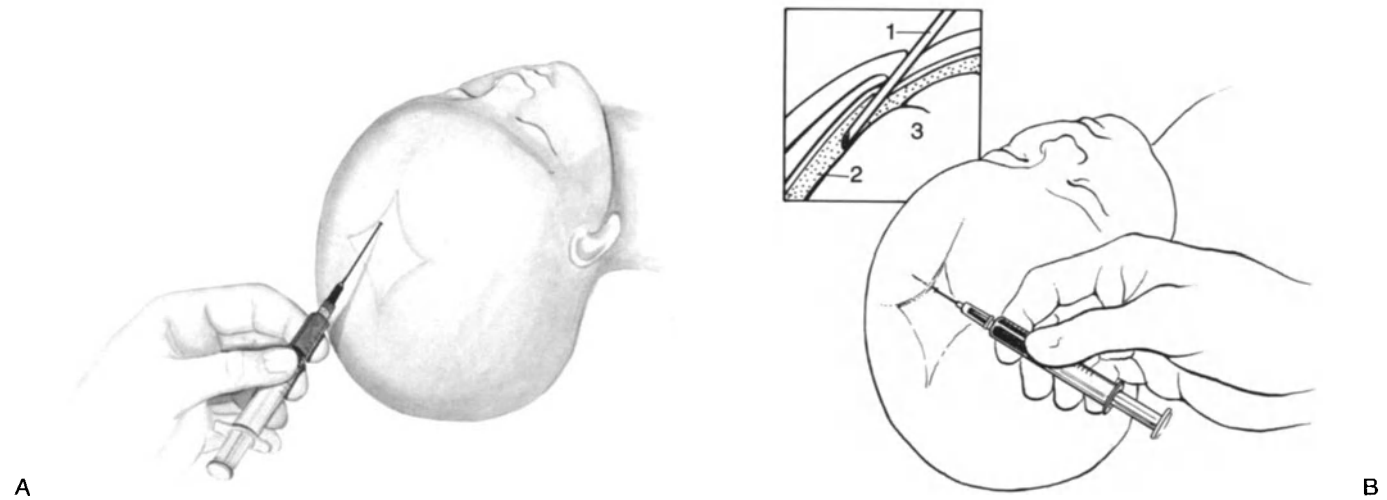


Figure 13.9. (A) Incorrect technique for performing subdural taps: the needle is directed perpendicular to the fontanel, a direction which increases risks of penetrating the brain or damaging a cortical bridging vein. (B) The correct technique: the needle is inserted through the skin and beneath the dura, at an oblique angle, so as to minimize risks of penetrating the brain. It is directed away from the cortical bridging veins. The inset shows the desired position of needle tip (*1*) in the subdural space (*2*), not entering brain (*3*)

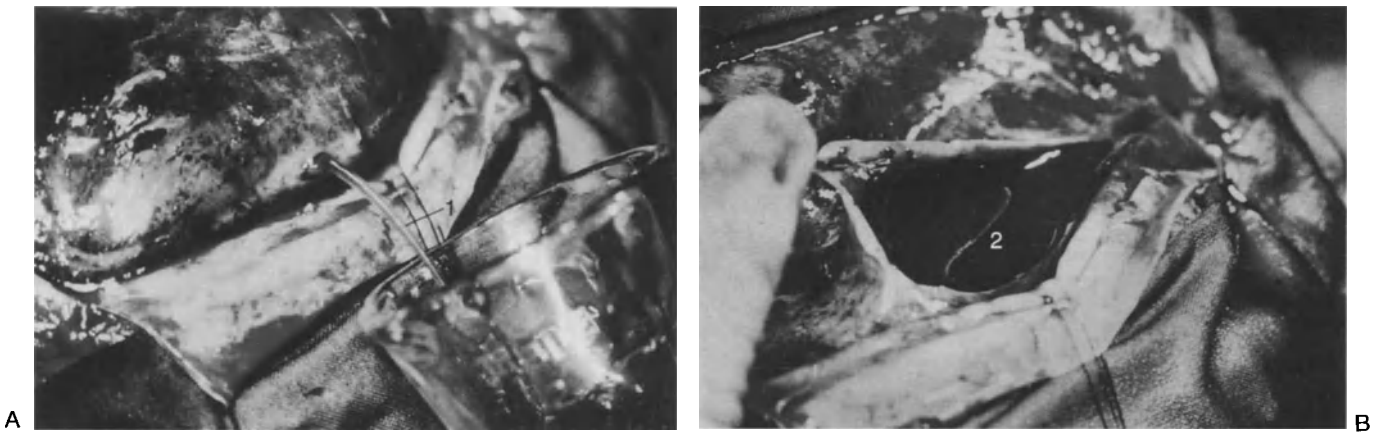


Figure 13.10. (A) The chronic subdural hematoma is subdural clot in varying stages of liquefaction from xanthochromic fluid (*1*), to (B) chocolate-colored fluid (*2*), both from the same child. After the xanthochromic fluid was drained, the dura was opened.

subdural taps to distinguish 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, burr 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 the author's opinion, both ineffectual and dangerous. The original concept that the 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.

The lack of understanding of the physiopathology of persistent collections of fluid in the subdural spaces is the reason for failure to find a successful mode of treatment.

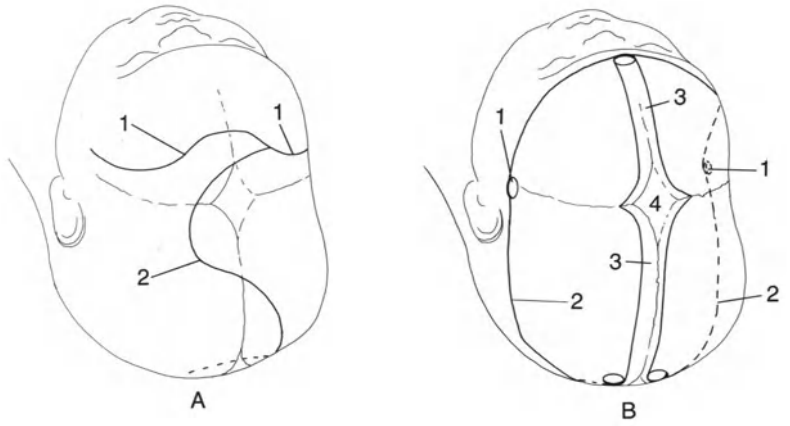


Figure 13.11. (A) The bifrontal (*1*) and parasagittal S-shaped (*2*) incisions are herein illustrated. (B) The burr holes (*1*) and craniotomy margins (*2*) are indicated. Note that a sagittal strip of bone (*3*) is left on either side of the sagittal suture and that the craniotomy margin skirts along the borders of the anterior fontanel (*4*).

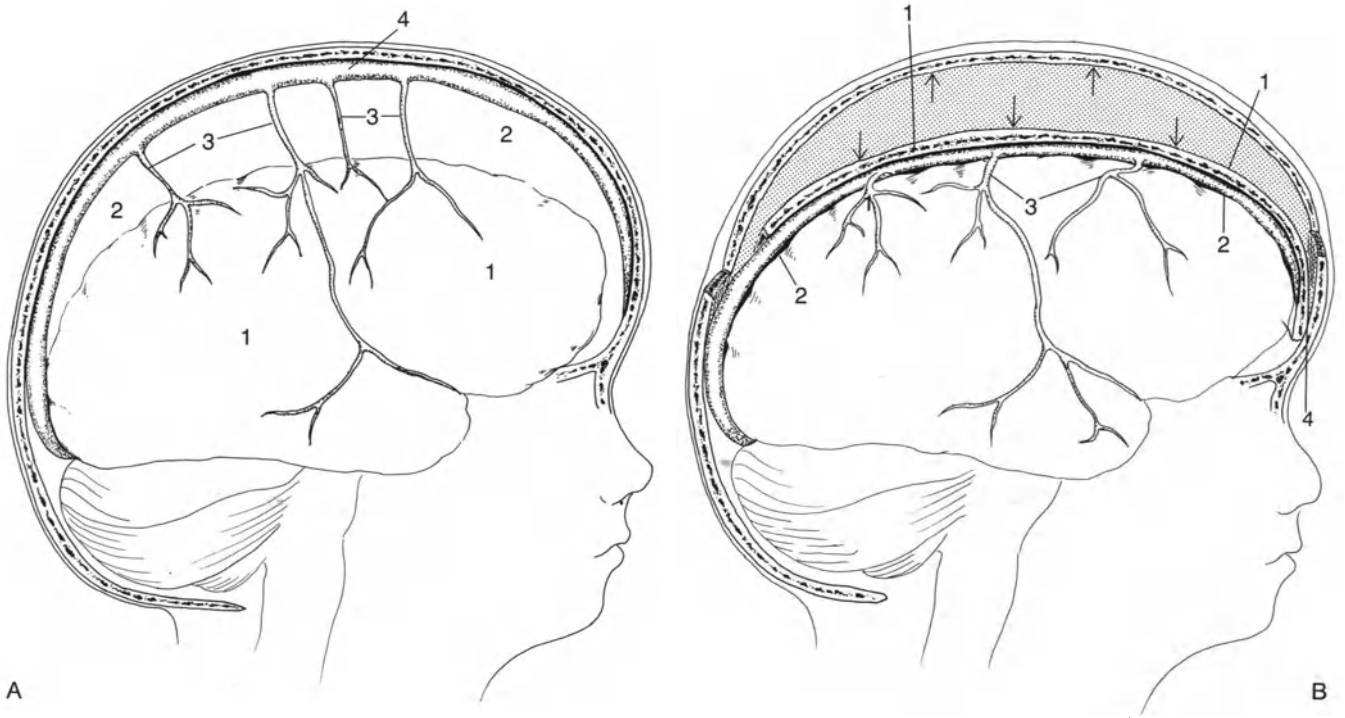


Figure 13.12. (A) This drawing represents schematically the diminution in volume of the cerebral hemisphere (*1*), the large subdural collection of fluid (*2*) within which one may identify the "hanging veins" (*3*) as they bridge across the subdural space to enter the superior sagittal sinus (*4*). The lowering of the SSS was designed to bring surgically the bridging veins into normal anatomic 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).

Pathogenesis of Chronic Subdural Hematoma

The etiological factors that contribute to chronicity of a subdural fluid in children remain obscure. I postulate that a possible mechanism is the progressive stretching and narrowing of the cortical veins that bridge the subarachnoid and subdural spaces to enter the sagittal sinus. This 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 leads to thrombosis of the "hanging veins." Narrowing and angulation of these veins results in elevated back pressure, favoring the formation of a transudate. The "hanging veins" likely diminish venous drainage into the superior sagittal sinus, triggering vasogenic brain edema. This would explain 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, the *lowering of the superior sagittal sinus* onto the cerebral hemispheres, bringing it to rest in its normal anatomic 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 intracranial pressure, characterized either by progressive enlargement of the head, bulging fontanel, splitting sutures, and/or progressive psychomotor retardation, with radiological evidence of progressive craniocerebral disproportion and characteristic cortical "hanging veins" are indications for surgery.

Operative Technique for Lowering the Superior Sagittal Sinus (SSS) (Figures 13.13 to 13.17)

The purpose of the procedure 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 bring it to fit the brain as snugly as possible, obliterating the subdural space and giving a normal course to the cortical bridging veins.

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 the subdural membranes.

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 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. Two Penrose drains are left in the epidural space for 24 hours.

Follow-up angiograms 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]

Burr Holes

In the very early stages of subdural fluid formation, simple burr holes (bilateral frontal) may suffice as a curative procedure. However, it is not uncommon for the fluid to reform, for the skin over the burr holes to bulge. Repeat drainage is attempted through the same burr holes, with or without insertion of drains into the subdural space. If the fluid reforms, one should proceed to angiography to learn whether "hanging veins" are present and, if so, plan to lower the superior

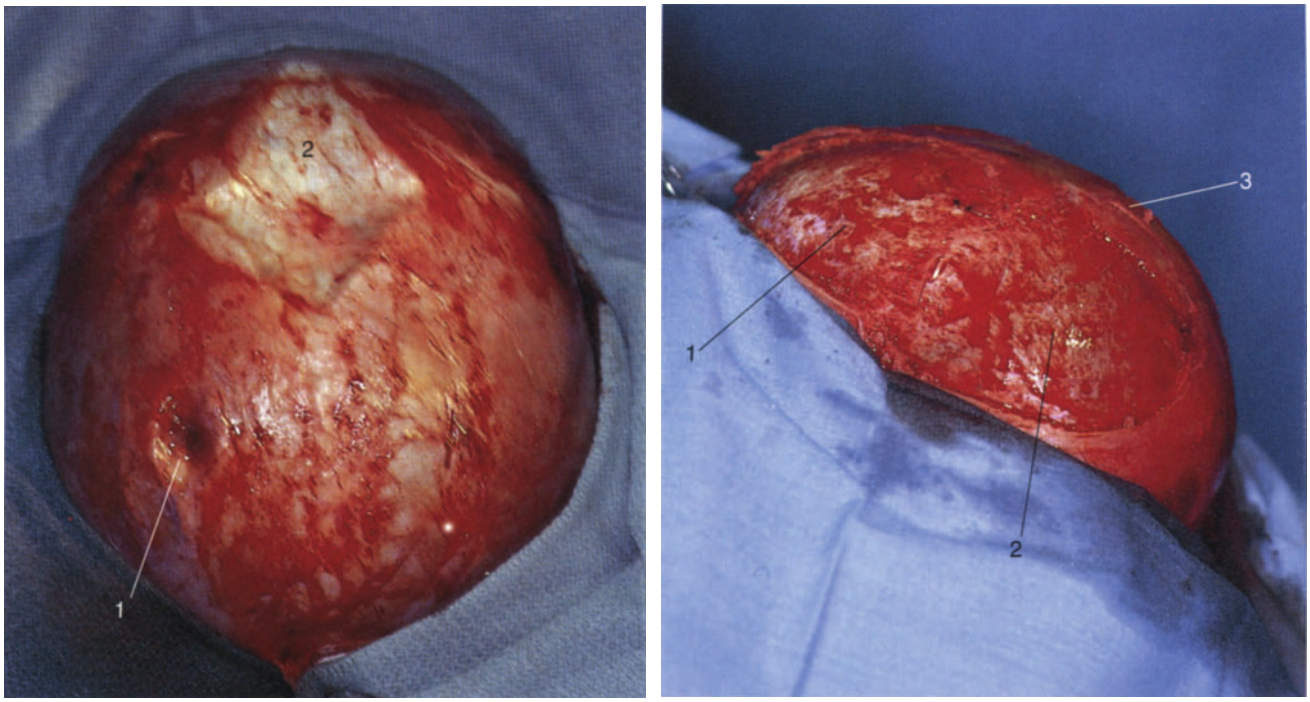


Figure 13.13. (A) After the scalp flap has been reflected, the burr holes (*1*) are made preparatory to lifting the bone flaps. Note the very large anterior fontanel (*2*), which is, paradoxically, sunken. In fact, it is not unusual for children with chronic subdural collections of fluid to suffer progressive increase in head size and to have a sunken fontanel. (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.

sagittal sinus as soon as possible. Some authors (K. Sato and S. Ishii) have told me that external subdural drainage, over prolonged periods of time, has been successful in curing the subdural and in permitting the brain to maintain its normal volume.

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.

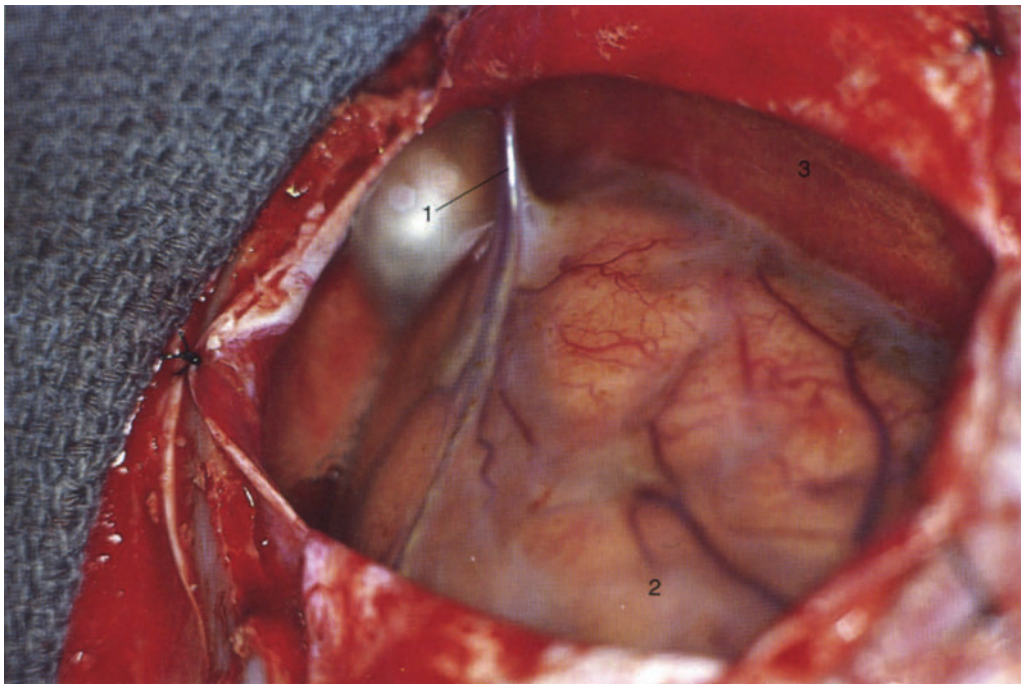
Membrane Resection

Resection of the membranes, which form over both the dura mater and the arachnoid, does not cure the child. The subdural fluid reforms within hours of bilateral frontoparietal craniotomy and extensive resection of the membranes, and soon thereafter proteinacious membranes reform.

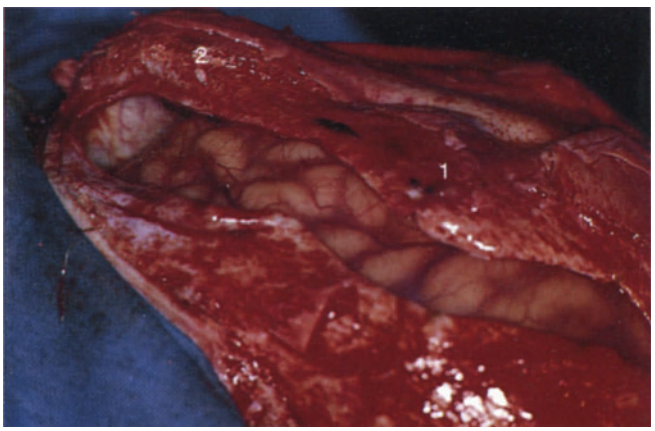
Cerebrospinal Fluid Leaks

Leakage of cerebrospinal fluid (CSF) into the periorbital tissue, directly through fractures of the mastoid, into the subGaleal space, are 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 petrous bone, the lateral surface of the body of the sphenoid, 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

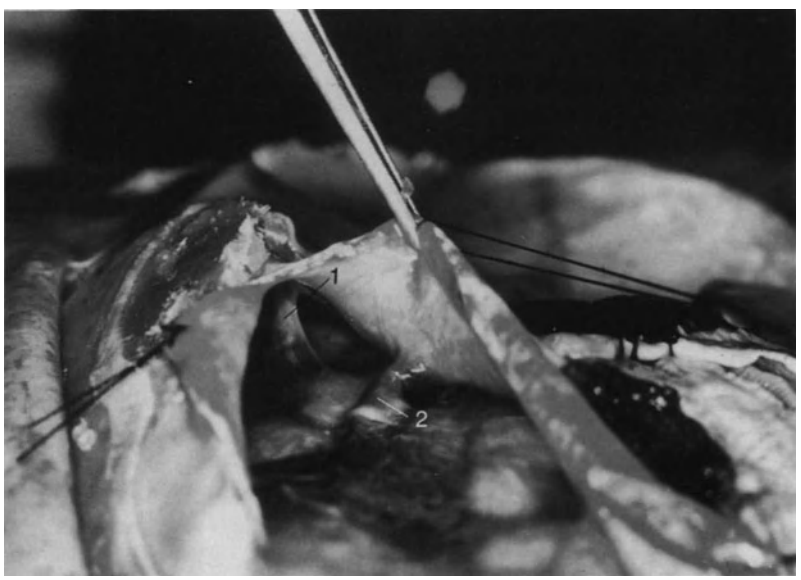


A

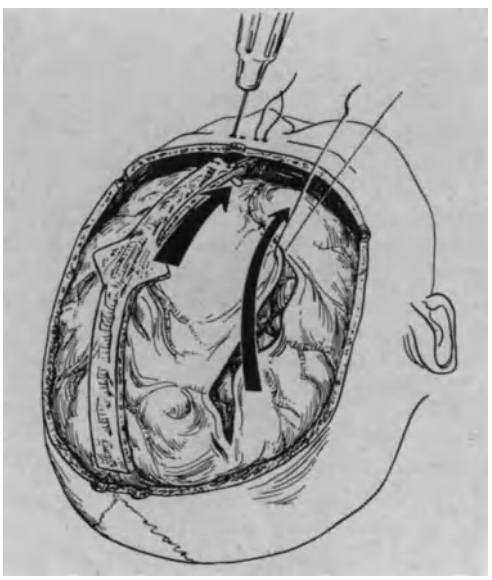


B

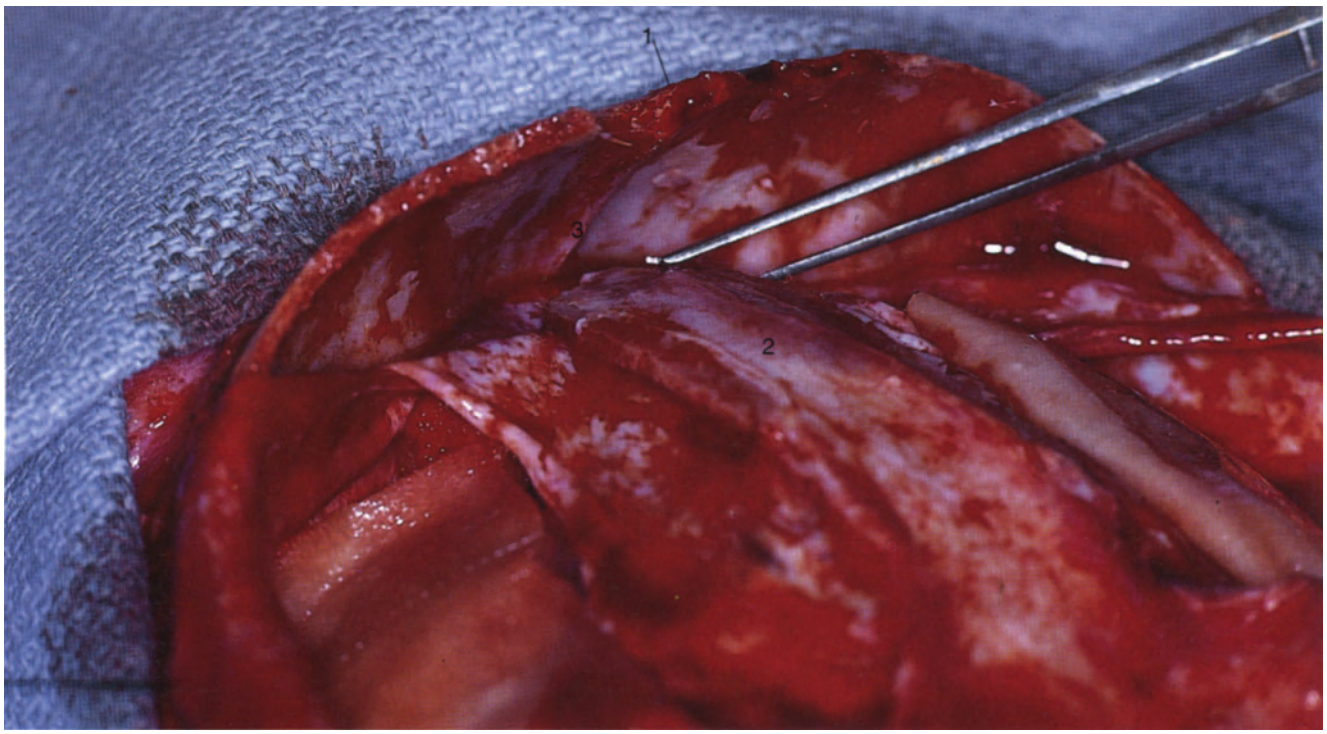
Figure 13.14. (A) This photograph puts into relief the "hanging veins" (*1*), the opalescent outer subdural membrane (*2*), and the falx cerebri (*3*). (B) The dura has been opened bilaterally, and the sagittal strut of bone over the metopic suture (*1*) freed from the glabella (*2*), allowing the superior 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 almost normal course of a bridging vein (*2*) posteriorly, where the dura and 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.



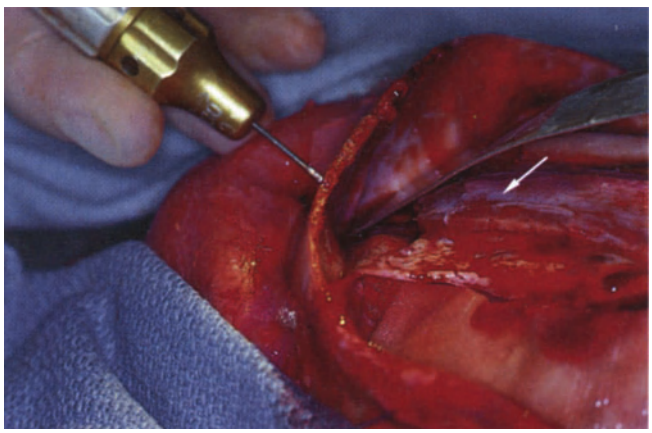
C



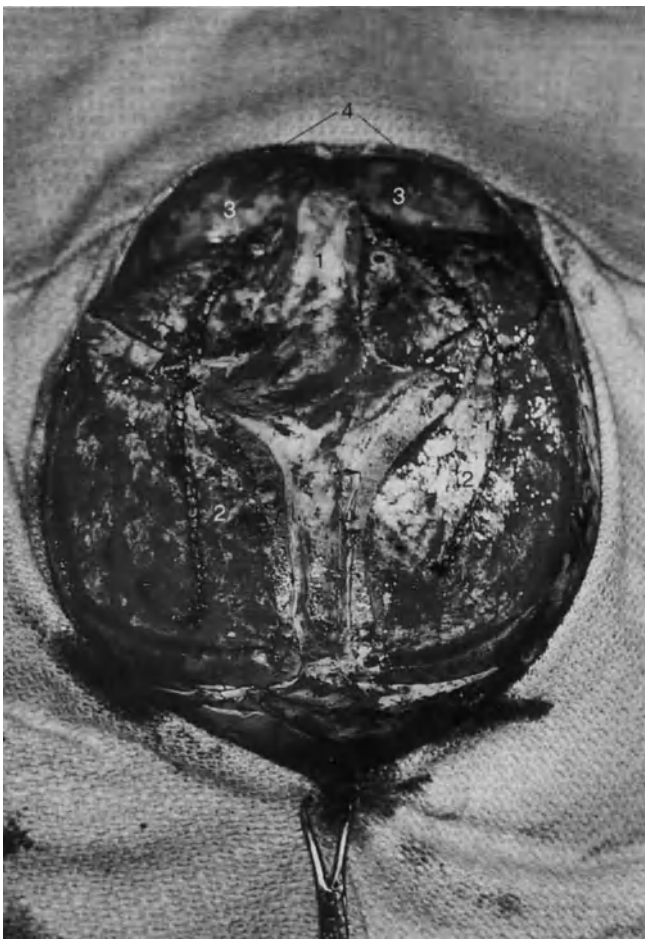
D



A



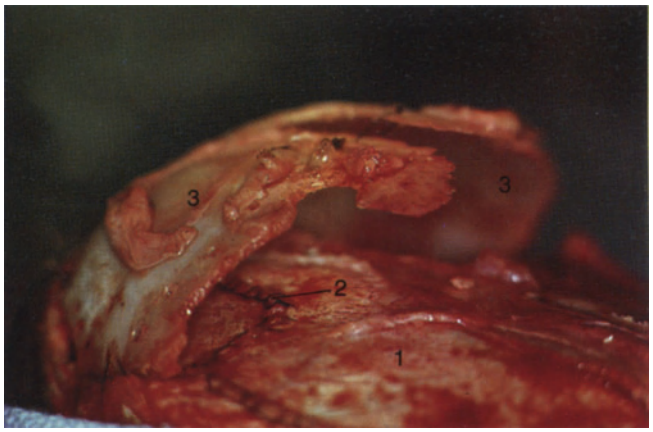
B



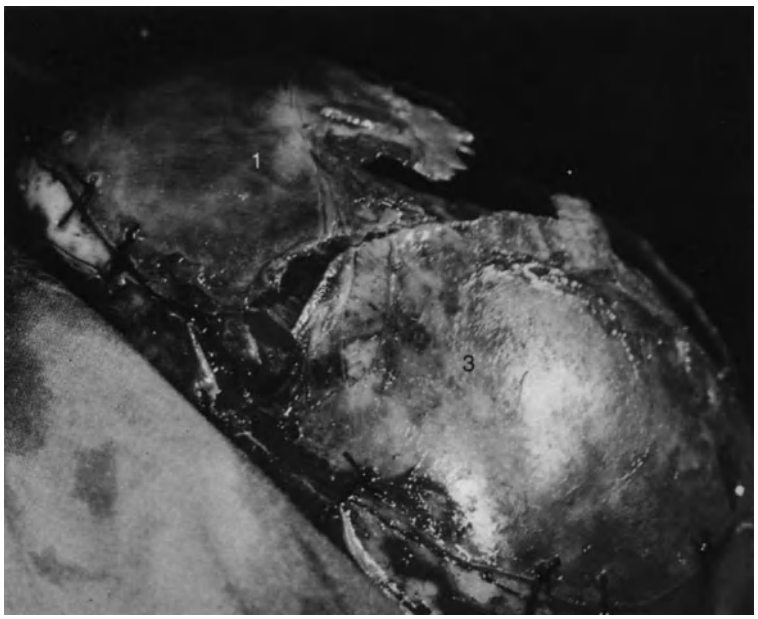
C

Figure 13.15. 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 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*). (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 moulding 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 surface 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 the 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 a Penrose drain left in the subGaleal space. Note that the contour of the head is quite acceptable.

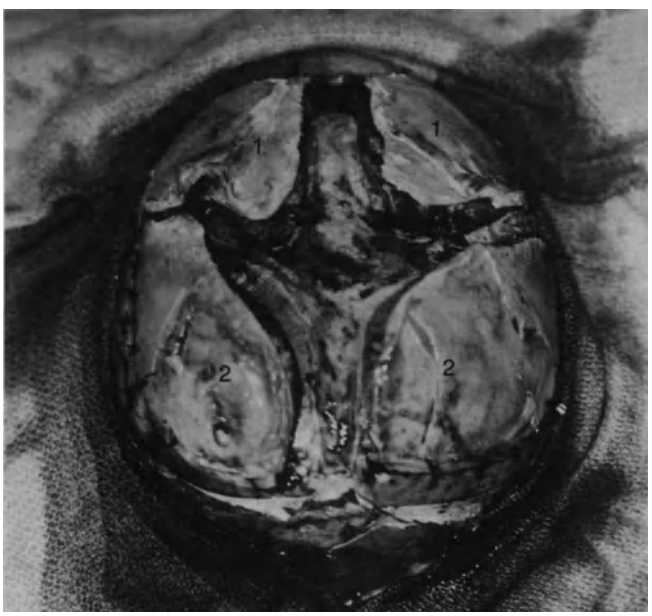


D

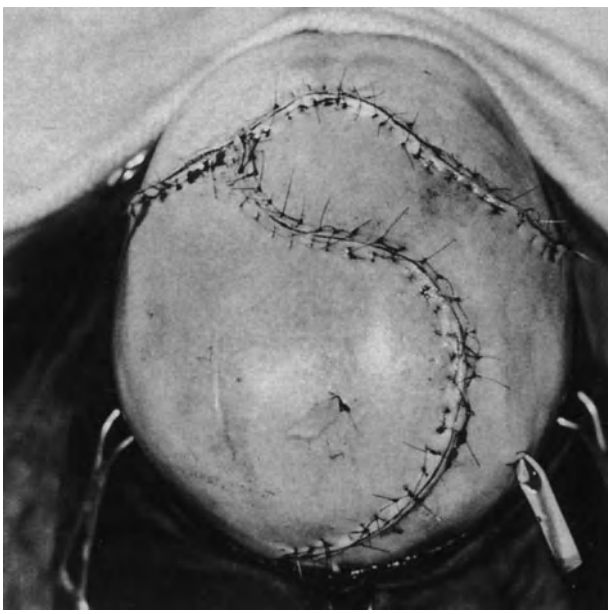


E

F



G


H

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 assurances 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 transphenoidal 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 are 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. The gliotic tissue is best removed with a laser, using 6 W of a defocused continuous wave beam. In the postoperative period, lumbar spinal drainage should be maintained for approximately two weeks, with the patient receiving prophylactic intravenous antibiotics. CTT scans will reveal whether parenchymal healing has occurred. If not, one may be obliged to insert a ventriculoperitoneal shunt.

Posttraumatic 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, long course of the cortical bridging veins within

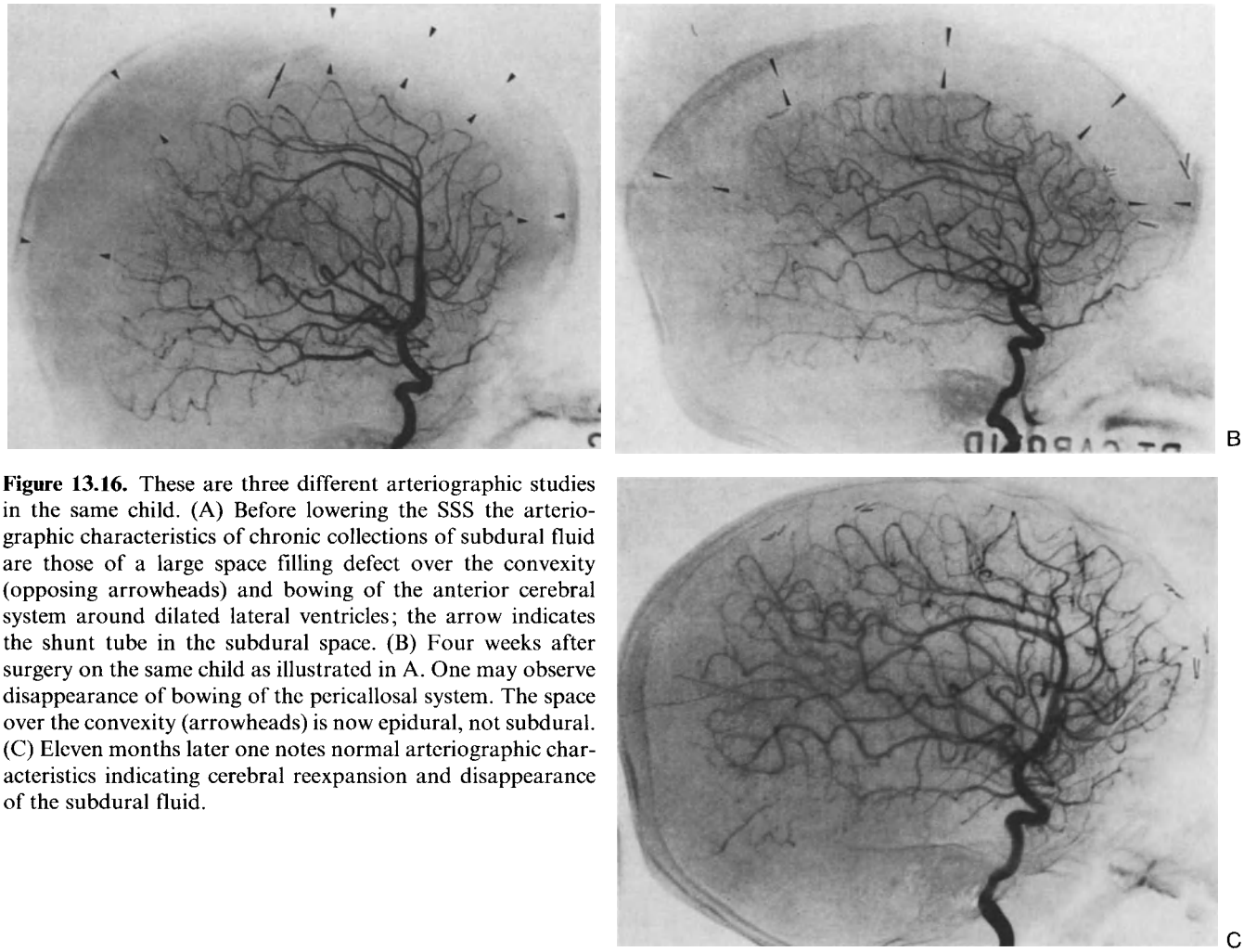


Figure 13.16. These are three different arteriographic studies in the same child. (A) Before lowering the SSS the arteriographic characteristics of chronic collections of subdural fluid are those of a large space filling defect over the convexity (opposing arrowheads) and bowing of the anterior cerebral system around dilated lateral ventricles; the arrow indicates the shunt tube in the subdural space. (B) Four weeks after surgery on the same child as illustrated in A. 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.

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 will not be considered. Because of the particular anatomical characteristics of the vault and base of the cranium, vascular injuries resulting from calvarial and basal structures will be 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 are not studied neuroradiologically with angiography, and those remaining in a 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.

The mechanics of vascular injury are the same as those for cerebral laceration—shearing forces, explosive forces, cavitation. These physical insults and stresses are indirect, resulting from different densities of individ-

ual 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 fontanels 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 Posttraumatic Cerebrovascular Injuries

The nature of injury varies almost directly with the age of the child: perinatal injuries are incurred by the fetus in his 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 his 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 two years of life are subject to being battered. 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 higher 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 neurologic dysfunction is identified. The so-called congenital giant 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, and so on 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 compaigns have been waged against use of the high chair and bassinet, so that this cause for injury has decreased remarkably. Unfortunately, despite the fact 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, he begins to fall regularly. Fortunately, the child instinctively falls by dropping to this buttock, so that head injuries from walking-falls are rare. However, the exploratory instincts of a toddler and his lack of coordination combine to render him particularly susceptible to falls from chairs, beds, tables, windows. Also, because of his low stature and total unawareness of danger, he suffers a high incidence of being struck by an automobile either in the streets in front of his home or, most tragic of all, while playing in the driveway of his own home. His high degree of mobility and rapid psychomotor development are responsible for his 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 his parents.

The first two years of life are those in which the child is particularly vulnerable to senseless beatings by his parents, siblings, 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 cause linear and eggshell skull fractures, explosive forces upon the membranous bones of the skull, 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 sagittal 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 to 10 mm.

Anatomy of Posttraumatic Cerebrovascular Injuries

So as both to put into relief the wide spectrum of vascular damages resulting from craniocerebral injury and

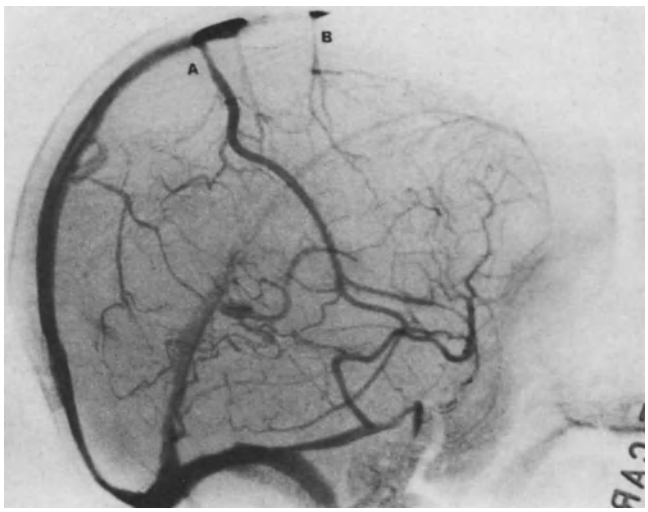
to present an anatomicoclinical correlation, two classifications will be presented: (1) anatomical compartments, and (2) bleeding sources.

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 extraordinarily 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 limit subperiosteal hematomas to the area immediately 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 from diploic bleeding.

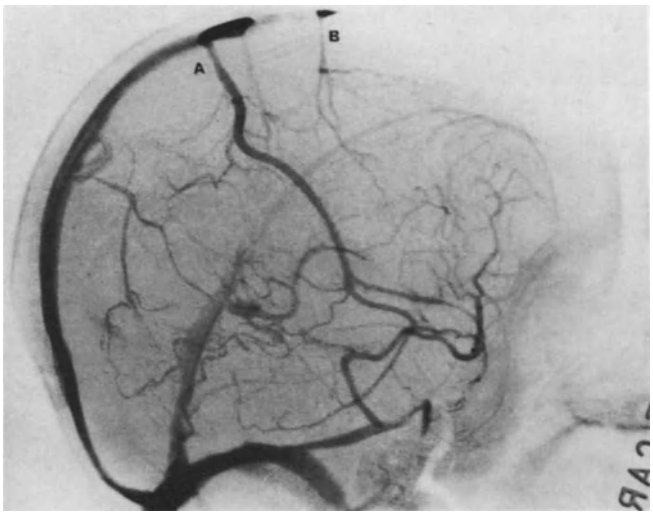
Collections of blood within the epidural space, with the exception of crushing and explosive injuries, do not cross the suture lines, since the outer layer of the dura, the suture, and periosteum are an anatomical continuum between membranous bones. Tears in the middle meningeal artery (anterior, middle, posterior branches) do occur during the first two 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, newborn 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 extent 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 it posteriorly (hairpin style) upon itself 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. Knowing that the child was injured in an automobile accident, either struck by or within the vehicle, suggests a deceleration injury, whereas battered children suffer acceleration injuries. 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).

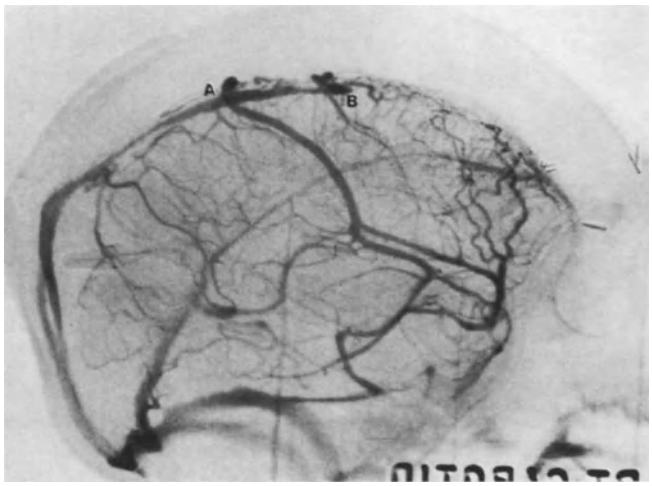
Subarachnoid bleeding is an expression of either regional cerebral contusion or, very rarely, torn arteries. The regional cerebral contusion results either from deceleration injuries, or contre-coup acceleration injuries that damage cortical vessels. Shearing and stretching forces may rent or tear the 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 aneurysms, over time, increase dramatically in size, developing into giant aneurysms or aneurysmal tumors. I doubt that the reported "congenital" aneurysms of



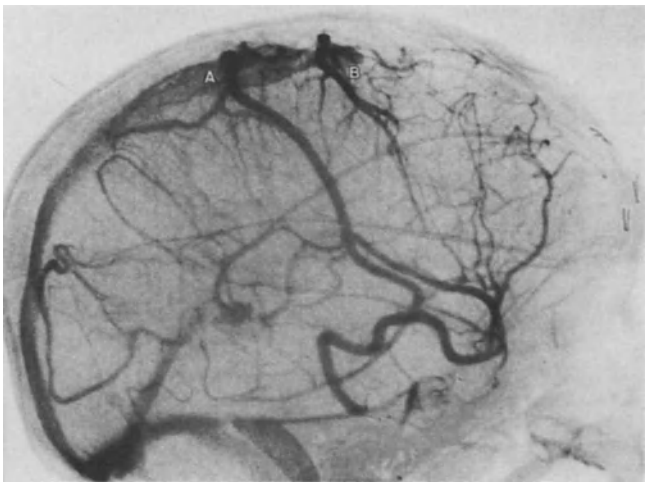
A



B



C



D

Figure 13.17. These are the venographic studies of the same child as shown in Figure 13.16. They reveal the venous characteristics of preoperative progression in volume of the subdural collection of fluid and postoperative return to normal. (A) One notes the subdural space-filling defect and the "hanging veins." It is of value for the reader to follow these hanging veins (labeled *A* and *B*) through this figure and the subsequent three. (B) 3 weeks after the study illustrated in A, one notes progression in size of the subdural collection of fluid and further stretching of the hanging veins. (C) 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. (D) This study was performed 11 months postoperatively. One notes an almost entirely normal venographic pattern.

the vertebral, posterior inferior cerebellar, and middle cerebral systems are anything other than posttraumatic (false) aneurysms. The fact that these aneurysms are most common on the vertebral basilar system is in support of their posttraumatic 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. Posttraumatic 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. Posttraumatic occlusion of the anteromedial and anterolateral perfor-

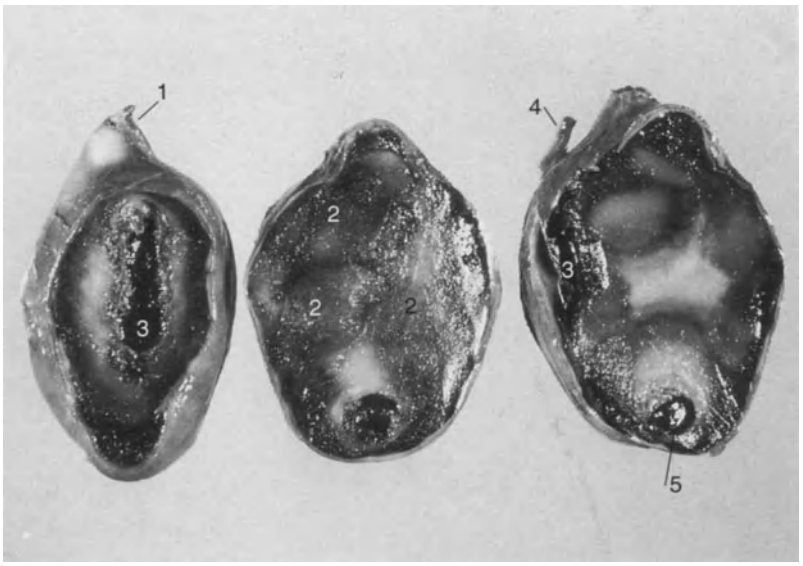


Figure 13.18. Anatomical specimen of a false aneurysm, the most common aneurysm of childhood. In essence, the false aneurysm is the fibrocollagenous capsule tamponading a rent in a vessel, but with a lumen at the center. One sees the parent vessel entering the false aneurysm (*1*), the lumen of the false 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.

ating systems, independent of occlusion of the internal carotid may very well occur. However, it has not been reported and I have never documented a case. Similarly, posttraumatic 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, whether hemorrhagic or occlusive, 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 very 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, interhemispheral-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 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, posttraumatic 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 invariably increase in size and cause brainstem compression or hemispheral 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 (Figures 13.18 to 13.22)

"There are three types of intracranial aneurysms: (1) mycotic, (2) arteriosclerotic, and (3) congenital. Perhaps a fourth or syphilitic group should be included..." (W. E. Dandy, 1944).[11]

During the past two score years, intracranial aneurysms have been the subject of rather intense study from therapeutic, statistical, and technical points of view.[12–18] The pathological characteristics of these an-

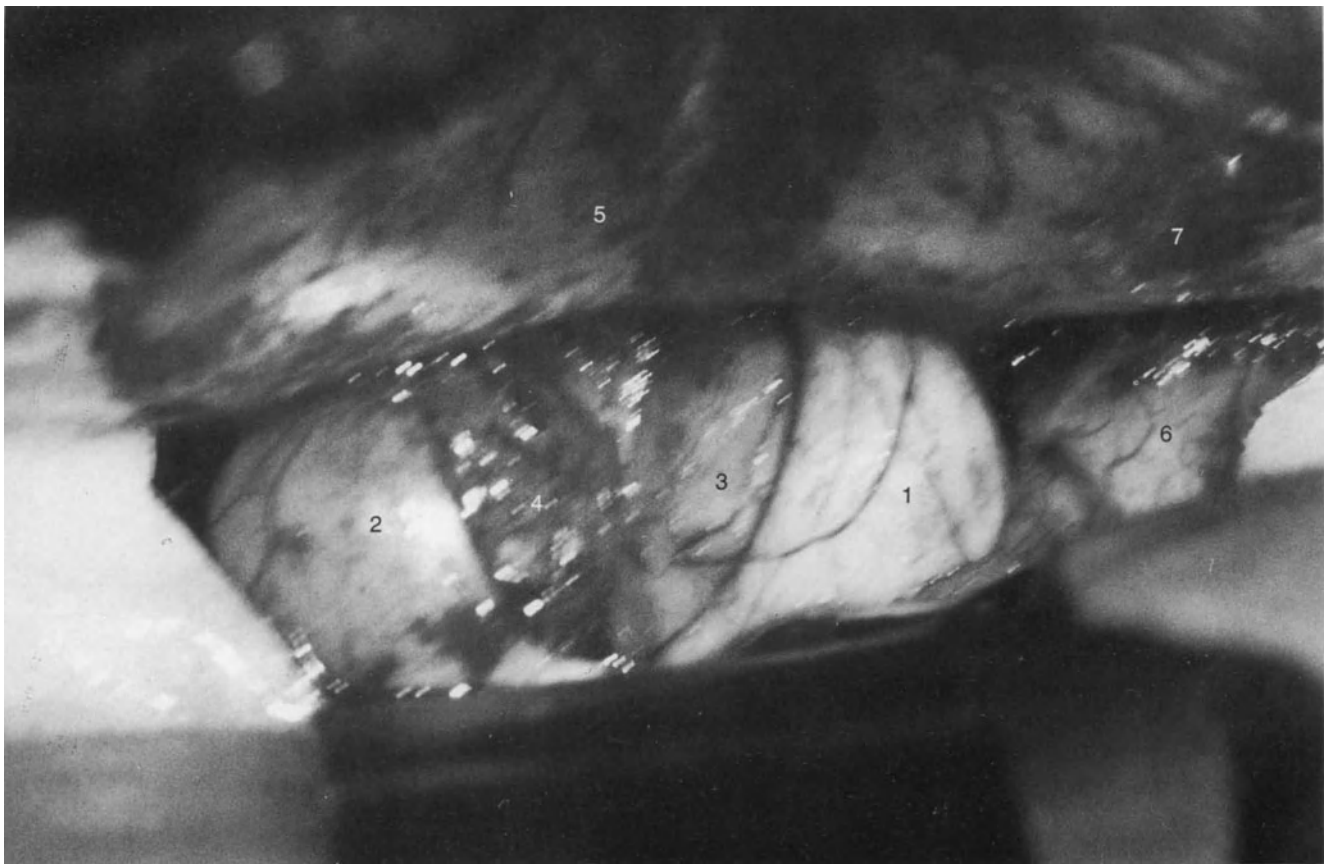


Figure 13.19. Operative photograph of bilateral internal carotid artery false aneurysms elevating and flattening the optic nerves. The right optic nerve (*1*) is more flattened than the left (*2*), and the right internal carotid false aneurysm (*3*) appears to be displaced laterally by a slightly larger left false aneurysm (*4*) which has 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.

eurysms were investigated morphologically,[12,19-24] biologically,[25,26] and hydrodynamically.[27,28] 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,[29] the extradural/intracranial portion of the internal carotid artery,[30,31] and intradural branches of the internal carotid artery, secondary to surgical manipulation,[32,33] or in association with recurrent meningiomas,[34] have been reported. McKissock reported on the recurrence of an aneurysm after he had resected it.[35] Already recognized aneurysms have been observed to increase in size over varying periods of time, with and without repeated subarachnoid bleeds.[36,37]

With the steady increase in direct surgical approach to intracranial aneurysms of the berry variety, and with the advent of 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.[32,38-42] 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 carotid 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[30] or carotid occlusion (something I have observed, and illustrated here in Figure 13.19). A variety of traumatic aneurysm has been described[43] in which an injury, generally a fracture of the base of the skull, has caused late hemorrhage and death. Birley and Trotter[31] reported a case of "trau-

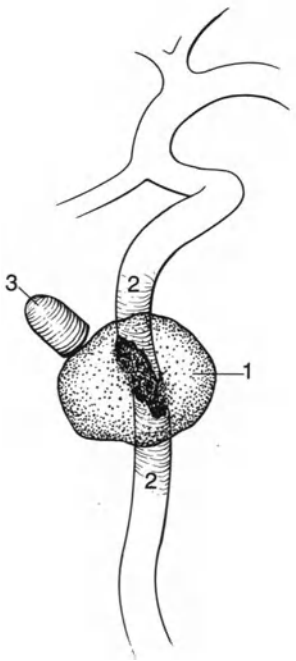


Figure 13.20. This a composite drawing of the operative findings in a patient with a posttraumatic false aneurysm (gunshot wound), demonstrating the fresh clot (*1*) encircling both severed ends of the internal carotid artery (*2*) and thereby permitting the passage of blood across the intervening space. One bullet (*3*) caused the tear.

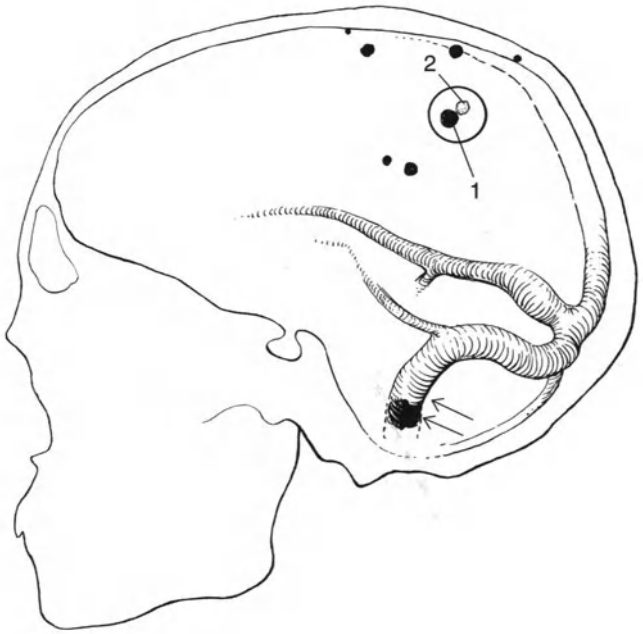


Figure 13.21. Composite drawing of a venous phase angiogram in a previously reported child who had suffered a buckshot wound to the head. The black dots indicate the pellets, (*1*) indicates a pellet which damaged a cortical artery, (*2*) indicates persistent filling of a cortical false aneurysm, and the arrows indicate the posttraumatic occlusion of the jugular bulb.

matic" aneurysm of the internal carotid (intracranial/extradural) artery, and Goald and Ronderos[44] reported the formation of a traumatic aneurysm on the intracavernous portion of the internal carotid secondary to perforation by a steel spring.

Finkemeyer[32] 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[45] described the formation of a "posttraumatic 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.[46–49]

Cressman and Hayes,[43] 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[37] 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,[42] reporting on the anatomy and angiography of vascular malformation, 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 continuous 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[36], reporting on 700 intracranial aneurysms, cited 18 cases which had repeat angiography after an interval ranging from two 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,[50] 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 stratifica-

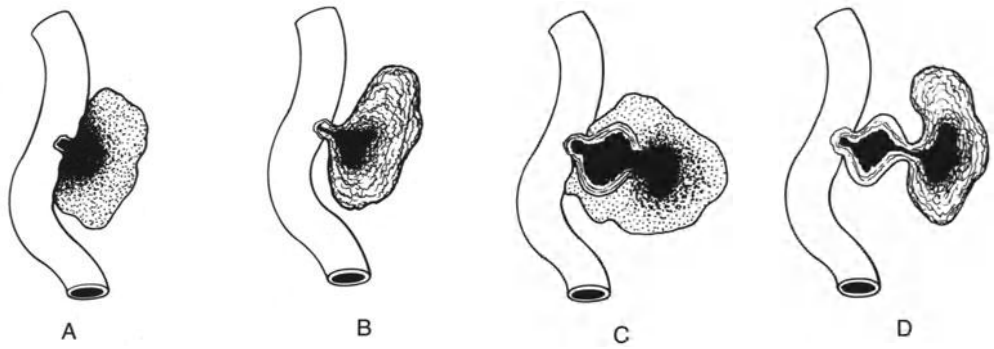


Figure 13.22. (A) This schematic drawing depicts a rent in an artery tamponaded by freshly formed clot. This is an *acute posttraumatic false aneurysm.* (B) The rent in the parent artery is tamponaded by fibrocollagenous tissue, which resulted from consolidation of the hematoma. This is a *chronic posttraumatic 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 aneursym 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.

tions 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.[51,52] 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 adoped.[11,16,45,51,53,54]

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.[36,55] We have had cases of a false aneurysm developing secondary to bleeding from a berry aneurysm and culminating in the formation of a true giant aneurysm, aneurysmal tumor.[56].

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. 13.22A).
   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. 13.22B).
2. Spontaneous
   a. Acute: consists of a ruptured berry, or arteriosclerotic, aneurysm and fresh hematoma tamponading the breach (Fig. 13.22C).
   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. 13.22D).
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. 13.18).

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 nor a fundus he may coat with plastic. 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.

One should make a distinction between false aneurysms and dissecting aneurysms[46,57,58] of the cerebral vessels. In dissecting aneurysms, almost always due to arteriosclerosis,[59-61] the intima is stripped from the media by a hemorrhage, with the resultant formation of a septum and a blind fundus. At times the dissection may progress to such an extent as to occlude the vessel. Poppen[18] reported on the surgical treatment of two of these.

Angiography does, to a reasonable degree of accuracy, permit the diagnosis of false aneurysm. The irregularity of outline of the aneurysmal surface, the slowness with which the aneurysmal cavity fills, and the presence of evidence of vascular deformity, suggestive of "hematoma" about the aneurysm, all make the possibility of a false aneurysm real indeed. The absence of vasospasm in the vicinity of the rent and the location of these "aneurysms" in sites where one normally does not encounter atherosclerotic or berry aneurysms serve as supportive evidence for the presence of a false aneurysm.

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[32] 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 and Raskind[62] 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[47] in their "iatrogenic cerebral cortical aneurysm" is strong supportive evidence that trauma may produce false aneurysms. The work of White and colleagues[63] offers an experimental basis for this conclusion.

Cranioplasty (Figure 13.23)

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 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 computed transmission tomography or nuclear magnetic resonance 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.

A

Figure 13.23. 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 moulded into the desired contour and cut the desired length prior to inserting them into the previously prepared pouch (*2*). (C) After all of 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. ►

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 a foreign substance.

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, 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 newborn, 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.

B

C

References

1. Henschen K: Diagnostik und Operation der traumatischen Subduraltung. Arch Klin Chir 99:67, 1912.
2. Gilles R: Deux observations d'hémorragie méningée chez le nouveau; guérison après ponction décompression dans l'outre; status ulter deux petits malades et considerations thérapeutiques. Rev Mens Gynecol Obstet Pediatr 7:465, 1912.
3. Peet MM, Kahn EA: Subdural hematoma in infants. JAMA 98:1851–1956, 1932.
4. Salmon JH: Subdural hematoma in infants. Clin Pediatr 10:597–9, 1971.
5. Ingraham FD, Matson DD: Subdural hematoma in infancy. J Pediatr 24:3–37, 1944.
6. Ransohoff J: Chronic subdural hematomas treated by subdural pleural shunt. Pediatrics 20:561–4, 1957.
7. Shulman K, Ransohoff J: Subdural hematomas in children. The fate of the child with retained membranes. J Neurosurg 18:175–80, 1961.
8. Collins WF, Pucci GL: Peritoneal drainage of subdural hematomas in infants. J Pediatr 58:482–5, 1961.
9. McLaurin RL, et al: Results of non-operative treatment in 15 cases of subdural hematoma. J Neurosurg 34:753–9, 1971.
10. Gutierrez FA, McLone DG, Raimondi AJ: Physiopathology and a new treatment of chronic subdural hematoma in children. Child's Brain 5:216–32, 1979.
11. Dandy WE: Intracranial Arterial Aneurysms. Ithaca, NY, Comstock Publishing, 1944, p 147.
12. Alpers BJ: Aneurysms of the circle of Willis: Morphological and clinical considerations. In Intracranial Aneurysms and Subarachnoid Hemorrhage. Springfield, IL, Charles C Thomas, 1965.
13. McCromich WF, Nofzinger JD: Saccular intracranial aneurysms. An autopsy study. J Neurosurg 22:155–9, 1965.
14. McDonald CA, Korb M: Intracranial aneurysms. Arch Neurol Psychiatry 42:298–328, 1939.
15. Pool JL, Potts DB: Aneurysms and Arteriovenous Anomalies of the Brain. New York, Harper and Row, 1965, p 463.
16. Housepian EM, Pool JL: A systematic analysis of intracranial aneurysms from the autopsy file of the Presbyterian Hospital. J Neuropathol Exp Neurol 17:409–23, 1958.
17. Hamby WB: Intracranial Aneurysms. Springfield, IL, Charles C Thomas 1952, p 564.
18. Poppen JL: Specific treatment of intracranial aneurysms. Experiences with 143 surgically treated patients. J Neurosurg 8:75–102, 1951.
19 Carmichael R: The pathogenesis of non-inflammatory cerebral aneurysms. J Pathol Bacteriol 62:1–19, 1950.
20. Forster FM, Alpers BJ: Anatomical defects and pathological changes in congenital cerebral aneurysms. J Neuropathol Exp Neurol 4:146–54, 1945.
21. Stehbens WE: Medial defects of the cerebral arteries of man. J Pathol Bacteriol 78:179–85, 1959.
22. Walker AE: The pathology and pathogenesis of cerebral aneurysms. J Neuropathol Exp Neurol 13:248–59, 1954.
23. Walker AE, Allégre GE: Histopathologie et pathogénic des aneurysmes artériels cérébraux. Rev Neurol 89:477–90, 1953.
24. Wilson G, Riggs HE, Rupp C: The pathologic anatomy of ruptured cerebral aneurysms. J Neurosurg 11:128–34, 1954.
25. Nystrom SHM: Development of intracranial aneurysms as revealed by electron microscopy. J Neurosurg 20:329–37, 1963.
26. Sawyer PN, Pate JW: Bioelectric phenomena as an etiologic factor in intravascular thrombosis. Am J Physiol 175:103–6, 1943.
27. Odom GL, Woodhall B, Tindall GT, Jackson JR: Changes in distal intravascular pressure and size of intracranial aneurysm following common carotid ligation. J Neurosurg 19:41–50, 1962.
28. Wright RL, Sweet WH: Intra-arterial pressure determination at the time of carotid occlusion. In Intracranial Aneurysms and Subarachnoid Hemorrhage. Springfield, IL, Charles C Thomas, 1956, p 324–47.
29. Kuhn RA, Kugler H: False aneurysms of middle meningeal artery. J Neurosurg 21:92–6, 1964.
30. Araki C, Handa H, Handa J, Yashida K: Traumatic aneurysm of the intracranial extradural portion of the internal carotid artery. J Neurosurg 23:64–7, 1965.
31. Birley JL, Trotter W: Traumatic aneurysm of the intracranial portion of the internal carotid artery. Brain 51:184–208, 1928.
32. Finkemeyer H: Ein säckchenförmiges Aneurysma der A. cerebri media als postoperative Komplikation. F bl Neurochir 15:302–5, 1955.
33. Taylor PE: Delayed post-operative hemorrhage from intracranial aneurysm after craniotomy for tumor. Neurology 11:225–31, 1961.
34. Raskind R: An intracranial arterial aneurysm associated with a recurrent meningioma. J Neurosurg 23:622–5, 1965.
35. McKissock W: Recurrence of an intracranial aneurysm after excision. J Neurosurg 23:547–8, 1965.
36. Bjorkesten GA, Troupp H: Changes in the size of intracranial arterial aneurysms. J Neurosurg 19:583–88, 1962.
37. Sadik AR, Budzilovich GN, Schulman K: Giant aneurysm of middle cerebral artery. A case report. J Neurosurg 22:177–81, 1965.
38. Allen EV, Barker NW, Hines EA Jr: Peripheral Vascular Diseases. Philadelphia and London, Saunders, 1962, p 1044.
39. Crompton MR: The pathology of ruptured middle cerebral aneurysms. Lancet 2:421–5, 1962.
40. Fabian G: Traumatisches Aneurysma der Carotisinterna in der Keilbeinhöhle. H.N.D. 6:42–5, 1956.
41. Kinmonth JB, Rob CG, Simeone FA: Vascular Surgery. London, Edward Arnold, 1962, p 501.
42. Sugar O: Pathological anatomy and angiography of intracranial vascular anomalies. J Neurosurg 8:3–22, 1951.
43. Cressman MR, Hayes GJ: Traumatic aneurysm on the anterior choroidal artery. J Neurosurg 24:102–4, 1966.
44. Goald HJ, Ronderos A: Traumatic perforation of the intracranial portion of the internal carotid artery with eleven day survival. J Neurosurg 18:401–4, 1961.
45. Lunn GM: False cerebral aneurysm of the vertebral artery from remote missle injury of the neck. Br J Surg War 1:258–60, 1947.

46. Hirsch JR, David M, Sachs M: Les aneurysms artériels traumatiques intracraniens. Neurochirurgie 8:189–201, 1962.
47. Overton MC, Calvin TH Jr: Iatrogenic cerebral cortical aneurysm: Case Report. J Neurosurg 24:672–5, 1966.
48. Pouyanne H, Leman P, Got M, Gouaze A: Aneurysme artériel traumatique de la méningée moyenne gauche. Rupture un mois après l'accident. Hématome intracérébral temporal. Intervention. Neurochirurgie 5:311–5, 1959.
49. Vaughn BF: Middle meningeal haemorrhage demonstrated angiographically. Br J Radiol 32:493–4, 1959.
50. Taveras JM, Wood EH: Diagnostic Neuroradiology. Baltimore, Williams and Wilkins, 1960, p 1958.
51. Blackwood W, McMenemey WH, Meyer A, Norman RM, Russel DS: Greenfield's Neuropathology, Baltimore, Williams and Wilkins, 1963, p 679.
52. Paillas JE, Bonnal J, LaVieille J: Angiographic images of false aneurysmal sac caused by rupture of median meningeal artery in the course of traumatic extradural hematoma: Report of 3 cases. J Neurosurg 21:667–71, 1964.
53. De Takats G: Vascular Surgery. Philadelphia and London, Saunders, 1959, p 726.
54. Krauland W: Zur Entstehung traumatischer Aneurysmen der Schlagadern am Hirngrund. Schweiz Z Pathol Bakteriol 12:113–27, 1949.
55. Jane JA: A large aneurysm of the posterior inferior cerebellar artery in one-year-old child. J Neurosurg 18:245–7, 1961.
56. Raimond AJ, Yashon D, Reyes C, Yarzagaray L: Intracranial false aneurysms. Neurochirurgia 11:219–33, 1968.
57. Glynn Le: Medial defects in the circle of Willis and their relation to aneurysm formation. J Pathol Bacteriol 51:213–22, 1940.
58. Norman RM, Urich H: Dissecting aneurysm of the middle cerebral artery as a cause of acute infantile hemiplegia. J Pathol Bacteriol 73:580–2, 1957.
59. Richardson JC, Hyland HH: Intracranial aneurysms. Medicine 20:1–83, 1941.
60. Scott GE, Neubuerger KT, Denst J: Dissecting aneurysms of intracranial arteries. Neurology 10:22–7, 1960.
61 Wolman L: Cerebral dissecting aneurysms. Brain 82:276–91, 1959.
62. Boldrey Eb: Cited by Raskind R: An intracranial arterial aneurysm associated with a recurrent meningioma. J Neurosurg 23:622–5, 1965.
63. White JC, Sayre GP, Whisnant JP: Experimental destruction of the media for the production of intracranial aneurysms. J Neurosurg 18:741–5, 1961.

Uncited References

Gutierrez FA, McLone DG, Raimondi AJ: Epidural hematomas in infancy and childhood. In Hoffman H, Epstein F, Raimondi (eds): Concepts in Pediatric Neurosurgery I. Basel, S Karger, 1981, p 188.
Gutierrez FA, Raimondi AJ: Acute subdural hematoma in infancy and childhood. Child's Brain 1:269–90, 1975.
Gutierrez FA, Raimondi AJ: Delayed onset of acute posttraumatic subdural effusion. Em J Dis Child 128:327–31, 1974.
Hahn Y, Raimondi AJ, McLone DG, Yamanouchi Y: Traumatic mechanisms of head injury in child abuse. Child's Brain 10:229–41, 1983.
Raimondi AJ: The neuroradiologic evaluation of craniocerebral injury in the newborn and infant. Minerva Pediatr 21:1251–63, 1969.
Raimondi AJ, Matsumoto S: Acute head injuries in infancy and childhood. Chicago Med School Q 25:60, 1965.

"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

Chapter 14

Congenital Anomalies

Congenital Anomalies Involving the Craniocerebrum and Craniocervical Junction

Synostotic Cranial Anomalies

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, invariably 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] though others[4] have found his 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 his own immunological responses and to establish his 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 before 3 months of age is much higher than those operated between 3 and 7 months of age.

Metopic (Including Frontonasal) Synostosis: Trigonocephaly (Figures 14.1 to 14.3)

Premature closure of the metopic suture results in a rather characteristic keellike 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, there

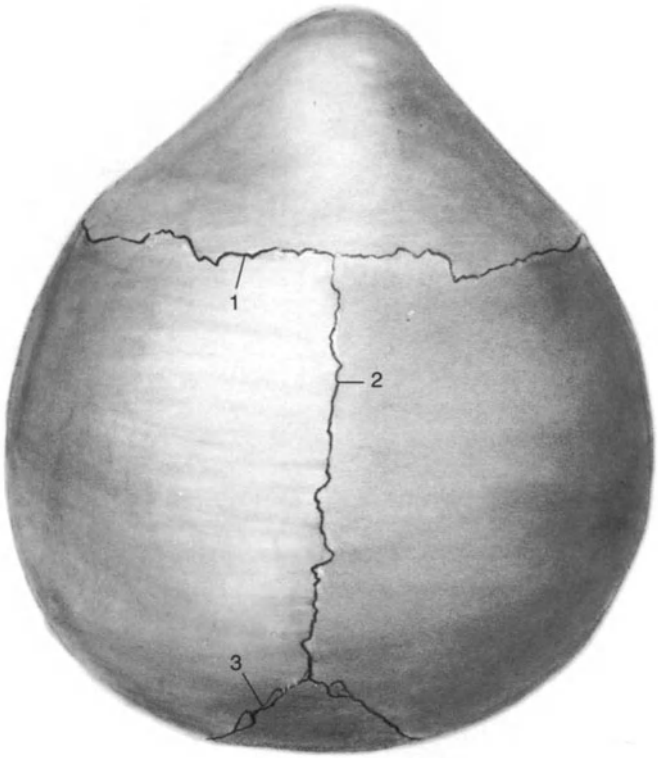


Figure 14.1. (A) Premature synostosis of the metopic suture (viewed form above). Though brain and skull growth in the longitudinal axis is much greater than normal, the resultant cramping of the frontal lobes displaces the brain posteriorly, across the line of the coronal suture (*1*). Because of functional sagittal (*2*) and lambdoidal (*3*) sutures, that portion of the skull posterior to the coronal suture expands circumferentially. This is shown diagrammatically in A. (B) A trigonocephalic skull after the scalp has been prepped and draped.

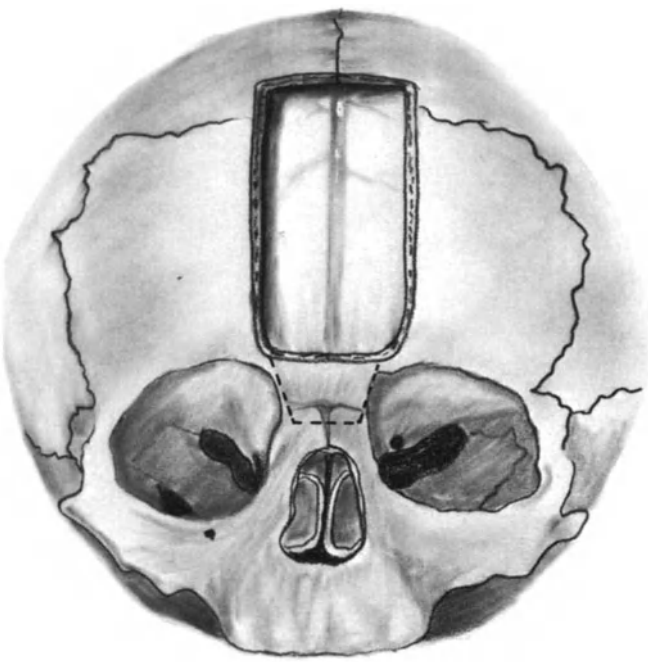


Figure 14.2. Resection of the synostosed metopic suture exposes the underlying anterior third of the superior sagittal sinus. Note that it extends into the sagittal, and across the coronal, sutures posteriorly, but that it is not carried anteroinferiorly across the frontonasal or into the nasal sutures (area within dotted lines). Consequently, there will always remain an observable hypotelorism. This limited procedure provides one with the illusion that the synostosis has been effectively dealt with because the keellike 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.

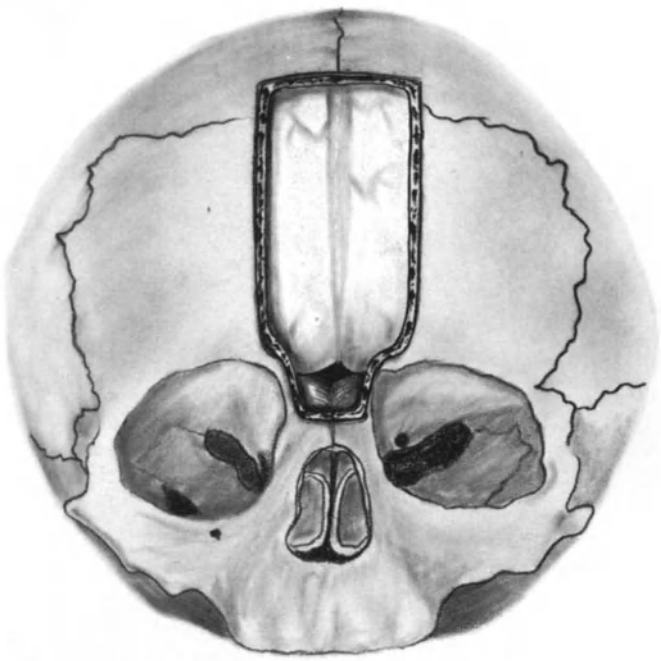


Figure 14.3. The only effective way to release completely the synostosed metopic suture is to extend the osteotomy across the frontonasal suture, and then to run it across the nasal suture. This releases the medial orbital rims from one another, allowing the subsequent brain growth to correct the hypotelorism.

is no protrusion posteriorly with ledge deformity of the occipital bone.

Surgical correction of metopic synostosis consists 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.

Sagittal Synostosis: Scaphocephaly

(Figures 14.4. to 14.12)

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 fontanel 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 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 he may identify coronal and lambdoidal sutures 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 cor-

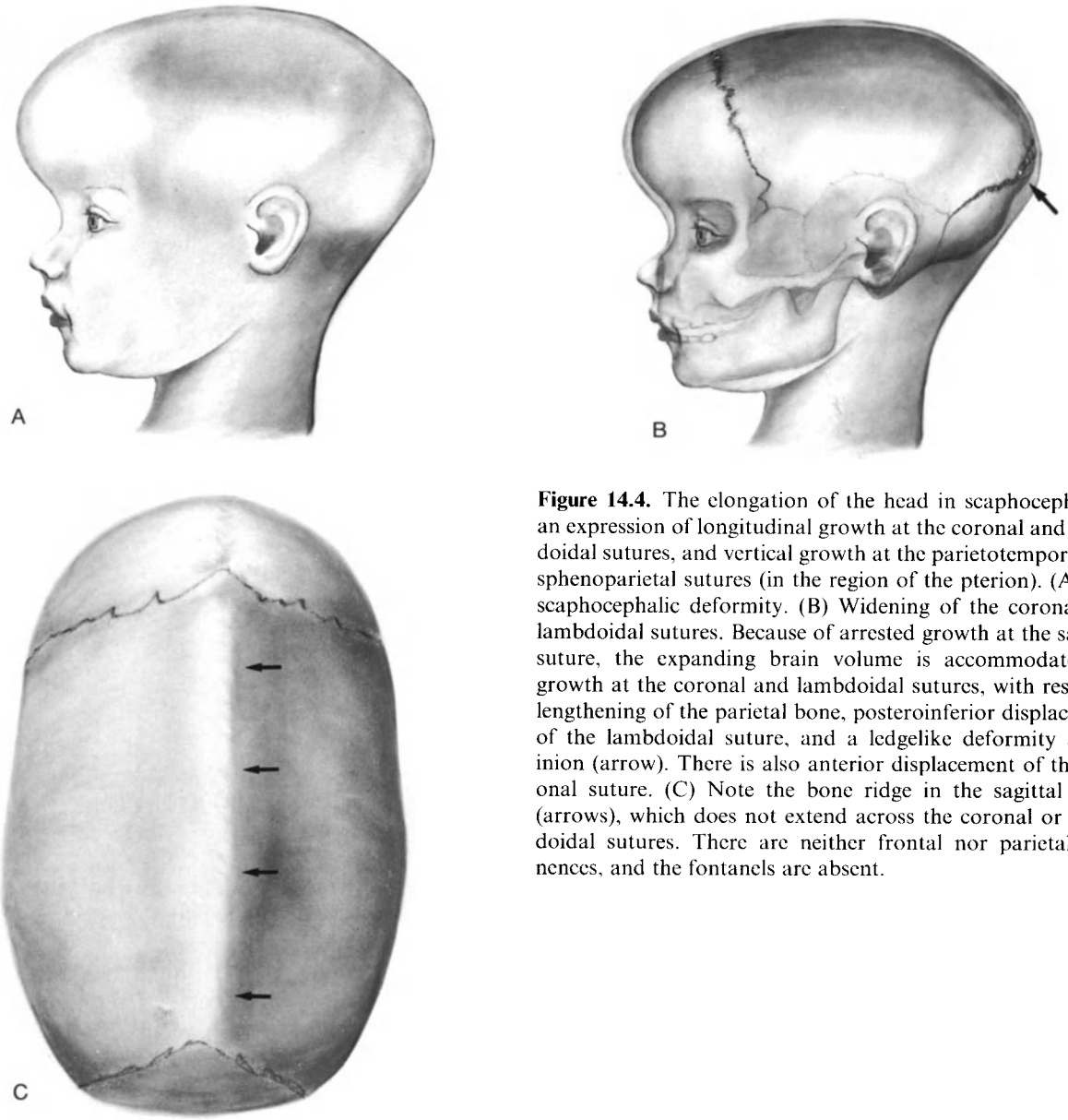
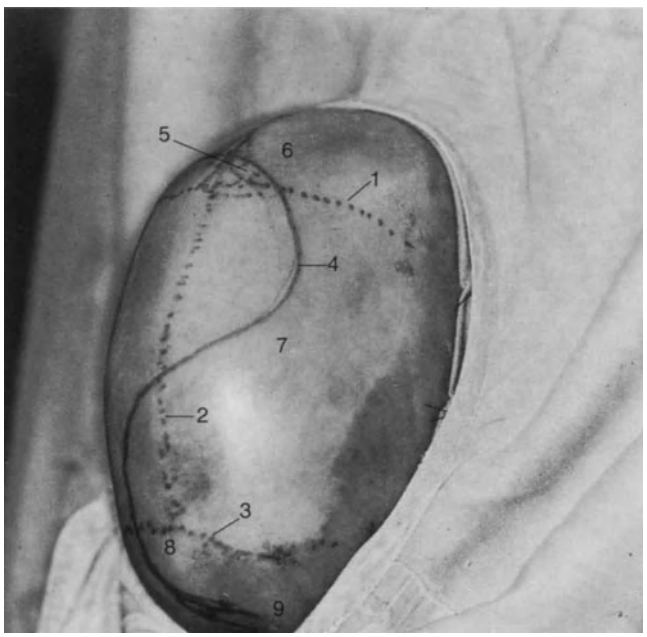


Figure 14.4. The elongation of the head in scaphocephaly is an expression of longitudinal growth at the coronal and lambdoidal sutures, and vertical growth at the parietotemporal and sphenoparietal sutures (in the region of the pterion). (A) The scaphocephalic deformity. (B) Widening of the coronal and 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 fontanels are absent.

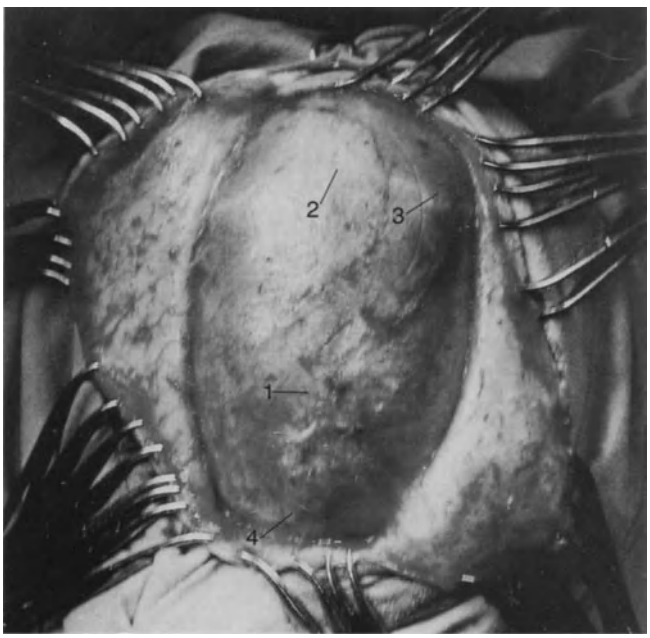
onal and lambdoidal sutures is performed with the periosteal elevator, after the periosteum has been coagulated with the bipolar cautery forceps and incised. Proper use of the periosteal elevator is to angulate its cutting edge, so that a corner may be insinuated between periosteum and skull, and then 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 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



A



B

Figure 14.5. (A) The scaphocephalic head has been draped and the coronal (*1*), sagittal (*2*), and lambdoidal (*3*) sutures marked off. The S-shaped skin incision (*4*) extends from right to left, passing along the border between the anterior fontanel (*5*) and the right frontal bone (*6*), and then over the right parietal bone (*7*). Posteroinferiorly, it extends behind the inion (*8*) to end over the squamosal portion of the left occipital bone (*9*). (B) The scalp flap has been reflected to either side, after the Galeal clips were applied. Note the hyperostosed sagittal suture (*1*), the anterior fontanel (*2*), the coronal suture (*3*), and the inion (*4*).

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 superior sagittal sinus (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 his 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.

I wish to note, however, that the incidence, severity, and multiplicity of synostosis varies from one ethnic group to another so that my experiences with caucasian, black, and oriental children in Chicago may be very different from what one would observe, for example, in Algeria.

Synostotic Craniofacial Anomalies

Gilles and Harrison, after treating a recessed malar compound fracture in an oxycephalic patient, reported

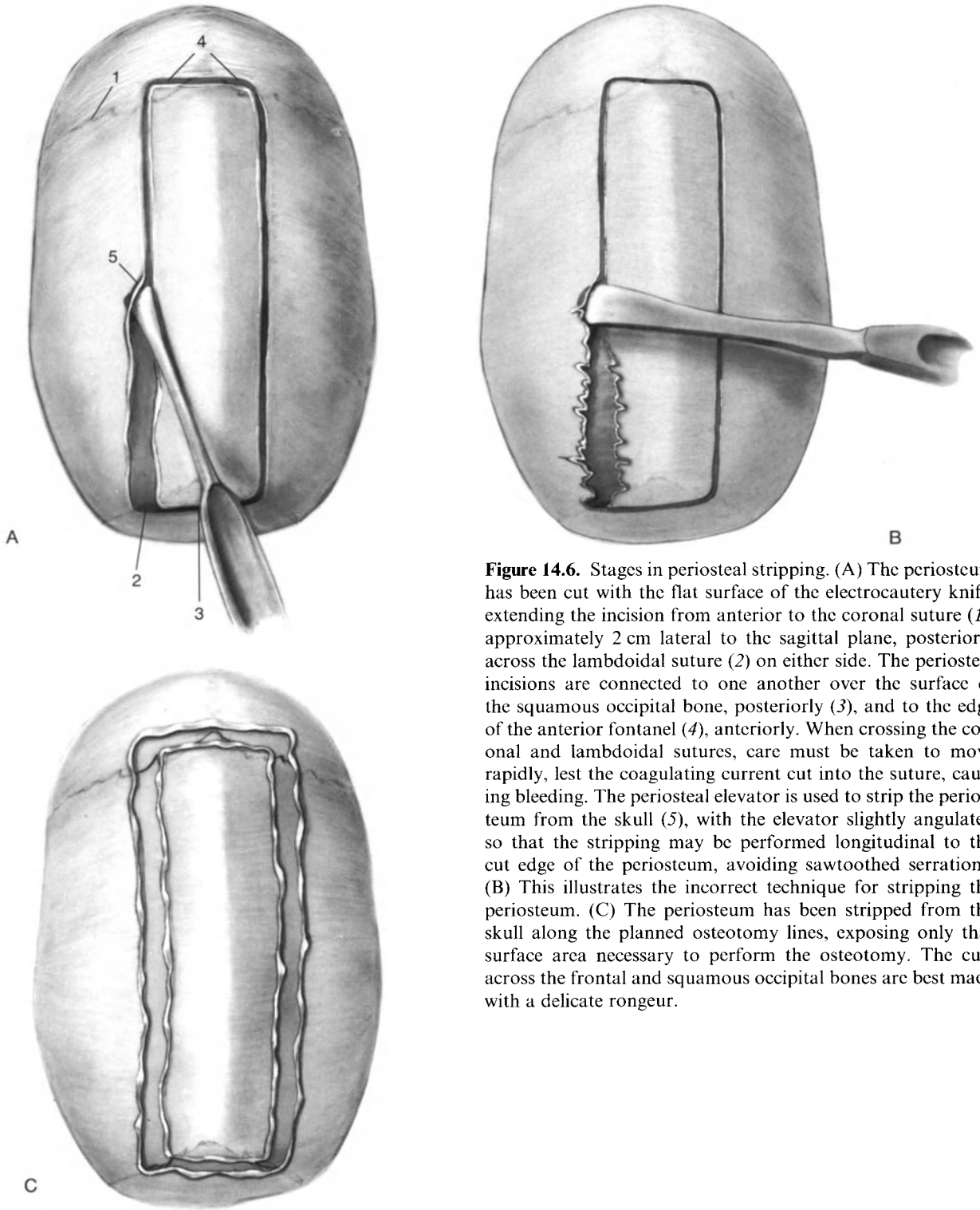
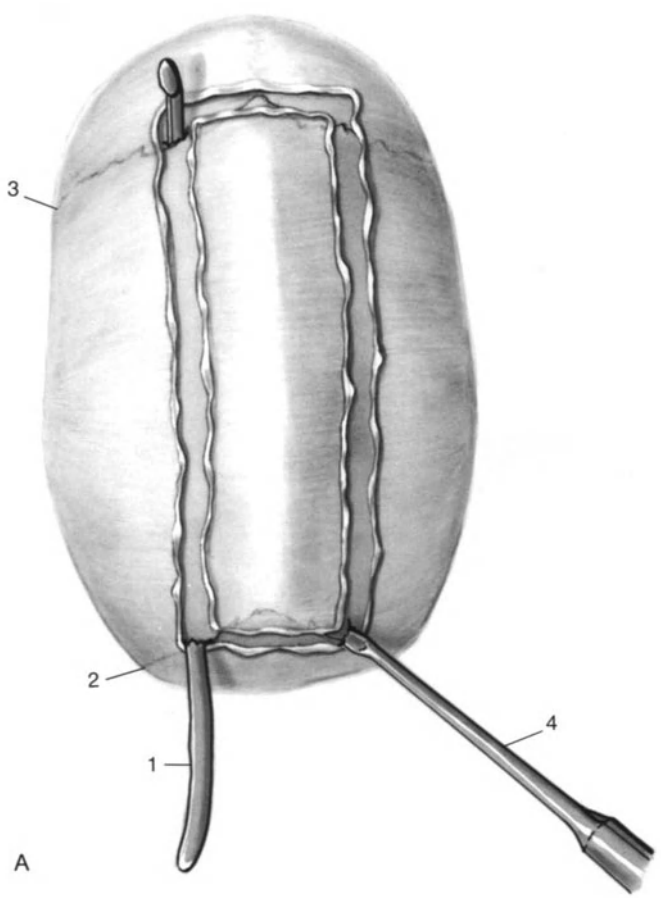


Figure 14.6. Stages in periosteal stripping. (A) The periosteum has been cut with the flat surface of the electrocautery knife, extending the incision from anterior to the coronal suture (*1*), approximately 2 cm lateral to the sagittal plane, posteriorly across the lambdoidal suture (*2*) on either 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 fontanel (*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.



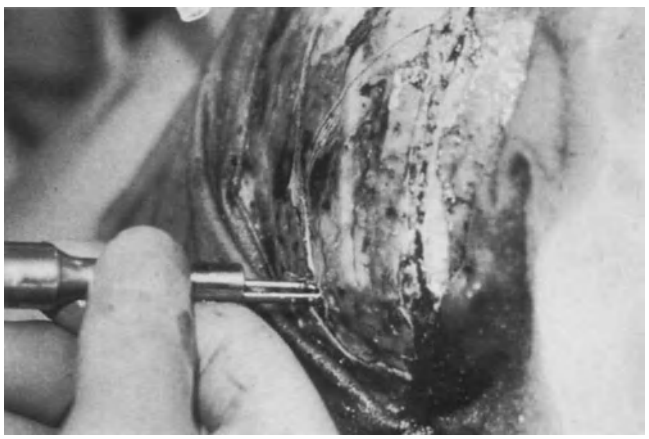


Figure 14.7. (A) The Gigli saw guide (*1*) has been passed from posterior to anterior, to separate the dura from the skull, before proceeding to use the craniotome to make the osteotomy. Note that the entrance into, and the exit from, the epidural space is at the lambdoidal (*2*) and coronal (*3*) sutures, respectively. No burr holes are made. Rather, the already described technique (Figures 4.3 and 4.4) is used to gain access to the epidural space, illustrated here by use of the curette (*4*) to open the suture. (B) Once the dura has been freed from the inner surface of the skull, the craniotome is used to perform the osteotomy.

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

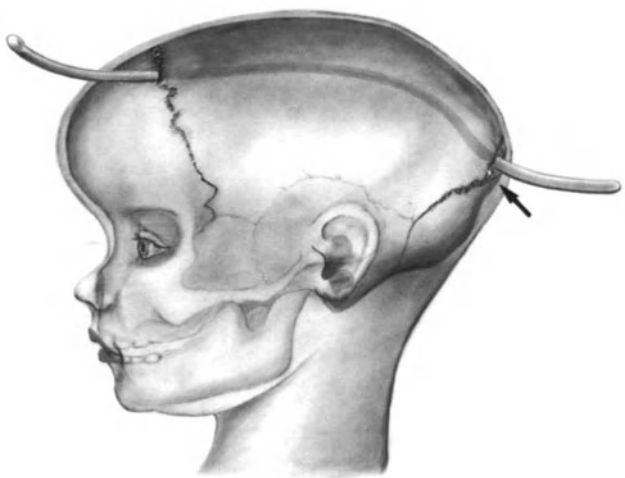
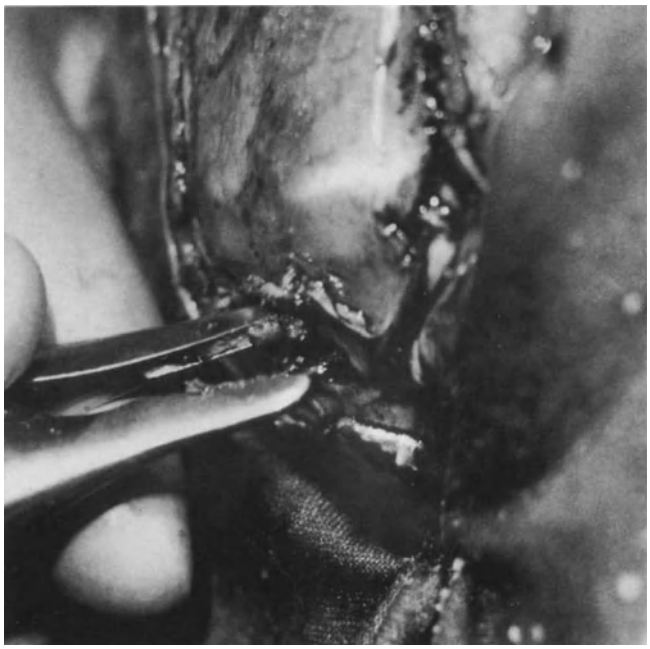
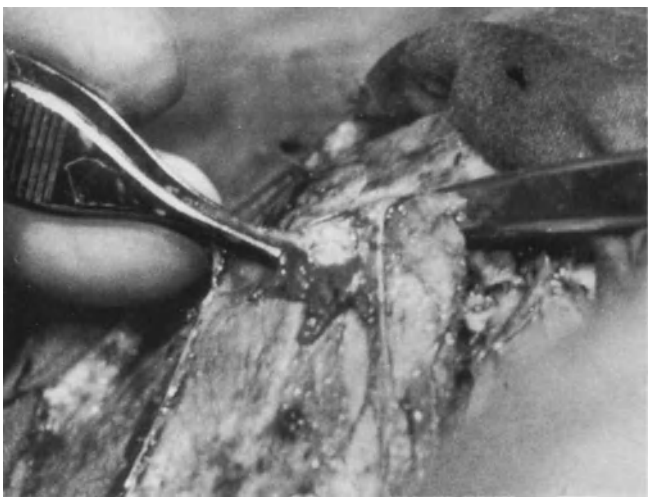


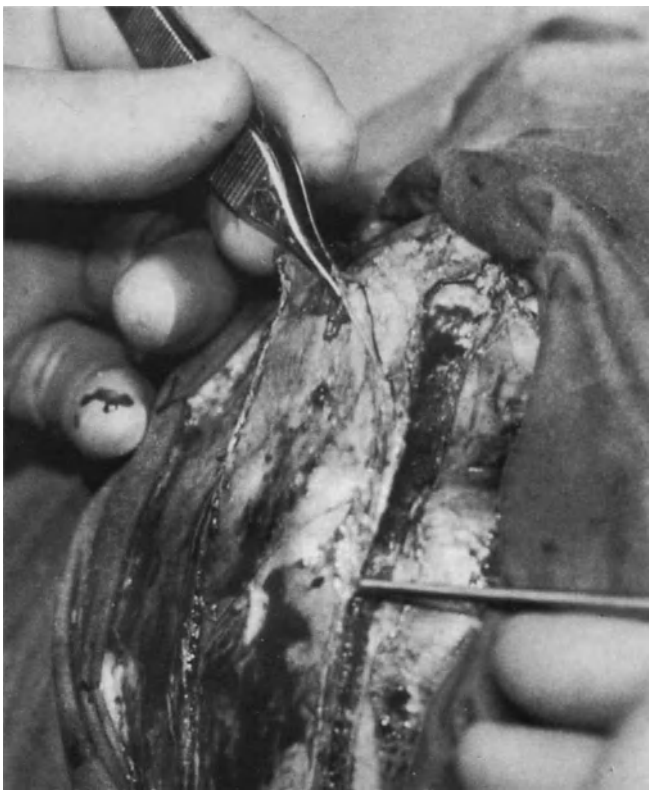
Figure 14.8. The inserted Gigli saw guide has been molded to fit the convexity of the brain and then passed from the posterior (arrow), lambdoidal suture, to the anterior coronal suture. Before removing the Gigli saw guide, mold the terminal end into a concavity so as to avoid cerebral depression.



A



B



C

Figure 14.9. (A) A rongeur is used to osteotomize across the suture line into the squamous occipital bone at the inion and then across the coronal suture bilaterally. (B) Heavy scissors are then used to cut the coronal suture from the parietal bone anteriorly, at the fontanel. (C) The now completely freed bone flap is stripped from the underlying dura and superior sagittal sinus surfaces by insinuating a Penfield dissector between bone and dura, as the flap is lifted away.

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

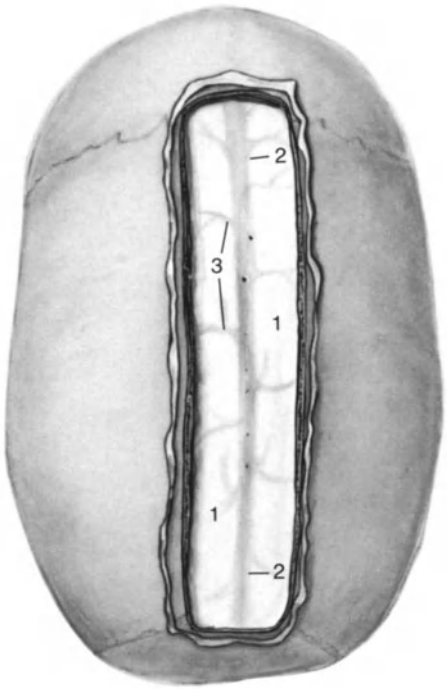


Figure 14.10. Anatomical representation to illustrate dural surface (*1*), superior sagittal sinus (*2*), and intradural portions of the bridging cortical veins (*3*) after the sagittal strip of bone has been removed.

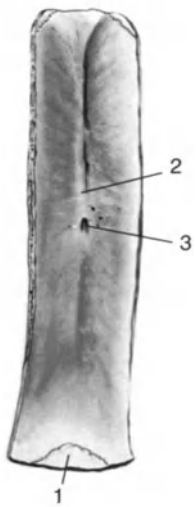


Figure 14.11. A wide strip of bone holding the hyperostosed superior sagittal suture has been removed from the underlying dura and the superior sagittal sinus (SSS). Note that the osteotomy has extended across the coronal suture anteriorly and the lambdoidal suture posteriorly (*1*). This strip of bone 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.

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 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; and
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, the reflection of a birfontal flap, preserving the orbital frontal band, measuring 2 cm in length 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, sparing the facial nerves. 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

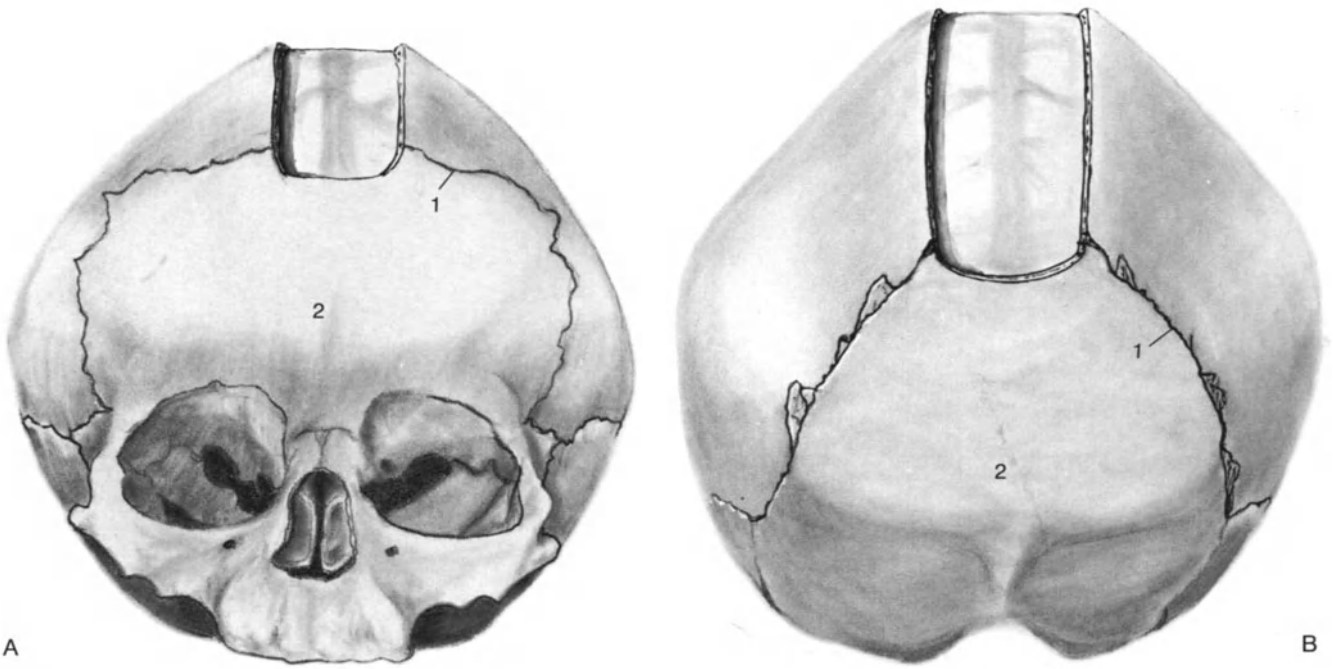


Figure 14.12. (A) Anterior extent of suturotomy across coronal suture (*1*) and into frontal bone (*2*). (B) Posterior extent of suturotomy across lambdoidal suture (*1*) and into squamous occipital bone (*2*).

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 Chapter 3. Instead, two parasagittal burr holes are placed, one to either side of the midline, just anterior to the coronal suture; two posterior frontal burr holes on the either side, 2 cm above the supraorbital rim and at the insertion of the temporalis muscle along the superior temporal line; two anteromedial burr holes, 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 form 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 crease 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.

Coronal Synostosis: Plagiocephaly

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 of the eye, so as to put stereopsis at risk. It is incorrect—incomplete—to consider this anomaly to be synonymous with 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 posterior-

ly 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 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 fontanel 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. For unilateral plagiocephaly, one must, therefore, always reconstruct the superior and lateral aspects of the orbit, and on occasion also reconstruct the medial aspect. For bilateral plagiocephaly, the corrective procedure involves both orbits, the ethmoid and nasal bones.

The various involvement of individual sutures, and the degree of deformity resulting from their synostosis, may not 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 CTT 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 my intention to present these anomalies as independent and distinct anatomicopathogenic entities.

Unilateral Plagiocephaly

Frontosphenoidal Synostosis (Figures 14.13 to 14.22). 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.

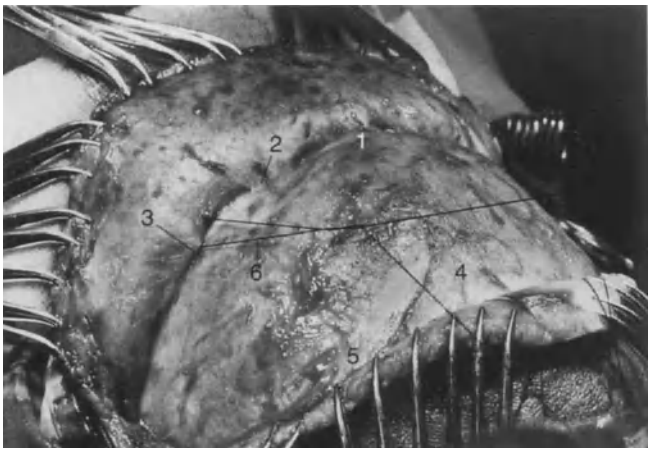


Figure 14.13. The scalp flap has been reflected, exposing the periosteum from over the frontal bone to a line running across the supraorbital ridge so that one may identify the glabella (*1*), the left supraorbital foramen (*2*), and the zygomatic process of the frontal bone (*3*). The posterior limb of the flap has been sewn out of the way, exposing the region of the anterior fontanel (*4*) and the line of the coronal suture (*5*). Note that the line of the coronal suture extends anteriorly, almost upon the zygomatic process of the frontal bone. Retention sutures, starting at the zygomatic process of the frontal bone (*6*), are now placed.

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 should be put in 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 and sulcus, zygomaticofrontal suture, pterional region and zygomatic arch. Posterior retraction of the scalp flap should be 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,

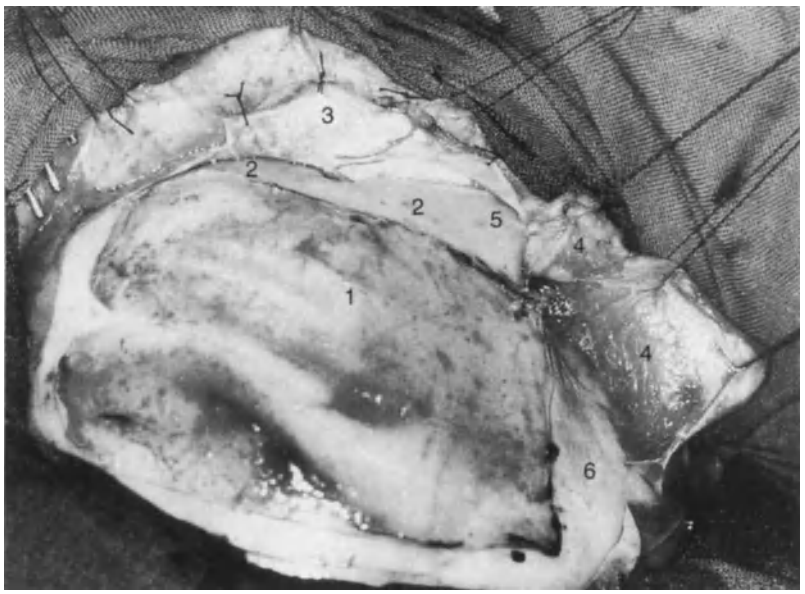


Figure 14.14. The periosteum has been incised, permitting one to reflect it from the planned osteotomy lines. It is left adherent to the frontal bone (*1*), but dissected from the supraorbital rim (*2*) and sewn out of the way (*3*). The periosteum and temporalis muscle (*4*) have been stripped from the zygomatic process of the frontal bone (*5*) and the bones of the pterional region (*6*).

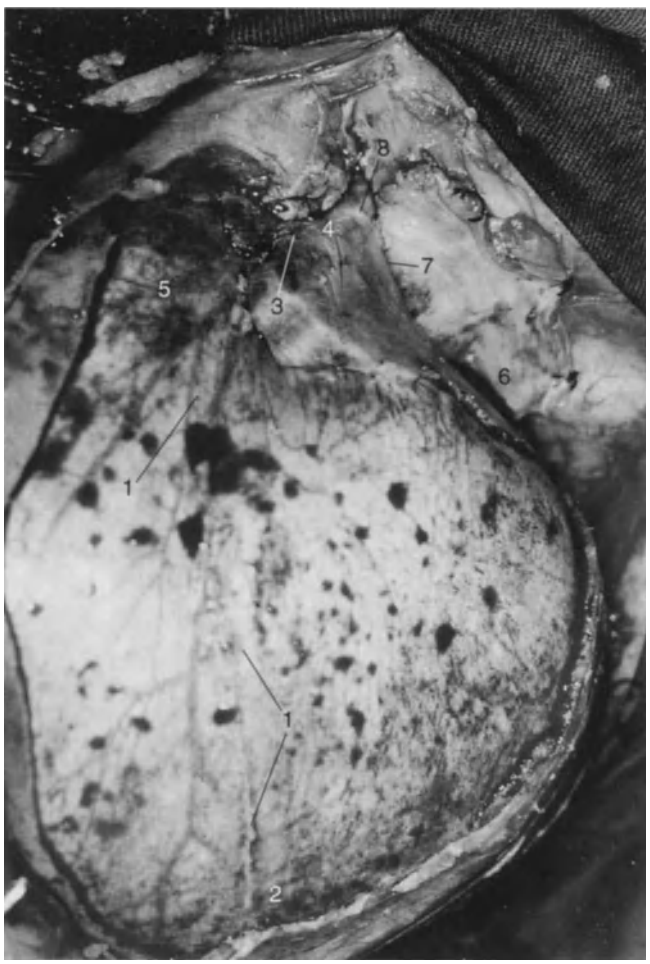


Figure 14.15. The left frontal flap has been reflected, permitting one to appreciate the almost vertical course of the coronal suture line (*1*) from the anterior fontanel (*2*) to the pterion (*3*), which has been displaced anterior to the zygomatic process of the frontal bone (*4*). Note that the temporal lobe (*5*) is so far anterior that it has come to rest lateral to the orbit. The periosteal dissection from over the orbital rim (*6*) has extended inferiorly, along the zygomatic process of the frontal bone, to the lateral surface of the orbit (*7*). One notes the fused zygomaticofrontal suture (*8*).

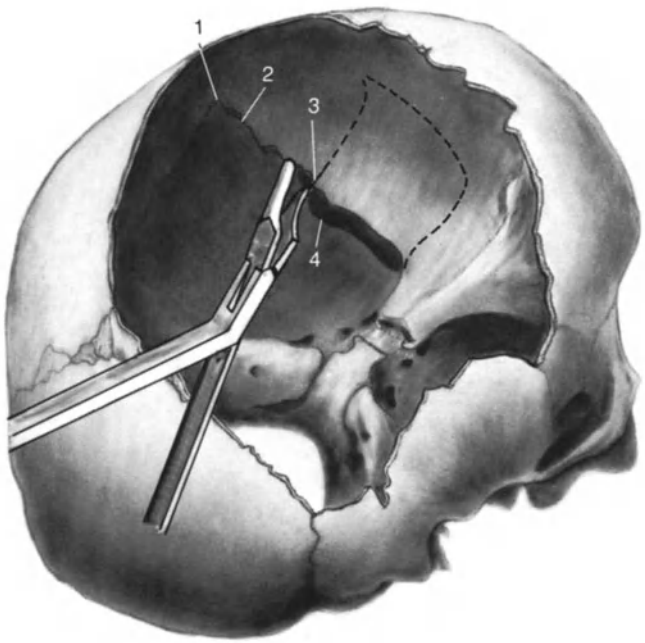


Figure 14.16. A 3-dimensional artistic representation of the inner surfaces of the neurocranium, illustrating the extension of the craniectomy, along the synostosed coronal suture (*1*) from the pterional region (*2*), superomedially, to the lesser wing of the sphenoid. At this point it is extended into 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.

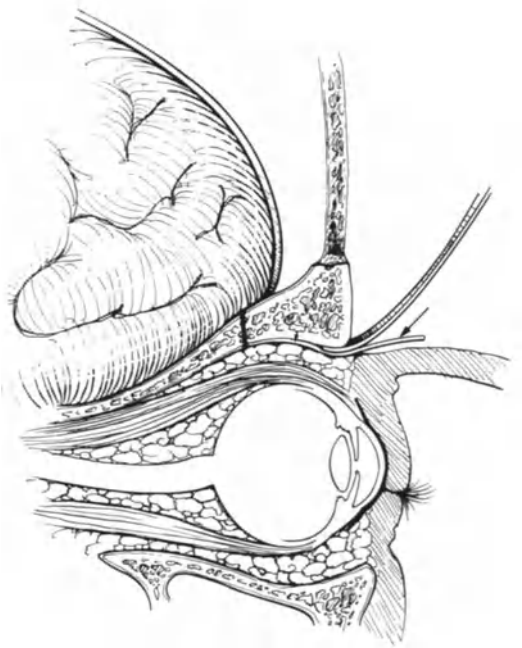


Figure 14.17. The floor of the anterior fossa is exposed by retracting the frontal lobes. The periosteum (arrow) is dissected from the roof of the orbit with a Penfield #3 dissector, separating the periosteum and periorbita from the orbital rim and roof.

as is the hyperostosed coronal suture from the region of the anterior fontanel to the pterion. The periosteum is incised by using the flat edge of the electrocautery blade, the periosteum is reflected over the supraorbital ridge, and the burr 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; and
4. posterosuperior to the pterion.

It is important to note that the *keyhole* burr 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 burr 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 between 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 minutes of the time the frontal flap and hyperostotic suture are removed. This is remarkable. The fluid ranges in color form 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, becom-

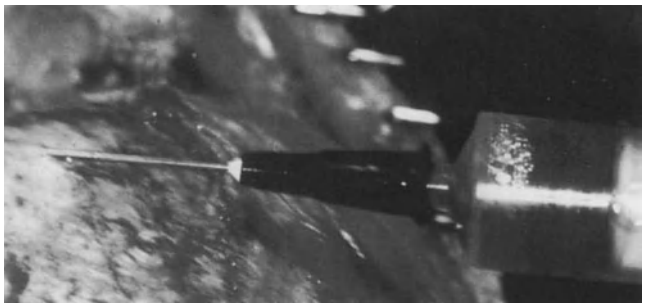


Figure 14.18. Fluid that accumulated in the subdural space, after the synostosed coronal suture and frontal bone were removed, is being aspirated.

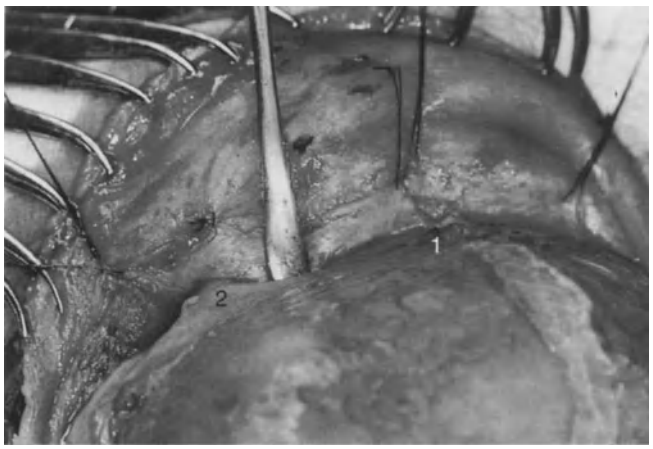


Figure 14.19. A Penfield #4 dissector is being used to separate the periorbita from the orbital surface of the roof of the left orbit. For orientation purposes, the glabella (*1*) and zygomatic process of the frontal bone (*2*) are indicated.

ing unfolded by the brain, after the synostosed sutures have been removed from the anterior fontanel 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 keellike deformity, without damaging the underlying dura or brain. One should connect the burr holes with a craniotome, not the Gigli saw, though it is safest to pass the Gigli saw guide from the posterosuperior burr 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. It is unwise to attempt to pass the saw guide along the line extending from the *keyhole* to the posteroinferior burr hole, since this would oblige one to cross the keellike hyperostosis. Rather, once the three osteotomies have been cut with the craniotome, it is safest to perform a suturectomy, rongeuring away the synostosed sutures from the anterior fontanel 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 burr, 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 entire 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 must be accomplished with great care, to avoid damaging the olfactory nerve and, more importantly, tearing dural sleeves from around the olfactory bulb outlets, risking creation of a cerebrospinal

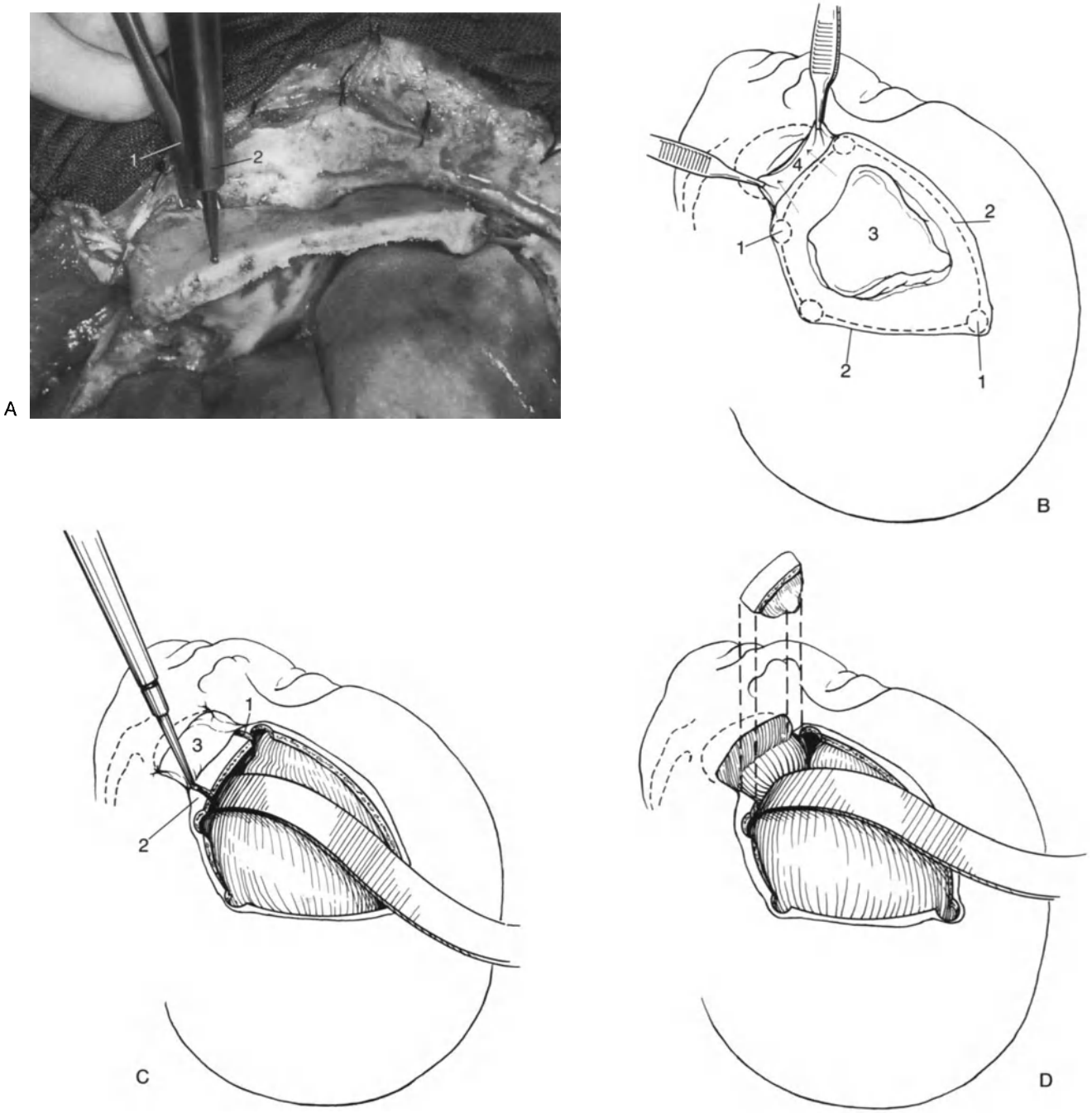


Figure 14.20. (A) A Penfield #3 dissector (*1*) has been inserted between the periorbita and the orbital surface of the roof of the left orbit. This provides protection against inadvertent perforation by the high-speed drill (*2*) as the osteotomy is being made. (B) The frontal burr holes (*1*) and craniotomy (*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.

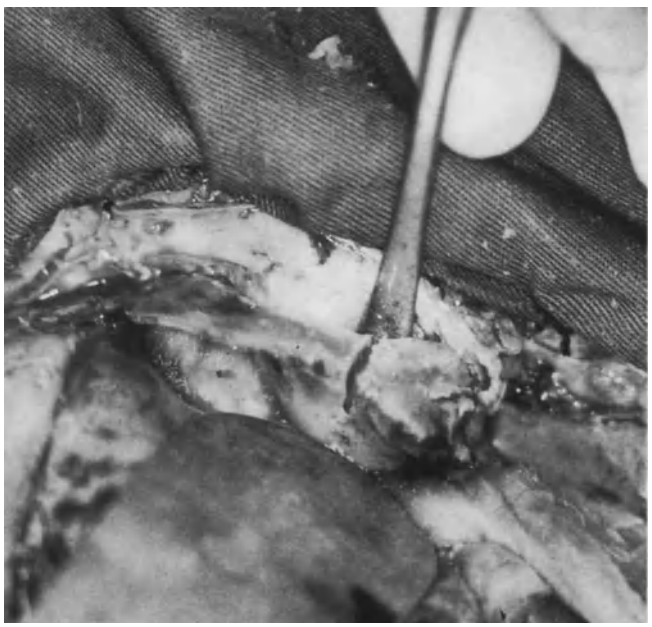


Figure 14.21. The Penfield #3 dissector has been inserted deep to the osteotomy line, affording protection to the periorbita and intraorbital contents.

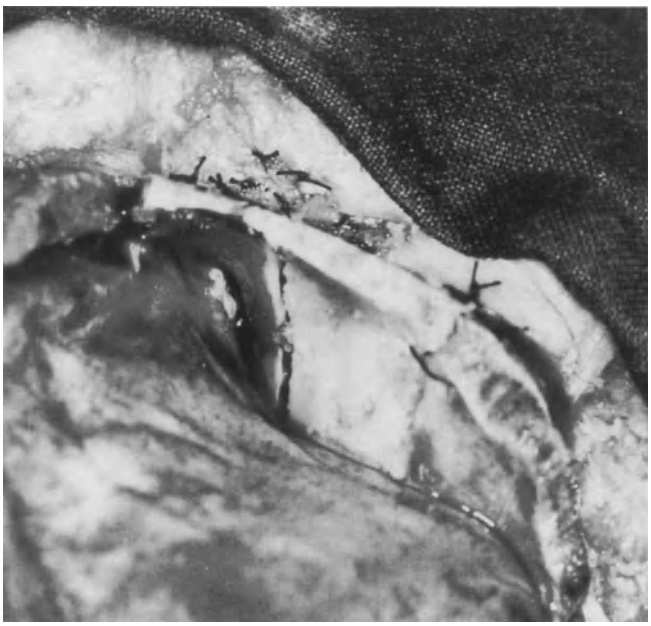


Figure 14.22. The medial and lateral osteotomy lines have been completed. They extend posteriorly along the orbital roof.

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.

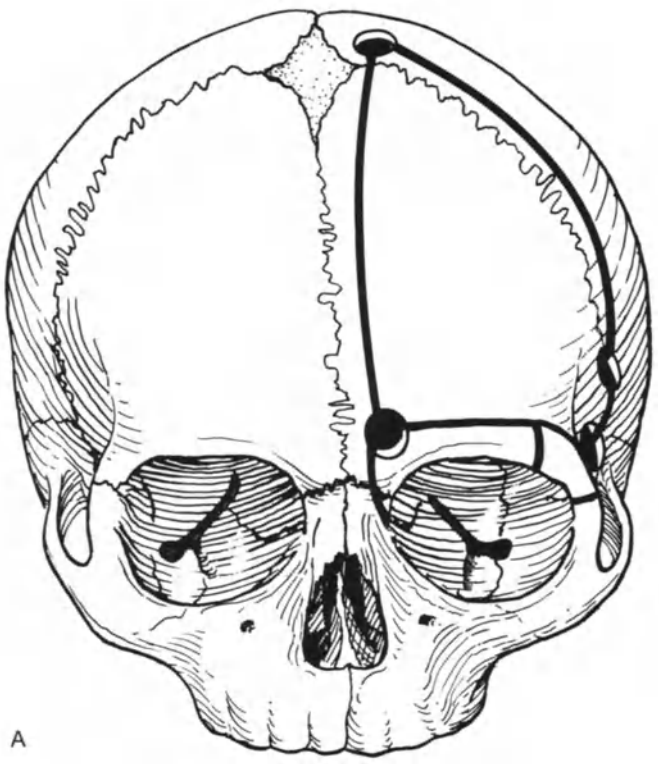
Periosteum is now dissected from the supraorbital ridge, from medial to lateral, taking care not to damage the lacrimal gland, since this occupies the most superolateral portion of the orbit. A Penfield #4 dissector, or an Oldberg periosteal elevator, is desirable for completing the separation of periosteum 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 orbital roof.

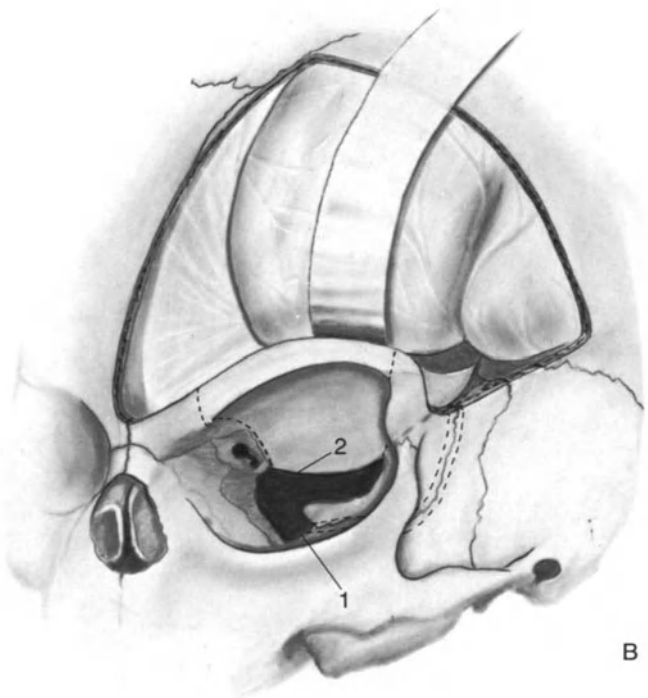
The Penfield #3 dissector is used to protect the intraorbital contents during osteotomy, medial and lateral, of the supraorbital rim and orbital roof. One now has fully exposed the orbital rim from just lateral to the glabella to the zygomatic process of the frontal bone, the periorbita from the region of junction of the periosteum to the superior orbital fissure.

Zygomaticofrontal Synostosis (Figures 14.22 and 14.23). 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 must be 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





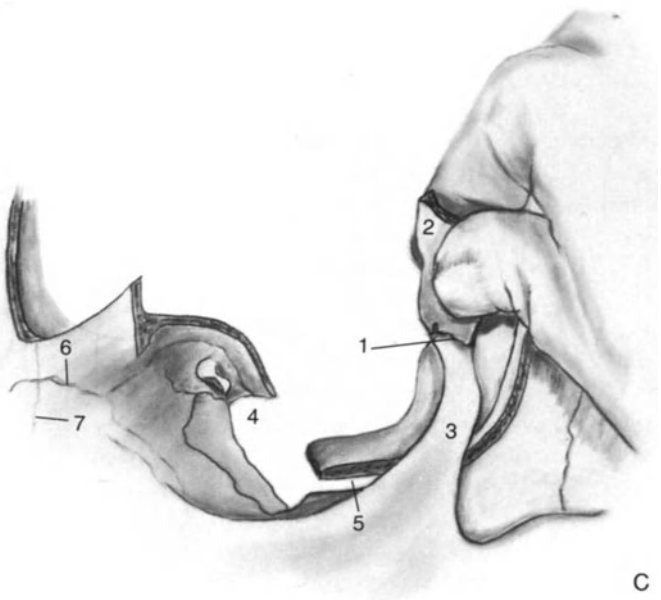


Figure 14.23. (A) The craniotomy lines and burr holes are illustrated for the reflection of the frontal flap, and the osteotomy lines for removal of the supraorbital rim are made. (B) After the frontal flap has been reflected, the exposed dura is seen and a spatula placed over the frontal lobe. The broken 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.

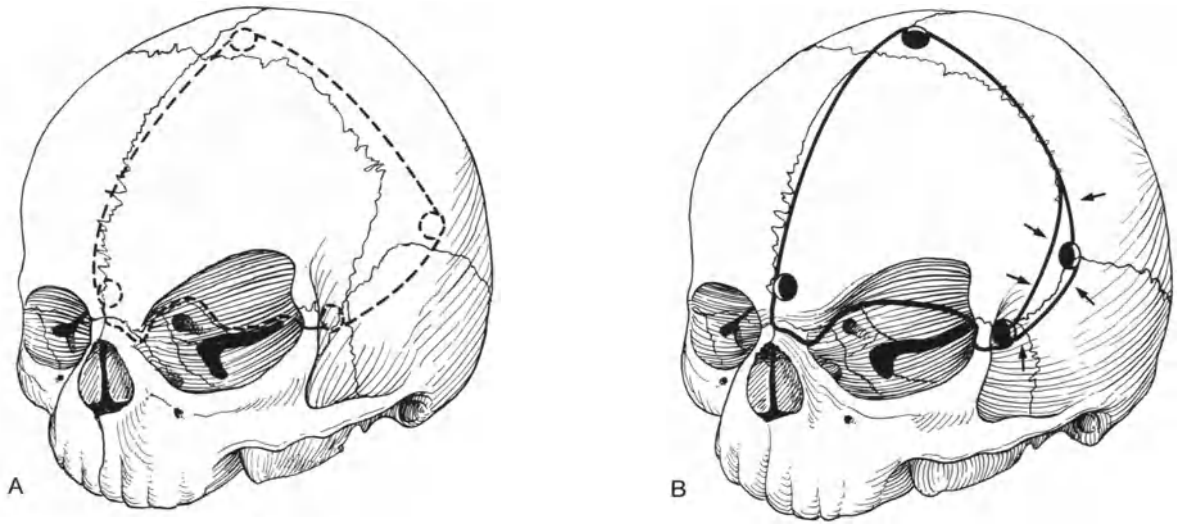


Figure 14.24. (A) Burr hole placement if one chooses to reflect the supraorbital rim, orbital roof, and lateral wall of the orbit in one piece (stage), after osteotomizing the frontoethmoidal suture and synostosed pterional sutures into the superior orbital fissure. The broken line indicates the osteotomy along the calvarium, into the orbital roof, and over the nasal bone. (B) Resection of the synostosed coronal/frontosphenosquamosal suture (arrows).

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 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 (Figures 14.24 and 14.25). *When the unilateral plagiocephaly involves the medial* (frontonasal, nasal, and frontoethmoidal) *as well as the lateral sutures,* then one must free completely the nasal and ethmoidal bones from the frontal bone. This must be done along the line of the orbital roof *and* at the supraorbital rim. Consequently, the placement of the inferomedial burr 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 standard 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) burr 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

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; and
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 must osteotomize the frontonasal, nasal, and frontoethmoidal sutures. Synostosis along these suture lines is responsible for deformity of the medial orbital rim and the hypotelorism.

If there is remarkable, unsightly, flatenning 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 *must not* be tightly anchored at the time of closure, they should either be loosely moored into position or just laid over the periorbita and the dura, allowing the growing brain to mold them.

Bilateral Coronal Synostosis: Plagiocephaly
(Figures 14.26 and 14.27)

Bilateral plagiocephaly may result in such severe exorbitism as to subject the sclerae and corneae to destruc-

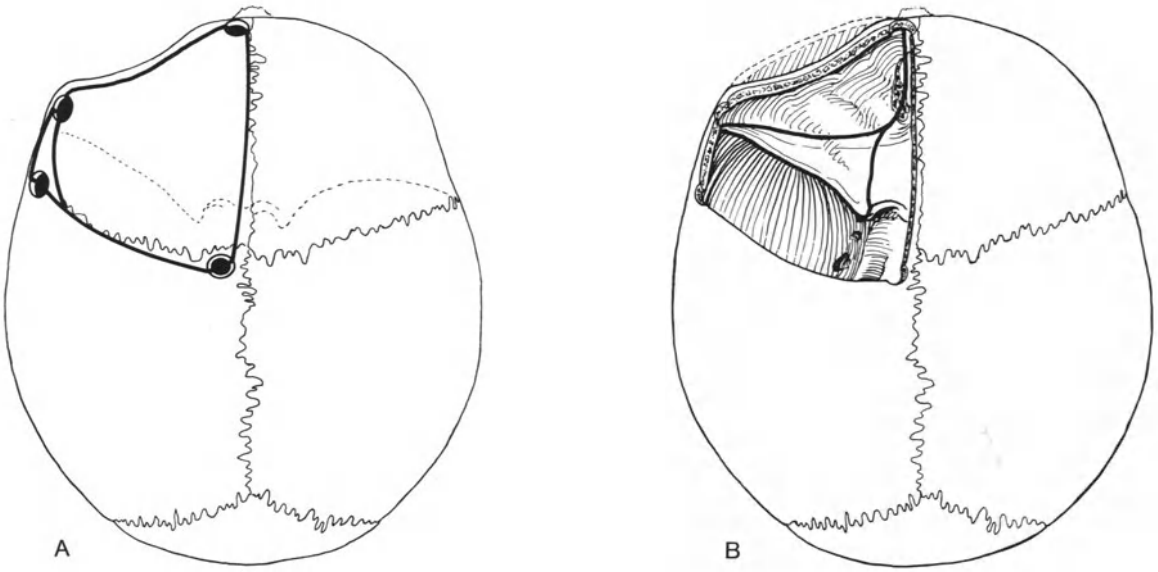


Figure 14.25. This view from the vertex illustrates the osteomies over the vault and along the floor of the anterior fossa. (A) The solid line indicates the osteotomy and illustrates the flattening of the frontal eminence. The broken line outlines the lesser wings and body of the sphenoid. (B) The frontal 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°.

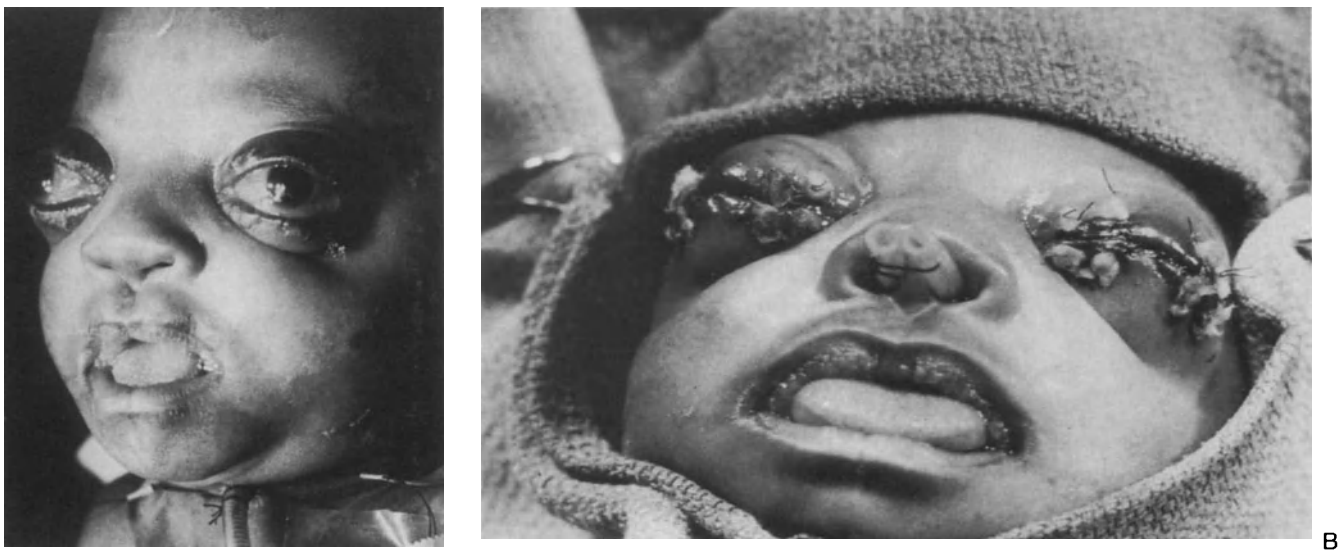


Figure 14.26. (A) The exorbitism which children with bilateral plagiocephaly suffer may be extraordinary. (B) In such instances, bilateral tarsorrphaphies are necessary.

tive 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 must not be undertaken without 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.

Figure 14.27. Stages in the performance of radical craniectomy in the young infant for bilateral plagiocephaly. (A) The periosteum has been stripped along the suture line, and a periosteal elevator inserted between the outer layer of the dura and inner surface of the skull, separating the two from one another. (B) At this age, it is not necessary to use either a craniotome or a Gigli saw. Mayo scissors suffice to perform the craniotomy. (C) The left frontotemporal craniotomy has been performed, revealing the remarkably tense underlying dura. (D) After the bilateral frontotemporal craniotomies have been performed, the periosteum and periorbita are separated from the supraorbital rim and orbital surface of the orbital roof, so one may proceed to resect these bony structures. (E) The dura is separated from the roof of the orbit, from lateral to medial, as far posteriorly as the lesser wing of the sphenoid and as far medially as the ethmoid bones. The preserved frontal nerve may be seen along the Galeal surface of the frontal scalp flap. (F) The orbital roofs, zygomatic processes, and pterional bones, are rongeured and discarded. (G) The frontal bone, with its supraorbital rims and zygomatic processes, has been removed, as have the roofs of the orbits and the synostosed pterional bones, extending the craniectomy into the superior orbital fissures bilaterally. One notes the metopic suture line (*1*), the frontal lobes (*2*), the periorbita contouring the orbital contents bilaterally (*3*). The scalp is simply closed over the dura, since the bone will regenerate completely at this age. ►

Hypertelorism (Figure 14.28)

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 an understanding of what the plastic surgeon is attempting to accomplish is immediate. "Ocular hypertelorism" was coined in 1924 by Greig,[20] "orbital hypertelorism" in 1972 by Tessier.[21] 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 lachrymal 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."

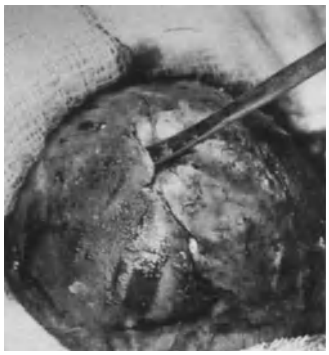
Reflection of a bifrontal flap for hypertelorism entails considerations that the crista galli may be remarkably hypertrophied, the dura may be deformed by an associated meningocele or meningoencephalocele, and that one may be able to preserve the olfactory nerves because the olfactory grooves are displaced 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.

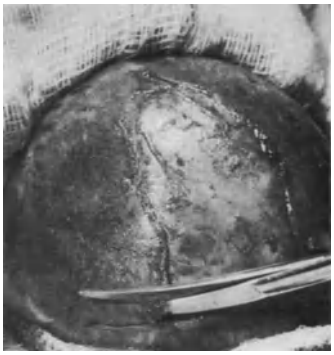
The steps in performance of craniofacial reconstruction for hypertelorism, the A-3 Tessier procedure, through the intracranial route as discussed here are taken from the 1972 publication by Tessier, Guiot, and Derome.[22]

Crouzon and Apert

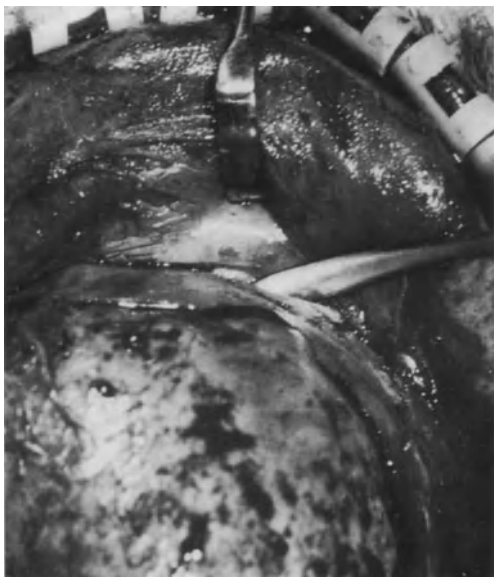
The term craniofacial dysostosis may be used to describe both Crouzon's and Apert's deformities, since, for all intents and purposes, the clinical condition and surgical considerations are the same. It still is not clear, however, in either of the two genetic entities what the relationship is between the cranial synostosis and the very characteristic facial deformities, though some authors[23] have postulated that such facial hypoplasias as facial stenosis, independent of the presence of retrusion of the maxilla, are pathogenically identical to craniostenosis.

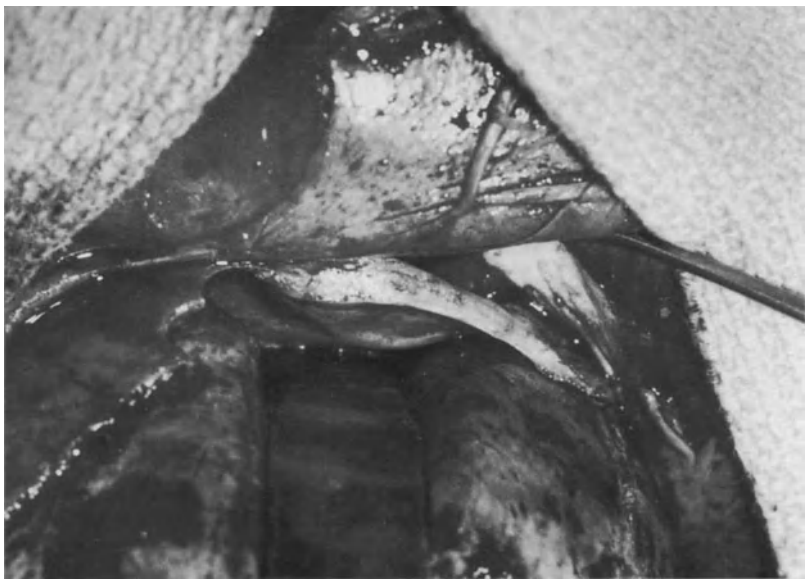
The aim of surgical treatment of craniofacial dysostosis is correction of the remarkable exorbitism, the malocclusion secondary to retrusion of the maxilla, the compressive obliteration of the nasal pharynx, and the recession of the frontal bone which is an expression of coronal synostosis.



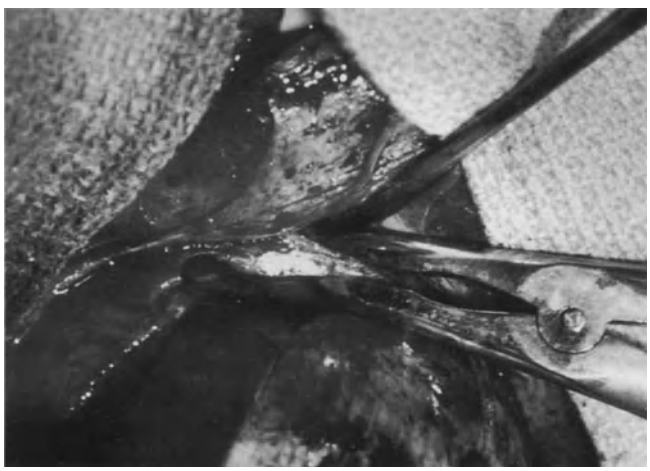


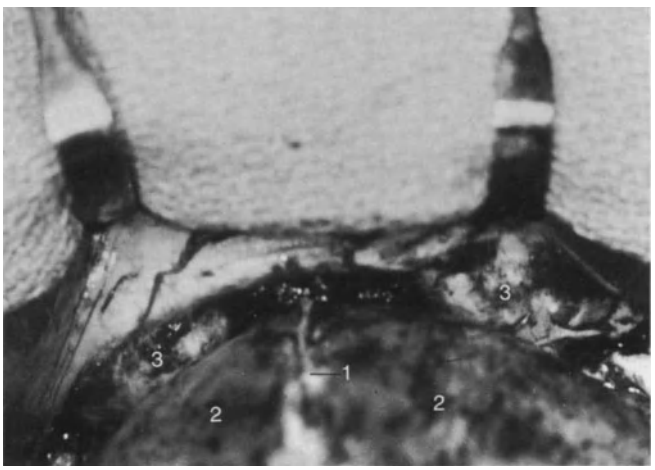
A C





D E





F G

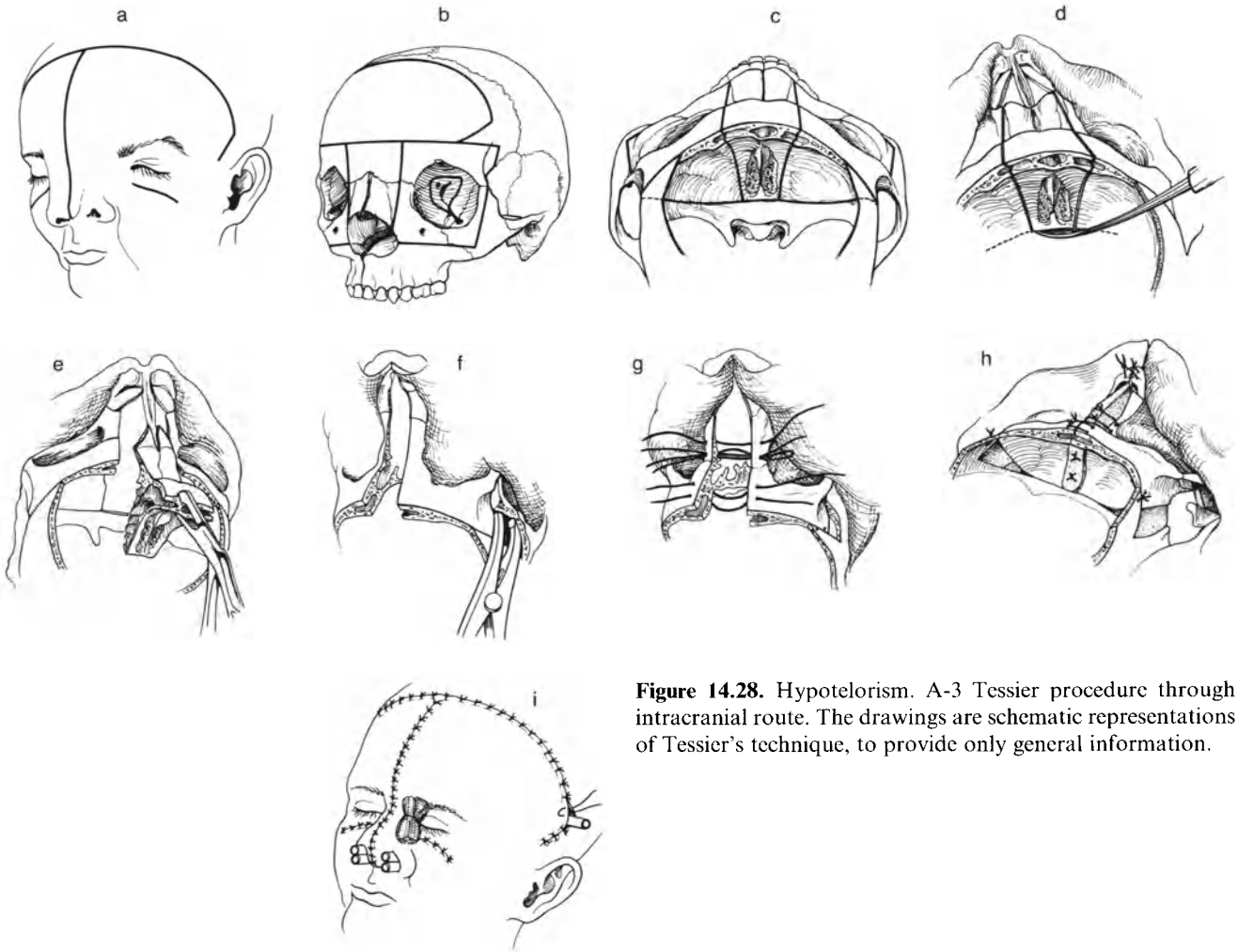


Figure 14.28. Hypotelorism. A-3 Tessier procedure through intracranial route. The drawings are schematic representations of Tessier's technique, to provide only general information.

In 1950 Sir Harold Gilles[6] was the first surgeon to perform a midface advancement for craniofacial dysostosis, 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[24] performed his first midface advancement, which was followed by a period of nine years of surgical experience. In 1967 Tessier[25] 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 entities 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[26] 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 Holtermüller and Wiedermann[27] 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 fontanel 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[28] 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 the remarkable bulging of the anterior fontanel 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 (Figures 14.29 to 14.31)

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 collicular 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 involves 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 to ostium. However, experience with the instrument and technique are wanting.

Cystoperitoneal Shunting

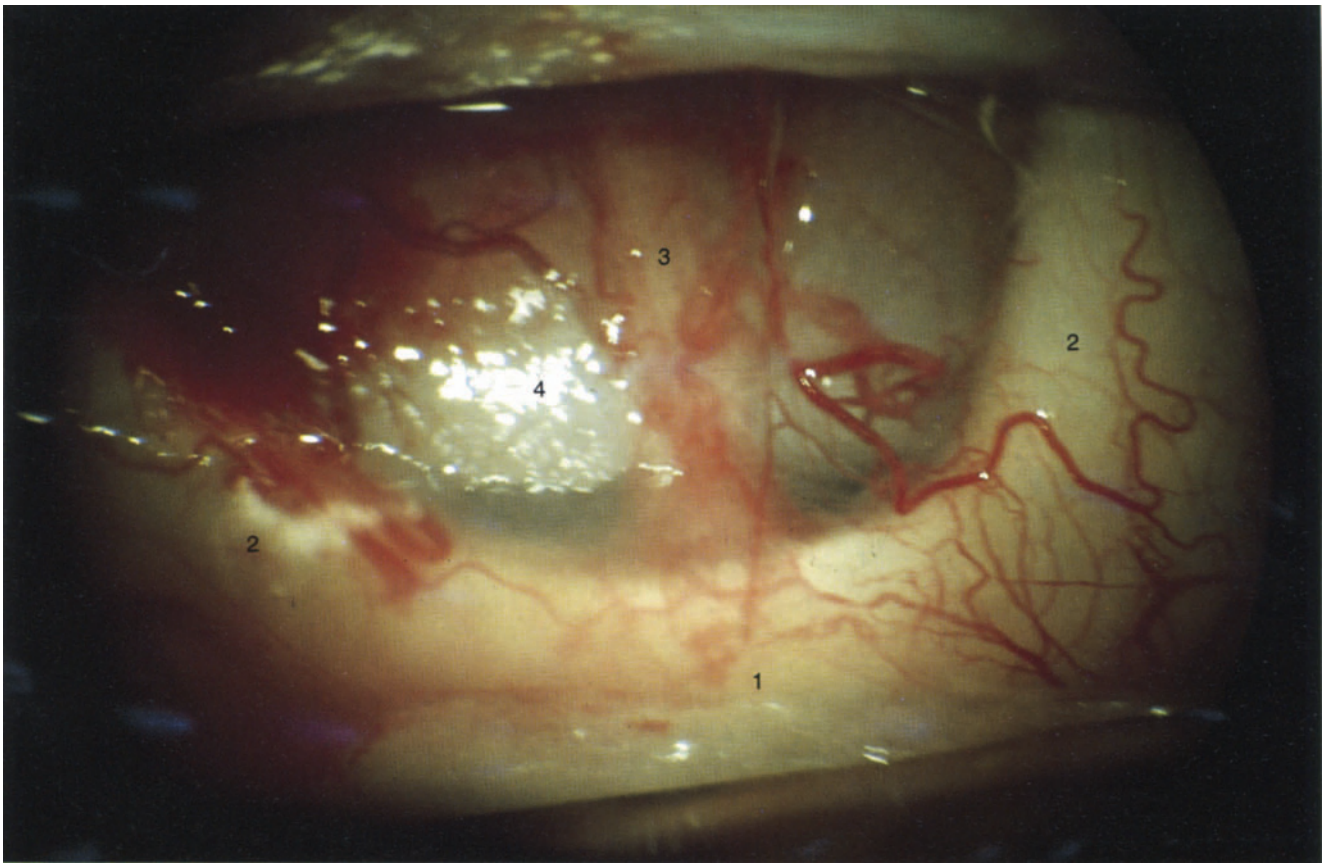
Cystoperitoneal shunts drain continuously the cerebrospinal fluid from ependymal or arachnoidal cysts, providing the possibility that the ostium may seal itself off spontaneously. The proximal end of the shunting system may be inserted into the cyst cavity either stereotaxically or through a ventriculoscope. Ultrasound is of great assistance for stereotaxic placement.

Chiari IV Malformation

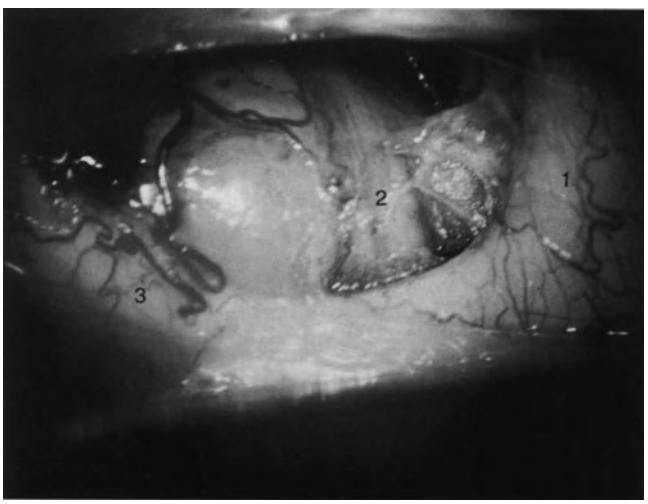
Aplasia of the entire cerebellum with the exception of the flocculonodular lobes results in the *ex vacuo* dilation of the IV ventricle, cisterna magna, and pontine cistern. There is no obstruction to the flow of cerebrospinal fluid. This primary dysplasia has no surgical treatment.

Lateral Arachnoidal Cysts (Figure 14.32)

It is difficult to state precisely whether cerebrospinal fluid-(CSF) containing cysts located over the cerebral



A



B

Figure 14.29. (A) 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. (B) The arachnoid has been taken from the right optic nerve (*1*) and the lateral surface of the pituitary stalk (*2*), but left intact between the pituitary stalk and the left optic nerve (*3*). Note that, despite the fact that the arachnoid has been coagulated to the right of the pituitary stalk, the cystic chamber on the left remains distended, thereby indicating that this arachnoidal cyst is composed of many smaller chambers, rather than one single compartment. The realization of this fact permits one to understand why resection of the arachnoid cyst is not always followed by a cure, and insertion of shunt tips into the cyst cavities does not always drain the entire cyst.

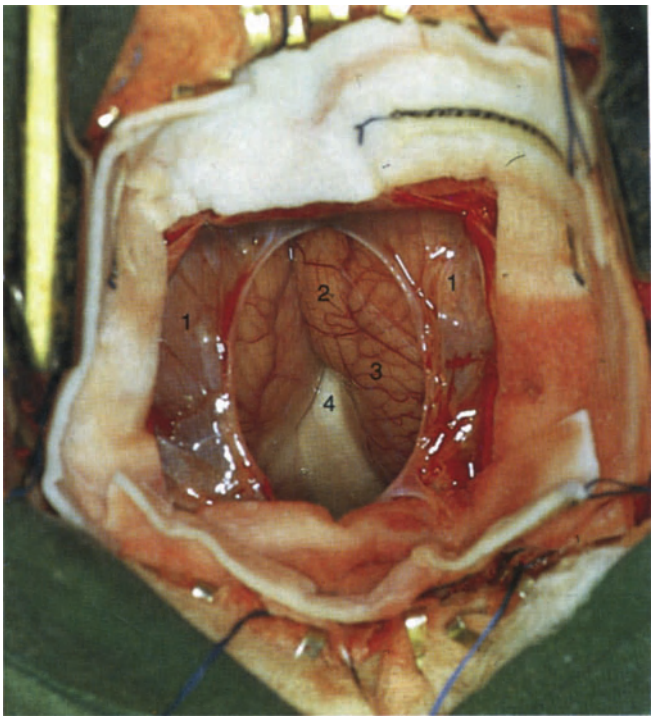


Figure 14.30. Arachnoidal cyst of the cisterna magna. One should distinguish between a large cisterna magna, which is not pathogenic, and cystic transformation of the cisterna magna, which is pathogenic. Here, a child with cystic transformation of the cisterna magna is illustrated. Note the very dense 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.

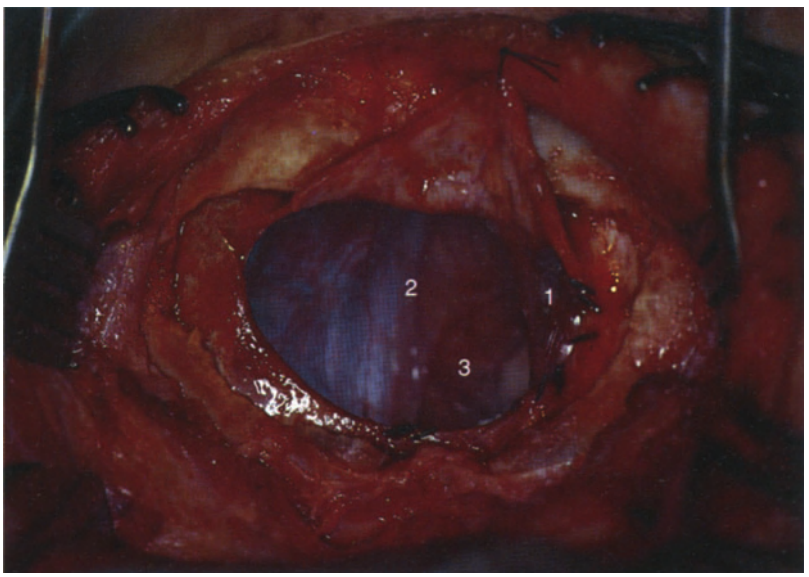
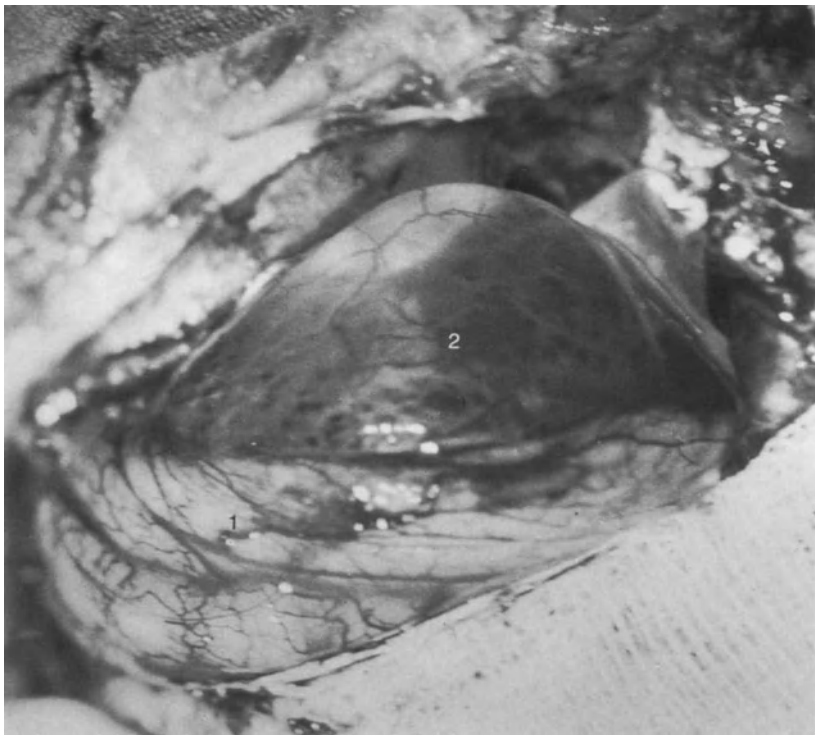
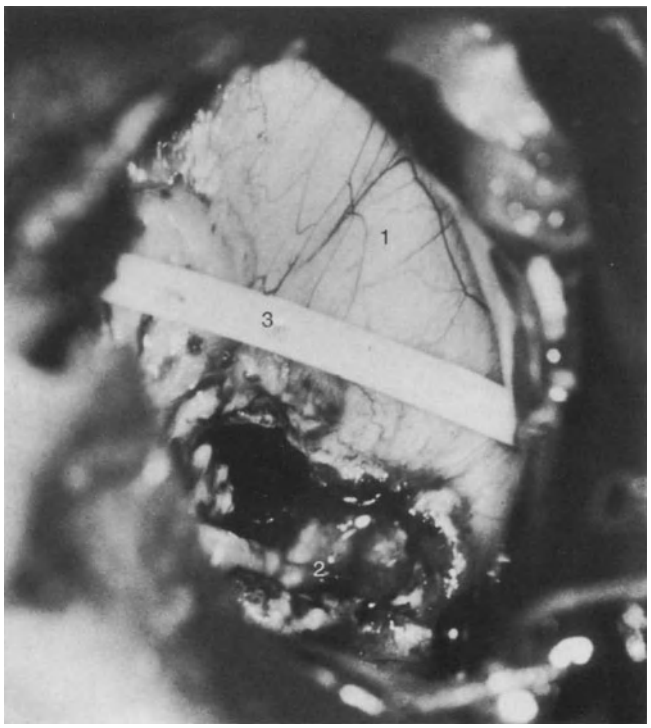


Figure 14.31. Arachnoidal cyst of the cisterna magna. In this child, different from the one in the previous figure, the arachnoidal cyst occupied almost the entirety of the posterior fossa. The posterior arachnoidal membrane (*1*) has been opened and partially resected. The very dense anterior membrane (*2*) is adherent to the cerebellar vermis in the midline, and the remarkably compressed and flattened cerebellar hemispheres on either side (*3*).



A

B



C

Figure 14.32. (A) 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*). (B) The cyst has been drained and its membrane (*1*) is being resected. The frontal lobe has fallen into a normal position (*2*). (C) After resection of the cyst membrane, one was able to look directly into the temporal horn. The ependymal surface of the temporal horn (*1*), choroidal fissure (*2*), and proximal end of the shunting system which had been inserted into the cyst (*3*) are shown.

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 space: 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 per emergency if the child develops a clinical picture of an increase in intracranial pressure. Otherwise, he 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, itractable seizures, or incapacitating episodes of headache.

Craniofacial Encephalomeningoceles

Ingraham and Swan,[29] 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 cerebrospinal fluid (CSF); to speak of *cranial encephalocele* (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[30] 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. The plastic surgeons use the grafts invariably in their repair of the basal defects, the 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[31] classification of craniofacial encephalomeningoceles 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[32] 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 endocranial 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.[29, 33, 34]

Blumenfeld and Skolnik[35] suggested not to treat asymptomatic small basal encephaloceles in neonates and infants. Robinson[36] 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 ap-

proach 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[32, 37, 38]. 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[39–41] instances. 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 sphenoorbital 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 sphenoorbital 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[42] 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 Encephalomeningoceles

Basal Craniofacial Encephalomeningoceles

Basal craniofacia encephalomeningocela result from defects in the clivus (basiocciput, sphenooccipital synchondrosis, basisphenoid), the sella turcia (body of the sphenoid), the planum sphenoidale, the ethmoid.

It is not unusual to observe radiolucencies within the sphenoid bones of the newborn and infant, though they disappear by the second or third year. These are residua of vascular channels, considered by some to be the embryologic and anatomic basis of congenital defects in the sphenoid bones, which result in craniopharyngeal encephalomeningoceles. Accordingly, Heinecke (1882)[43] identified the sphenoorbital 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,[44] 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. 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 Encephalomeningoceles

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[45] and Virchow,[46] where the masses bulged through the nares and mouth.

Sphenoethmoidal Encephaloceles. Sphenoidal encephaloceles present in the posterior nasal cavity, extending through the interval between the sphenoid and ethmoid bones, at the planum sphenoidale.

Sphenoorbital 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 Encephaloceles (Figure 14.33)

The encephalocele extends through the ethmoid bone and into the nasal cavity posteriorly.

Frontal Ethmoidal Encephaloceles: 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

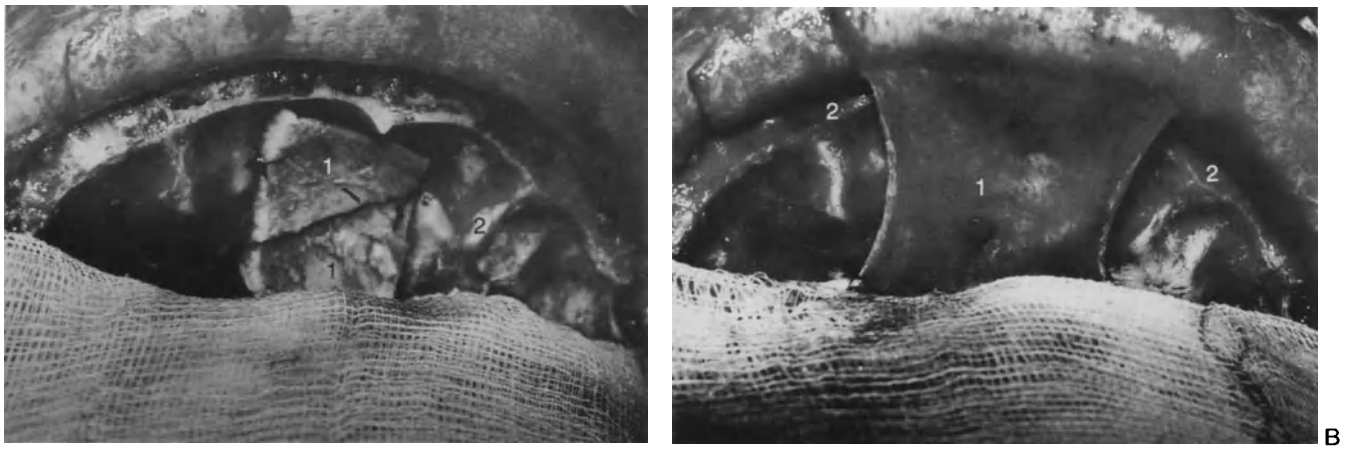


Figure 14.33. After the encephalocele has been resected, bone grafts should be lain into the defect and the dura reconstructed and/or grafted. (A) The defect in the ethmoidal bone is filled with bone grafts (*1*). The orbital roofs (*2*) are intact. (B) The prepared periosteal flap (*1*) is then brought over the frontal bone and bone grafts, before being sewn to the dura posterior 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.

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 encephalocele 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 encephalomeningoceles previously described and the cranioschisis and cranioencephalomeningocele that are described later.

It was not possible to provide any information concerning the geographic distribution of this particular congenital anomaly before the work of Suwanwela and coworkers,[34] since this classification had not yet been suggested and since 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 eight-year period they treated 100 cases of sincipital (frontoethmoidal) encephalomeningocele, but only 1 temporal, 3 vertex, 11 occipital, and 35 spinal meningoceles, stating that the incidence of sincipital encephalomeningocele is one in 5000 living people in Thailand. They do not give information concerning incidence of the anomaly among live births or stillbirths.

At birth, the sincipital encephalomeningocele 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 encephalomeningocele 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 encephalomeningocele (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 encephalomeningocele 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 encephalomeningocele and reconstruct the craniofacial anomaly.

Nasal Orbital Encephalomeningocele

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, 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 frontoorbital encephalomeningocele 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. Take care to 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 encephalomeningocele is not bilateral. The dura is separated well behind the posterior rim of the frontoethmoidal defect(s), and then the dural covering of the encephalomeningocele 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 encephalomeningocele 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, so that it will 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 encephalomeningocele 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 Encephalomeningocele

(Figure 14.34)

The encephalomeningocele 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 superiorly, 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 encephalomeningoceles. Because of the superior displacement of the nasal bone and the midline location of the encephalomeningocele, 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. 14.35).

Nasal Frontal Encephalomeningocele

The extrusion of the nasofrontal encephalomeningocele 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 are displaced inferiorly and lateralward. The nasal septum, cartilage, and bones have normal relationships with one

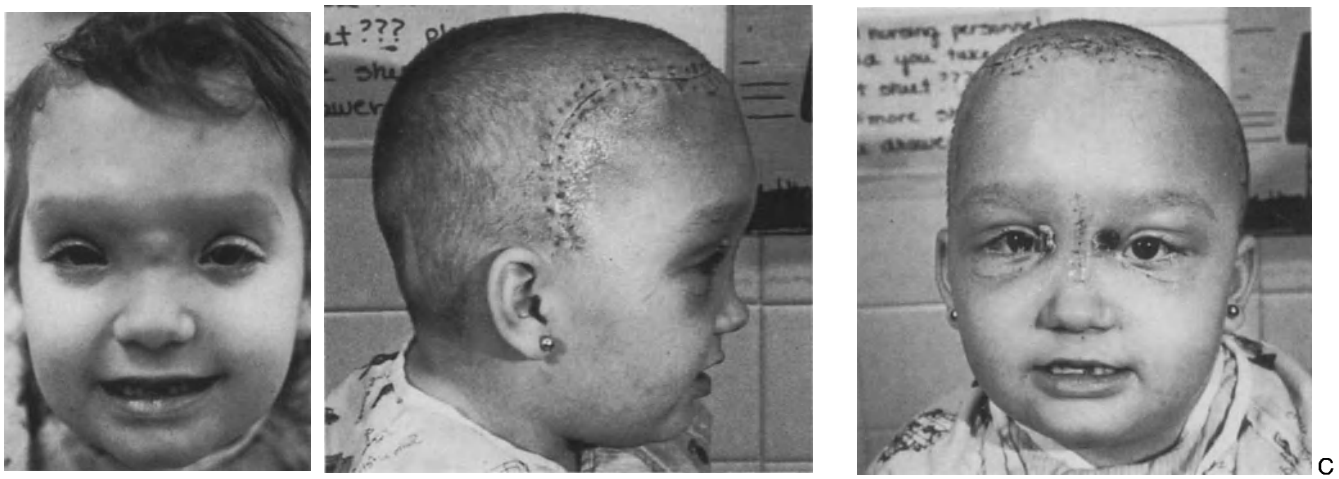


Figure 14.34. (A) Preoperative nasal ethmoidal encephalocele. (B and C) Postoperative results. Note that the procedure was performed by a combined intracranial and nasal-frontal approach.

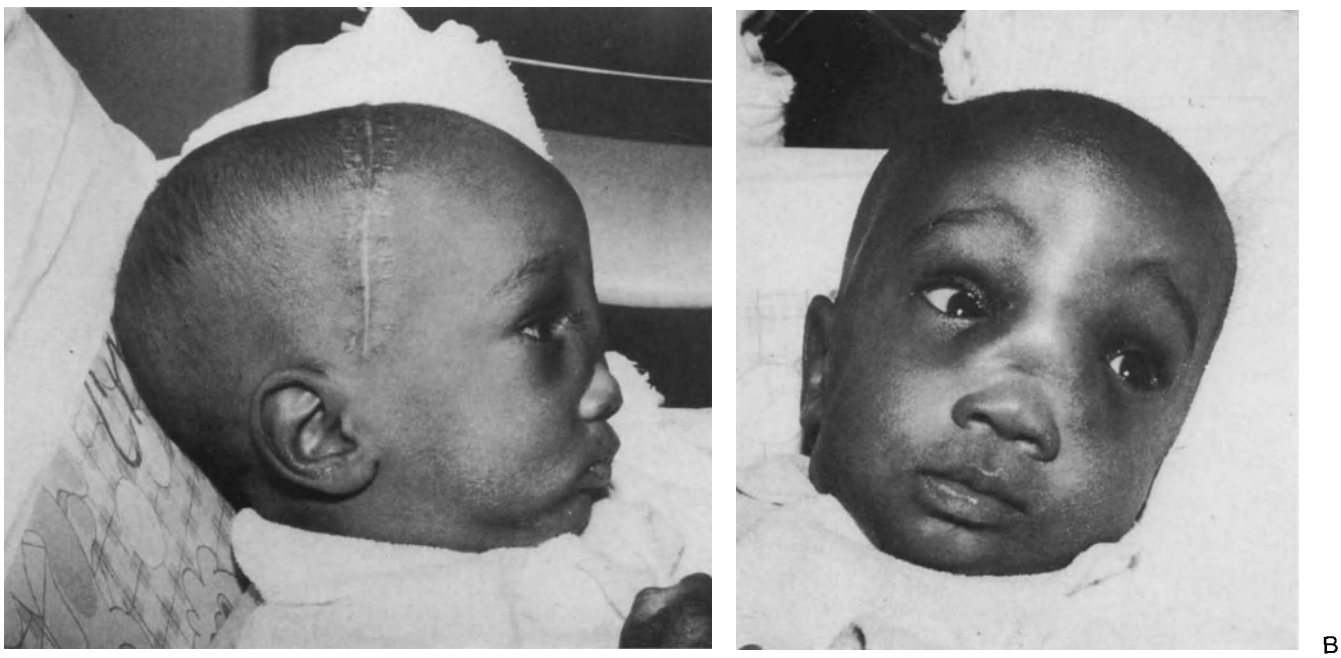


Figure 14.35. Postoperative result of a repair of the nasoethmoidal subgroup of sincipital (frontal ethmoidal) cranioencephalocele. (A) The repair was performed by the intracranial route, and all of the encephalocele was removed, (B) The dura is repaired and the bony defect is grafted with autogenous bone. Reconstructive nasal surgery still needs to be performed.

another and the frontal processess of the maxilla. Thus, the encephalomeningocele 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 encephalomeningoceles, 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 encephalomeningocele. 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 encephalomeningocele subgroups tend to become less severe over the months and early years following resection of the encephalomeningocele 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 encephalomeningocele, or in two stages, the orbital reconstruction is performed only after the encephalomeningocele 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. I have never seen a case of acrania.

Cranial Encephalomeningocele

Cranial encephalomeningoceles 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 fontanel, interparietal, posterior fontanel, and occipital (cervical) encephalomeningoceles. As the craniofacial encephalomeningoceles, 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 encephalomeningocele 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 through lachrymal gland, or, more likely, 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 either 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.

Interfrontal Encephalomeningocele

Interfrontal encephalomeningocele 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 inferiormedial 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 mini-

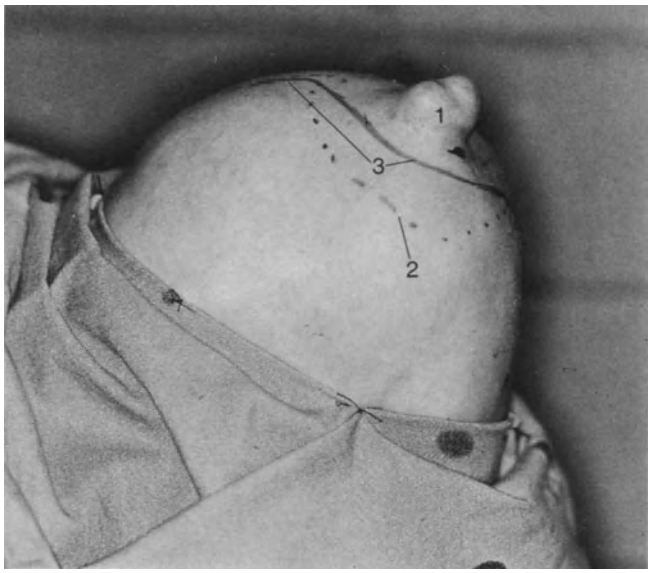


Figure 14.36. This is an interparietal encephalomeningocele that was operated on because the underlying brain was intact (as determined by cerebral angiography and pneumoencephalography). The encephalomeningocele (*1*) is bilobed and quite small, whereas the bony defect (*2*) is large. An S-shaped incision (*3*) was used to expose the sac and stalk of the encephalomeningocele.

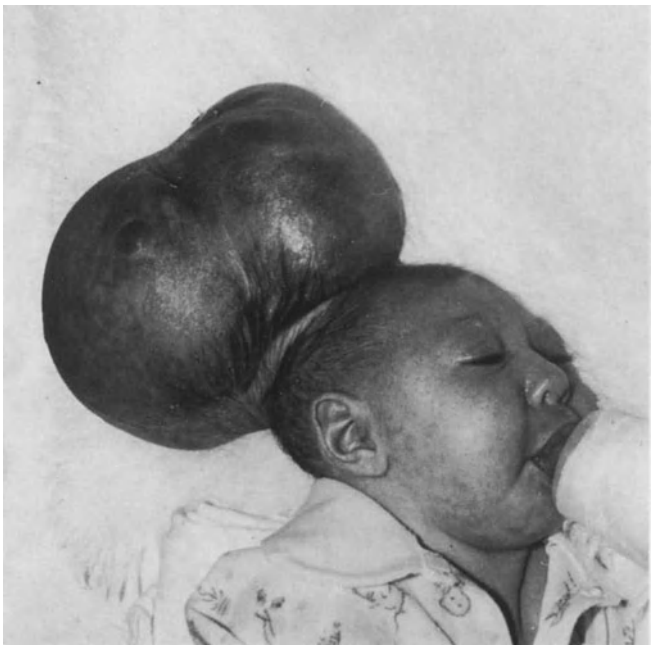


Figure 14.37. This child has an enormous interparietal encephalomeningocele, which contained most of both cerebral hemispheres, the anterior and middle cerebral arteries. This type of encephalocele is not a surgical entity.

mally 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 Fontanel Encephalomeningocele

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 encephalomeningocele, with the exception of consideration of implantation of a bone graft.

Interparietal Encephalomeningocele

(Figures 14.36 and 14.37)

The anterior and posterior fontanels represent, respectively, the anterior and posterior extremities of the defect, which is bordered laterally by the parietal bones. These defects invariably 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.

Surgical considerations should be limited to serving humane, social, and psychological purposes.

Posterior Fontanel Encephalomeningocele

(Figure 14.38)

Posterior fontanel encephalomeningoceles 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.

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 encephalomeningoceles. 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

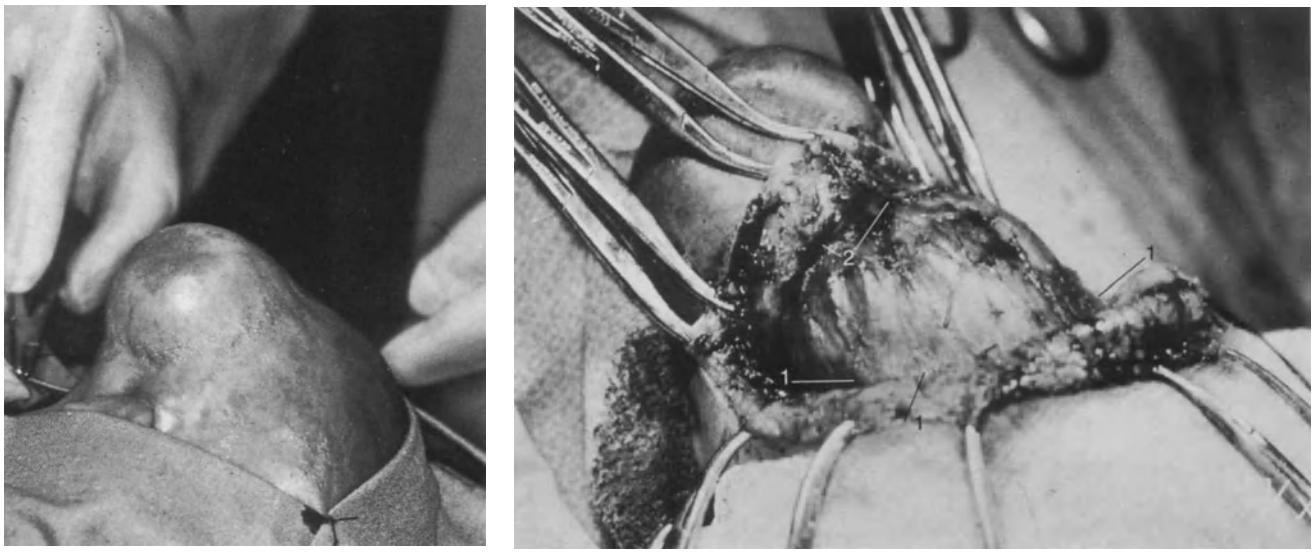


Figure 14.38. Encephalomeningocele of the posterior fontanel. (A) Note that this posterior fontanel encephalomeningocele is completely covered by normal skin. (B) The base of the dural sac (*1*) is quite broad in another child with a posterior fontanel encephalomeningocele which was bilobed, with a constriction (*2*) at its midportion.

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 with cerebral angiography, 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 dysgenic brain in the cranial cavity.

Craniocerebral Disproportions: Chiari Malformations

Actually, it was Cleland,[47] in 1882, who first gave a detailed description of what is now referred to as the Chiari II malformation. Arnold[48] 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 course, 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 the 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:

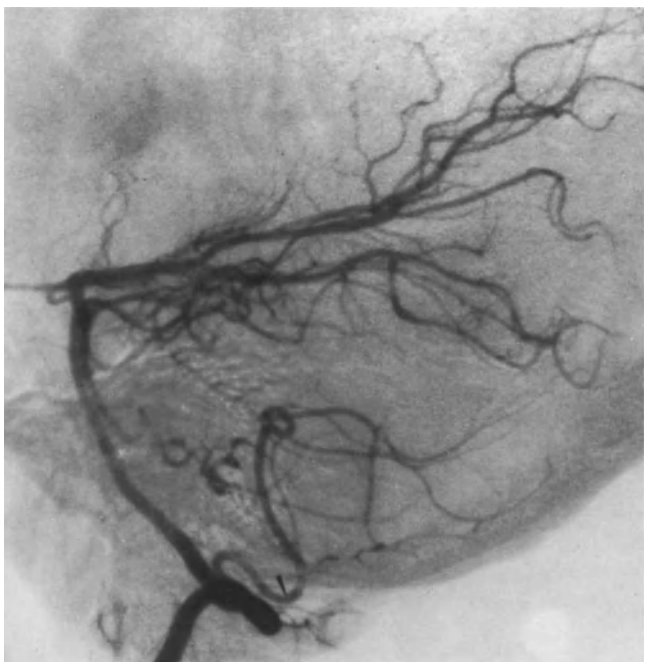
Chiari I: characterized by variable displacement of the tonsils into the upper cervical canal

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

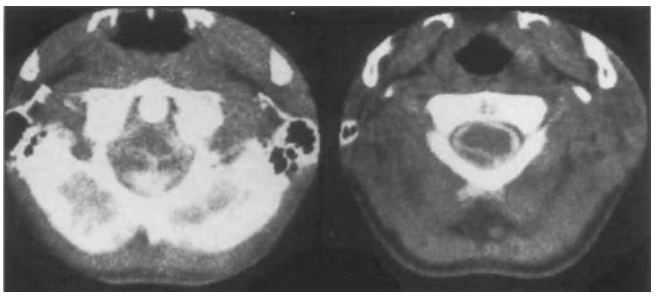
Chiari III: downward displacement of the medulla oblongata and herniation of portions of the cerebellum and, in some instances, the hindbrain into a high cervical or suboccipital meningocele

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

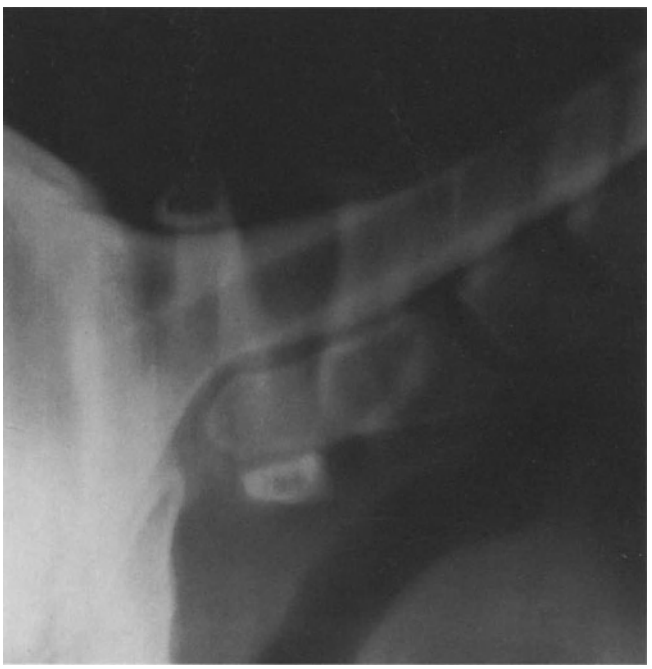
Any discussion of surgery to treat either the Chiari I



A



B



C

Figure 14.39. (A) Chiari I, anatomical variant. The caudal loop (arrowhead) of the posterior inferior cerebellar artery (PICA) is located inferior to the rim of the foramen magnum. (B) This is a CTT of a child with Chiari I malformation. Notice the displacement of the tonsils into the upper cervical canal (Courtesy Thomas Naidich). (C) Myelography illustrates in sagittal section the extension of tonsils below the foramen magnum (Courtesy Giorgio Belloni).

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, medulla spinalis.

Chiari I "Malformation" (Figure 14.39)

The Chiari I "malformation" is not a malformation! It is 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. Its diagnosis is either angiographic or CTT. It may, consequently, be encountered in normal and asymptomatic children as well as an individual who suffers from hydrocephalus and syringomyelia. If metrizamide cisternography in a patient who has cerebellar tonsils below the level of the foramen magnum reveals ample spaces around the medulla and a normal cisterna magna, one must look elsewhere for the causative factors or conclude they cannot be identified. By definition then, a Chiari I anatomical variant is a neuroradiological diagnosis, more specifically a vertebral angiographic or CTT 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. This is an anatomical variant, not a pathogenic factor, because the location of the caudal loop of PICA, in and of itself, cannot cause neurological deficit. Similarly, the tonsils may be located within the foramen magnum and not be pathogenic. Certainly, decompressing the foramen magnum, along an arc no greater than one fourth of its circumference, in a child with normal cisterna magna and perimedullary cisterns, cannot be expected to be therapeutic.

Figure 14.40. Chiari II malformation. A large portion of the squamous occipital bone has been resected (*1*), as have the arches of C1 (*2*)–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.

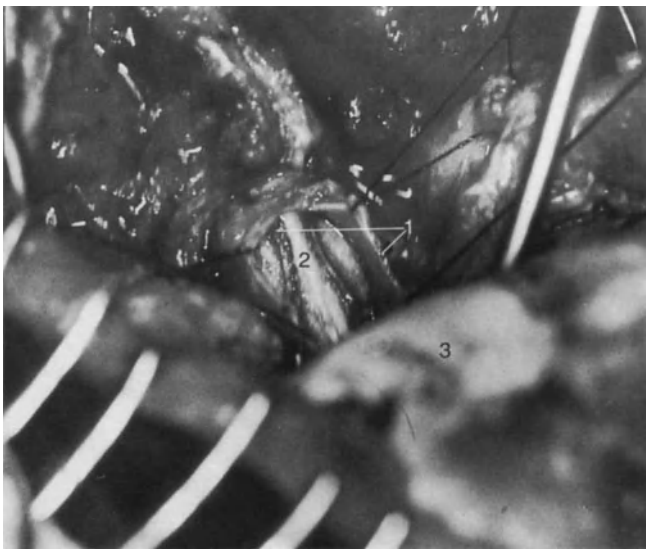


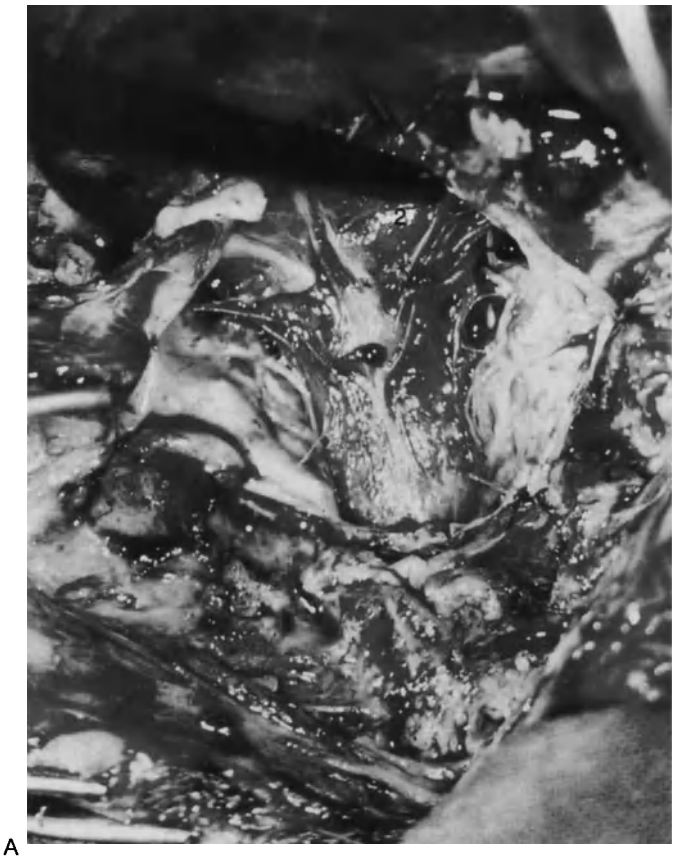
Figure 14.41. This is another child with a Chiari II malformation on whom cervical laminectomy and opening of the dura (*1*) was performed. The child is prone. One has an excellent view of the spinal cord (*2*), but the ledge of the squamous occipital bone (*3*) obstructs completely the surgeon's line of vision to the rim of the foramen magnum, and impairs access to the cerebellar hemispheres. Cephalad is inferior.

Chiari II Malformation (Figures 14.40 to 14.44)

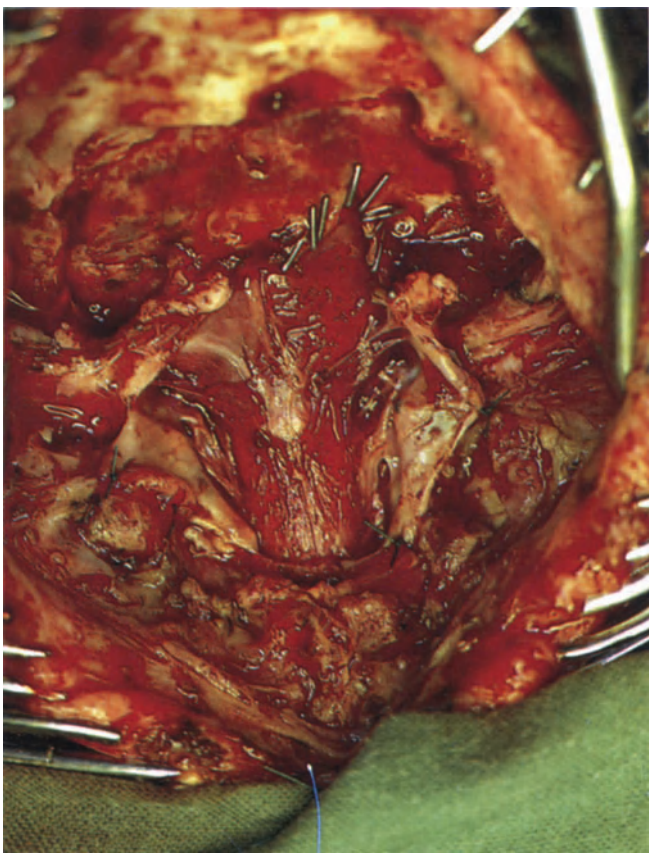
The Chiari II malformation is an extraordinarily complex and variable anomaly, one which is still neither understood nor has an effective surgical treatment or palliation. Attractive though it may be to the neurosurgeon, compression of the neural structures within the posterior fossa and foramen magnum 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 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 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 grey 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 side, 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 defi-



A



B

Figure 14.42. (A) This is still another child with a Chiari II malformation, on whom resection of C1 and C2 was performed to decompress the underlying cervical spinal cord (*1*). The cerebellar tonsils (*2*) are seen at the level of the rim of the foramen magnum. The photograph was taken from the caudal end of the patient, along a line of vision perpendicular to that of the surgeon, putting into relief the difficulty or impossibility for the surgeon to visualize adequately the squamous occipital bone and foramen magnum. Cephalad is superior. (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.

cits. 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 unacceptable 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 procedure.

If one undertakes to perform a decompressive procedure for treatment of the Chiari II malformation (a procedure I discourage), he should be alerted that (1) the squamous portion of the occipital bone 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 neu-

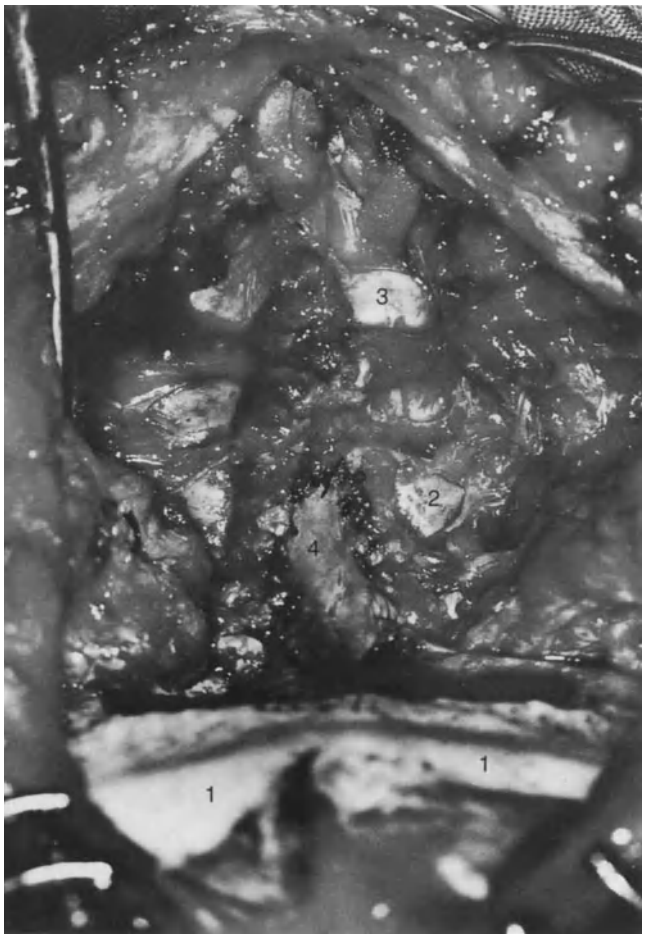


Figure 14.43. The ledge-like squamous occipital bone (*1*), and resected arches of C1 (*2*) and C2 (*3*) are illustrated, as is the patch graft (*4*) of fascia used to cover the underlying cervical cord after the dura had been opened to decompress the cord. Cephalad is inferior.

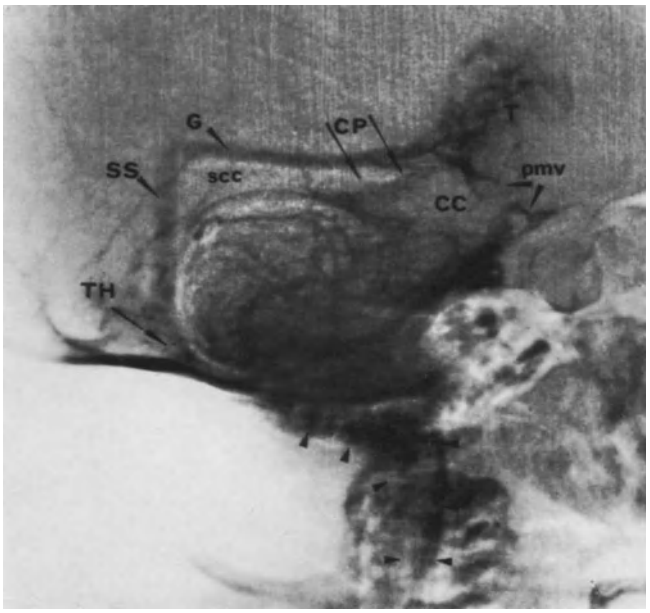


Figure 14.44. Posterior fossa venographic study of the same child illustrated in Figure 14.43. Note the staining of the posterior two thirds of the thalamus (*T*), in order to appreciate fully the anteroinferior displacement of the thalamus so typical of aqueductal stenosis. The perimesencephalic vein (*pmv*) is well shown as is the hypertrophy of the crus cerebri (*CC*). The collicular 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 a right angle. 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 cerbellar hemispheres and the hemispheral 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.

tral. After the skin incision has been made and the muscles dissected from the squamous occipital bone and the desired posterior cervical arches, self-retaining retractors maintain adequate exposure of the bony structures. Because of the "horizontalization" of the squamous occipital bone, 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 spine, however, is complete.

After the periosteum has been stripped from the squamous occipital bone and the posterior arches of the cervical vertebrae, one may perform the craniectomy and laminectomy most precisely with the use of power driven diamondhead burrs, extending the craniectomy on each side to the most medial aspect of the condyles of the occipital bone. 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 oblon-

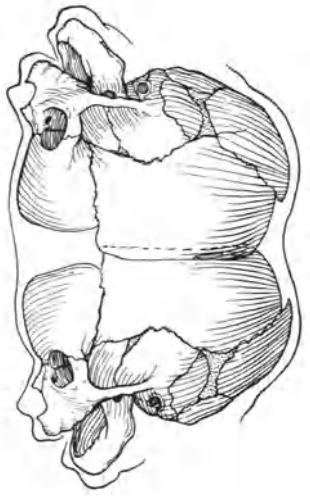


Figure 14.45. Röentgenograms of the skull made at the age of 5 days. Note absence of bony septum and overriding of parietal bones.

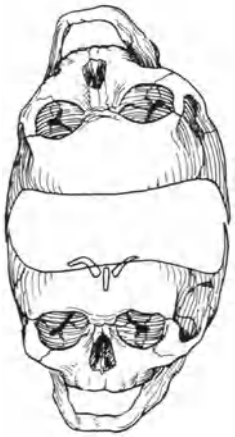


Figure 14.46. Pneumocephalograms made at the age of 6 weeks. Air was injected into lumbar subarachnoid space of R.D.B. The subarachnoid air is confined to one brain. Note the height of the III ventricle between lateral ventricles. (There was questionable agenesis of the corpus callosum.)

gata 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 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.

The muscle and the skin are closed, as already described for suboccipital and cervical procedures.

Separation of Craniopagus Twins

The following is a transcription of the paper "Surgical Separation in Craniopagus Twins" by H. Grossman, O. Sugar, and P. Greely.[49]

On Sept. 16, 1951, craniopagus twins were born in Rock Island, Ill. The record of birth and course during the first six weeks has been adequately described by Durr.[50] 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 Oct. 2, 1951. They were in a conjoined state of *craniopagus parietalis* [Figure 14.45]. 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 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 third interspace. It was considerably louder in intensity in R.D.B. 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 face. 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 [Figure 14.46] by the lumbar route disclosed separate subarachnoid spaces and indicated a dural septum between the two brains.

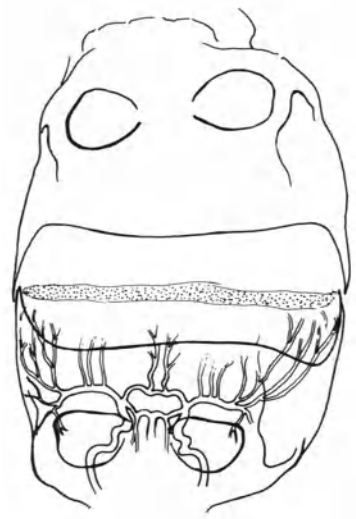


Figure 14.47. Angiogram in R.D.B. at age 5 months. Iodopyracet (Diodrast) in the middle cerebral artery distribution appears to cross over to the opposite hemisphere, but the midline dye is actually from the anterior cerebral arteries. No dye appears in the cerebrum of R.L.B.

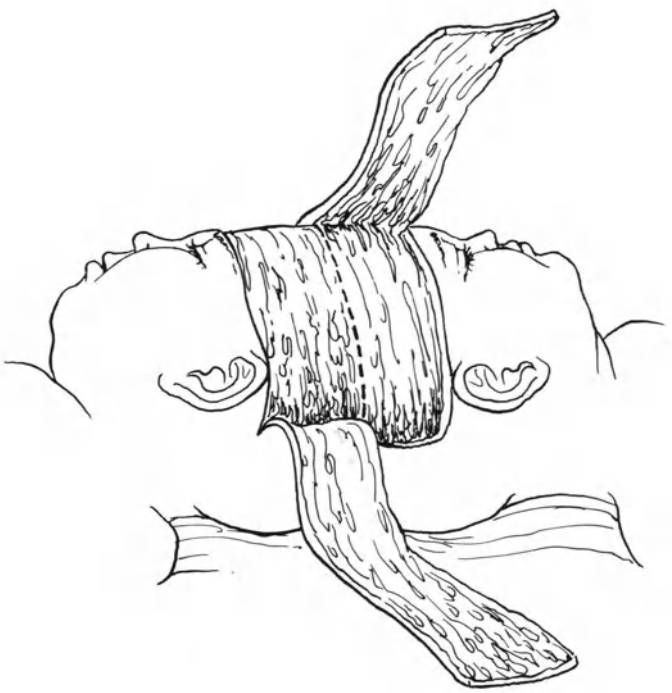


Figure 14.48. Pedicle flaps, which were replaced after the first attempt at separation on Nov. 26, 1952.

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 intraarterial 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. [Figure 14.47] 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 fontanel at age 6 weeks, the sinus could not be found. Even through a midline anterior burr 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.

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 as 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 May 14, 1952 [Figure 14.48]. After three stages, the scalp flaps were completely divided around their peripheries. During the next five 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 Nov. 26, 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 $12^1/_2$ hours. 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

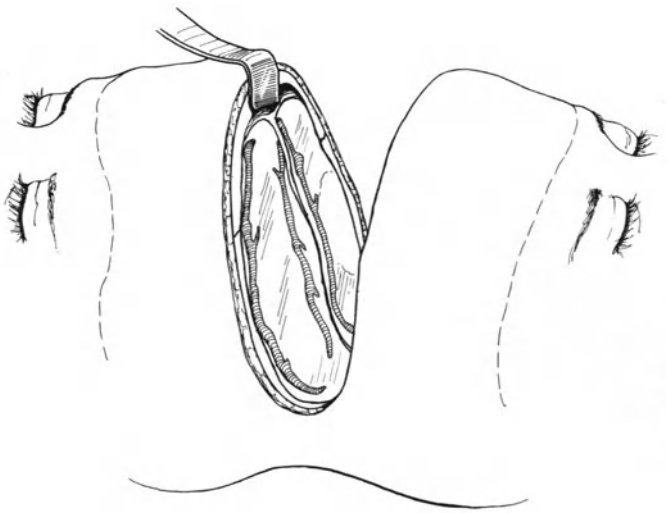


Figure 14.49. Artist's drawing of findings during final separation (age 15 months).

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 six and a quarter hours, 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 hours.

Final separation

The final separation was made on Dec. 17, 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 [Figure 14.49]. 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

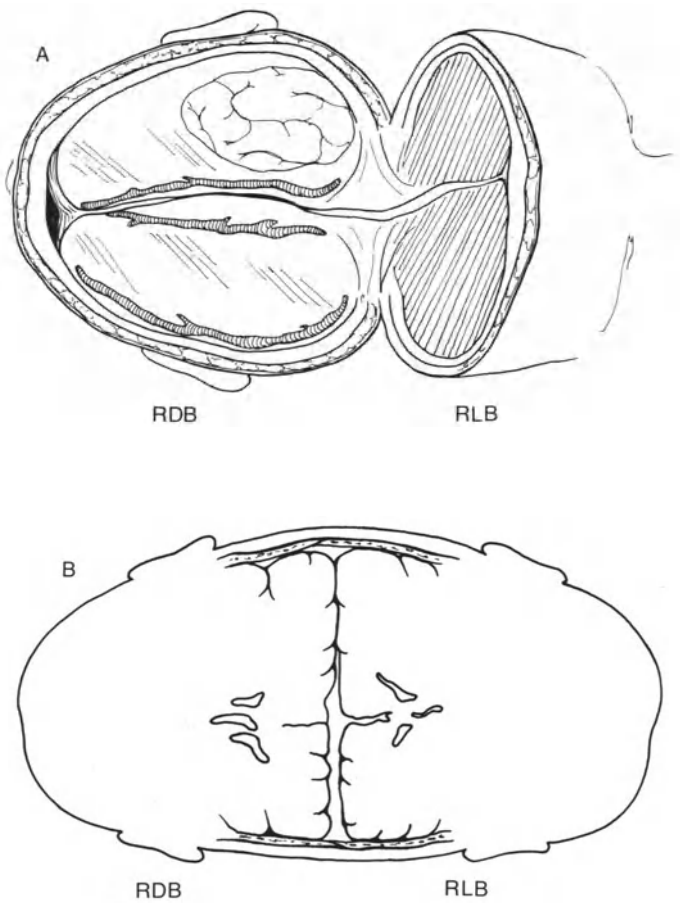


Figure 14.50. (A) Diagram of arrangement of abnormal dural venous sinuses as disclosed by operation. Note the dural defect over the right hemisphere of R.D.B. (B) Reconstruction of arrangement of dural venous sinuses, seen in coronal section. Note the apparent absence of the falx in R.D.B.

three in number [Figure 14.50]; 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, $10^1/_2$ hours 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 burdern, so this, too, was omitted and we were content to use sterile aluminum foil to cover the top of each head before putting 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 $12^1/_2$ hours. R.D.B. received 2,500 cc of blood and R.L.B. 3,500 cc 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 eight hours 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 of body weight per day. No evidence of systemic toxicity was discovered, although the antibiotic was given until his death 34 days after separa-

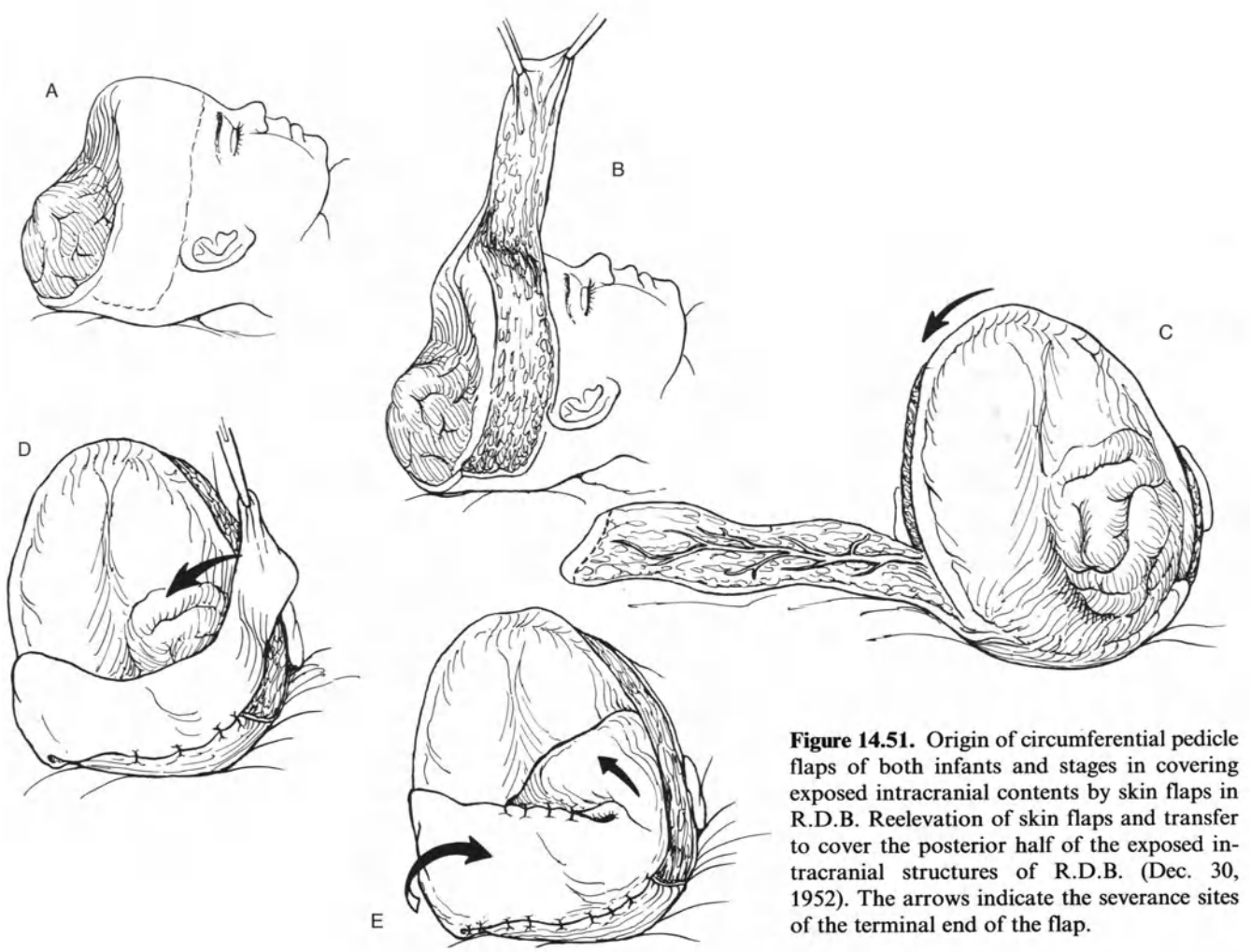


Figure 14.51. Origin of circumferential pedicle flaps of both infants and stages in covering exposed intracranial contents by skin flaps in R.D.B. Reelevation of skin flaps and transfer to cover the posterior half of the exposed intracranial structures of R.D.B. (Dec. 30, 1952). The arrows indicate the severance sites of the terminal end of the flap.

tion. 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 to 50,000 units of trypsin in 2 to 3 cc diluent at pH 7.4) every six to eight hours. On several occasions, Alevaire (a solution of glycerine, 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 post-operative days, and thereafter nutrition was maintained by feedings through a # 19 polyethylene nasogastric tube. UP to 4,500 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 Jan. 19, 1953, and he died on Jan. 20, 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 fourth postoperative day, he was irritable and refused oral feedings. There was an odor from the head dressing characteristic of a *Ps. 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 Dec. 30, 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 [Figure 14.51]. 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 [Figure 14.52].

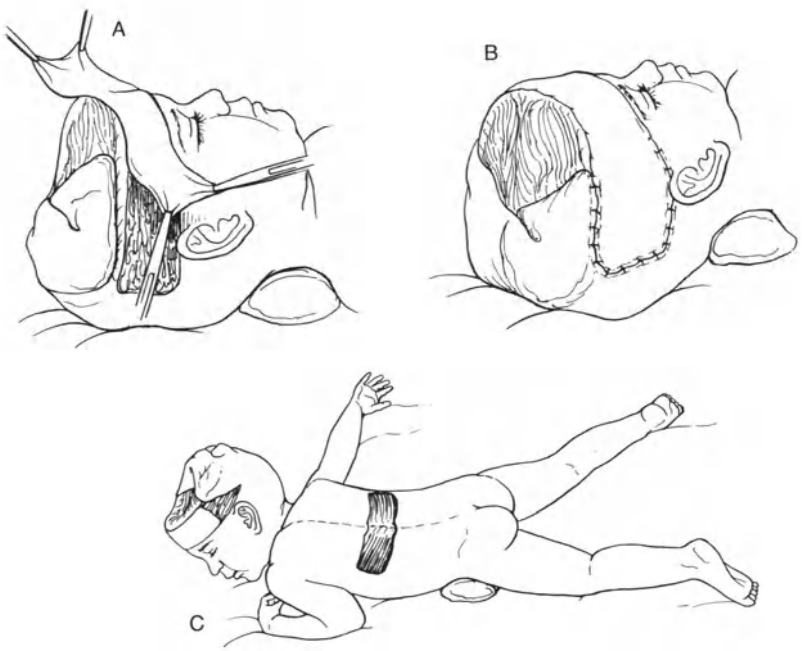


Figure 14.52. Covering of exposed donor site of pedicle flap with split-thickness graft from the back of the pedicle flap (Jan. 6, 1953).

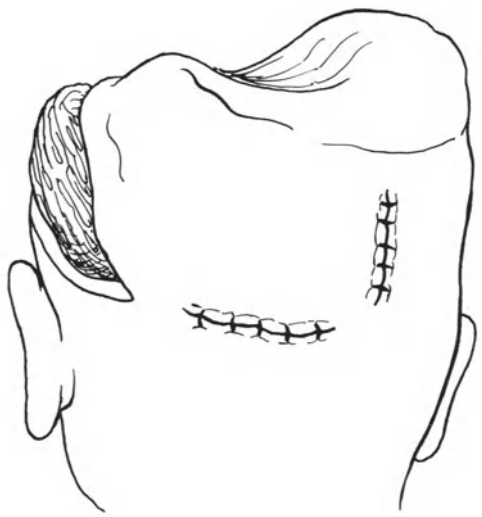


Figure 14.53. Extension of the proximal end of the full-thickness flap by delay of circulation to the base (Jan. 17, 1953).

On Jan. 17, 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 [Figure 14.53]. On Feb. 10, 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 [Figure 14.54]. Donor sites were covered in each case by split-thickness grafts, some of which had to be revised later.

On March 11, the "dog ear" in the flap was divided in its central portion. The anterior part of the flap was reelevated 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 first month, but the astereognosis still continues, although less marked than before. There are no hyperactive stretch reflexes on the left, nor is there any loss of common sensibility. There are intermittent Babinski signs on both sides, which are 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 three 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.

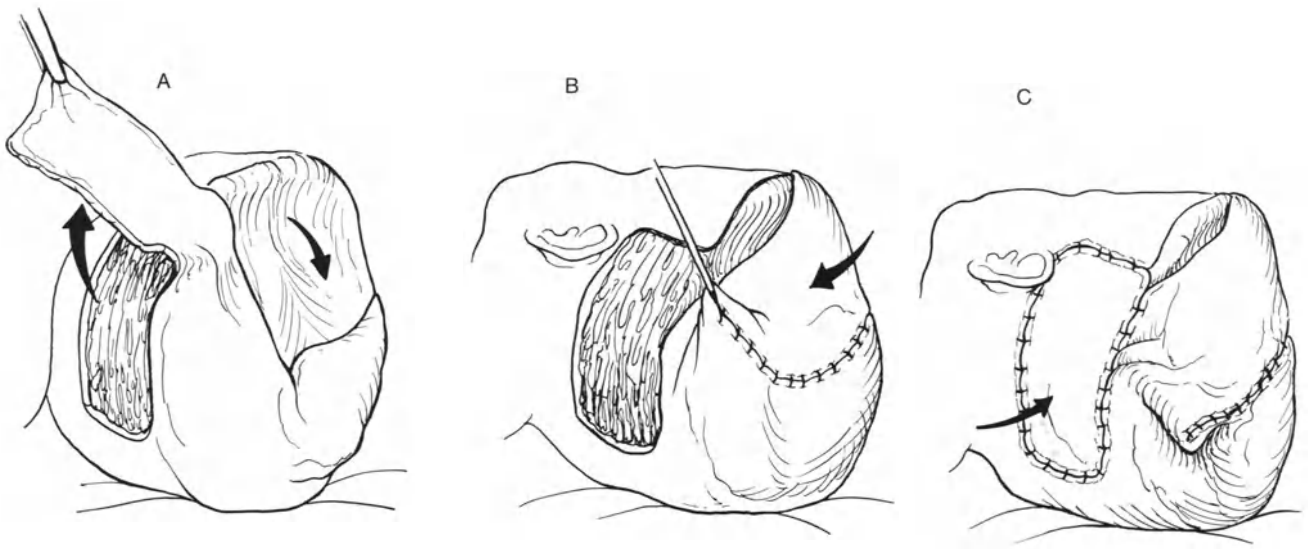


Figure 14.54. Completion of the rotation of the full-thickness graft to cover the remaining exposed intracranial structures (Feb. 10, 1953). The donor site is covered by a split-thickness graft (March 1, 1953).

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 this 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 for his permission to transcribe this classic report.

Vertebrospinal Congenital Anomalies

The matter of vertebrospinal traumatology has already been discussed. Suffice it to repeat here that this is a most unusual event in infancy and childhood.

Vertebrospinal congenital anomalies, 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, meningomyelocele, meningocele; lipomeningocele, lipomeningomyelocele; some hamartomas (lipoma, dural fibrolipoma, leptomyelolipoma, anterior spina bifida, 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 almost invariably blends imperceptibly into another hand, in many instances such as lipomeningomyelocele and hamartomas, several may coexist.

Before undertaking an operative procedure, one asks

himself what specifically he 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[51] *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 medullaspinalis, removing it affords similar, but not identical, beneficent results as removing a neuronoma 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 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)[52]; Lebedeff (1881)[53]; von Recklinghausen (1886)[54]; Morgagni (1761)[55]; Patten (1953)[56]; James and Lassman (1962)[57]; Ingraham and Matson (1954)[58]; Talwalker and Dastur (1970)[59]; Emery and Lendon (1973)[60]; Rokos (1973)[61].

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 through all children born with a sack on their 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[62] is recommended:

Myelocele (Figures 14.55 to 14.59)

Failure of closure of the neural tube (myeloschisis) resulting in a flattening of the spinal cord. The 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 lays 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.* This confusion of two distinctly different dysraphic states has led some authors to suggest that it is possible to reconstruct the unclosed spinal cord in meningomyelocele. 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. 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 ar-

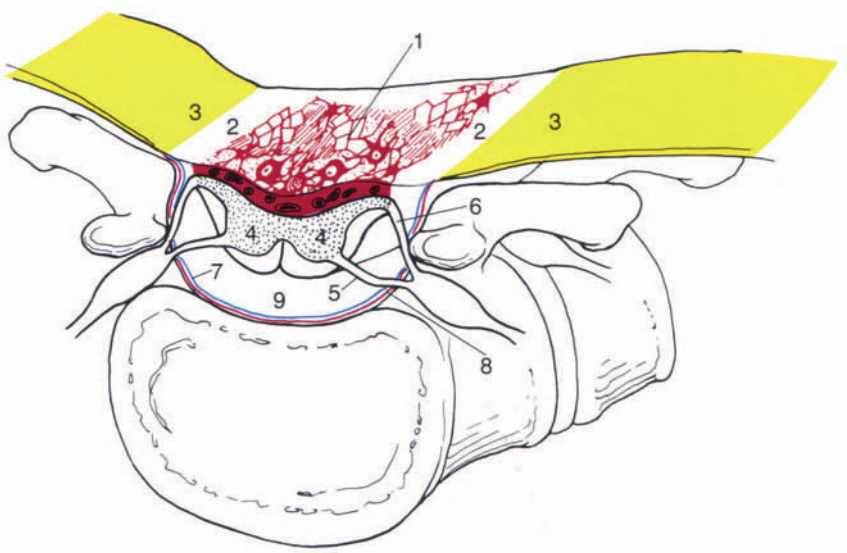


Figure 14.55. Simple myeloschisis (myelocele). Its surface (*1*) is a composite of glial scar, ectopic neurons, patches of ependyma, and degenerative changes. This is the medullovasculosa. It is bordered by a zone of tissue devoid of neural or epithelial tissue (*2*) which, in turn, is surrounded by 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*).

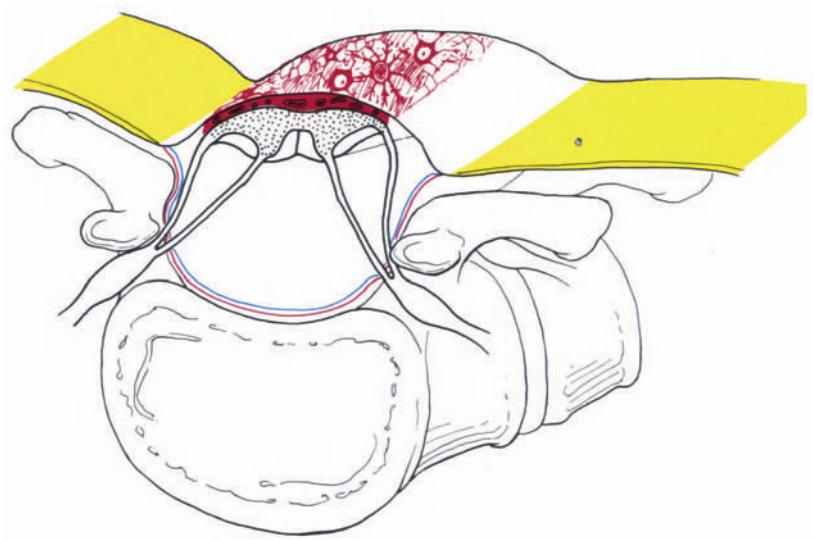


Figure 14.56. Cystic myeloschisis. This is a myelocele floating on an arachnoidal cyst, which separates it from the ventral surface of the spinal canal.

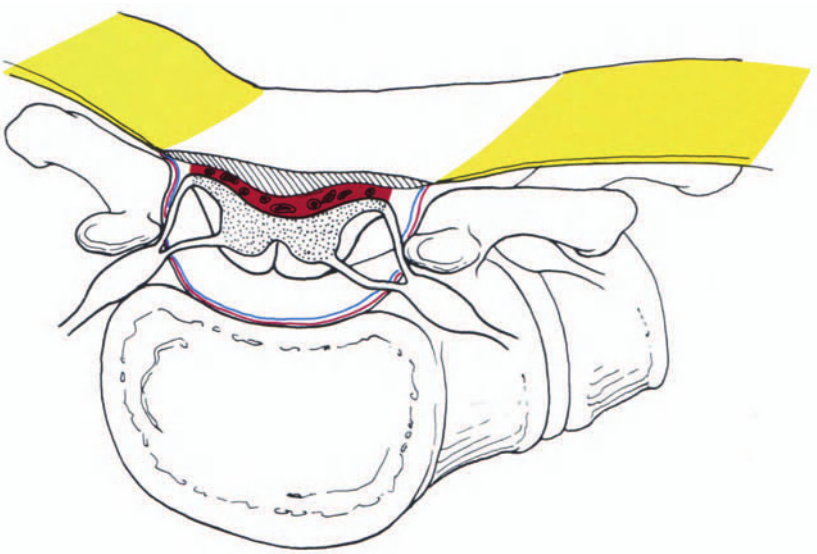


Figure 14.57. Scarred myelocele. The posterior surface of myeloschisis, simple or cystic, may be covered with granulation tissue, which is organized and epithelialized. This is a *scarred myelocele*, an entity easily confused with meningomyelocele.

eas 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 (Figures 14.60 to 14.65)
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. 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.

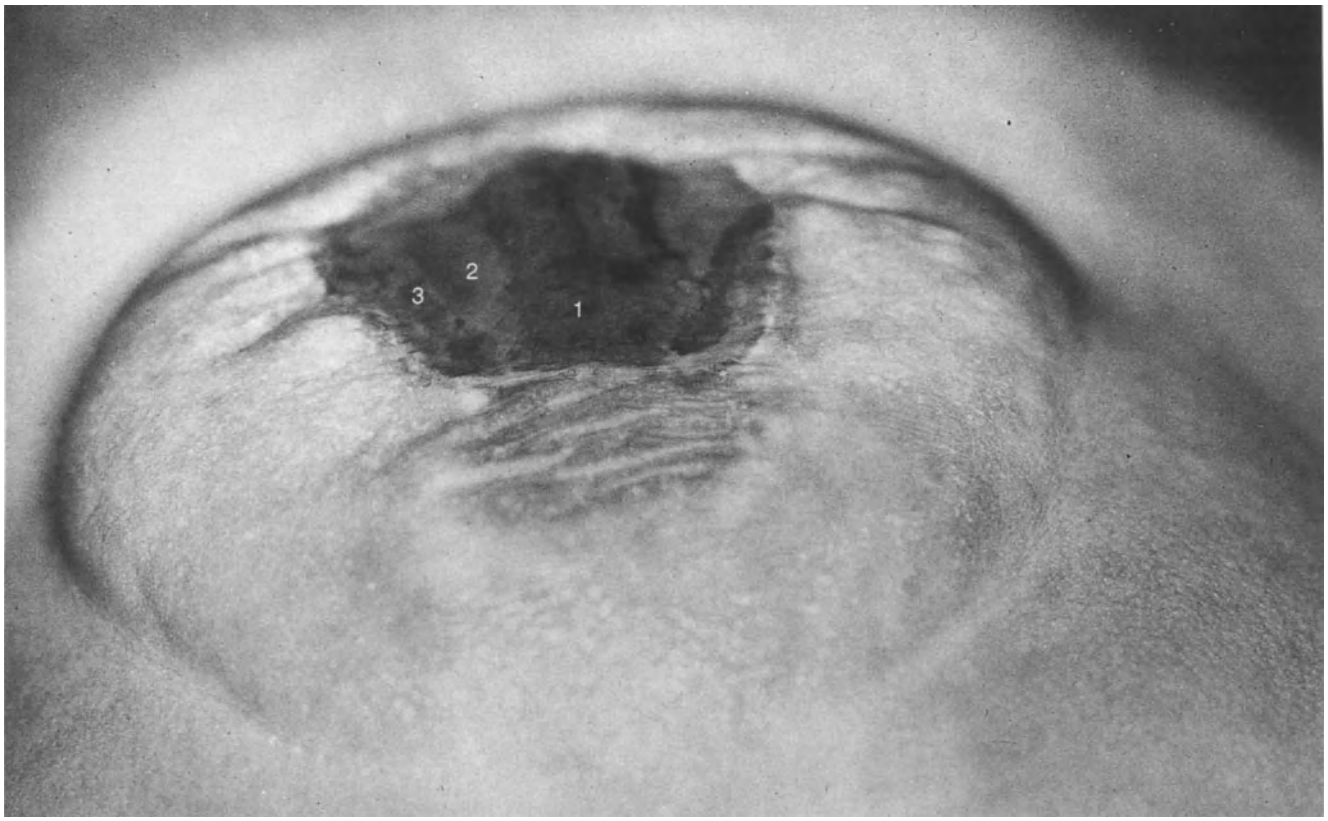
An incision, eliptical in form, is extended along the midline from superior to inferior, to the base of the meningocele, where it is bifurcated to either side of the pedicle, taking care to leave enough skin for the closure. At the inferior portion of the pedicle, the incisions from either side of the 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 his 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 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) on dysgenic (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 a 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.

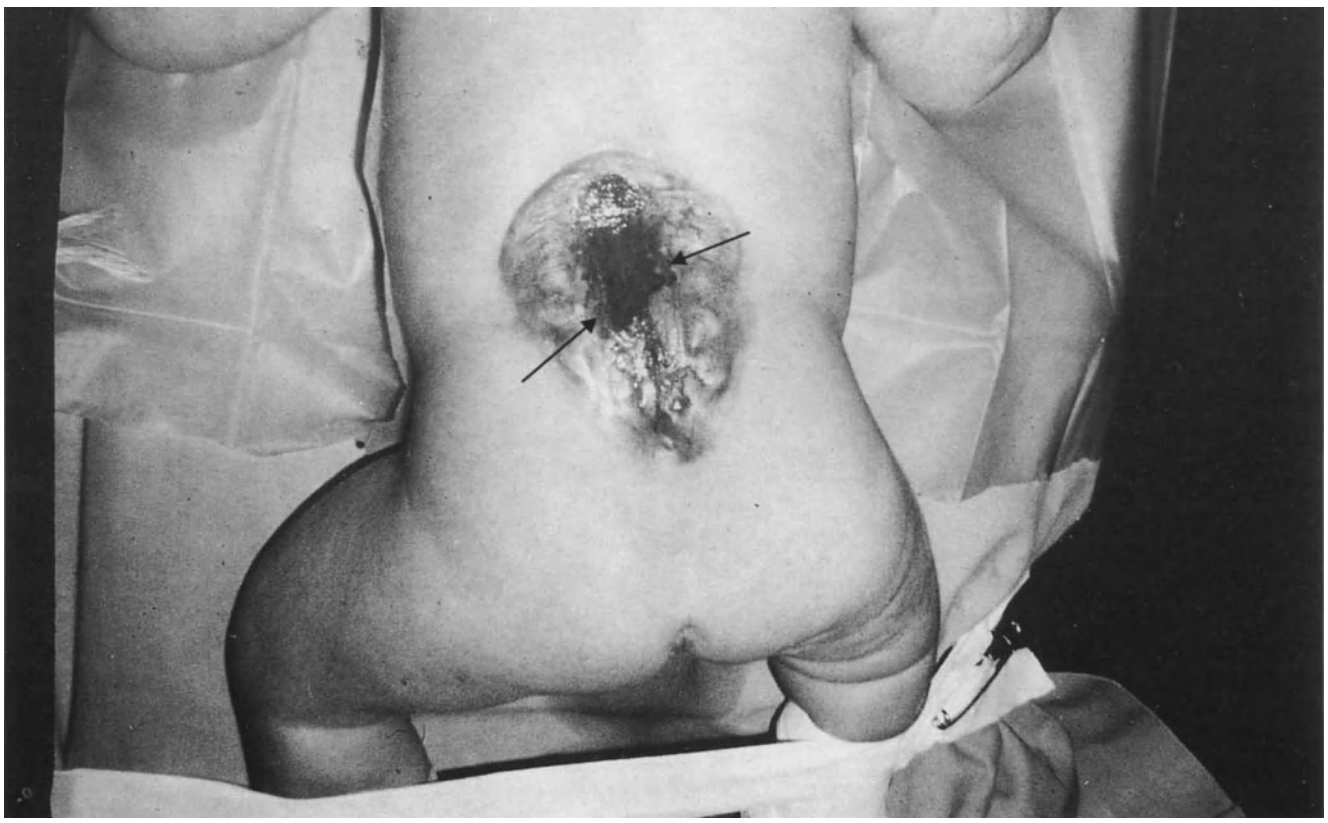
One of the two fashioned dural trapdoor openings is then laid snugly over the arachnoid and sewn to the inner surface of the dura on the opposite side, maintaining the normal dural circumference, using 4-0 or 5-0 sutures. If it is necessary to imbricate the dura at the superior and inferior durotomies 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 then 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

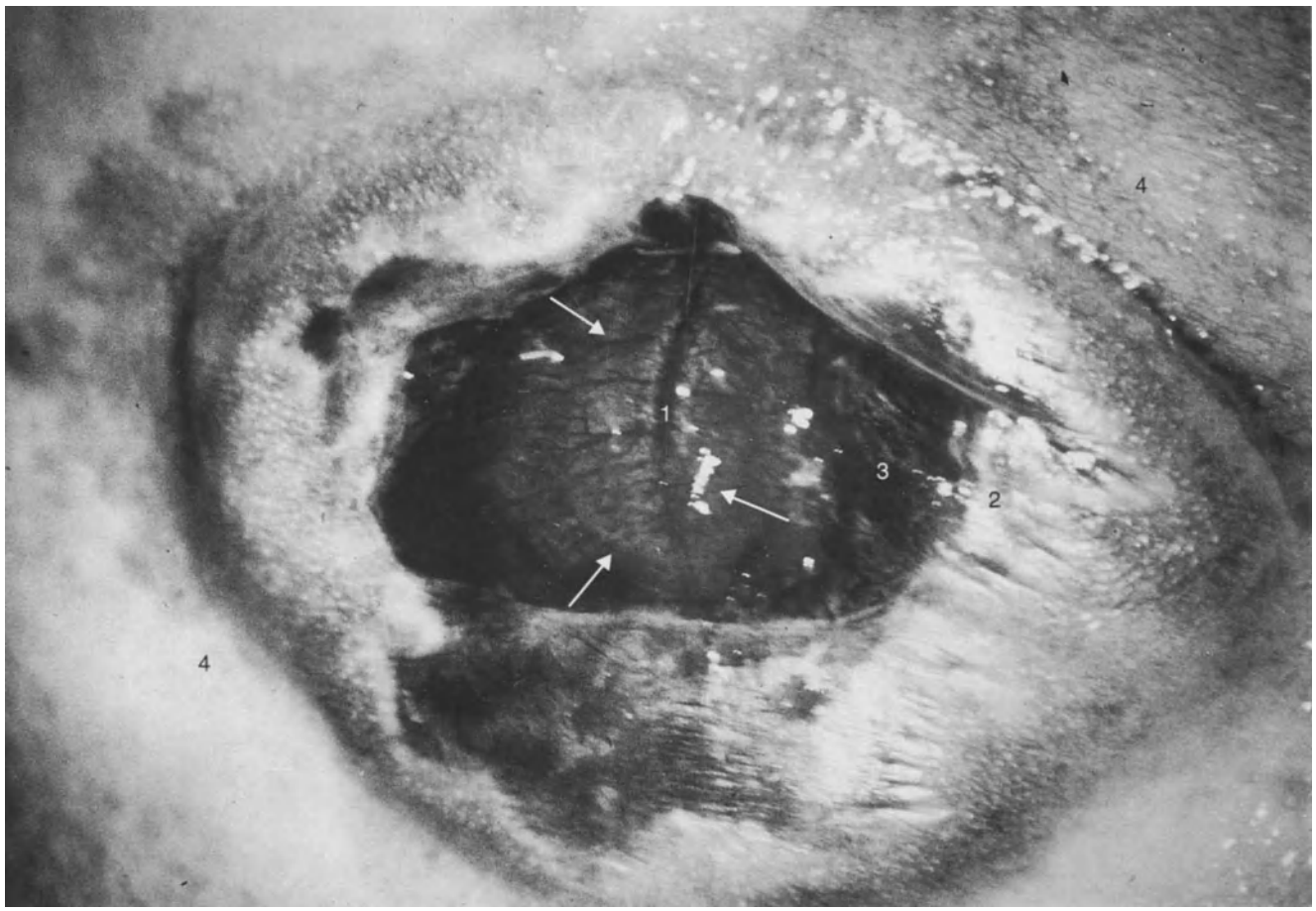
Figure 14.58. A nonscarified myelocele (A) is illustrated with a scarified myelocele (B) to permit one to appreciate how scarification converts the appearance of a myelocele into a dysraphic anomaly which is visibly almost indistinguishable from the meningomyelocele. (A) The myelocele (*1*) and its zona 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. ►



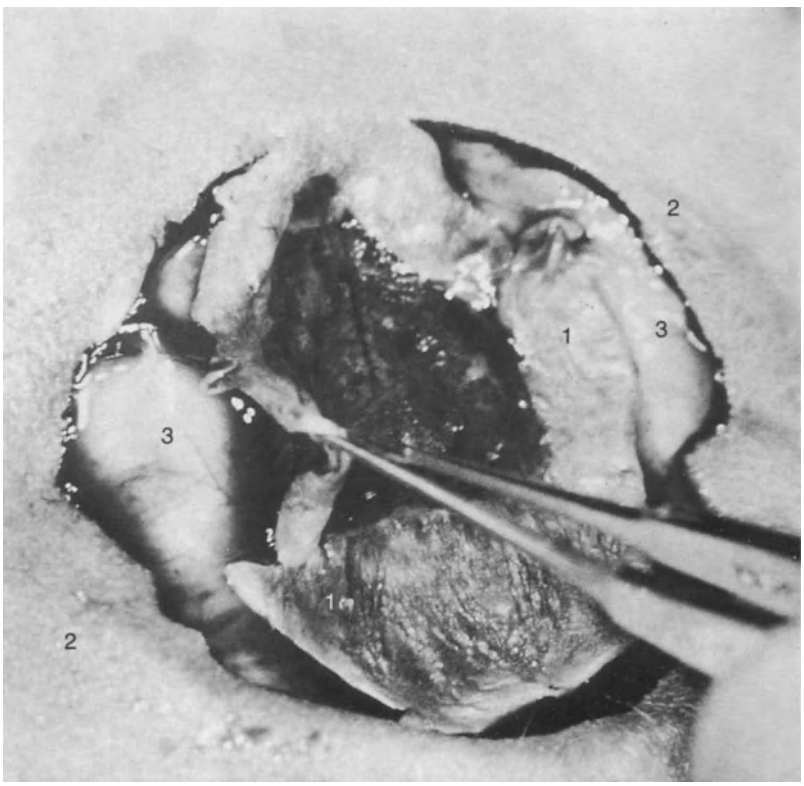
A



B

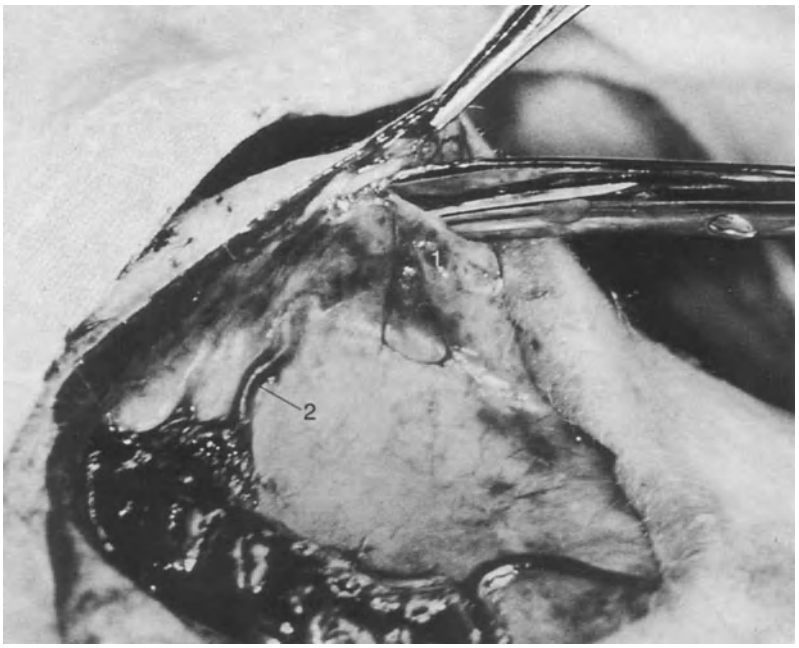


A

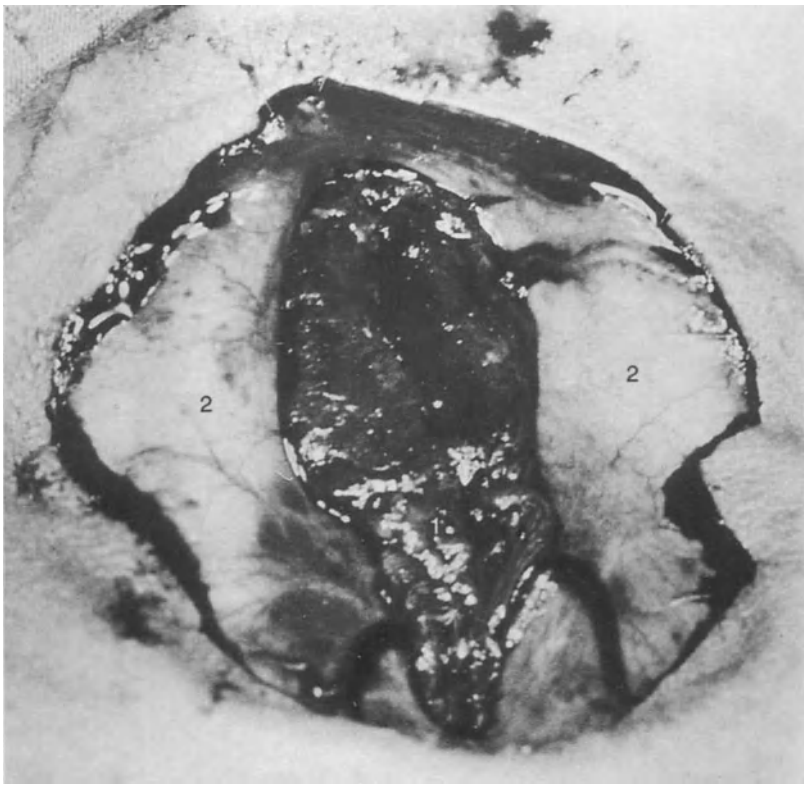


B

Figure 14.59. (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



C



D

time, one picks up the zona epitheliosa before proceeding to incise it perpendicular to the margin of the zona neurovasculosa. (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.

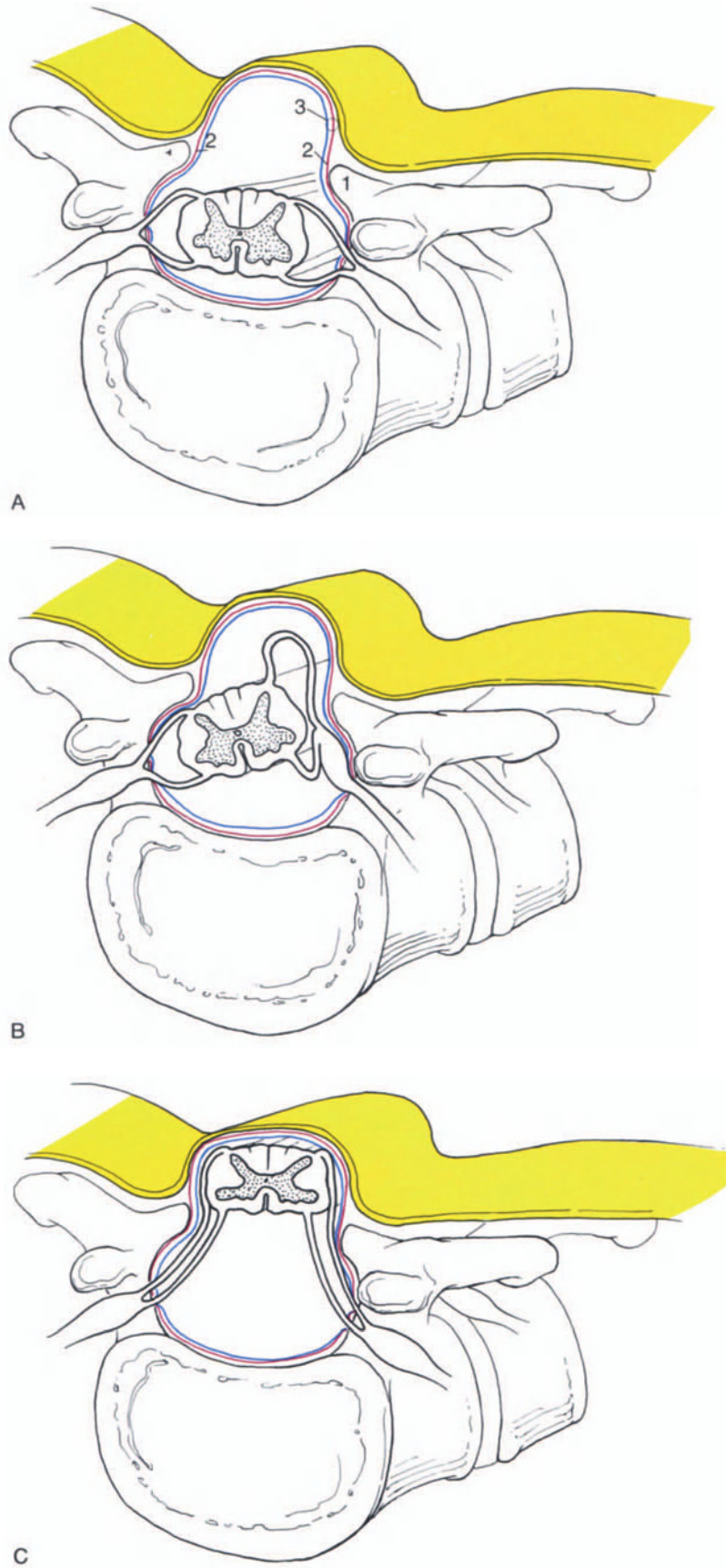
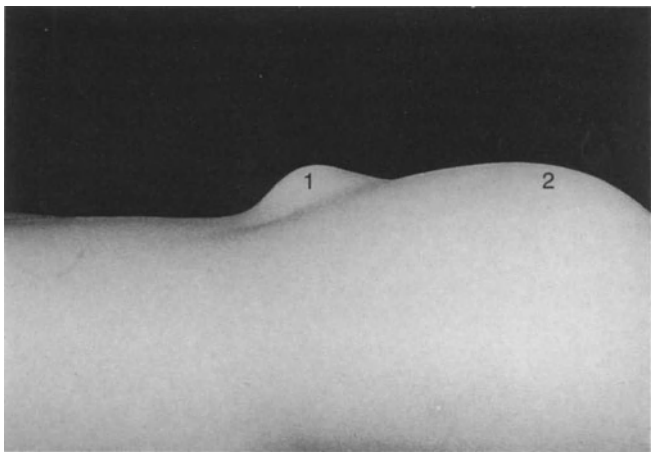
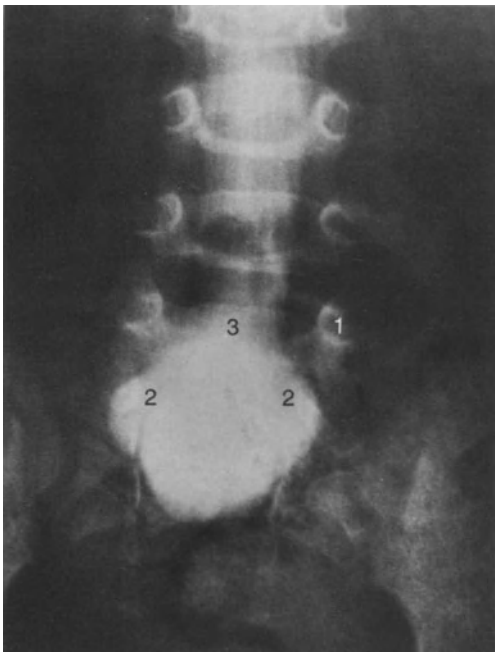


Figure 14.60. Simple meningocele. (A) The small defect in a single arch (*1*) borders the neck of the hernia (*2*), through which dura and arachnoid (*3*) herniate. The sac is a simple meningocele, extending through the neck, dorsal to normal cord and roots. (B) When there is a wide defect in a single 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.



A



B

Figure 14.61. (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) The myelogram reveals that the meningocele neck is limited to the 5th lumbar vertebra (*1*). Two nerve roots (*2*) are tethered at the hernia neck (*3*). This is one of the two types of "tethering meningocele, the *spinal root varitey.*"

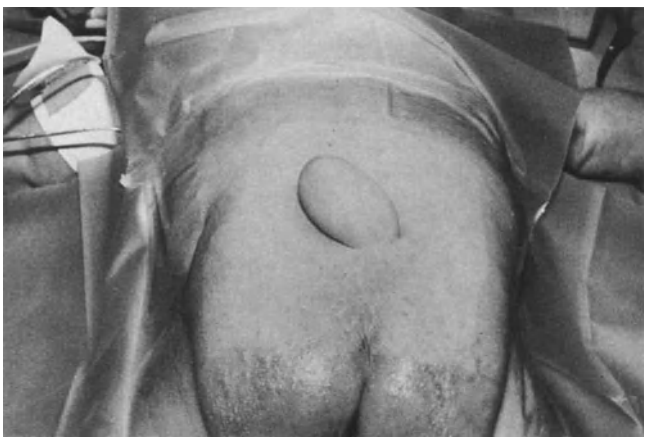


Figure 14.62. This child has a relatively large, fingerlike meningocele located at the lumbosacral junction.

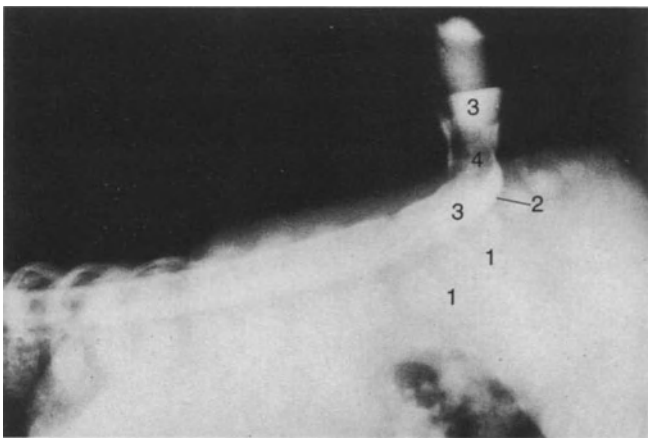


Figure 14.63. Myelography reveals that the laminae of two vertebrae (*1*) are dysraphic, permitting exit of the neck (*2*) of the meningocele (*3*). The spinal cord (*4*) is tethered at the neck. This is the spinal cord *variety* of tethered meningocele, the type illustrated diagrammatically in Figure 14.60B.

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 meningmyelohydrocele has three layers within the hernial 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

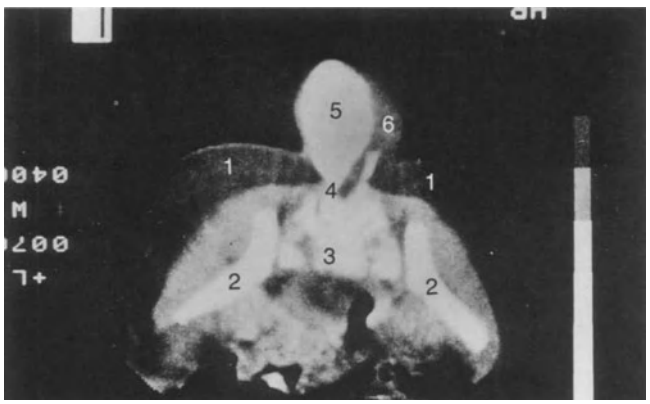


Figure 14.64. This CTT study illustrates the buttocks (*1*), iliac crests (*2*), first sacral vertebra (*3*), and the very narrow neck (*4*) of the meningocele (*5*). A small lipoma (*6*) was attached to the tethered cord, and lies along the surface of the meningocele.

Figure 14.65. *Spinal cord* variety of tethered meningocele. The spinal cord (*1*) is stretched and adherent to the neck (*2*) of the hernia sac inferiorly, to the reader's left. The dura (*3*) has been reflected superiorly. Visible nerve roots are thickened.

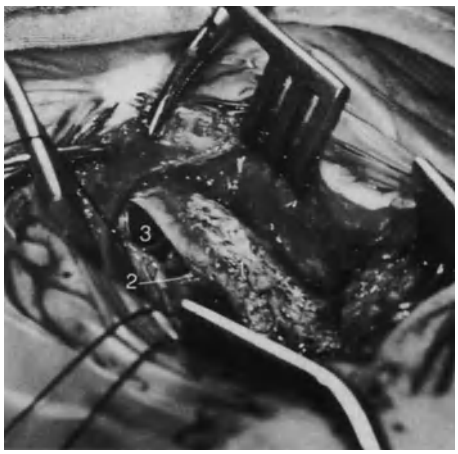
the spinal canal and onto one another. This seals cerebrospinal fluid within the spinal canal and bacteria and foreign substances from it.

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 his 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 (Figure 14.66). 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*, he 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

A

B



C

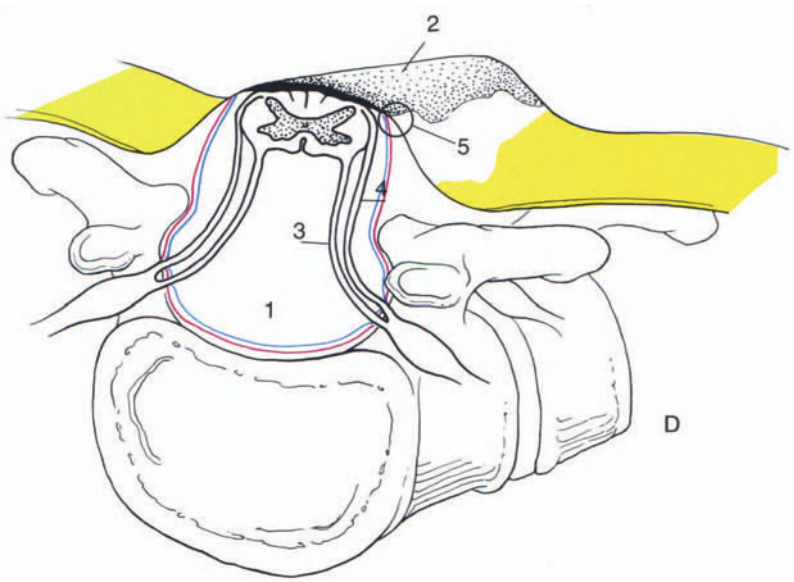


Figure 14.66. Meningomyelocele: the placode, the arachnoid cyst elevating it, and the attachment of neural elements (in this case spinal cord and nerve roots) to it. (A) The elevated placode (*1*) has been dissected from the dura matter (*2*), exposing thickened arachnoid along its base (*3*). (B) Another child with meningomyelocele. The epithelialized placode (with its ectopic ganglion cells, degenerated and rudimentary glial and neural elements, and hemorrhagic granulation tissue) has been removed and discarded. The underlying arachnoidal cyst bulges into the field. (C) After the cyst has been opened, one may identify the spinal cord (*1*), a thickened nerve root (*2*), and the cystic space ventral to the cord (*3*). (D) 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*).

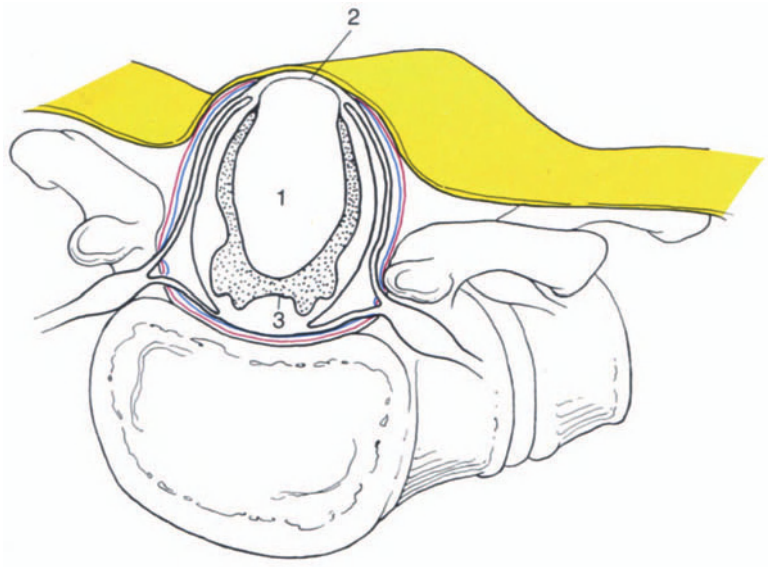


Figure 14.67. Meningomyelohydrocele. The spinal cord has blown out around the enormously dilated central canal (*1*), so that the dorsal cord (*2*) is converted into a membrane and the ventral cord (*3*) flattened. The dorsal and ventral roots are in relatively normal position.

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 surrounding placode, separating this from the full-thickness skin. When the epithelial membrane and centrally placed placode are elevated, one will see dysgenic nerve roots adherent to the undersurface of the placode. The epithelialized tissue and dysgenic 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 dysgenic 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 (Figures 14.67 and 14.68). 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, he 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 paraspinal muscle fascia, is 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. 14.69), 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,

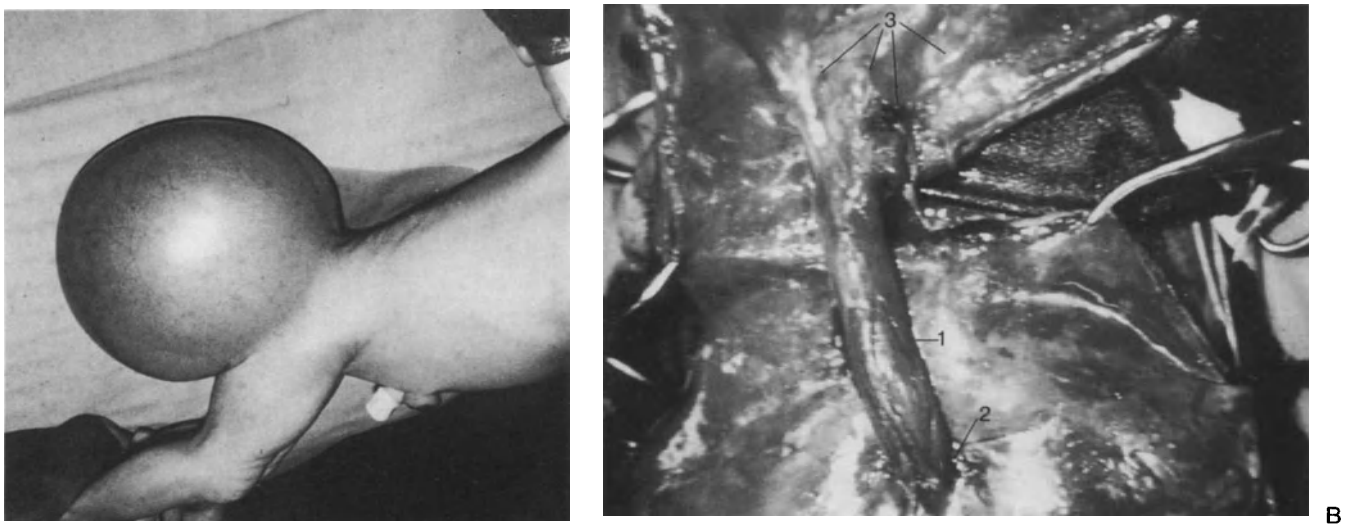


Figure 14.68. Meningomyelohydrocele. (A) The meningomyelohydrocele is located over the sacrum and buttocks. This child did not have an accompanying cloacal extrophy, something which is more obvious because of its grotesqueness than its incidence. (B) In this child, the spinal cord (*1*) is herniated through the neck (*2*) and tethered to the sac (*3*) of the hernia.

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 (Figure 14.70)

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. This 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.

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 he 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, fi-

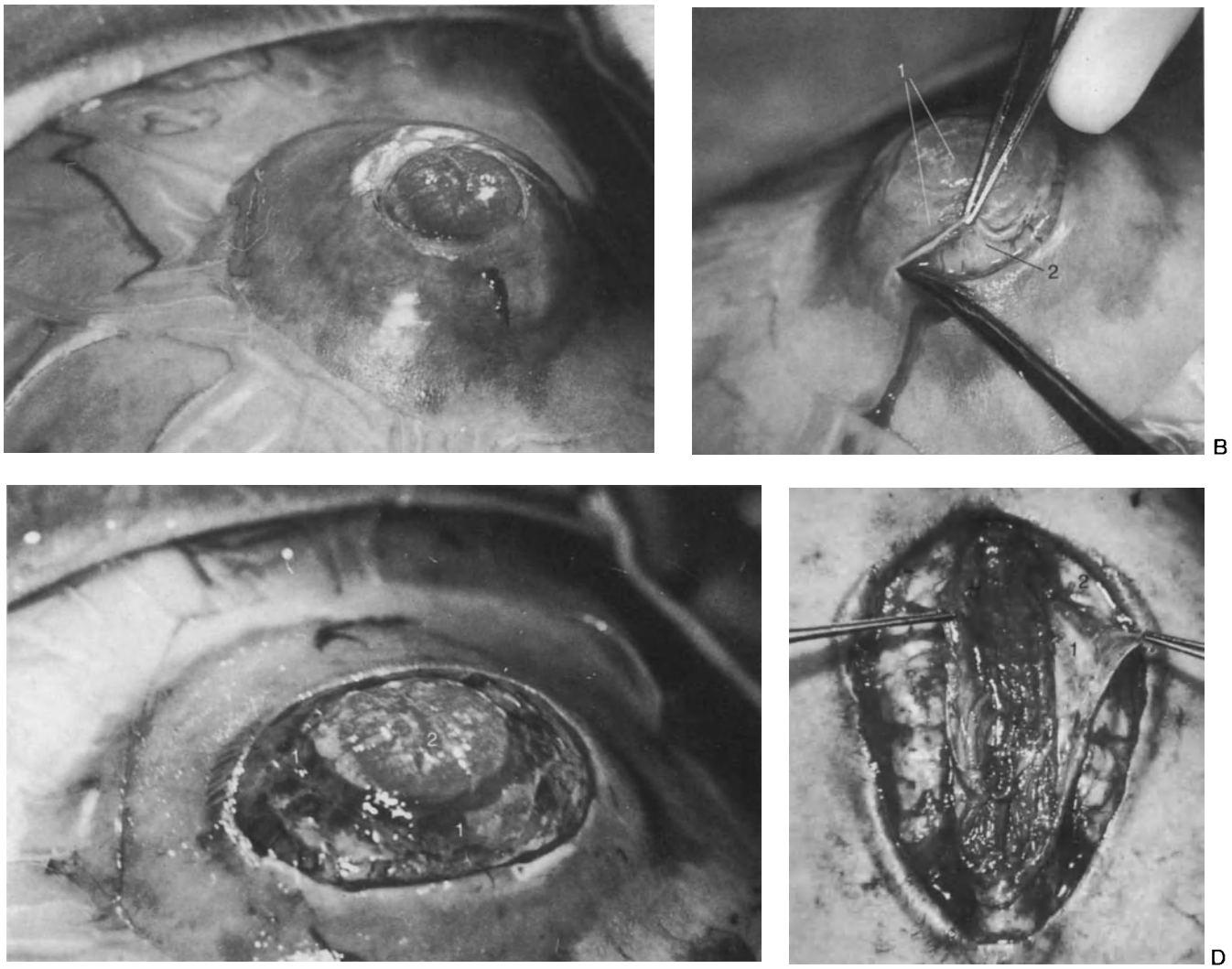


Figure 14.69. Stages in the closure of a myelocele, meningomyelocele, meningomyelohydrocele, and tethered (spinal cord or nerve root) meningocele. All four of these dysraphic states have in common herniation of meninges and neural elements into a sac composed of meninges and epithelial tissue which vary 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*). (C) The nondescript tissue has been completely removed, the cyst wall (*1*) preserved, along with the unclosed spinal cord (*2*). This child has a myelocele, so the cord is left intact. If this were a child with a meningomyelocele, the exposed surface of the placode would be separated from the underlying neural tissue and discarded. If it were a meningocele with the spinal cord tethered to the hernia sac, the epithelial and dysgenetic surfaces of the sac would be separated from the spinal cord so that the latter could be preserved. (D) After the neural elements have been allowed to fall back into the ventral surface of the spinal canal, the dura (*1*) is dissected from the paravertebral fascia (*2*) on either side, mobilizing enough to permit one to imbricate it. The dura is closed without constricting the underlying cord and paravertebral fascia mobilized on both sides, with the fascia from the right being brought across the dorsal surface of the dura and sewn to the base of the fascial pedicle on the left (*5*). Subsequent to this, the left fascial flap is imbricated onto the one on the right.

brous band running from the ventral to the dorsal surfaces 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 osteocartillagenous spur, the surgeon should remove it with the finest tipped rongeur available, biting away one or two mm fragments at a time. The disadvantage of using a high-speed drill, even one with a diamond burr, to do this rests in the necessity to retract 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, so one need not be concerned. After the diastematomyelic spur has been removed, the operative procedure is considered completed. There is no need to open the dura mater.

Figure 14.70. 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 2 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.

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 (Figures 14.71 to 14.74). 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 esthetic or neurological. If the child is perfectly normal neurologically, the indication for surgery is esthetic. If a neurologic deficit becomes apparent, however, surgery is to be recommended. Some neurosurgeons, especially at the present time, are recommending elective resection of the lipoma even in children without neurological 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.* For the present, consequently, 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 "prophylactically." The author prefers to observe asymptomatic children, and to operate on only those who develop clinical signs or symptoms.

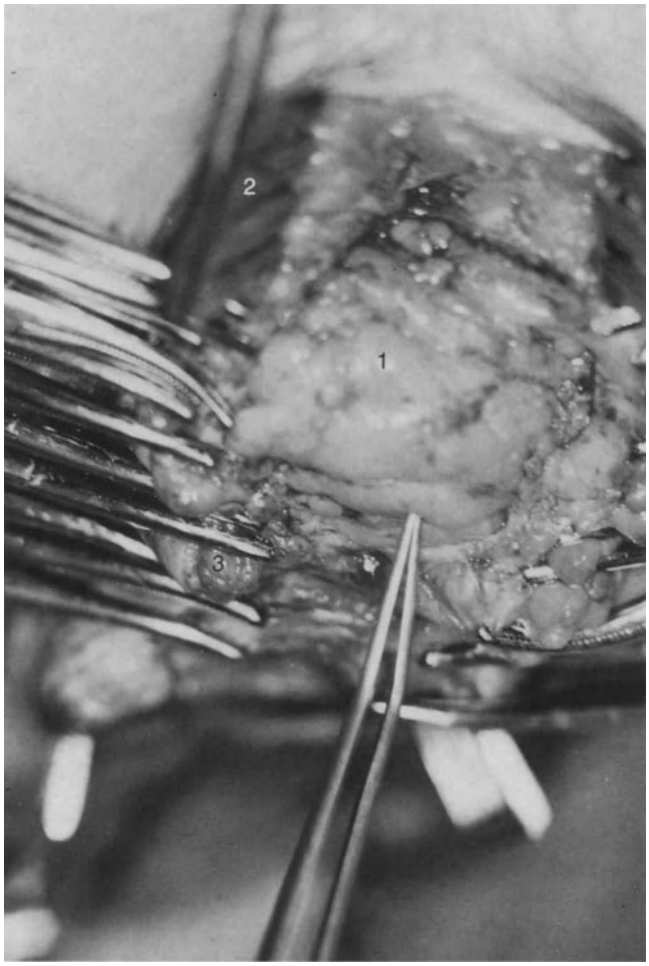


Figure 14.71. Lipoma. The lipoma expands to either side of the dura, within both the epidural and subcutaneous compartments. The subcutaneous expansion may be within the midline or, as in this case, lateral to it. In fact, the lipoma (*1*) is located between the skin (*2*) and muscle mass (*3*) of the child's left buttock.

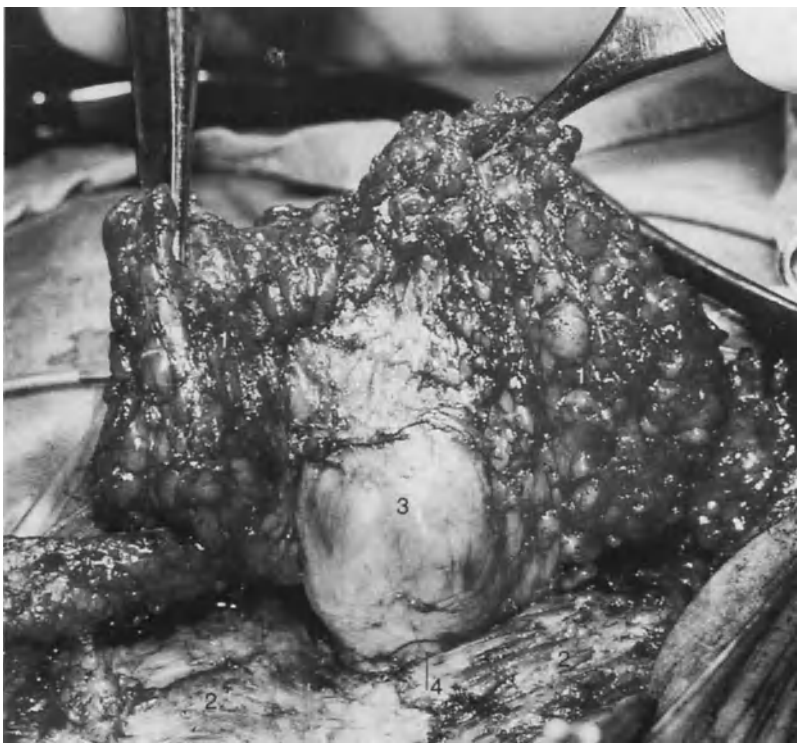


Figure 14.72. After the skin is incised and the subcutaneous portion (*1*) of the lipoma dissected from the underlying fascia (*2*), one may identify the extension of a lipomatous bundle (*3*) from the subcutaneous lipoma through the fascia at the neck of the hernia (*4*).

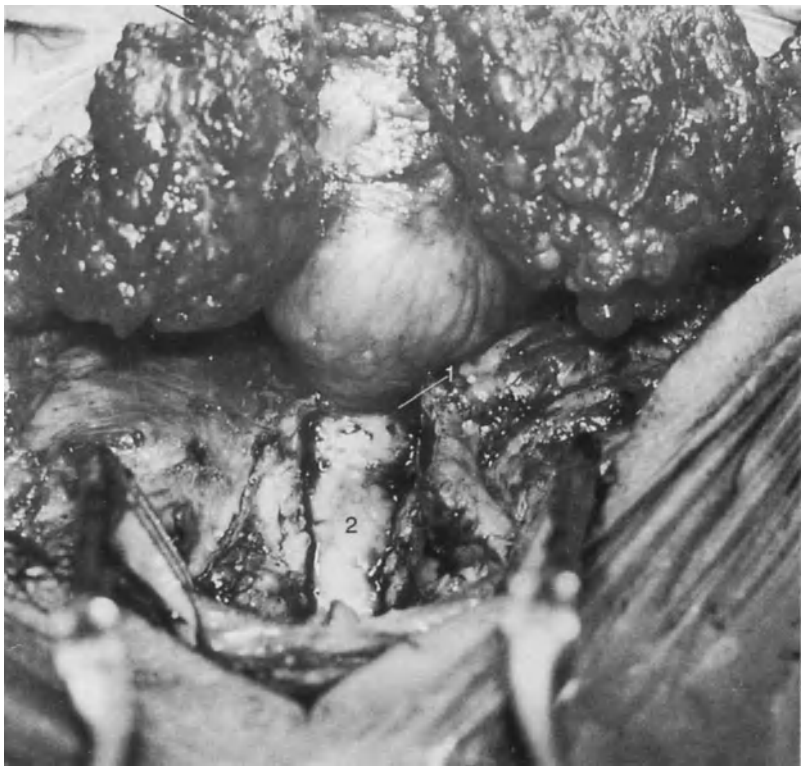


Figure 14.73. A laminotomy (not a laminectomy!) should be performed over 2 or 3 vertebral levels superior to the neck (*1*) of the lipoma. This uncovers the spinal canal, exposing the dural extension of this lipoma (*2*).

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 recommended 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 fron 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 opposite reasons. It is a matter of surgical judgment and familiarity: what works well for one surgeon is not necessarily incorrect 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

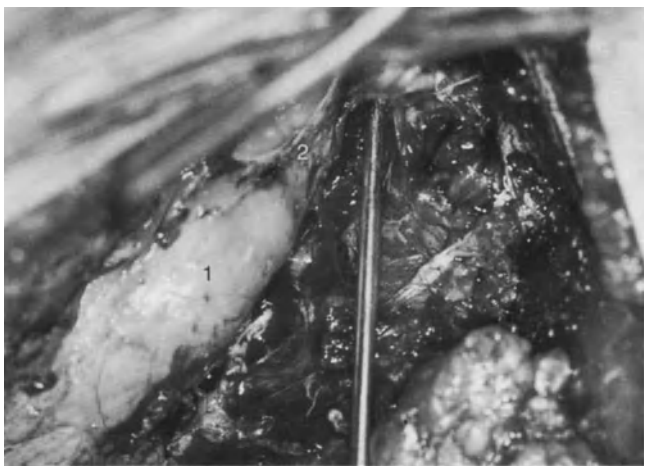


Figure 14.74. One may follow the intraspinal lipoma (*1*) to the point of its entry into the conus medullaris (*2*). Although all lipomas do not extend into the conus medullaris, the majority do. It is this which renders the efficacy of prophylactic surgery of lipoma questionable: one cannot safely remove all the lipoma from the spinal cord, and fatty tissue varies in volume with age.

Figure 14.75. 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*).

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 telescopes the fingerlike extension of lipoma fron 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 diagreement 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 pre-operatively 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. Intraoperative somatosensory evoked potentials may be helpful, but are generally of little value.

Once one has removed all of the lipoma he is *comfortable* with removing (this, and only this, should represent the end point of his 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

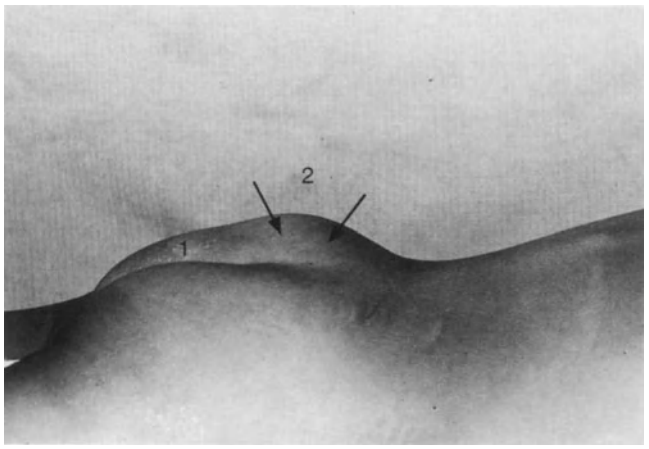


Figure 14.76. Dural fibrolipoma. The child's buttock (*1*) is to the left of the bulge from the dural fibrolipoma (*2*). After the skin has been opened, one immediately encounters the subcutaneous lipoma. Dural fibrolipomas may be adherent to the dura or, as in this case, remote to it.

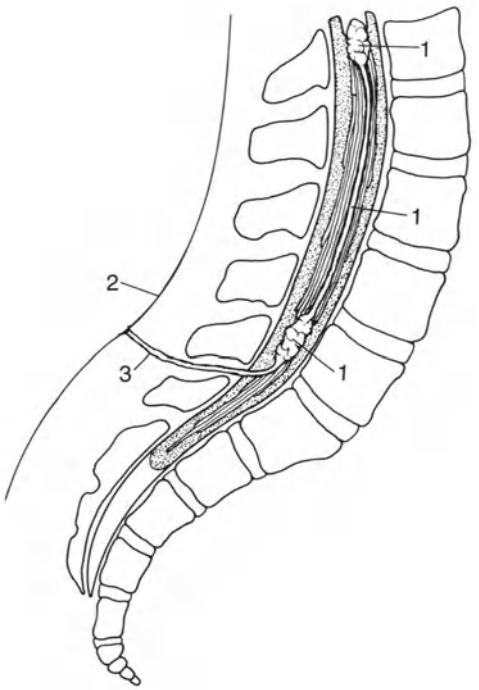


Figure 14.77. Dermal hamartomas (*1*) are connected to the skin (*2*) either by a solid or tubular stalk (*3*).

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, subcutaneous connective tissue dermal scar.*

Leptomyelolipoma (Figure 14.75). The simple lipoma is extremely difficult to distinguish from the leptomyelolipoma, 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.

Dural Fibrolipoma (Figure 14.76). 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, seprating 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 for them to serve. Lemire and colleagues[63] 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. I have chosen to use the anatomically more detailed, and surgically valuable, classification, which has evolved from the works of James and Lassman[57], and Karch and Urich.[64]

Dermoid Hamartoma (Figures 14.77 to 14.81)

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.

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,

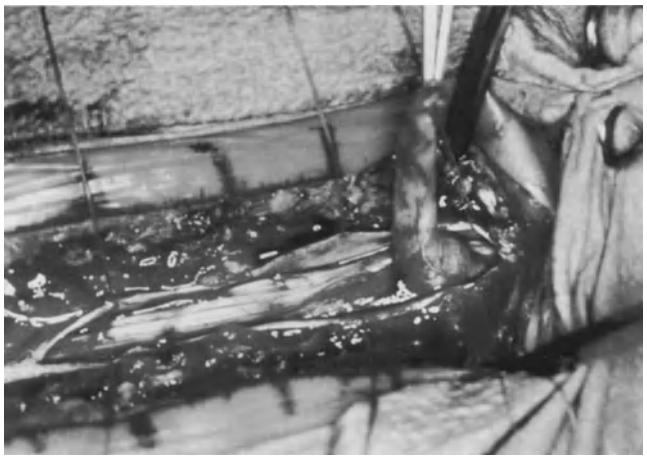


Figure 14.78. The dermal hamartoma may extend into the subarachnoid space either as fibrous bands, stalks, tubes, or (as in this case) lipomatous masses.

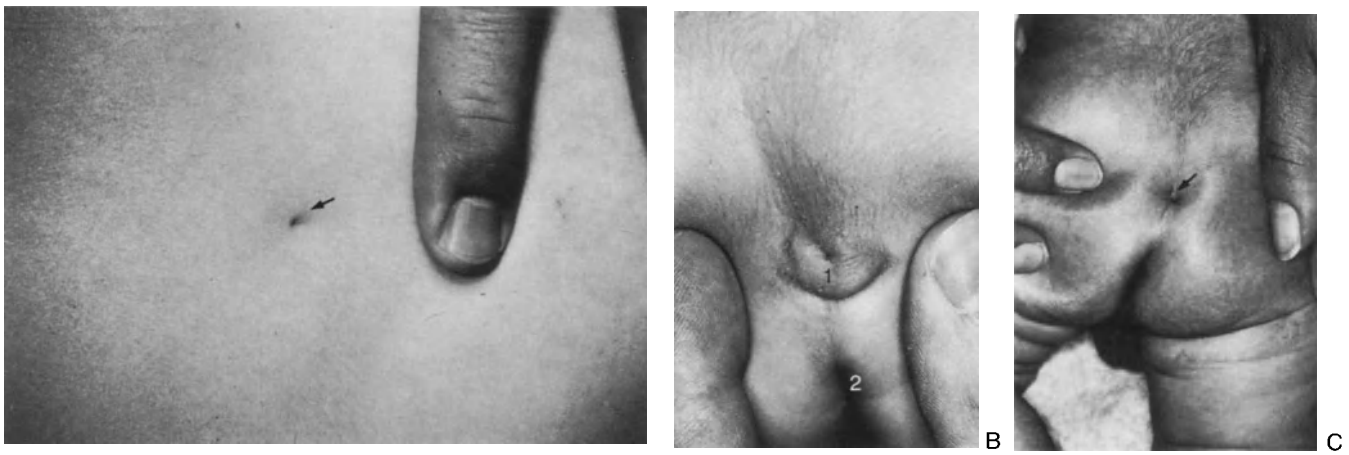


Figure 14.79. Dermnoid sinus tracts, extending through the skin, muscle, posterior vertebral arch or interspinous ligaments, dura, and into either the subarachnoid space or neural elements (cord and roots) are very variable in surface appearance. They are, however, perforations of the skin and, therefore, 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.

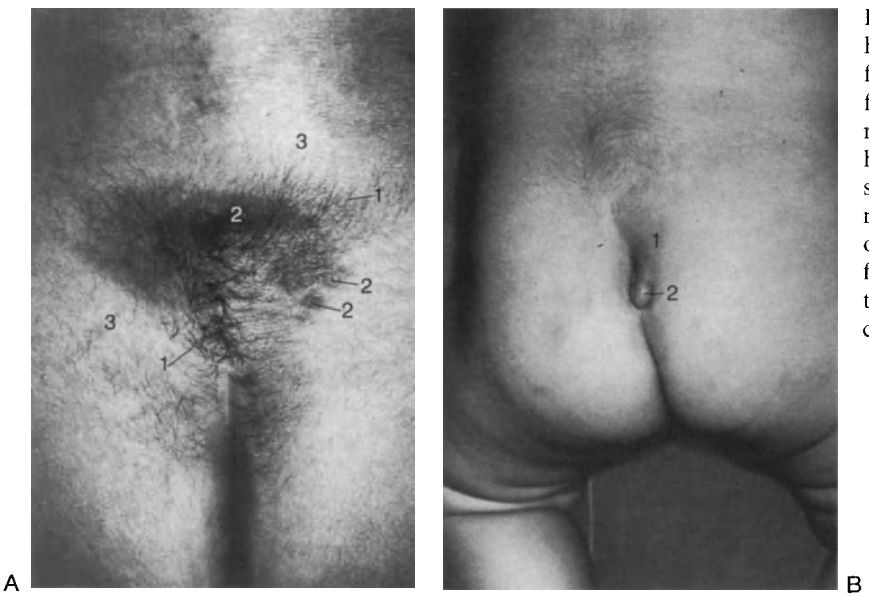
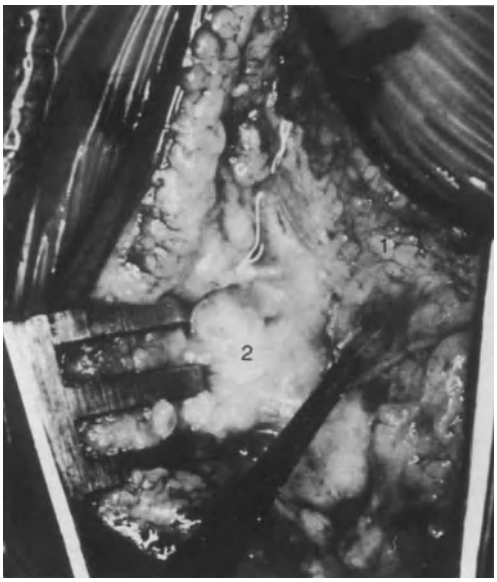
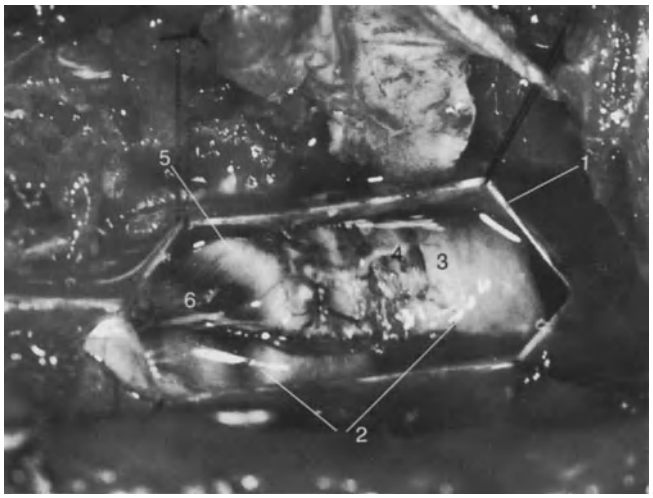


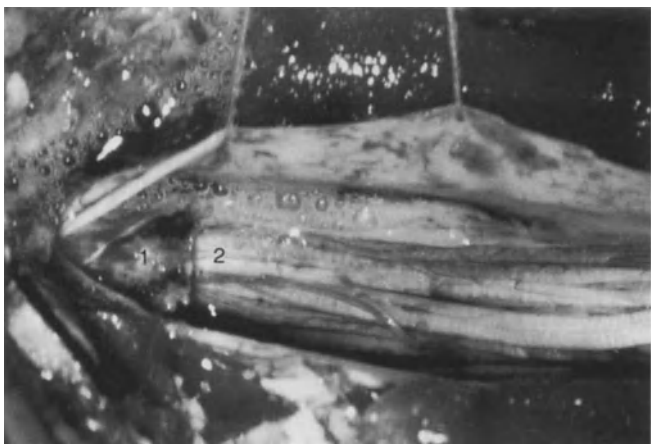
Figure 14.80. Though dermoid sinus tracts have a hole within the skin as a *sine qua non* for their diagnosis, dermoid tumors extending from the skin and into the spinal canal or cord may be associated with a subcutaneous mass, hypertrichosis, hemangioma, digitlike extensions of skin. (A) Hypertrichosis (*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 excresence of skin (*2*) in this child.



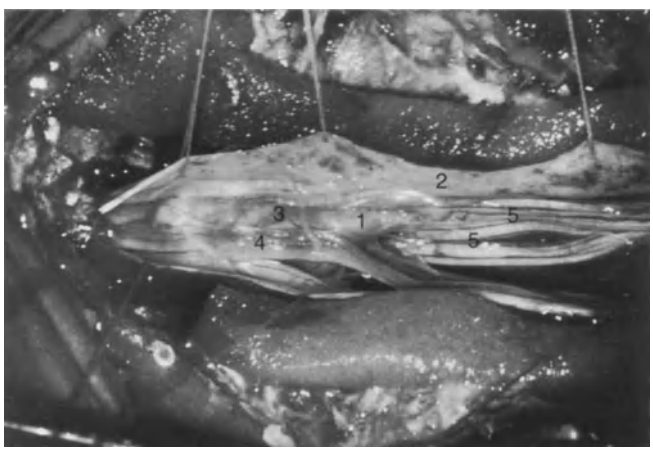
A



B



C



D

Figure 14.81. The technique for removing dermoid tumors is no different from that for removing other tumors of the spinal cord, nerve roots, spinal canal, meninges, posterior spinal arch. One characteristic of dermoid tumors, however, is that they very often involve all tissue elements from the skin into the spinal cord and/or roots. Their dissection becomes progressively more tedious and dangerous as the tumor expands. Another characteristic of dermoid tumors is that they consist of dysgenetic tissue, so that one may find it difficult to separate normal from abnormal tissue within the tumor or the structures bordering it. This figure illustrates some of these points. (A) Fatty (*1*) and pearly white (*2*) components of this dermoid tumor extend from the subcutaneous area into the region of the unclosed posterior vertebral arch. (B) The dura (*1*) has been opened but the arachnoid (*2*) is intact. Pearly white tissue (*3*), with a vascularized surface (*4*), occupies the superior portion of the subarachnoid space. The tumor invades a nerve root (*5*), increasing greatly its size, and obstructs the subarachnoid space, producing a complete block and a subarachnoid cyst (*6*). (C) In this child the dermoid tumor (*1*) extends ventral to the cauda equina (*2*). (D) One notes that the dermoid tumor (*1*) extends through the dura mater (*2*), directly into the conus (*3*) and nerve roots (*4*). It does not involve all elements of the cauda equina (*5*).

Figure 14.82. Ventral meningocele. The ventral meningocele is an hamartoma. After the dura (*1*) has been opened, the radicular elements of the cauda equina (*2*) are identified and found to have been splain over a cystic mass occupying the ventral aspect of the spinal canal (*3*).

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 almost invariably 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 (Figures 14.82 to 14.86)
The endodermal hamartomas are 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 intraabdominal (retroperitoneal) mass is of significant size, the general surgeon should remove it through a laparotomy.

Nondysraphic Spinal Cord Anomalies and Congenital Tumors

Hydromyelia
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.

If the hydromyelia is severe, irrespective of its association with spinal dysraphism, it should be treated. The recommended treatment consists of laser penetration of the spinal cord, so as to establish a fistulous tract between the hydromyelic central canal and the subarachnoid spaces. The arachnoid should be opened approximately 1.5 to 2 cm from the point of laser fenestration and then, after the fistula has been made, a watertight closure of the arachnoid performed.

Figure 14.83. The communication (*1*) between the anterior (intraabdominal) 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.

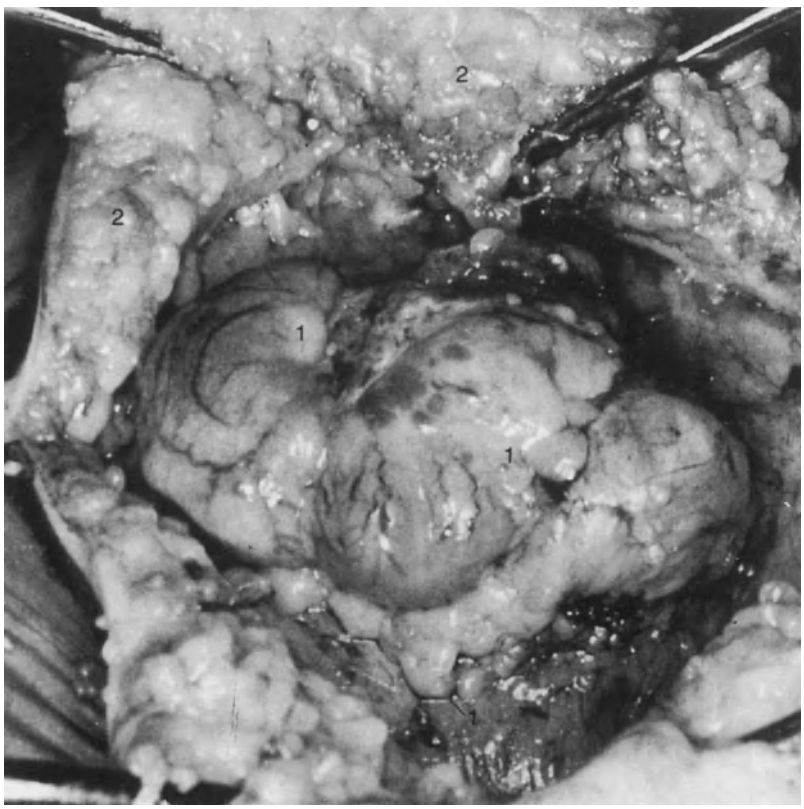


Figure 14.84. Entodermal hamartomas consist of herniation of bowel through the dysraphic vertebrae and into the subcutaneous compartment. Here, one notes bowel (*1*) occupying all of the subcutaneous space. There is lipoma (*2*) adherent to the under surface of the subcutaneous compartment.

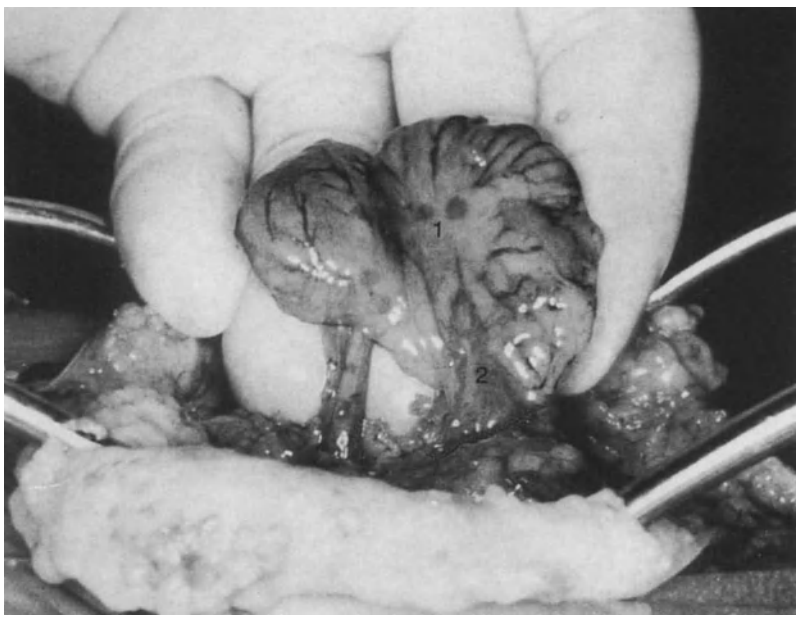


Figure 14.85. One is able to identify the rudimentary bowel (*1*) and its mesentery (*2*).

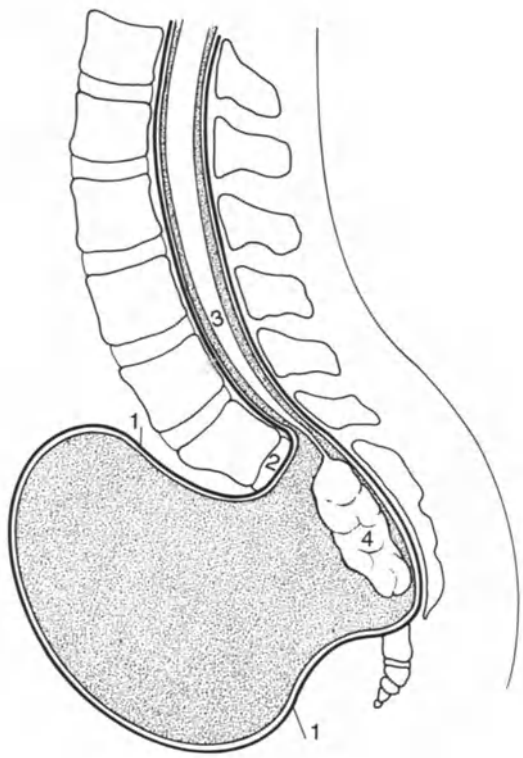


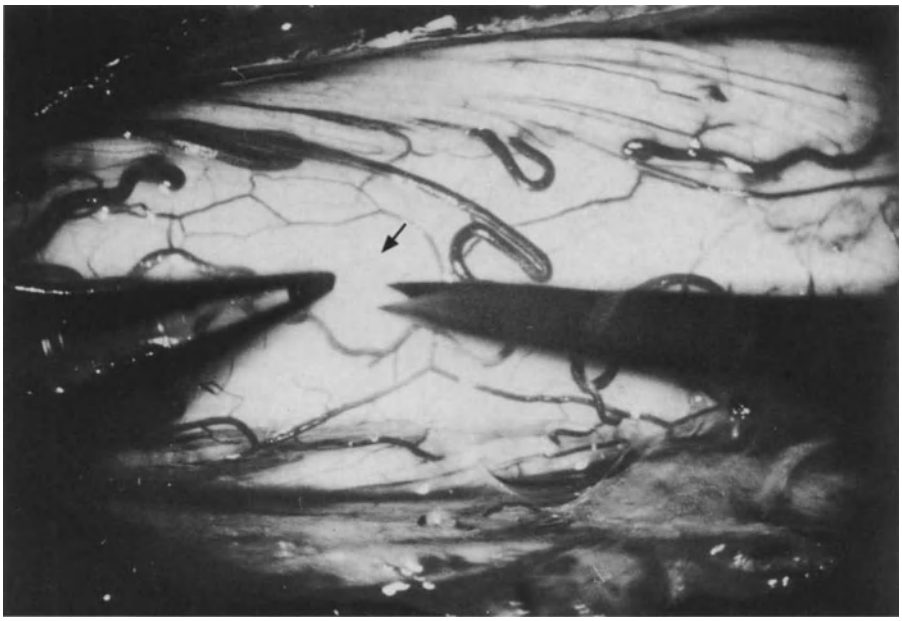
Figure 14.86. A ventral meningocele (endodermal hamartoma) (*1*), illustrating the bony (*2*), neural (*3*), and hamartomatous (*4*) components.

Syringomyelia (Figures 14.87 to 14.89)
The operative treatment for syringomyelia is the same as for hydromyelia.

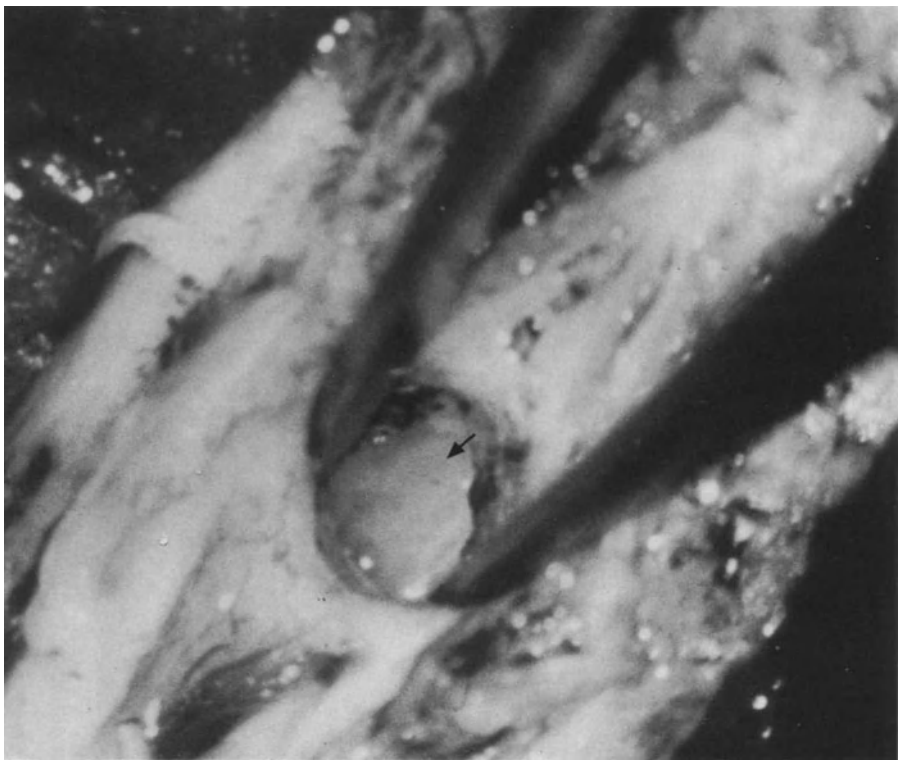
The Tethered Cord (Figure 14.90)
In its broadest sense, one could use the term "tethered" to indicate any bridling of the spinal cord or nerve roots, which results form a band of mesenchymal tissue extending form the spinal cord to the dura mater. Thus, a fibrotic *filum terminale,* extending from the 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 arch may also cause tethering. A common cause of tethering may be scar tissue binding the spinal cord and nerve roots of a previously operated meningomyelocele or lipomatou 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 to reoperate 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 seriously ques-



A



B

Figure 14.87. Syringomyelia. Whether idiopathic or traumatic, the surgical treatment for 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.

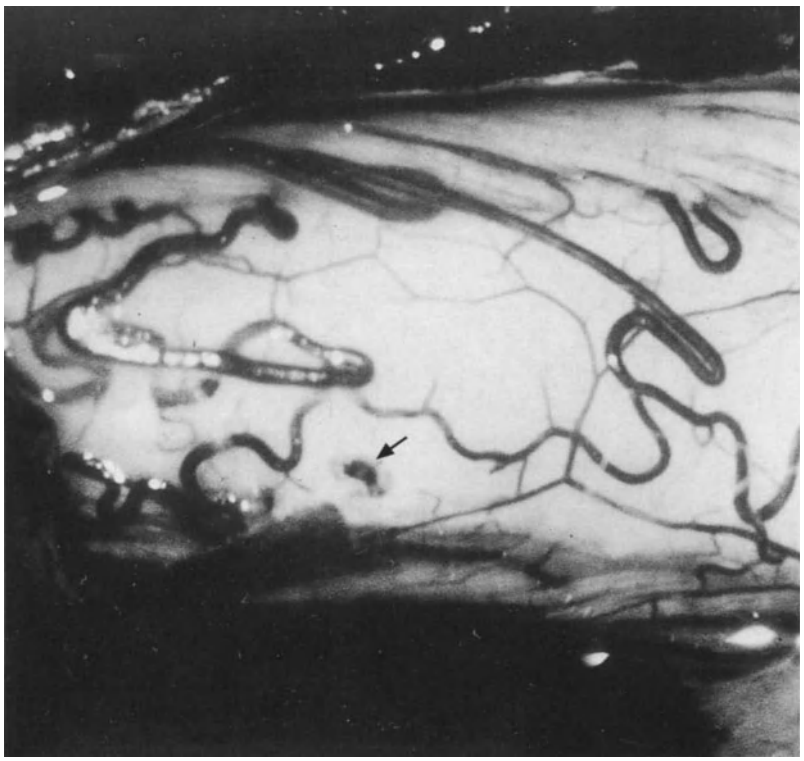


Figure 14.88. If the laser is available, it is much more precise and effective in producing marsupialization between the syringomyelic cavity and the pial surface of the cord. In this illustration the laser is being used to perforate the pial surface and underlying wall of the syringomyelic cavity (arrow).

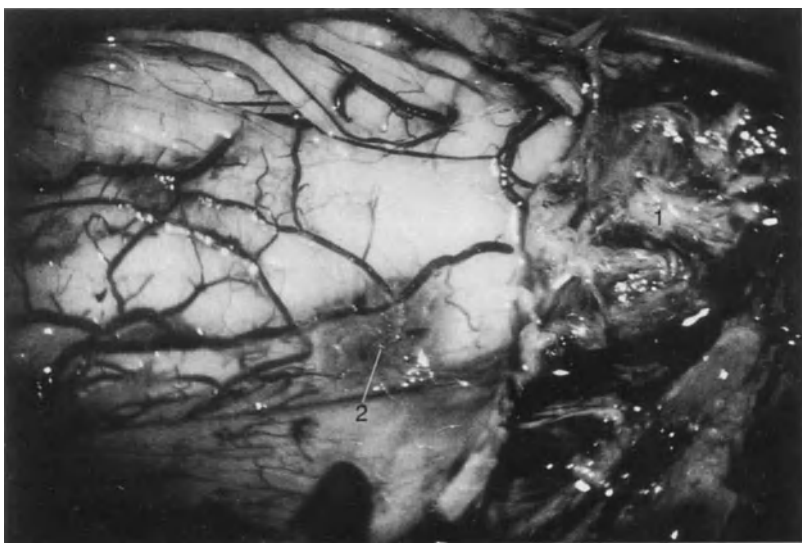


Figure 14.89. This patient suffered posttraumatic syringomyelia. The glial hypertrophy and scar formation (*1*) are seen at the right, and the pial wall of the posttraumatic syringomyelic cavity (*2*) at the center. A laser is being used to perforate the pial wall and enter the cavity.

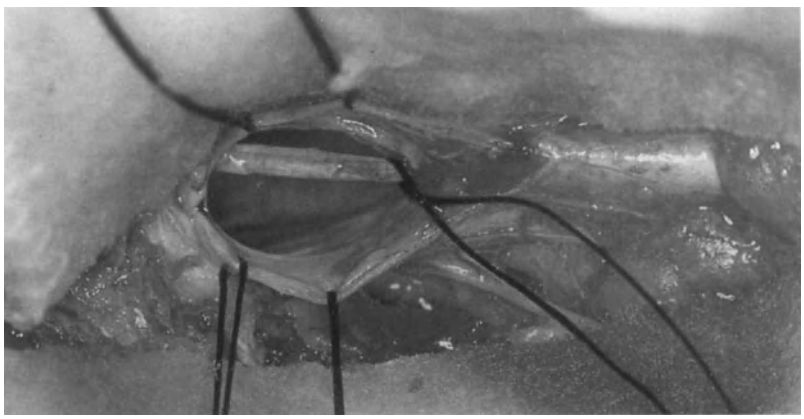


Figure 14.90. Tight thick filum at operation in child with tethered spinal cord. Courtesy of Dr. H. Hoffman.

tioned, 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.

References

1. Davis CH Jr, Alexander E Jr, Kelly DL Jr: Treatment of cranial synostosis. J Neurosurg 30:630–6, 1969.
2. Shillito J Jr, Matson DD: Cranial stynostosis: A review of 519 surgical patients. Pediatrics 41:829–53, 1968.
3. Ingraham Fd, Alexander E Jr, Matson DD: Clinical studies in craniosynostosis: Analysis of 50 cases and description of method of surgical treatment. Surgery 24:518–41, 1948.
4. Paul RP, Sugar O: Zenker's solution in the surgical treatment of cranial synostosis. J Neurosurg 36:604–7, 1972.
5. Anderson FM, Johnson FL: Cranial synostosis: A modification in surgical treatment. Surgery 40:961–70, 1956.
6. Giller HD, Harrison SH: Operative correction by osteotomy of recessed malar maxillary compound in a case of oxycephaly. Br J Plast Surg 3:123, 1950.
7. Tessier P: Anatomical classification of facial, cranio-facial and latero-facial clefts. J Maxofac Surg 4:69–92, 1976.
8. LeFort R: Experimental study of fractures of the upper jaw. Part III. In McDowell F (ed): The Source Book of Plastic Surgery. Baltimore, Williams & Wilkins, 1977.
9. Converse JM, Smith B: An operation for congenital and traumatic hypertelorism. In Troutman RC, Converse JM, Smith B (eds): Plastic and Reconstruction Surgery of the Eye and Adnexa. Washington, DC, Butterworths, 1962.
10. Tessier P: Orbital hypertelorism: II. Definitive treatment of orbital hypertelorism by cranio-facial osteotomies. Scand J Plast Reconstr Surg 7:39–58, 1973.
11. Tessier P: The scope and principles, dangers and limitations, and the need for special training in orbito-cranial surgery. In Hueston JT (ed): Trans Fifth International Congress on Plastic and Reconstructive Surgery. Butterworths, Melbourne, 1971, p 903.
12. Munro IR: Orbito-cranio-facial surgery: The team approach. Plast Reconstr Surg 55:170–6, 1955.
13. Rougerie J, DeRome P, Anguez L: Craniostenoses et dysmorphies craniofaciales. Principes d'une nouvelle technique de traitement et ses résultats. Neurochirurgie 18:429–40, 1972.
14. Edgerton MT, Jane JA, Berry Fa, Fischer JC: The feasibility of craniofacial osteotomies in infants and young children. Scand J Plast Reconstr Surg 8:164–8, 1974.
15. Tessier P: Treatment of facial dysmorphisms in craniofacial dysostosis (DCF). Crouzon and Apert diseases. Total osteotomy of the facial massif. Sagittal displacement of the facial massif. Neurochirurgie 17:295–322, 1971.
16. Lannelongue M: De la craniectomie dans la microcéphalie. CR Acad Sci Paris 110:1382–5, 1890.
17. McLaurin RL, Matson DD: Importance of early surgical treatment of craniosynostosis. Review of 36 cases treated during the first six months of life. Pediatrics 10:637–52, 1952.
18. Anderson FM, Johnson FL: Craniosynostosis. A modification in surgical treatment. Surgery 40:961–70, 1956.
19. Raimondi AJ, Gutierrez FA: A new surgical approach to the treatment of coronal synostosis. J Neurosurg 46:210–4, 1977.
20. Greig DM: Hypertelorism: A hitherto undifferentiated congenital craniofacial deformity. Edinburgh Med J 31:560, 1924.
21. Tessier P: Orbital hypertelorism. I. Successive surgical attempts. Material and methods. Causes and mechanisms. Scand J Plast Reconstr Surg 6:135–55, 1972.
22. Tessier P, Guiot G, Derome P: Orbital hypertelorism. II. Definite treatment of orbital hypertelorism (ORH) by

craniofacial or by extracranial osteotomies. Scand J Plast Reconstr Surg 7:39–58, 1973.
23. Delaire J: La croissance des os de la voûte du crane. Principes généroux. Rev Stomat 62:518–26, 1961.
24. Tessier P: Dysostoses craniofaciales osteotomies totales de la face. Trans 4th Int Congr Plast Reconstr Surg Amsterdam, Excerpta Medica, 1969.
25. Tessier P: Osteotomies totales de la face. Syndrome de Crouzon. Syndrome d'Apert. Oxycephalies. Scaphocephalies. Turricephalies. Ann Chir Plast 12:273, 1967.
26. Tessier P: The definitive plastic surgical treatment of the severe facial deformities of craniofacial dysostosis. Crouzon's and Apert's diseases. Plast Reconstr Surg 48:419–42, 1971.
27. Holtermüller K, Weidermann HR: Kleeblattschädel Syndrom Med Monatsschr 14:439–46, 1960.
28. Turner PT, Reynolds AF: Generous craniectomy for Kleeblattschädel anomaly. Neurosurgery 6:555–8, 1980.
29. Ingraham DF, Swan H: Spina bifida and cranium bifidum. I. A survey of five hundred and forty-six cases N Engl J Med 228:559–63, 1943.
30. Fenger C: Basal hernias of the brain. Am J Med Sci 109:1–17, 1895.
31. Charoonsmith T, Suwanwela C: Frontoethmoid encephalomeningocele with special reference to plastic reconstruction. Clin Plast Surg 1:27–47, 1974.
32. Lewin ML, Schuster MM: Transpalatal correction of basilar meningocele with cleft palate. Arch Surg 90:687–93, 1965.
33. Matson DD: Neurosurgery: Infancy and Childhood, ed 2. Springfield, IL, Thomas, 1969, p 68.
34. Suwanwela C, Sukabote C, Suwanwela N: Frontoethmoidal encephalomeningocele. Surgery 69:617–25, 1971.
35. Blumenfeld R, Skolnik EM: Intranasal encephaloceles. Arch Otolar 82:527–31, 1965.
36. Robinson RG: Anterior encephalocele. Br J Surg 45:36–40, 1958.
37. Avanzini G, Crivelli G: A case of sphenopharyngeal encephalocele. Acta Neurochir 22:205–12, 1970.
38. Bulinska H: A case of sphenoid meningocephalocele. Otolar Pol 26:719–23, 1972.
39. Lau BP, Newton TH: Sphenopharyngeal encephalomeningocele. Radiol Clin Biol 34:386–98, 1965.
40. Pollock JA, Newton TH: Encephalocele and cranium bifidum. In Newton TH, Potts (eds): Radiology of the Skull and Brain—The Skull. St Louis, Mosby, 1973, pp 634–47.
41. Wiese GM, Kempe LG, Hammon WM: Transsphenoidal meningohydroencephalocele. J Neurosurg 37:475–8, 1972.
42. Choudhury A, Taylor JC: Primary intranasal encephalocele: Report of four cases. J Neurosurg 57:552–5, 1982.
43. Heinecke W: Die chirurgischen Krankheiten des Kopfes. Stuttgart, Enke 1882, p. 252. Cited in Fenger C: Basel hernias of the brain. Am J Med Sci 109:1–17, 1895.
44. Gisselsson L: Intranasal forms of encephalomeningocele. Acta Otolaryngol 35:517, 1947.
45. Lichtenberg G: Congenital tumour of the mouth, involving the brain and connected with malformations. Trans London Soc Pathol 18:250–2, 1866–1867.
46. Virchow RLK: Die krankhaften Geschwulste. Berlin, Hirschwald, 1863, vol 1, p 188.
47. Cleland J: Contribution to the study of spina bifida, encephalocele, and anencephalus. J Anat Physiol 17:257, 1882.
48. Arnold J: Myelocystic Transposition Nongewebsheimen und Sympodie. Beitr Pathol Anat 16:1, 1894.
49. Grossman H, Sugar O, Greely P, Sadove MS: Surgical separation in craniopagus. JAMA 153:201–7, 1953.
50. Durr SP: Craniopagus twins. JAMA 150:93, 1952.
51. Sharrard WJW, Zachary RB, Lorber J, Bruce AM: A controlled trial of immediate and delayed closure of spina bifida cystica. Arch Dis Child 39:18–22, 1963.
52. Duckworth T, Sharrard WJ, Lister J, Seymour N: Hemimyelocele. Dev Med Child Neurol Suppl 16:69–75, 1968.
53. Lebedeff A: Über die Entstehung der Anencephalie mit Spina Bifida bei Vögeln und Menschen. Virchows Arch Pathol Anat Physiol 86:263–98, 1881.
54. Recklinghausen F von: Untersuchungen über die Spina bifida. Teil 2: Über die Art und die Entstehung der Spina bifida ihre Beziehung zur Rückenmarks- und Darmspalte. Virchows Arch Pathol Anat Physiol 105:296–332, 1886.
55. Morgagni GB: De Sedibus et Causis Morborum, (transl. Alexander B), London, Miller and Cadell, 1769.
56. Patten BM: Embryological stages in the establishing of myeloschisis with spina bifida. Am J Anat 93:365–95, 1953.
57. James CCM, Lassman LP: Spinal dysraphism. The diagnosis and treatment of progressive lesions in spina bifida occulta. J Bone Joint Surg 44:828–40, 1962.
58. Ingraham FD, Matson DD: Neurosurgery in Infancy and Childhood. Springfield, IL, Thomas, 1954.
59. Talwalker VC, Datsur DK: Meningoceles and meningomyeloceles (ectopic spinal cord). Clinicopathological basis of a new classification. J Neurol Neurosurg Psychiatry 33:251–62, 1970.
60. Emery JL, Lendon RG: The local cord lesion in neurospinal dysraphism (meningomyelocele). J Pathol 110:83–96, 1973.
61. Rokos J: Pathogenesis of congenital malformations of the central nervous system with special reference to spina bifida and Arnold-Chiari malformation. MD Thesis, University of Birmingham, England, 1973.
62. Greenfield JG: Greenfield's Neuropathology, ed 3. Chicago, Year Book Medical Publishers, 1976, p 361.
63. Lemire RJ, Loeser JD, Leech RW, Ellsworth CA Jr: Normal and Abnormal Development of the Human Nervous System. Hagerstown, MD, Harper and Row, 1975.
64. Karch SB, Urich H: Occipital encephalocele: A morphological study. J Neurol Sci 15:89–112, 1972.

Uncited References

Cerullo LJ, Raimondi AJ: The neuroradiological evaluations of the Arnold-Chiari malformations. In Proceedings of the Symposium on Myelomeningocele, Cincinnati, Ohio. Grune & Stratton, New York, 1977.

McLone DG, Raimondi AJ, Sommers MW: The results of early treatment of 100 consecutive newborns with myelomeningocele. Kinder Chirurgie Hippokrates Verlag Stuttgart 117–121, 1981.

Mullan S, Raimondi AJ: Respiratory hazards of the surgical

treatment of the Arnold-Chiari malformation. J Neurosurg 19:675–8, 1962.

Raimondi AJ: Aqueductal stenosis. In Concepts in Pediatric Neurosurgery II. Basal, S. Karger, 1982, p 45.

Raimondi AJ, Gutierrez FA: A new surgical approach for the treatment of coronal synostosis. J Neurosurg 46:210–14, 1977.

Raimondi AJ, Samuelson G, Yarzagaray L, Norton T: Atresia of the foramina of Luschka and Magendie: The Dandy-Walker cyst. J Neurosurg 31:202–16, 1969.

Raimondi AJ, Sato K, Shimoji T: The Dandy-Walker Cyst. Basal, S. Karger, 1983.

Soare PL, Raimondi AJ: Intellectual and perceptual motor characteristics of treated myelomeningocele children. Am J Dis Child 131:199–204, 1977.

Soare PL, Raimondi AJ: Quality of survival in treated myelomeningocele children: A prospective study. Decision making and the defective newborn. In Proceedings of a Conference on Spina Bifida and Ethics. Charles C Thomas, 1978, p 68.

"... 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

Chapter 15

Hydrocephalus

Introduction

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 cranio-cerebral size did not have particularly clear ideas as to where beneath the skin the fluid present in hydrocephalus was located[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 that, "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 Augsburgs 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[3] 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 interhemispheral fissure, or over the corpus callosum, in the 10 children between the ages of 2 and 15 years that he studied

at necropsy. The notable absence of subdural hygroma, subdural hematoma, 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 cerebral spinal fluid accumulates within the ventricular system, dilating the ventricles and increasing head size.

Hydrocephalus has been considered simply to consist of an accumulation of cerebral spinal 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.

By and large, it was assumed that external hydrocephalus and subdural effusions were identical anatomic 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. Computerized transmission tomography has recently provided us with additional information: cerebral spinal fluid may accumulate in pathological volumes within the subarachnoid spaces, in the absence of ventricular dilation, and it may pass freely across the arachnoidal membrane into the subdural spaces. Consequently, the term external hydrocephalus was expanded to include accumulations of cerebral spinal 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 inspite 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 CSF. Changes in intracranial pressure occur during the early stages of sleep and during REM sleep, they are episodic and rapid in onset and cessation, therefore expressive of the fact that alterations in production or absorption of cerebral spinal fluid are not the causative factors. Rather, alterations in cerebral flood flow (CBF), mediated by cerebral vaso-dilation and induced by neuronal or humoral mechanisms, resulting from cerebral vaso-dilation 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 to substitute the concept of "time-related pressure variations" for the older one of "level of pressure" in patients with defective ICP control mechanism.

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 cerebral spinal 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 cerebral spinal 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.

Thus, subarachnoid space or ventricular dilation occur as a result of intermittent increases in extraparenchymal cerebral spinal 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 his 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

It has, therefore, become necessary to consider critically whether hydrocephalus is characterized by: 1) an increase in intraventricular pressure; 2) an increase in intraventricular cerebral spinal fluid volume; 3) increased, or increasing, ventricular size. With the observations which the CTT 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, hormonal or humoral causes for alterations in cerebral flood flow.

We still *do not* have 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 anatomo-pathological state of inverse relationships between cerebral spinal fluid (CSF) volume and parenchymal bulk. We look upon CSF as a cushion, not a system for transporting cells, hormones, metabolites; not, as I 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:

CSF: sulcal, cisternal, ventricular, peri-neural, central canal, spinal;

Extracellular: white matter, gray matter, areas without blood brain barrier;

Intracellular: ganglionic, glial, perivascular glial cell;

Axo Myelinic: intra-axonal, myelogenous;

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 organo-electric 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 organo-electric concentrations and gas partial pressure both over time and in different anatomic 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 secondary 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 anatomo-pathologic 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 break-down products from liquefaction of a pyogenic exudate and blood reactive elements.

By hydrocephalus, one means a *pathological increase in intracranial cerebral spinal 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 states 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 classification of external and internal, with the latter subdivided into communicating and obstructive subgroups. During the past 15 years, the additional category of constrictive (as in the Chiari II malformation) hydrocephalus was added. 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 cerebral spinal 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 cerebral spinal 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 cerebral spinal fluid within the ventricular system, allowing it to pass from one ventricular chamber to another, and from the IV ventricle into the cisterns. The circulation of cerebral spinal 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 an obstruction to the flow of fluid into the basal cisterns. Obstructive hydrocephalus, on the other hand, was characterized by a 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 in-so-far 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 3 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.

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, results 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 patho-anatomical architecture, has been considered to be aqueductal occlusion. The distinction between non-tumorous aqueductal occlusion and the rarely encountered membraneous occlusion may be made in the living patient only after appropriate neuroradiologic studies: combined positive contrast III ventriculography and pneumoencephalography with the patient in the sitting position.

Such terms as forking, atresia, and gliogenous occlusion, are of purely historical value and, consequently, the reader is urged not to consider them distinct etiopathoanatomic entities. Forking, atresia, gliogenous occlusion, stenosis, aqueductal occlusion, 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 etiologic 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 alter-

ations. Anatomists, neuropathologists, clinicians, 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, unfortunately, were at odds with one another.

Berengarius[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."[5] 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."[6] According to Baker's review,[7] another anatomist coincidentally and confusedly 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[8] and Russell in 1949[9] came to two diammetrically opposite conclusions concerning the etiology of aqueductal occlusion. Bonnevie, studying primarily the experimental animal, concluded that the aqueductal occlusion was a consequence of the hydrocephalus, not its cause! Thereafter, Russell, studying human 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),[10] which suggested a distinction between communicating and non-communicating hydrocephalus on the basis of intraventricular injections of phenol-sulphonphthaleine, which may be recaptured normally from the urine in 10 to 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.

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 preceeds the patho-anatomical 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.[11] demonstrated, in experimental animals with congenital hydrocephalus, that aqueductal stenosis and occlusion result from periaqueductal edema. Johnson and Johnson,[12] in their studies of injecting myxovirus into newborn hamsters reported a short period of inflammation in which viral particles were limited to the ependyma 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.

Russel found it difficult to reconcile the absence of pyogenic exudate in the periaqueductal subependymal glia—her evidence for the maldevelopmental, non-inflammatory, 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 grey 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, candida albicans, have all been reported to cause aqueductal stenosis and hydrocephalus with pathological changes resulting in the ependyma, and proliferation of glial cushions resulting in gliogenous stenosis. Ventriculitis, as a concommitant 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 likely are a consequence of hydrocephalus, not the primary cause. Grüneberg[13] 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 with-holding 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.[14] postulated that hydrocephalus was secondary to over-secretion 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.

Kesterson and Carlton[15] induced aqueductal stenosis in weaning mice by dietary substitution of hydroziene 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 1970's the causal relationship between viral infection of the brain, the induction of hydrocephalus in experimental animals, and aqueductal stenosis were postulated with Johnson, Johnson, and Edmunds[16] 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.

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 hypo-vitaminosis A, hyper-vitaminosis A, delurium, 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[17] to have hydrocephalus which was secondary to aqueductal stenosis was subsequently reported by Bonnevie in 1943[8] to suffer generalized hydrocephalus first and aqueductal stenosis second!

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 damaging the ependymal cells in the periaqueductal glia of the aqueduct of Sylvius, leaving intact lateral, III, and IV ventricle ependyma. One is hard-put to explain 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 described by Raimondi et al.[18] in their study of patients with the Dandy Walker cyst and confirmed by Jakurbowski and Jefferson.[19]

Occlusion of the foramina of Luschka and Magendie 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 antero-superior 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.

As of 1942, when Taggart and Walker[20] reported three patients with atresia of the foramina of Luschka and Magendie, only six previous cases had appeared in the literature, and only two of these cases had undergone surgical intervention. In the majority of published cases since then, the diagnosis was made either at operation or at post-mortem examination. Today, however, the literature contains no case of atresia of the foramina of Luschka and Magendie diagnosed ante mortem in a child under six weeks of age.

The treatment suggested consists of cyst resection after posterior fossa exploration, or shunting from the lateral or III ventricle. In 1956, Matson[21] 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."

There has been a good deal of controversy concerning the definition of the embryologic chronology as well as the description of the specific pathological changes associated with this particular clinical entity. Taggart and Walker maintained that failure in development of the foramina of Luschka and Magendie, in the fourth 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[22] 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[23] 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,[24] 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.,[25] in a review of their pathological material, came to the same conclusion. Gardner[26] 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 remarkable example of the impermeability of the rhombic roof.

I 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 complete occlusion of the foramina of Luschka and Magendie.

Increase in head size and dolichocephaly are the most commonly observed signs in this syndrome. The occurrence of a soft and sunken fontanel in patients with an enlarged head is particularly noteworthy, as is hypertelorism. Dilation of the IV ventricle and associated dysgenesis of the cerebellar vermis are essential features of the Dandy-Walker syndrome, and pneumoencephalography demonstrating both of these pathologic characteristics essential to the precise diagnosis. Cerebral angiography and/or CTT scanning are not reliable in distinguishing the Dandy-Walker syndrome from an arachnoidal cyst of the posterior fossa or a 'large cisterna magna'. A shunting procedure, whether cystoperitoneal or ventriculoperitoneal, or both, is the treatment of choice. Suboccipital craniotomy with resection of the cystic membrane does not afford the child reasonable chances of cure or palliation. The outcome of treatment of patients with Dandy-Walker syndrome is to be considered unfavorable with regard to psychomotor development, very likely because of associated cerebral anomalies. These unfavorable results are not indicative of unsuccessful surgical management of the problem of intracranial pressure or hydrocephalus.

Treatment for Dandy-Walker syndrome is a shunting procedure, either cystoperitoneal, ventriculoperitoneal, or both. The cystoperitoneal shunt results in an increase in the size of the cerebellum and a reduction in the size of the ventricular cyst.

The outcome of treatment is not favorable in terms of IQ (2/3's 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.

Children with a craniovertebral anomaly such as basilar impression or platybasia, as well as children who have meningomyelocele associated with the Chiari II malformation, may develop a particular form of hydrocephalus which I prefer to call *constrictive hydrocephalus*. 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 pathologic changes within it which result in occlusion of the aqueduct. Many of these children, as well as

all those who develop hydrocephalus, do not have anatomic 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, 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 non-existent or only a potential cavity.

It has long been thought that such *intraventricular tumors* as the choroid plexus papilloma of the lateral ventricle are capable of producing increased amounts of CSF, and that, accordingly, this type of tumor causes communicating hydrocephalus of the hypersecretory variety. Certainly, it is ture that children with choroid plexus papilloma show a clinical picture of both the space occupying lesion and hydrocephalus. In addition, a large majority of these children require ventricular shunts after the choroid plexus papilloma has been totally removed. The reasons for this are not clear. 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.

Milhorat, et al.[27] have shown that the rate of cerebral spinal fluid formation in a 5 year old child who had undergone bilateral choroid plexectomy for communicating hydrocephalus during infancy was 0.35 ml/min, plus minus 0.02 standard deviation: well within normal limits. This failure of choroid plexectomy as a treatment to hydrocephalus had been extensively documented in the world literature previously. Consequently, the cerebral spinal 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, medulloblastoma, may be associated with permanent hydrocephalus ... after the tumor (whether obstructing or not) was totally removed.

Hydrocephalus and Infratentorial Tumors

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. Changes in cerebral blood flow that result from an increase in ICP and ventricular dilation must be considered in the pathogenetic cycle of events. 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, communicating, or hypersecretory.

Increased intracranial pressure and papilledema associated with *intraspinal tumors* are now well documented, though the causes remain unknown. 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, significant elevation in CSF protein level as single and definitive causative factors.

Though it is quite uncommon, hydrocephalus may occur as a complication of an *arteriovenous fistula.* 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 produce 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 cerebral spinal 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 not been reported to cause communicating hydrocephalus.

Genesis of Parenchymal Destruction

Irrespective of the etiology, pathogenesis, or type of hydrocephalus, micro and macro vascular 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, palor 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 dimunition 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, were contradictory.[28] They stated 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[29] and De,[30] 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. The variability of observations of cerebral damage in experimentally induced hydrocephalus is well known, and probably results from lack of uniformity of species, of technique, and of reproducibility. In the hydrocephalic process, it is possible to postulate a sequence of events leading to irreversible brain damage. After the initial, remarkable dilation of the subarachnoid spaces and cisterns, ventricular dilation occurs and is, in turn, followed by periventricular lucency, cerebral edema, obliteration of the subarachnoid spaces, progressive cerebral edema, aqueductal occlusion. The progressive cerebral edema involves first the white matter, but soon extends to the grey matter. Only in the very late, terminal, stages of hydrocephalus does edema occur within the basal ganglia.

Transependymal CSF perfusion, a compensatory mechanism which certainly occurs also in the normal state, is a response to high intraventricular hydrostatic or pulse pressure. The intraventricular cerebral spinal 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 them, causing their caliber to diminish, and resulting in changes in CBF.

The first vascular changes in the development and progression of hydrocephalus consists 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" pattern disappears, the edema and atrophy of the periventricular white matter follow, and ischemia occurs. The end result is the following sequence of events, leading to irreversible brain damage: 1) accumulation of intraventricular CSF under either elevated hydrostatic pressure or increased pulse pressure; 2) increased transependymal flow; 3) parenchymal vascular compression; 4) ischemia; 5) increased extracellular fluid and communication between intraventricular and extracellular fluid; 6) cellular disruption and tissue destruction. The onset of irreversible brain damage becomes obvious with the identification of 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 (before 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. In fact, a significant increase in caliber of the cerebral arteries after ventricular drainage, as evidenced by follow-up with cerebral angiography, suggests that the increase in intracranial pressure associated with progressive hydrocephalus is transmitted from the ventricles by the extracellular space to compress, deform, and displace the cerebral vasculature and, therefore, results in dimin-

ished cerebral blood flow. These changes are compounded by the progressive cerebral edema, and may be directly correlated with clinical observations that the earlier a hydrocephalic child is shunted, providing the shunt remains functional, the greater the chances for the patient to attain normal intellectual and motor function.

Diagnosis

As early as 1918, Walter Dandy[31] 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 1920's Thorotrast enjoyed a brief reign of ventriculographic popularity, only to be abandoned when it was found to cause serious late sequellae. 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, Heinberger et al. studied the dosage, relative toxicity, and radiopacity of intraventricular injections of Conray 60 in animals and, subsequently, in over 100 patients.[32] 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, 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[33] reported preliminary angiographic studies on hydrocephalic infants, and in 1959 Fauré and Gruson[34] described the arteriographic characteristics of such specific tumors as the craniopharyngioma. Paillas,[35] et al. 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 fontanel 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, in 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[36].Computerized transmission tomography (CTT) simplified dramatically and added immeasurably to the needs of diagnosing hydrocephalus in general, and to evaluate with precision the individual stages of the development of the hydrocephalic process. Specifically, one became able to identify hydrocephalus, to distinguish between a communicating and obstructive variety, to learn that dilation of the subarachnoid spaces preceeds the appearance of periventricular edema and that this latter preceeds ventriculomegaly. Of more importance still, one obtains the means of distinguishing reversible from irreversible hydrocephalus and of outlining 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.

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 cerebral spinal fluid being formed; 2) diverting the cerebral spinal fluid outside of the ventricular system and subarachnoid spaces so that it may be absorbed elsewhere in the body; 3) increasing the amount of cerebral spinal 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 use of hypertonic agents, carbonic anhydrase inhibitors, steroids, etc., have not been successful in diminishing the amount of cerebral spinal fluid formed. No drug has been used to increase the amount of cerebral spinal fluid absorbed. The treatment today, as when the first shunting procedure was performed in the late ninteenth century, consists of deviating cerebral spinal 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[37] 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[38], developed techniques to connect the ventricles with either the subarachnoid or subdural spaces. Ballance compared the merits of gold and platinum to glass. Cerebral spinal fluid was first shunted from the lumbar subarachnoid space to the peritoneal cavity by Ferguson in 1898;[39] from the lateral ventricle, using a small caliber rubber catheter into the peritoneal cavity by Kausch in 1908,[40] and into the circulatory system by Payr in 1908.[41] Haynes, in 1913, drained CSF from the cisterna magna into the cranial sinuses.[42]

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.[43] 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,[44] 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[45] stressed the indications for use of the ureter, Ransohoff[46] for the pleural cavity, and Harsh[47] 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 problems 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 jugulo-cardiac 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.[48] and inflammatory pseudotumor of the mesentery by Keen and Weitzner.[49] 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, intestinal perforation. The intracranial complications consist of shunt tubing lost in the ventricle, subdural hematoma, unsuccessful ventricular cannalization, intraventricular or intraparenchymal hemorrhage, brain abscess, hemiparesis. The general complications consist of stitch infection, wound break down, electrolytic imbalance, 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—s. epidermides and s. aureus. The only exceptions occur in the meningomyelocele children, who tend to develop gram negative ventriculitis. The incidence of shunt infection is much higher in those newborn, especially with meningomyelocele, who are shunted during the first weeks of life ... so that it is advisable to postpone surgery to the fourth 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 non-operatively 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 congenital disease was based. Progressive increase of head size, bulging fontanel, 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 to 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, hydrancephaly, 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 I.Q. 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 I.Q. and severity of hydrocephalus 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 Treatment

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, 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.

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 deviated either from 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.

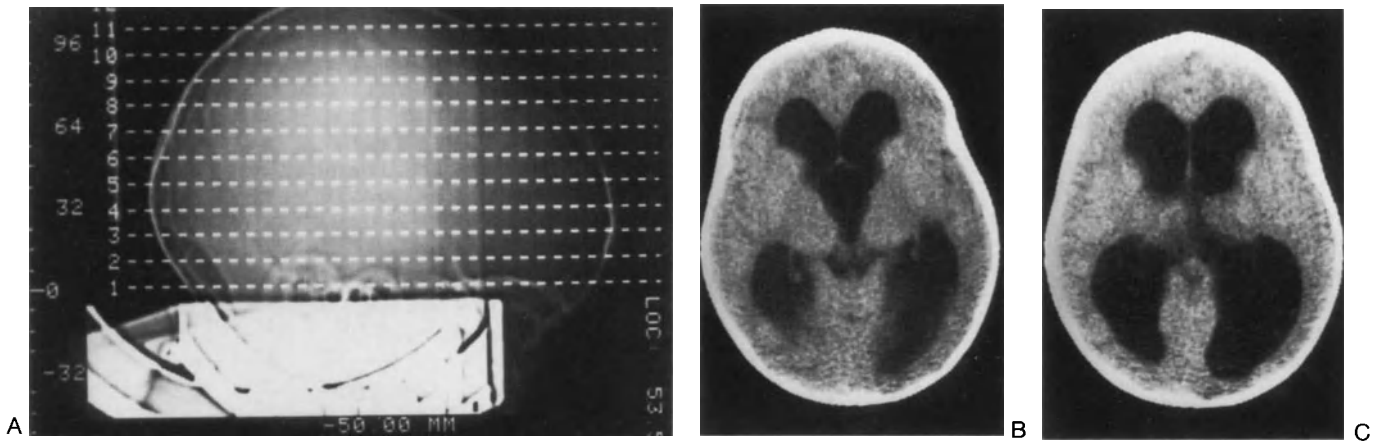
The indications and technique for external ventricular drainage have been described in the chapter on infections.

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 (Figure 15.1 to 15.3)
The location of the occipital horn should be plotted out on a CTT scan, through coronal 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, the base of the mastoid. (One should not use only external indicators, such as the parietal eminence, the midline, the external occipital protrusion, 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 which is orthogonal to the trunk). Settling for an obliquely positioned head may prove to be dangerous.

Either a semilunar or, preferably, horseshoe-shaped incision is then made, taking care that it be 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 twist drill or a 10-nm burr 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 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



A B C

Figure 15.1. The insertion of a cannula into the lateral ventricle should be planned with the use of a CTT 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 interval) plotted out, permit one to identify a known landmark or to cross reference with a single CTT 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.

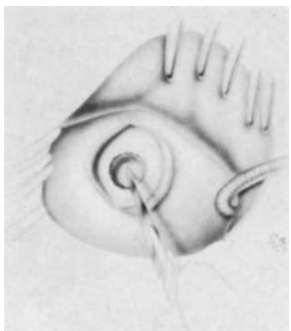
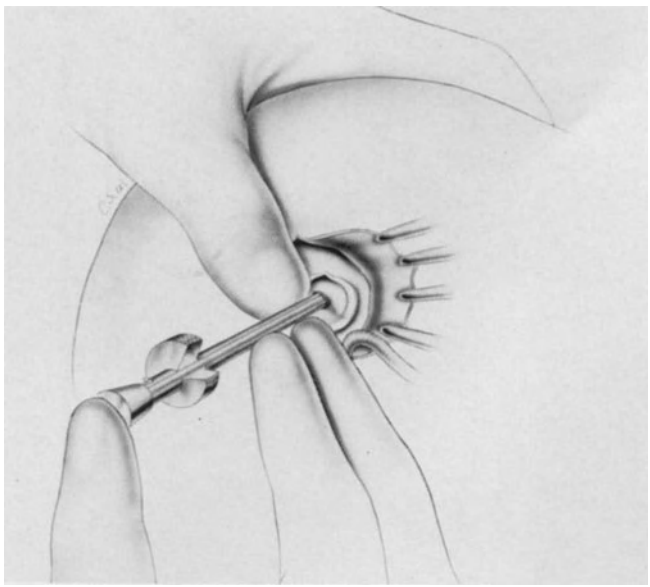


Figure 15.2. Making a cruciate incision in the dura, it is advisable to ascertain that the incision is large enough to permit the trocar free passage, for if it is small the surgeon may strip the dura from the skull as he inserts the trocar. This may result in a post operative epidural hematoma.

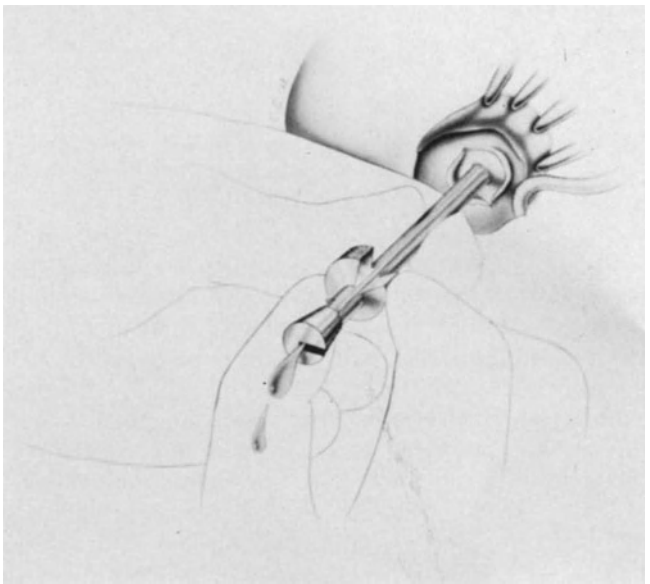
inserted through the barrel of the cannula into the ventricle. The cannula is removed and the distal end of the shunt tubing inspected to document flow of cerebrospinal fluid. Air pockets may obstruct the flow of fluid, so that it is often necessary to irrigate with 2–3 cc of saline. When free flow is 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.

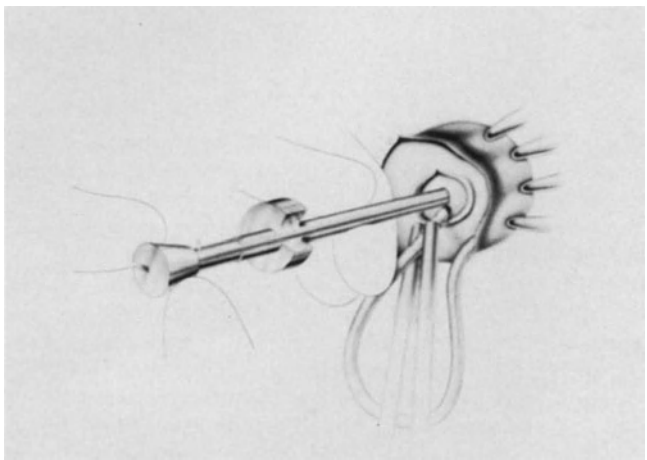
The multiperforated, flange-tipped ventricular catheter is subject to greater adhesions of choroid plexus, rendering it potentially more prone to complications than the smooth ventricular tip which has perforation openings. The use of a rigid obturator, placed in the shunt lumen, to permit insertion 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, 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.



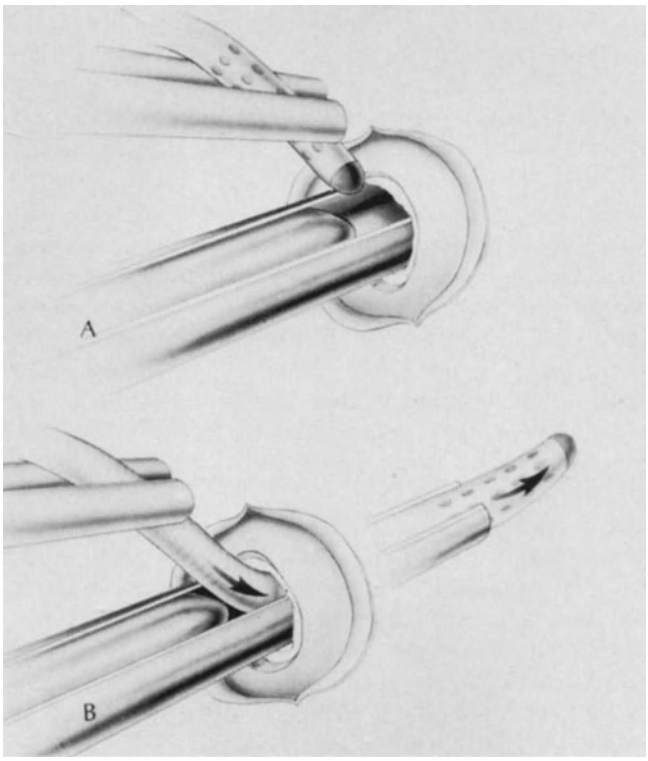
A



B



C



D

Figure 15.3. (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*) so as to facilitate threading the ventricular tip of the Uni-Shunt into the ventricle along the barrel of the split trocar (*B*).

Frontal Horn Cannulation

Coronal reconstruct CTT 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 CTT 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.

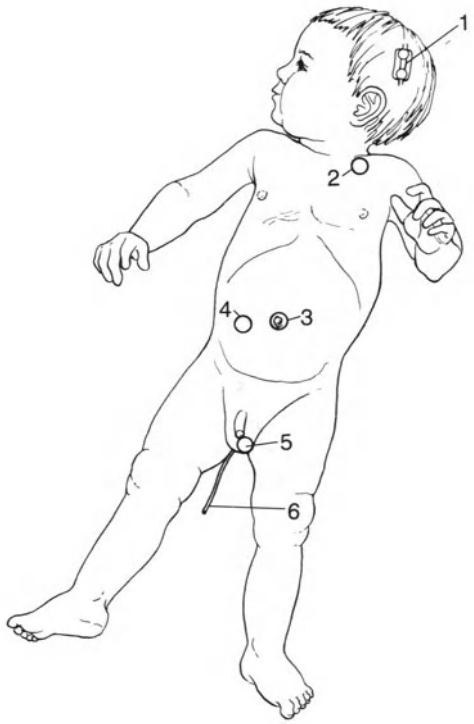


Figure 15.4. Areas of extrusion, disconnection, breakage, colonization, and occlusion along the path of the ventriculoperitoneal shunt. At the cranial end (*1*) one may observe disconnection of the shunting system if it is a three-piece unit (but not if it is a Uni-Shunt), extrusion of either a three-piece 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 puts into evidence 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 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 intraabdominal portion, may become coated with a proteinaceous material, which effectively seals the slit valve, obstructing drainage.

Shunts

Ventriculoperitoneal Shunt (Figures 15.4 to 15.22)

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 in-

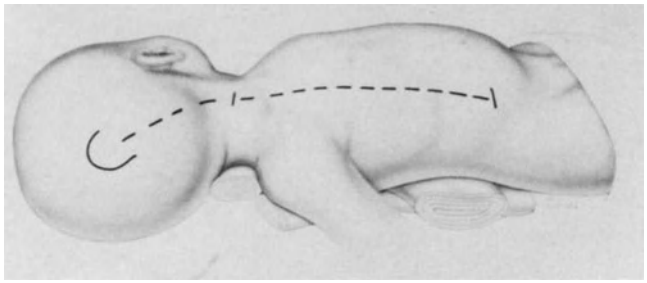


Figure 15.5. Recommended position for the patient. The tract for passage of the shunt through the subcutaneous space is indicated by the broken line, the skin incision by the intact lines.

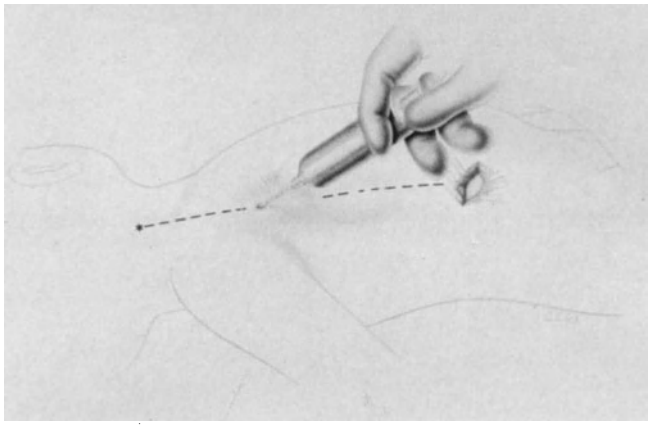


Figure 15.6. Normal saline is being injected into the subcutaneous space so as to separate the skin from the body wall. This permits an atraumatic and easy passage of the guide.

traabdominal 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 intraabdominal 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 under general anesthesia without allowing them to lighten, 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 Figure 15.5 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, 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.

After the incisions have been made and clamps put on either the subcutaneous tissue or the Galea, normal

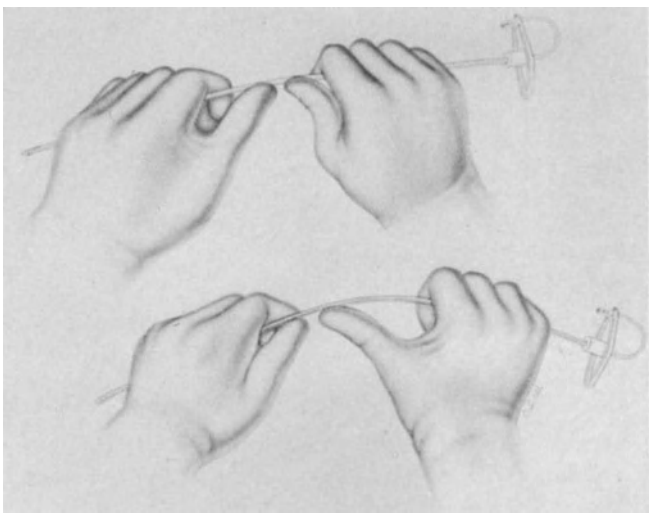


Figure 15.7. Molding of the subcutaneous guide.

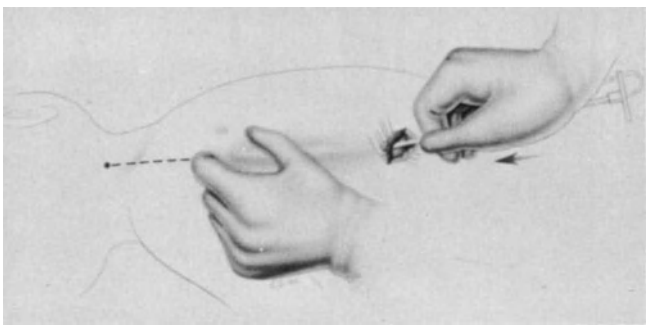


Figure 15.8. Passage of the guide through the subcutaneous space.

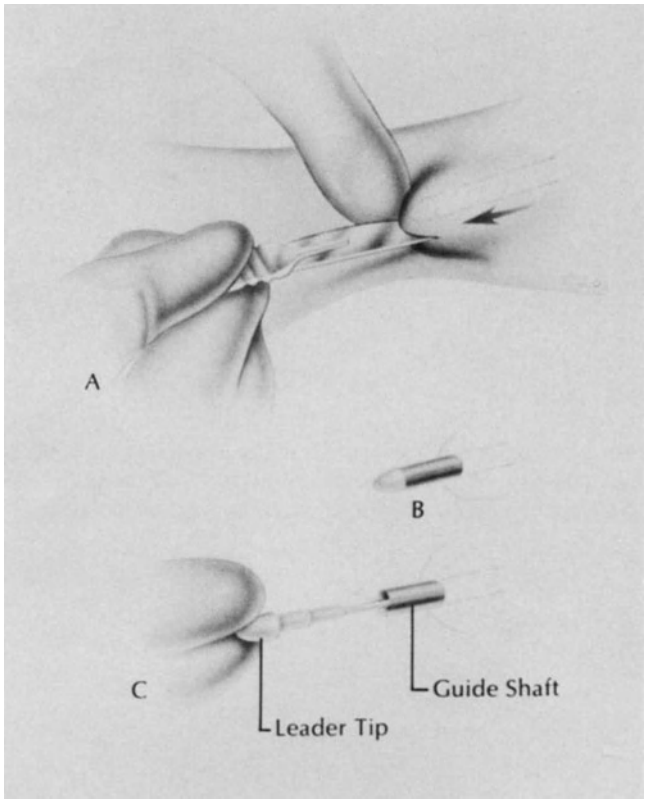


Figure 15.9. (A) The skin incision is being deepened, so as to permit exit of the leader tip and guide. (B) The leader tip and guide shaft have exited. (C) The leader tip is being pulled through the guide shaft.

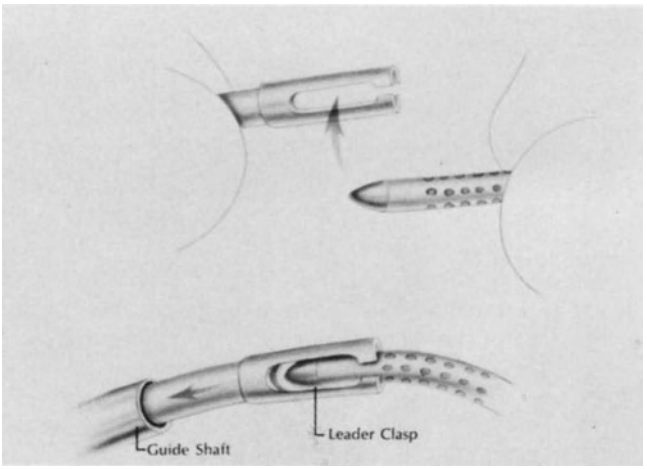


Figure 15.10. The ventricular end of the shunt is being readied for insertion into the clasp of the leader. Below, it has already been nestled into the clasp and the leader is being drawn through the guide shaft, pulling the shunt into the subcutaneous space.

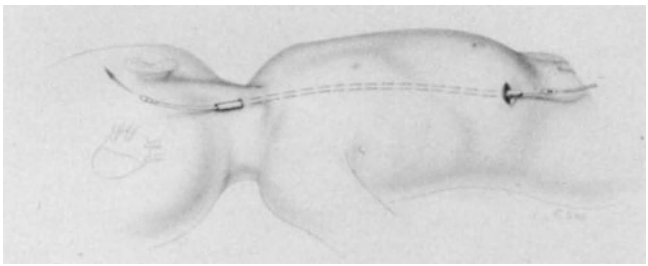


Figure 15.11. The guide rests in the subcutaneous space and the shunt is within it. When the guide shaft is withdrawn, the shunt will rest within the subcutaneous space.

saline is injected into the subcutaneous space, along the proposed shunt path, separating the subcutaneous connective tissue from the abdominal wall, the thoracic cage, 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 Figure 15.6, 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 both hands, and curving it to the desired form between the thumbs. This should be done before inserting

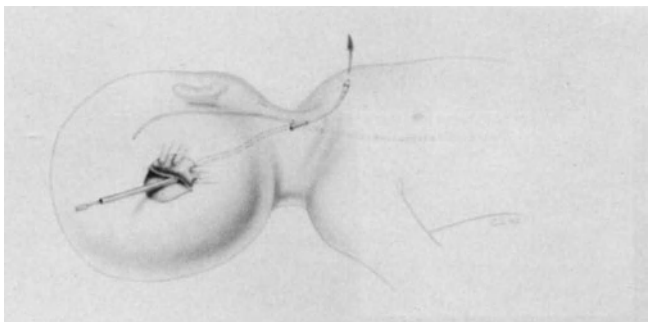
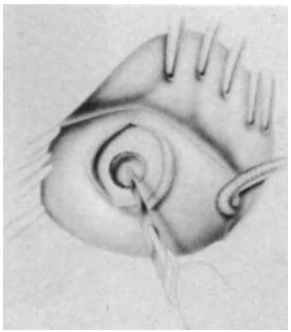


Figure 15.12. Passage of the guide and leader tip through the subcutaneous space from the parietal eminence to the supraclavicular space.

Figure 15.13. Making a cruciate incision in the dura.



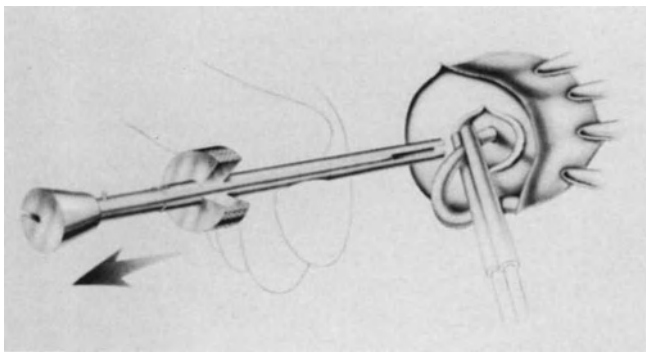


Figure 15.14. With the ventricular tip of the shunt in the ventricle, the barrel of the split trocar is withdrawn as the shunt is held securely in place.

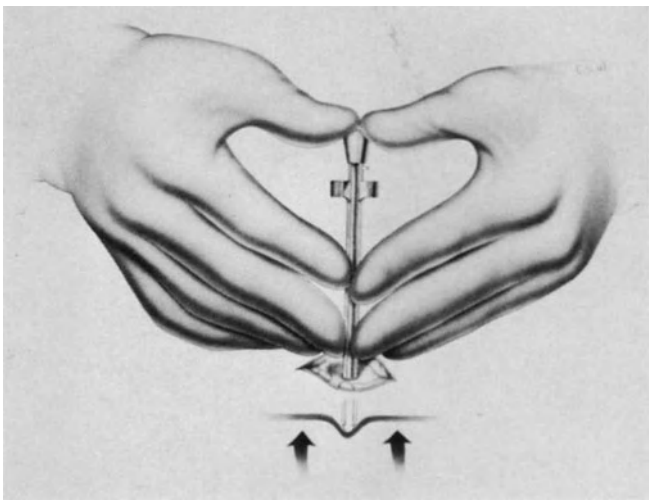


Figure 15.15. The trocar is brought against the peritoneum, exciting the guarding reflex so as to tighten the peritoneum and facilitate puncture into the peritoneal cavity. The peritoneum must be rigid before puncturing it, lest intraabdominal vessels and viscera be damaged!

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 an 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.

Once the tip and guide have been advanced through the supraclavicular opening, the leader tip is withdrawn for a distance of 3 to 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.

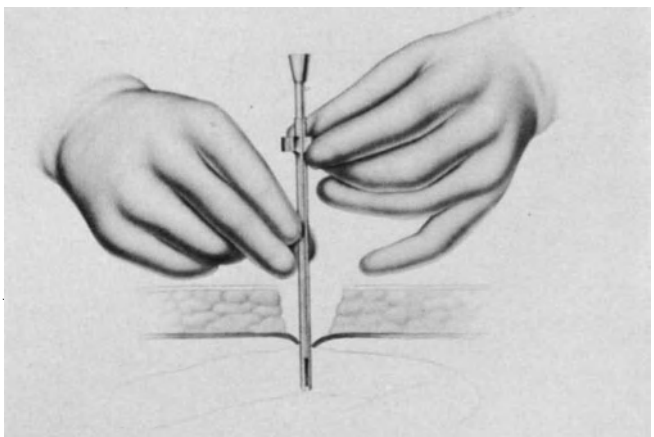


Figure 15.16. The peritoneal cavity is punctured and the obturator is being removed, leaving the barrel of the split trocar in place.

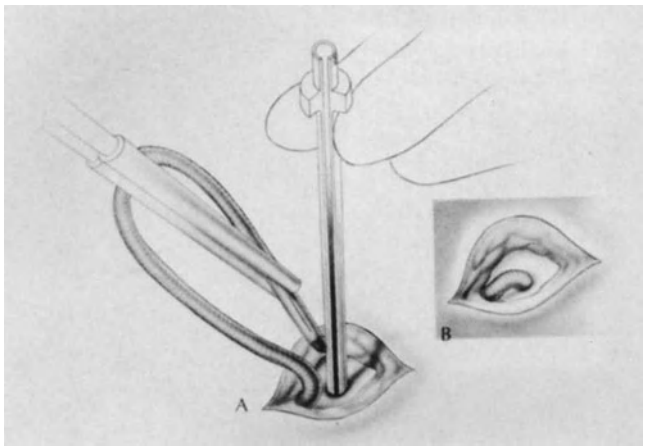


Figure 15.17. (A) The peritoneal end of the shunt is being threaded into the barrel of the trocar for passage into the peritoneal cavity; (B) then the barrel is removed.

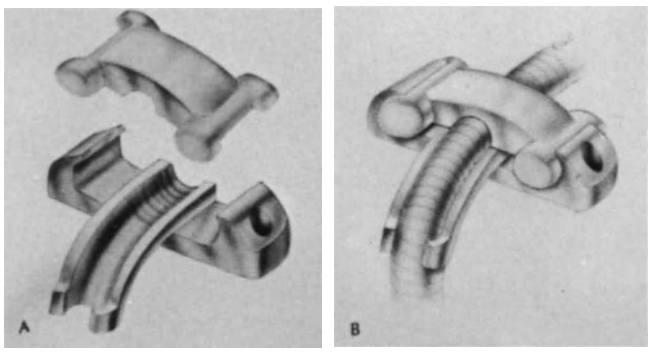


Figure 15.18. (A) The two disarticulated pieces of the clip; (B) locked into one another over the shunt.

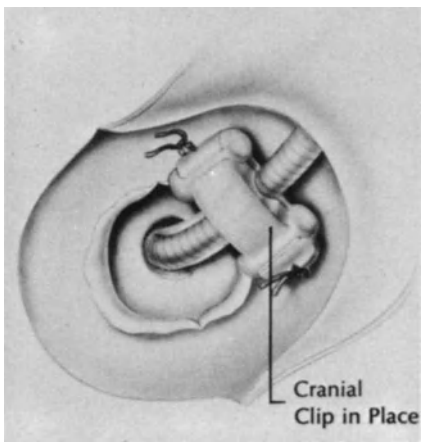


Figure 15.19. Anchored lock-clip locking shunt into place at cephalic end.

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 an # 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 a Cushing forceps, provided 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.

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" consits 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," gently stretch the tubing and ease the catheter into the clip. The "slip-clip" is then anchored to the abdominal musculature.

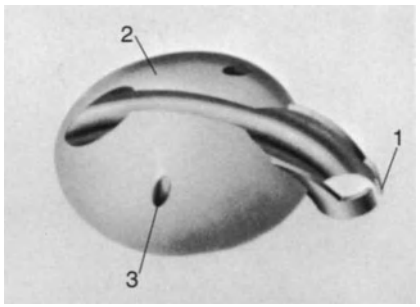


Figure 15.20. Slip-clip (abdominal end). The clasp (*1*) directs the distal end of the shunt tubing into the peritoneal cavity. The groove (*2*) is smaller than the external circumference of the Uni-Shunt, so it holds it snuggly in place. The perforations on either side (*3*) are for passage of suture material for anchoring of the slip-clip to the abdominal wall.

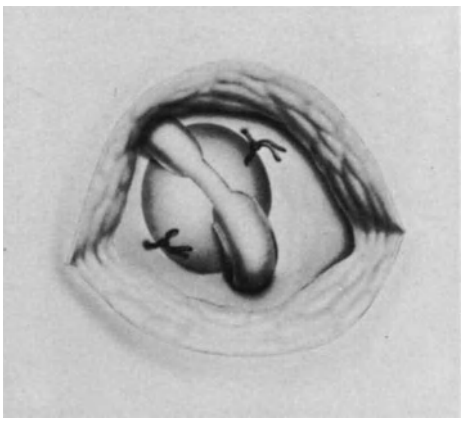


Figure 15.21. Anchored slip-clip holding shunt in place at abdominal end.

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 cor-

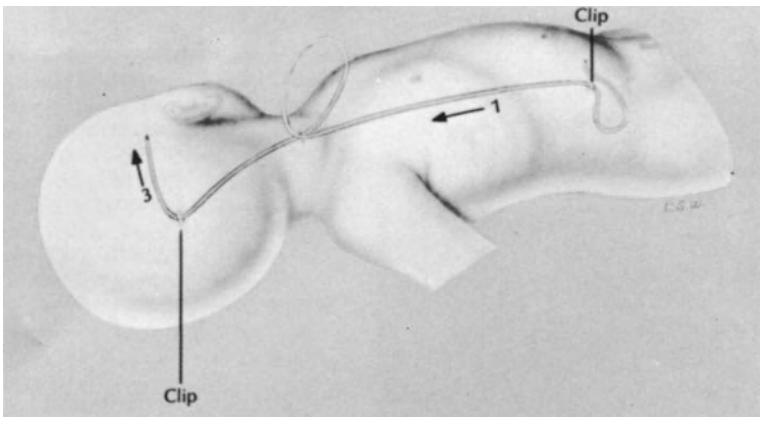


Figure 15.22. Four steps for inserting shunt if one uses split trocar for ventricular end.

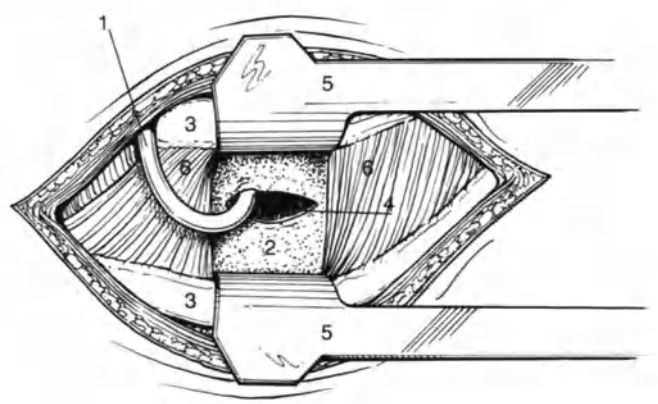


Figure 15.23. Ventriculopleural shunt insertion. At the time of insertion of the distal end of the shunt tubing (*1*) through the parietal pleura (*2*) one should spread the ribs (*3*) and ask the anesthesiologist to collapse the lung (*4*), in order to allow the shunt tubing to pass freely into the 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.

rect 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, 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 (Figure 15.23)

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[50] 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 are inserted into the 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.[51]

Ventriculogallbladder Shunt (Figure 15.24)

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/hr, 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 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 to 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 before water or cerebrospinal fluid may move across it at adequate absorptive volumes.

In 1958, Newman, Hoen, and Davis[52] treated a case of communicating hydrocephalus by shunting the cerebrospinal fluid into an isolated segment of ilium, demonstrating absorption over a period of ten 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[53] 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 three months, and seven were alive two months, five months, twelve months, and two years later.

Yarzagaray (L. Yarzagaray, unpublished observations) reported a six-year follow-up of 50 children with hydrocephalus: thirty-five cases of Chiari types II and III; eleven cases of aqueductal stenosis; three cases of Dandy-Walker; and one case of sarcoma of the meninges. Of this number, 32 patients had previous infections: twenty with ventriculitis, and twelve 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 an "hydrops" appearance: enlarged with thick walls, the peritoneum covering it was edematous. He reported no obstructions (intraoperative cholangiography).

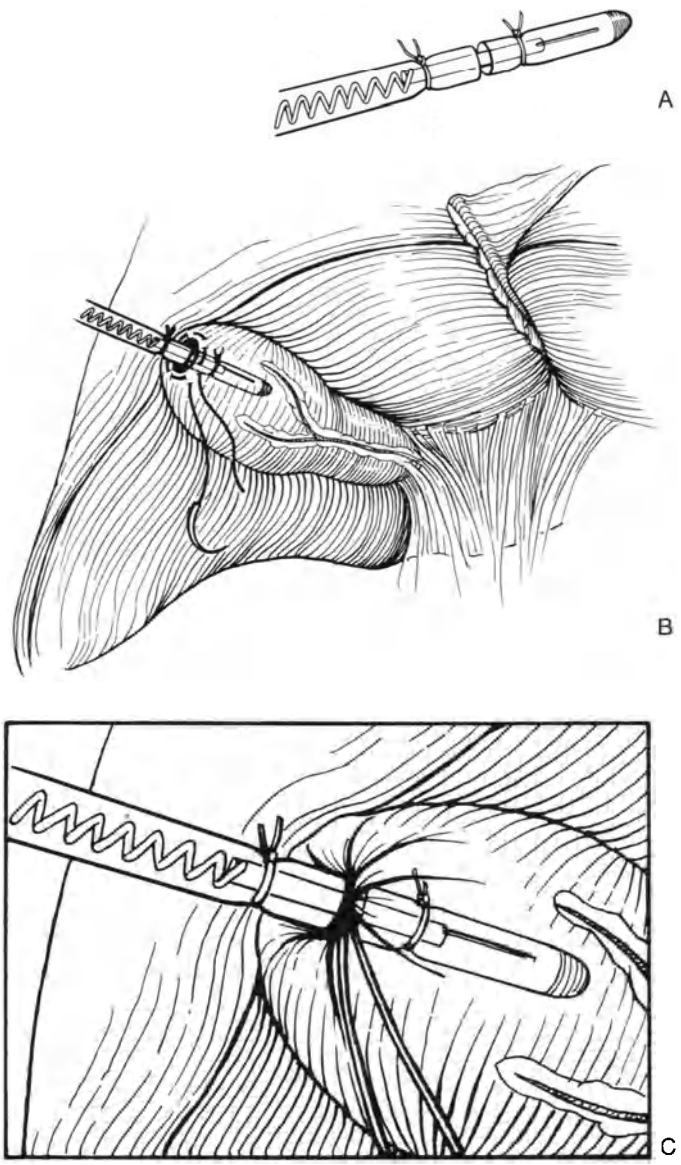


Figure 15.24. Ventriculogallbladder shunt insertion. (A) About 2 cm of the distal (slit) valve and spring catheter are prepared to receive a straight connector. After this is inserted and secured with a 4-0 suture, the proximal portion of the spring catheter is mounted over the straight connector and 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.

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 recon-

nected to the spring catheter with a straight connector. The distal portion is secured with 2-0 suture material. (The only purpose for 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 a 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. I have had two cases of bile ventriculitis, one fatal within less than 48 hours.

Ventriculoamniotic Shunt:
J. T. Brown Technique[54] (Figure 15.25)

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 "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[55] and primates.[56] Most recently, percutaneous ventriculoaminotic shunts have been placed in human fetuses under sonographic direction.[57, 58]

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.

The surgical team consits 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.

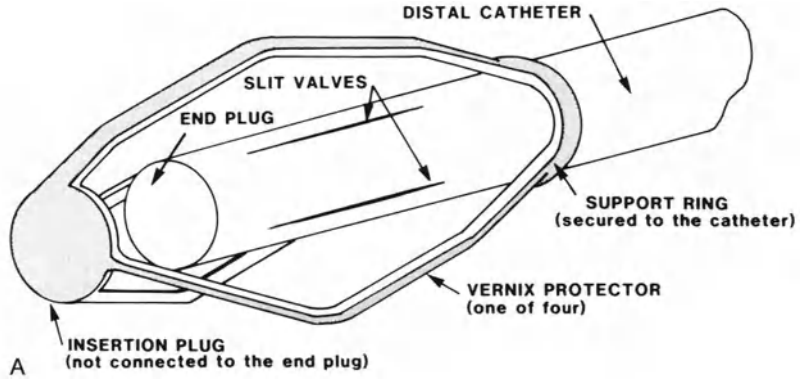
After the maternal abdomen is prepped and draped in a standard fashion, a sterile (draped) sonographic probe is used to locate the precise position of the fetus' head. Not surprisingly, the fetus is often in a breech presentation. A vertex presentation, however, does not preclude placement of a ventriculoamniotic 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- 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.

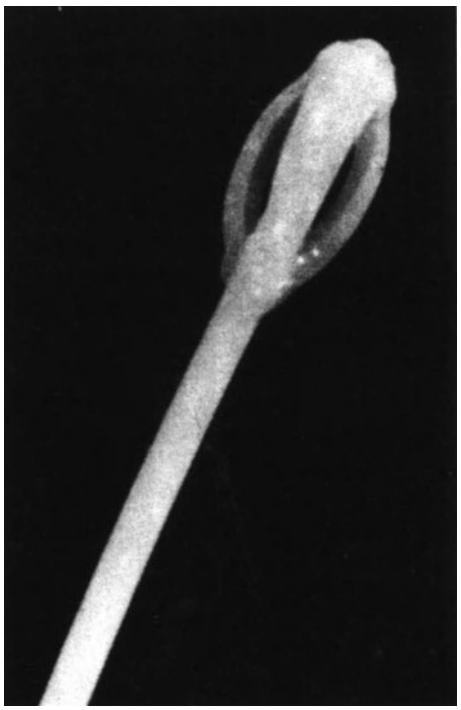
The site of trocar penetration into the cerebral ventricle often varies, but usually occurs at or just in front of the coronal suture. Alternatively, one may insert the trocar just behind the parietal eminence directing it anteromedially. The right hemisphere is chosen for penetration whenever possible.

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.

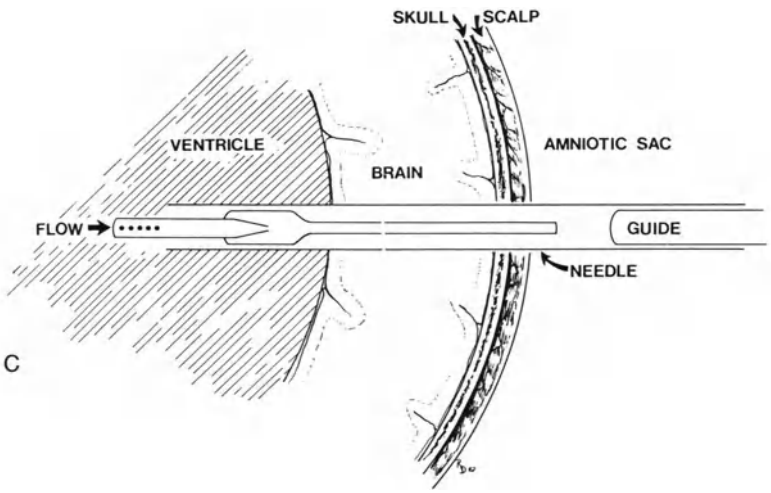
Figure 15.25. (A) Diagrammatic representation of vernix protector. (B) Distal (amniotic) end of new fetal shunt with vernix protector. (C) Ventriculoamniotic shunt driven down trocar by blunt stylet. (D) Twelve-gauge trocar with sharp stylet encountering fetal cranium. (E) Proximal (ventricular) end of new fetal shunt with ventricular flange. (F) Newly proposed fetal shunt.



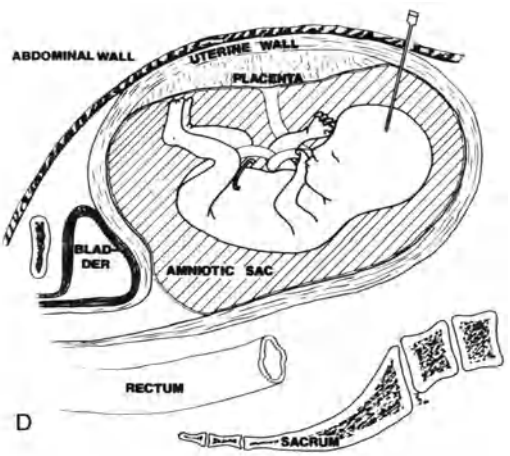
A



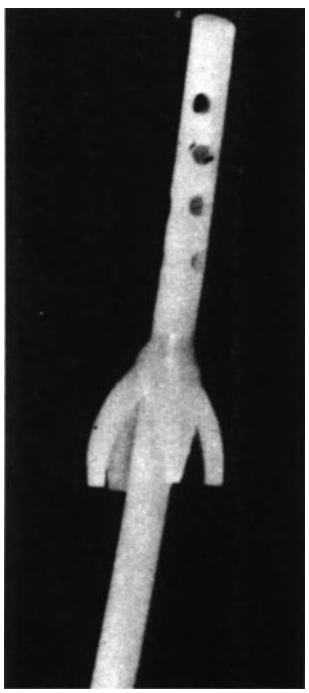
B



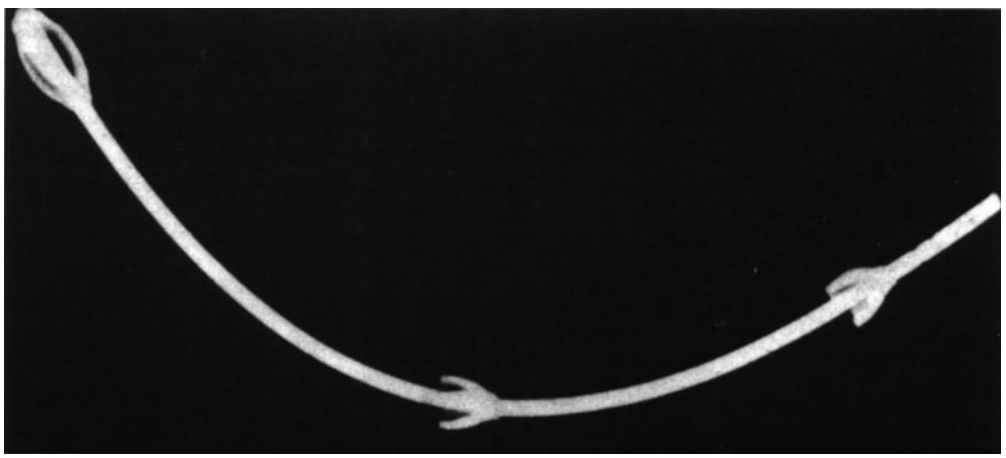
C



D



E



F

After cerebrospinal fluid is vented, the Newkirk* ventriculoamniotic shunt 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 2- or 3-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 lu-

* Denver Surgical Developments, Inc., 6851 Highway 73, Evergreen, Colorado.

men 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 3-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 (Figure 15.4)

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 first 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 get 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; and
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 forcè!"* 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 should immediately be placed over the ostium of the shunt tract to prevent draining of the ventricle. The shunt tip should then be 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 should be 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 should 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-cc 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 minutes before proceeding to reinsert the distal tip into the absorptive cavity to assure himself 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 to 5 minutes, 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 minutes, opening the shunt tubing every 2 or 3 minutes 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 cc of 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 either (in the subcutaneous tissue, the peritoneal cavity, the cerebrospinal fluid flowing through the tubing), he 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.

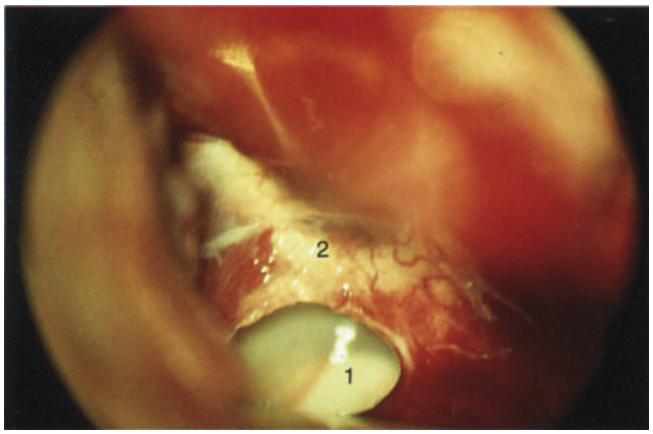
Intracranial Shunting

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.

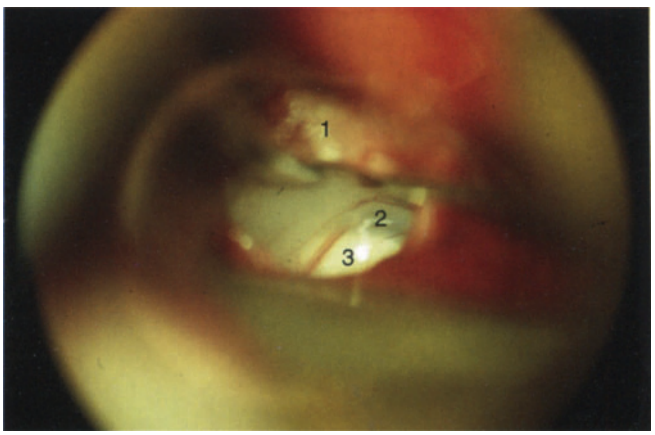
Ventriculoventricular Shunting

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, these procedures are generally unsatisfactory so that sooner on later one finds it necessary to proceed to a shunting operation.



A



B

Figure 15.26. 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.

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 (Figure 15.26)
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, between 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 one or more years, both the lamina terminalis and the posteroinferior floor

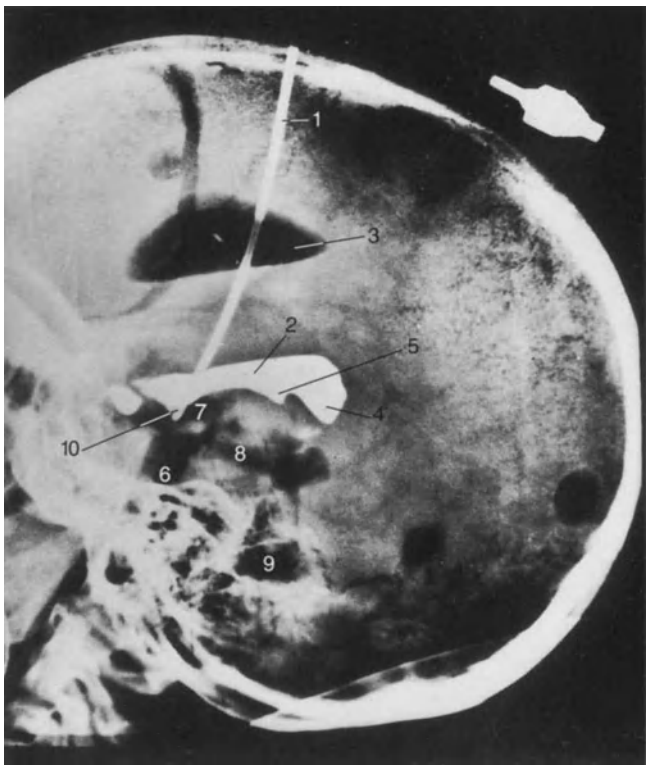


Figure 15.27. Stereotaxic, transventricular, III ventriculostomy. A catheter (*1*) has been inserted into the III ventricle (*2*) through the right lateral ventricle (*3*). One notes the positive contrast media within the inferior III ventricle and, within the chamber, the suprapineal (*4*) and pineal (*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 interperduncular cistern, establishing a III ventriculostomy.

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
(Figure 15.27)
The closed technique for III ventriculostomy may be performed through a burr 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 CTT, 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.[59]

It is my 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.

The Torkildsen Procedure (Figure 15.28)
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 burr 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 (Figures 15.29 and 15.30)
The IV ventriculocisternostomy procedure has been used in vain in attempts to cure cystic transformation

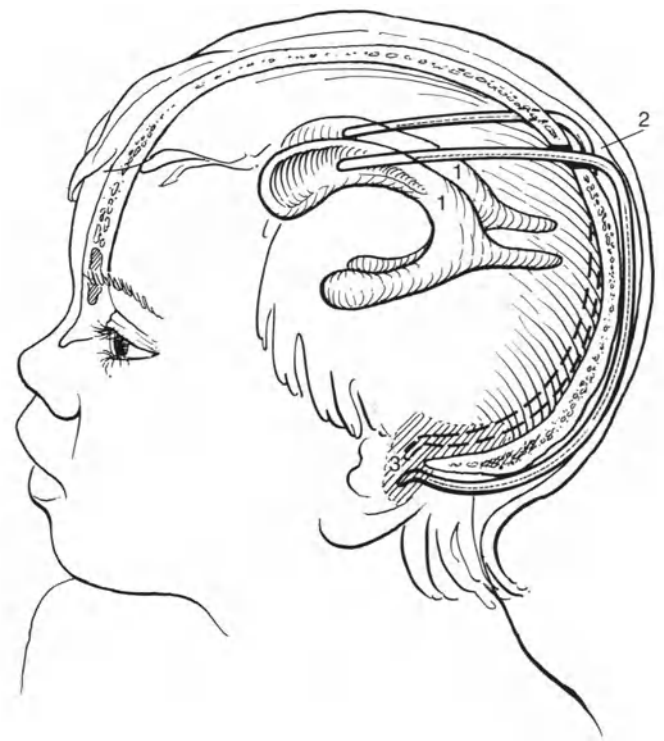


Figure 15.28. Drawing of the skull with the head placed obliquely to illustrate placement of 2 catheters into the lateral ventricles (*1*) and their passage through the subcutaneous space (*2*) down to the cisterna magna (*3*).

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.

Basic Structure of Shunt Systems

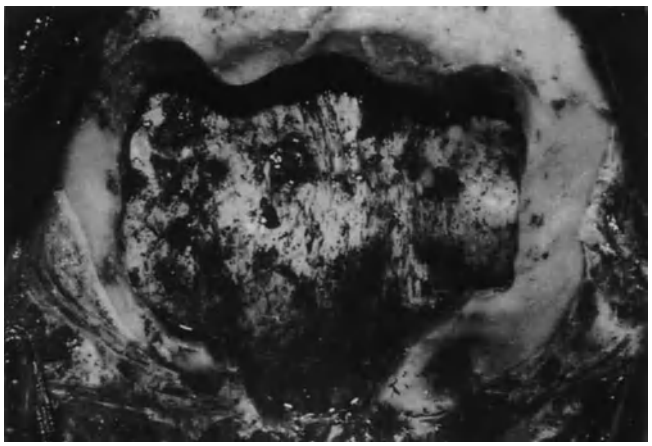
The basic structure of the first valve regulated shunt systems (Fig. 15.31) used consisted of three functional parts: (1) ventricular catheter, (2) reservoir/valve, and (3) peritoneal/atrial catheters. Subsequently, improvements evolved into two-piece functional systems, joining proximal and distal catheters at a single connector, and then into a one-piece system to provide maximum simplicity and efficiency.

1) Ventricular catheters: Fig. 15.32—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.

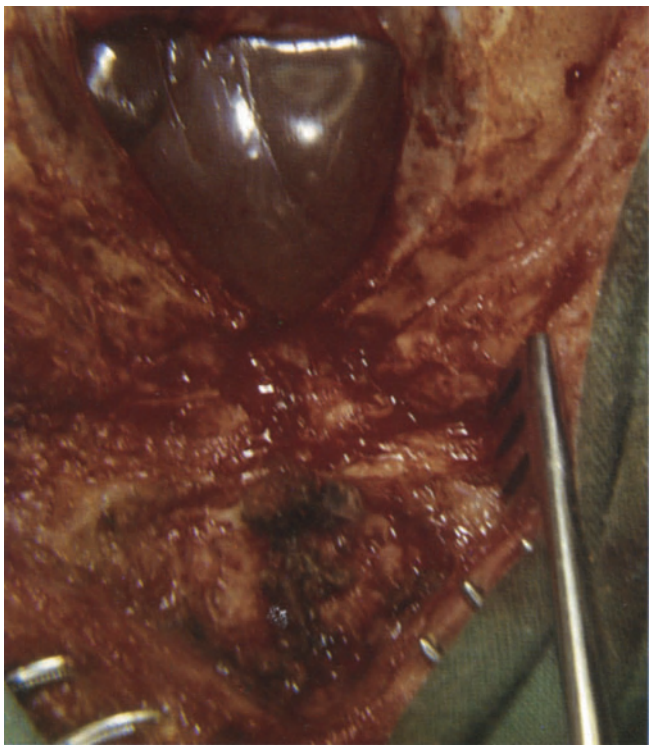
Fanged catheters, designed to be advantageous in that they are not obstructed by the choroid plexus or the ventricular walls, are liable to become *unremovable* at the time of revision. The X-ray marked, at one centimeter intervals, catheters are convenient for insertion into the ventricle.

2) Flushing devices—Theoretically flushing devices are classified into two types: i.e. one which functions as reservoir (Fig. 15.33) and the other which, in addition to the above function, has a pressure-controlling valve (Fig. 15.34). The hoped for objectives of 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.

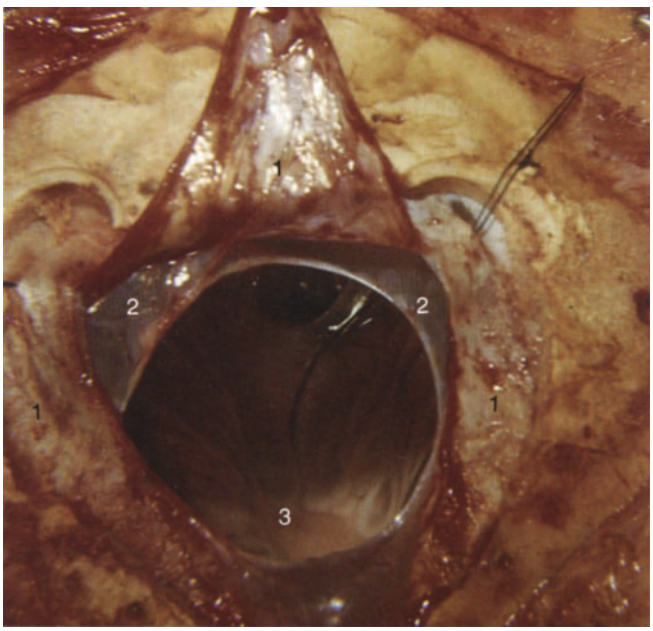
For the flat bottom type device, the twist drill hole fits well, but, with the burr hole type device, better fixation of the reservoir can be achieved; consequently, pressure upon the skin may be reduced. In neonates and infants, since the placement of a reservoir may cause tension in the skin and eventually lead to skin necrosis, it is desirable to use a right angle tube, connec-



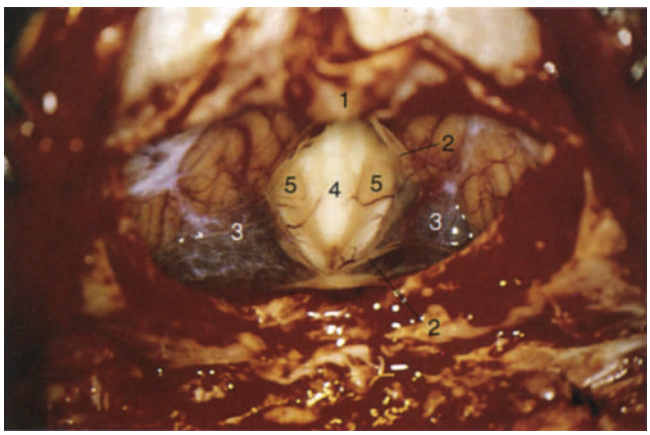
A



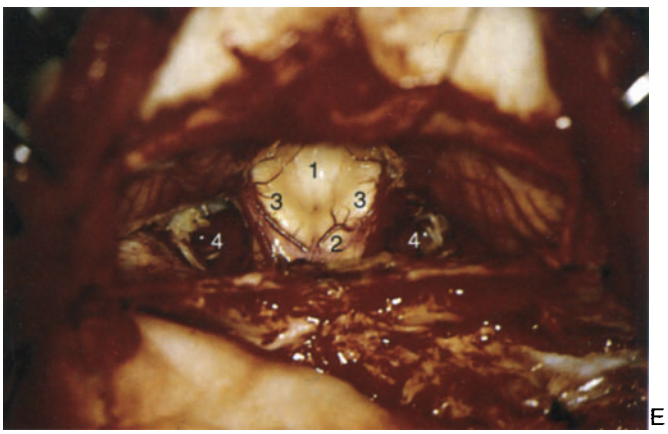
B



C



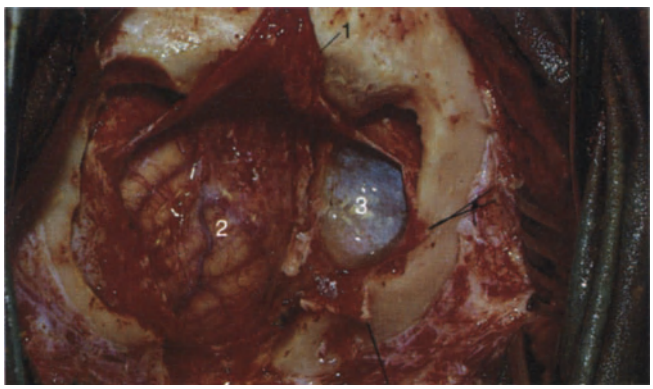
D



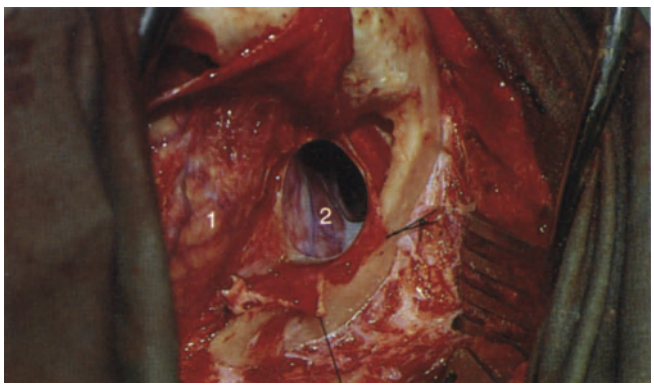
E

Figure 15.29. 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 cisterns 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) This child had had lateral ventriculoperitoneal shunting performed at birth. Her hydrocephalus was compensated until the time of this surgery, when her shunt obstructed and the surgeons chose to perform a suboccipital craniotomy and "freeing of adhesions between the cerebellum and brainstem so as to establish communication between IV ventricle and basal cistern." The informative observation here is the extraordinary thickness of the skull, something which one commonly observes in children shunted at birth, with long-standing com- ►

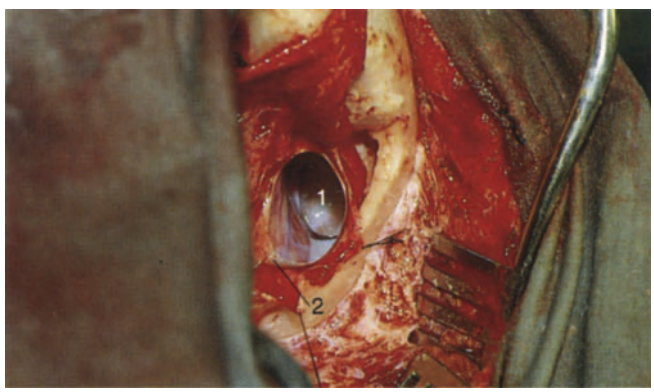
pensated hydrocephalus. (B) The suboccipital craniotomy and reflection of the dura mater have been performed, revealing the posterior inferior aspect of the cystic IV ventricle (arrows). (C) After the dura mater (*1*) has been reflected superiorly and the posterior inferior aspect of the encysted IV ventricle (*2*) opened, one looks directly at the floor of the IV ventricle (*3*) and the enormously dilated aqueduct of Sylvius (*4*). (D) 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. (E) The colliculus facialis (*1*) and 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.



A



B



C

Figure 15.30. Though unusual, the encysted IV ventricle may expand lateral to one or the other cerebellar hemispheres, rather than in the midline along the vallecular opening into the void left by the dysplastic 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 (*3*) which has expanded lateral to the right cerebellar hemisphere and into the region of the lateral recess 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*).

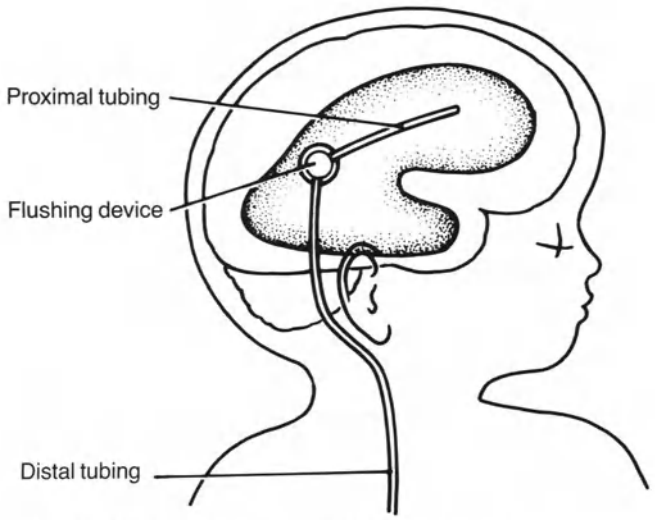


Figure 15.31. Basic structure of CSF shunt system.

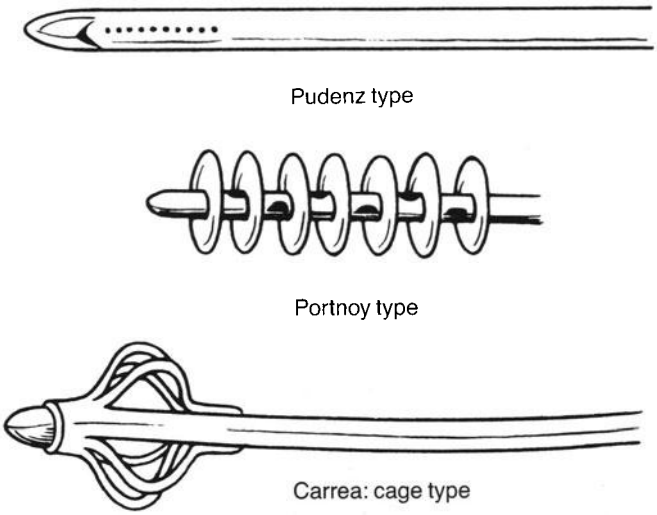
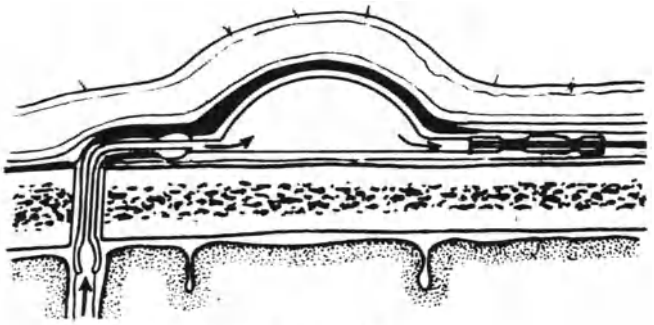


Figure 15.32. Various types of ventricular catheter.



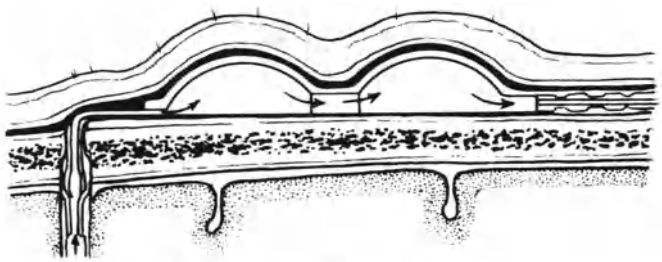


Figure 15.33. Various types of flushing reservoir.

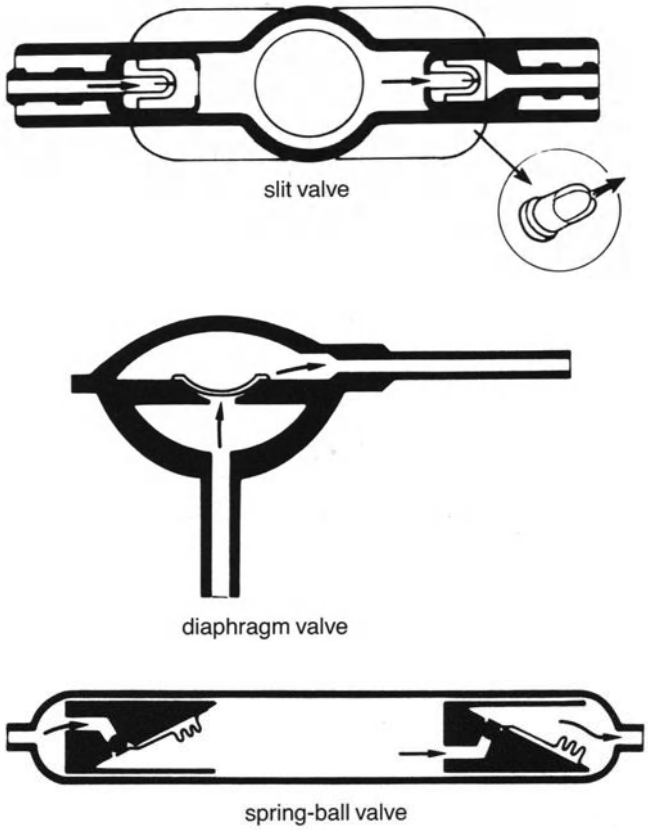


Figure 15.34. Various types of pressure-controlling valves.

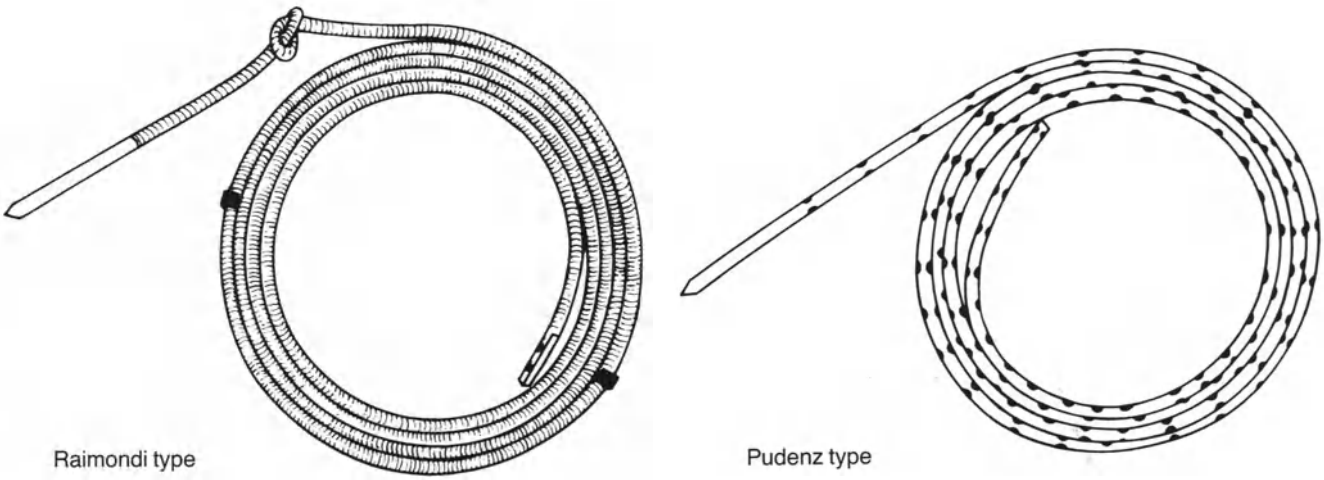


Figure 15.35. Various types of peritoneal catheters.

tor, or preferably, Unishunt so as to avoid the use of a reservoir. Recently, a reservoir 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. 15.35—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. 15.36) 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.

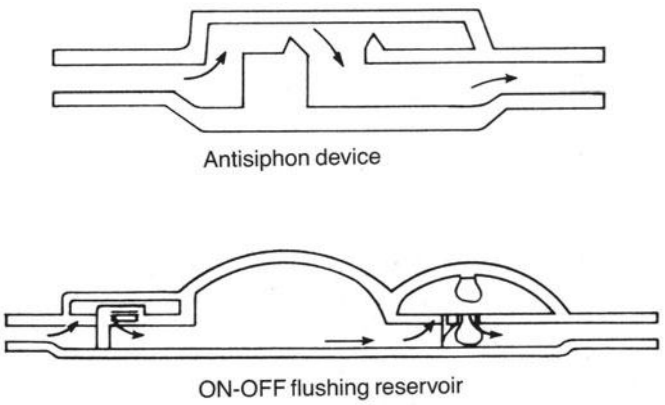


Figure 15.36. Other devices.

Characteristics of the Flow Rate

The drained CSF volume (ml) per minute is shown in the pressure-flow rate (resistance) curve (Fig. 15.37). 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 (Approx. 0.35 ml/minute) and the CSF pressure (approx 40 to 110 mm H_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. 15.38). The different curve profiles depending on various systems, are ascribable to differences in the pressure control valve, and, on the basis of Poiseuille's law (Fig. 15.39), in the diameter and length of catheters used in each system.

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

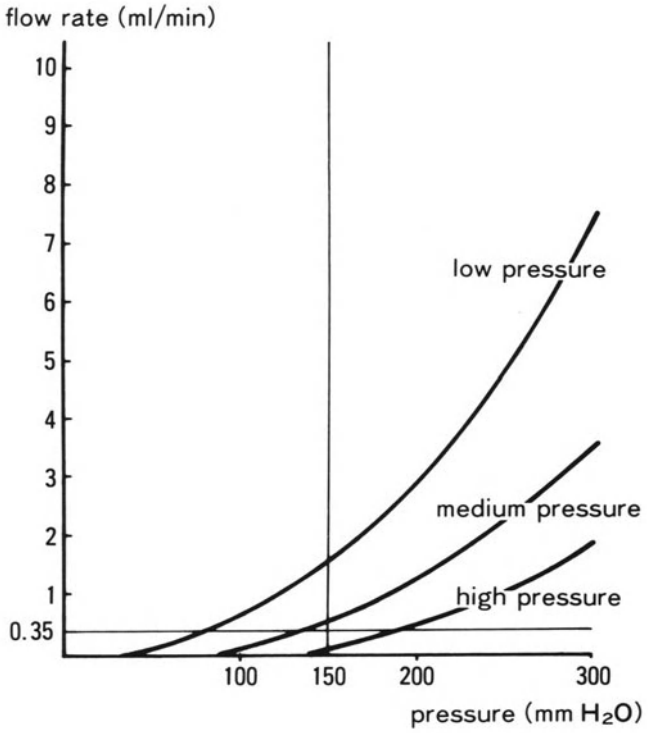


Figure 15.37. Pressure–flow rate curve.

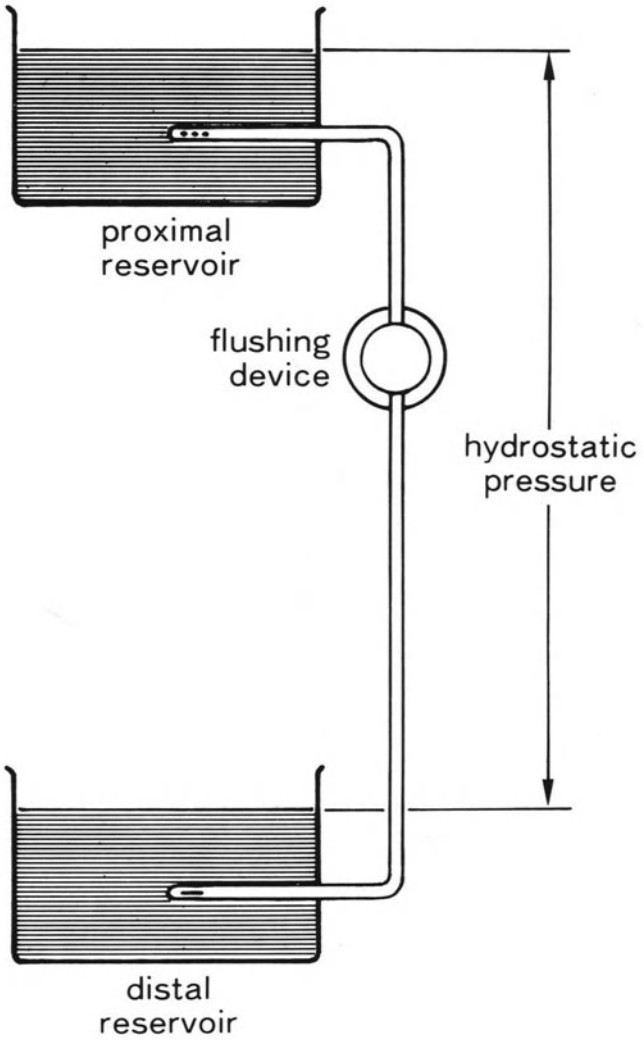


Figure 15.38. Measurement of the flow rate of shunt systems.

$$F = \Delta P \left(\frac{\pi}{8}\right)\left(\frac{1}{n}\right)\left(\frac{r^4}{l}\right)$$

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 15.39. Poiseuille's law.

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 anatomo-physiology 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. 15.34). 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. 15.40). 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

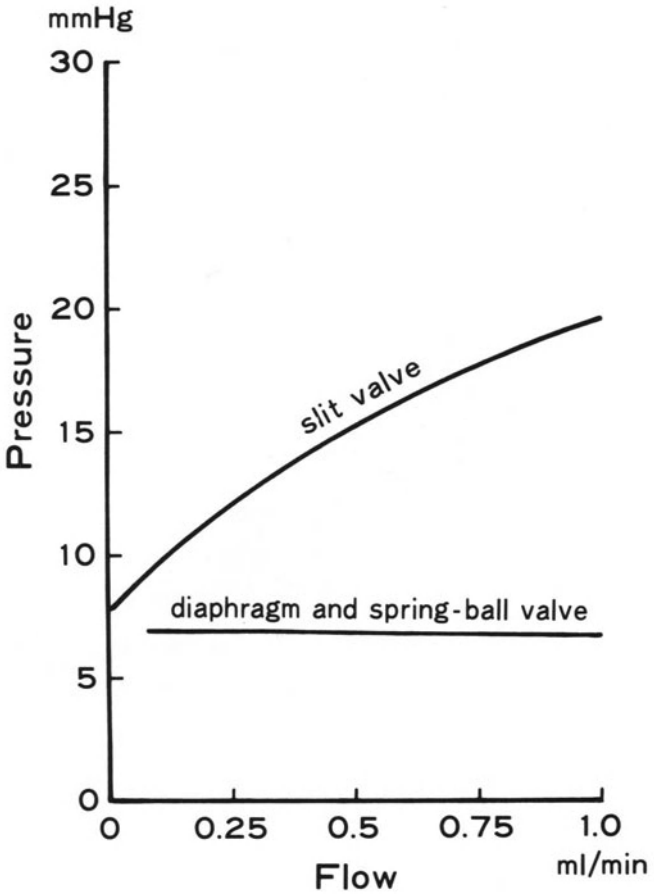
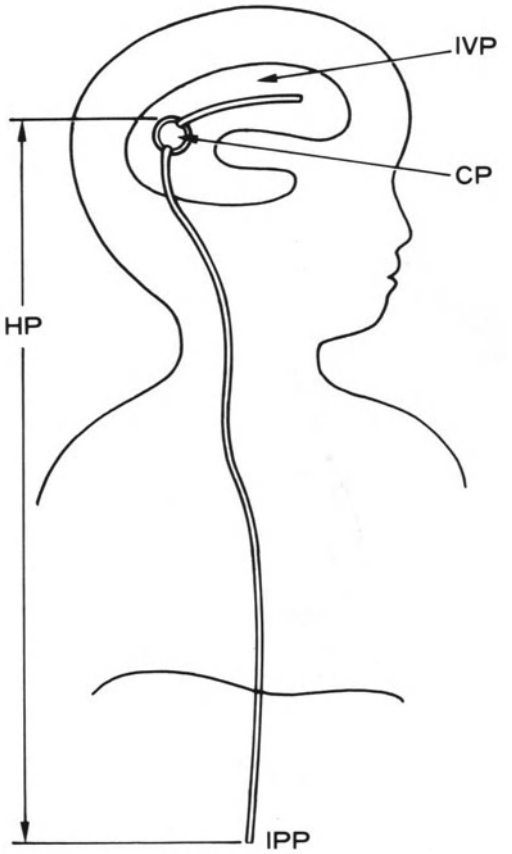


Figure 15.40. Types of valves and pressure–flow curves.



P = IVP + HP − (IPP + CP)

P = perfusion pressure
IVP = intraventricular pressure
CP = valve closing pressure
HP = hydrostatic pressure
IPP = intraperitoneal pressure

Figure 15.41. Factors determining perfusion pressure of CSF shunt system.

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. 15.41).

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.

References

1. Morgagni: Cited by Whytt, reference # 3.
2. Vesalius: De humani corporis labrica librorum epitome. Joannis Opporini, Baselieae, 1543.
2. Whytt R: Observations on the dropsy in the brain. Edinburgh J Balfour, 1768.
4. Berengarius: cited by Baker, reference # 7.
5. Sylvius: cited by Baker, reference # 7.
6. Arantius: cited by Baker, reference # 7.
7. Baker F: The two Sylviuses. An Historical Study. Bul John Hopkins Hosp 20:329–339, 1909.
8. Bonnevie K: Hereditary hydrocephalus in the house mouse. K. Maniefestation of the hy-mutation after birth and embryos 12 days old or more. Skr Norske Vidensk Adac Oslo 1 Mat—kat, KI No 4, 1943.
9. Russell DS.: Observations on the pathology of Hydrocephalus. Spec Rep Ser med Res Council No 265, 1949.
10. Dandy WE, Blackfan KD: Internal hydrocephalus. An experimental clinical and pathological study. Am J Dis Child 8:406, 1914.

11. Raimondi AJ, Clark SJ, McLone DD: Pathogenesis of aqueductal occlusion in congenital murine hydrocephalus. J Neurosurg 45:66–77, 1976.
12. Johnson RT, Johnson KP: Hydrocephalus following viral finection. The pathology of aqueductal stenosis developing after experimental mumps virus injection. J Neuropath Exp Neurol 27:591, 1968.
13. Gruneberg H: Two new mutant genes in the house mouse. J Genet 45:22–28, 1943.
14. Millen JW, Woolam DH Lamming GE: Hydrocephalus associated with deficiency of vitamin A. Lancet 2:1234, 1953.
15. Kesterson JW, Carlton WW: Histopathologic and enzyme histochemical observations of cuprizone—induced brain oedema. Experimental and molecular pathology 15:82–96, 1971.
16. Johnson RT, Johnson KP, Edmonds ES: Virus induced hydrocephalus development of aqueductal stenosis after mumps infection. Science 157:1066–7, 1967.
17. Clark DB: Hydrocephalus. In Textbook of Pediatrics, 9th ed, WE Nelson, VC Vaughan and RJ Mckay (eds), Philadelphia, Saunders, 1969.
18. Raimondi AJ, Samuleson GS, Yarzagaray L, Norton T: Atresia of the foramina of Luschka and Magendie. The Dandy-Walker cyst. J Neurosurg 31:202–216, 1969.
19. Jakurbowski J, Jefferson A: Axial enlargement of the third ventricle and displacement of the brain stem in benign aqueduct stenosis. Journal of Neurol Neurosurg and Psychiat 45:114–123, 1972.
20. Taggart JK, Walker AE: Congenital atresia of the foramens of Luschka and Magendie. Archs Neurol Psychiat 48:583–612, 1942.
21. Matson DD: Prenatal obstruction of the fourth ventricle. Am J Roentg 76:499–506, 1956.
22. Brodal A, Hauglie-Hannssen E: Congenital hydrocephalus with defective development of the cerebellar vermis (Dandy-Walker Syndrome). Clinical and anatomical findings in two cases with particular reference to the so called atreasia of the foramen of Luschka. J Neurol Neurosurg Psychiat 22:99–108, 1959.
23. Benda CE: The Dandy-Walker syndrome or the so called atresia of the foramen Magendie. J Neuropath exp Neurol 13:14–29, 1954.
24. Gibson JB: Congenital hydrocephalus due to atresia of the foramen of Magendie. J Neuropath exp Neurol 14:244–262, 1954.
25. D'Agostino AN, Kernohan JW, Brown JR: The Dandy-Walker syndrome. J Neuropath exp Neurol 22:450–470, 1963.
26. Gardner WJ, Abdullah AF, McCormack LJ: The varying expressions of embryonal atresia of the fourth ventricle in adults. J Neurosurg 11:591–607, 1957.
27. Milhorat TH: Failure of choroid plexectomy as a treatment for hydrocephalus. Surg Gynecol Obstet 193:505, 1974.
28. Penfield W, Elvidge AR: Hydrocephalus and the atrophy of cerebral compression. In Penfield W (ed) Cytology and Cellular Pathology of the Nervous System, New York, Hocher, 1932. Vol. 3, pp 1203–1217.
29. Hassler O: Angioarchetecture in hydrocephalus. An autopsy and experimental study with the aid of microangiography. Acta Neuropathol (Berl) 4:65–74, 1964.
30. De SN: A study of the changes in the brain in experimental internal hydrocephalus. J Path Bact 62:197, 1950.
31. Dandy WE: Extirpation of the choroid plexys of the lateral ventricles in communicating hydrocephalus. Ann Surg 68:569–579, 1918.
32. Heimberger RF, Kalsheck JE, Campbel RL: Positive contrast cerebral ventriculography using water—soluble media. Clinical evaluation of 102 procedures using methyl glucamine iothalamate 60 per cent. J Neurol Neurosurg Psychiat 29:281–290, 1966.
33. Tolosa E: L'exploration arteriographique dans l'hydrocéphalie infantile. Sem Hôp Paris, 27:2401–2403, 1951.
34. Fauré C, Burson B: L'exploration radiologique des craniopharyngiomes de l'enfant (A propos de 170 observations). Ann Radiol 2:197–228, 1959.
35. Paillas JE, Bonnal J, Berard-Bachic H: Les angiomes A-V du cerveau chez l'enfant. Presse med 66:525, 1958.
36. Raimondi AJ: Pediatric Neuroradiology, Saunders, Philadelphia, 1972.
37. Davidoff LM: Hydrocephalus and hydrocephalus with meningocele. Their treatment by chorid plexectomy. Surg Clin N Amer 28:416, 1948.
38. Sutherland GA, Cheyne WW: The treatment of hydrocephalus by intracranial drainage. Brit MJ 2:1155, 1898.
39. Ferguson AH: In Davidoff, LJ Treatment of Hydrocephalus, Arch Surg 18:1737, 1929.
40. Kausch W: Die Behandlung des Hydrocephalus der kleinen Kinder. Arch f Chir 87:709, 1908.
41. Payr E: Drainage der Hirnvertrikel mittlest frei transplantirtor Blutse: Bemerkungen über Hydrocephalus. Arch f klin chir 87:801, 1908.
42. Haynes IS: Congenital internal hydrocephalies: its treatment. Drainage of the cisterna magna into the cranial sinuses. Ann Surg 57:449, 1913.
43. Ingraham FD, Sears, RA, Woods, RP, Bailey, OT: Further studies on the treatment of experimental hydrocephalus. Attempts to drain the cerebrospinal fluid into the pleural cavity and the thoracic duct. J Neurosurg 6:207, 1949.
44. Nulsen FE, Spitz, E.G.: Treatment of hydrocephalus by direct shunt from ventricle to jugular vein. Surg Forum 2:399, 1952.
45. Matson DD: Neurosurgery of Infancy and Childhood, 2nd ed. Charles C. Thomas, Springfield, 1969.
46. Ransohoff J, Shulman K, Fishman RA: Hydrocephalus. A review of etiology and treatment. J Pediat 56:399, 1960.
47. Harsh GR III: Peritoneal shunt for hydrocephalus utilizing the fimbria of the fallopian tube for entrance into the peritoneal cavity. J Neurosurg 11:284, 1954.
48. Sakoda TH, Maxwell JA, Brackett, C.E.Jr.: Intestinal volvulus secondary to a ventriculoperitoneal shunt. Case Report. J Neurosurg 35:95–96, 1971.
49. Keen Weitzner WW: Surgery of the lateral ventricles. Verhandl d x internat med Kongr III Chirurgie, Berlin, p. 108, 1891.
50. Ransohoff J: Ventriculo-pleural anastomosis and treatment of midline obstructed masses. J Neurosurg 11:295–301, 1954
51. Hoffman HJ, Hendrick EB, Humphreys RP: Experience with ventriculo-pleural shunts. Child's Brain 10:404–13, 1983.

52. Neumann CG, Hoen TI, Davis DA: The treatment of communicating hydrocephalus by the absorption of cerebral spinal fluid by the mucosa of an isolated segment of ileum (modified ileo-entectropy). Surg Forum 705, 1958.
53. Smith GW, Moretz WH, Pritchard WL: Ventriculo-biliary shunt. A new treatment for hydrocephalus. Surg Forum 701, 1958.
54. Brown JT: Intrauterine shunting for hydrocephalus. In Pachi A, Calisti A, Astrei G (eds): Prenatal Diagnosis and Surgical Treatment of Congenital Malformations. Padova, Italy, Piccin/Butterworths, 1983 p 231.
55. Cambria S, Cambria M, Gambardella G, Cardia E, Labianca M: Idrocefalia sperimentale su feto in utero. Parte IV. Mini-uni-shunt ventricolo amniotico: Note technich Chir Patol Sper 27:369–71, 1979.
56. Michejda M, Hodgen GD: In utero diagnosis and treatment of non-human primate fetal skeletal anomalies. I Hydrocephalus. JAMA 246:1093, 1981.
57. Clewell WH, et al.: A surgical approach to the treatment of fetal hydrocephalus. N Engl J Med 306:1320, 1982.
58. Frigoletto FD, Birnholz JC, Greene MF: Antenatal treatment of hydrocephalus by ventriculoamniotic shunting. JAMA 248:2496, 1982.
59. Raimondi AJ: Pediatric Neuroradiology p 261. Philadelphia, Saunders, 1972.

Uncited References

DiRocco D, McLone DG, Shimoji T, Raimondi AJ: Continuous intraventricular CSF pressure recording in hydrocephalic children during wakefulness and sleep. J Neurosurg 42:683, 1975.
Gutierrez FA, Raimondi AJ: Peritoneal cysts: A complication of ventriculoperitoneal shunts. Surgery 79:188–92, 1975.
Hahn Y, Raimondi AJ: Ventriculoperitoneal shunting with one piece spring catheter: Technical note. Yonsei Med J 17:157–62, 1976.
Mori K, Raimondi AJ: An analysis of external ventricular drainage as a treatment for infected shunts. Child's Brain 1:243–50, 1975.
Raimondi AJ: A critical analysis of the clinical diagnosis management and prognosis of the hydrocephalic child. In Schulman I (ed): Chicago, Yearbook Medical Publishers, 1971, vol 18, p 265.
Raimondi AJ: Hydrocephalus and the congenital anomalies associated with it: Angiographic diagnosis. Semin Roentgenol 6:111–25, 1971.
Raimondi AJ, Guiterrez FA, Jones RR, Winston SR: Cystic cavum veli interpositi associated with normal or low pressure hydrocephalus. Child's Brain 1:291–305, 1975.
Raimondi AJ, Matsumoto S: A simplified technique for performing the ventriculoperitoneal shunt. J Neurosurg 26:357–60, 1967.
Raimondi AJ, Robinson JS, Kuwamurak: Complications of ventriculoperitoneal shunting and a critical comparison of the three-piece and one-piece systems. Child's Brain 3:321–42, 1977.
Raimondi AJ, Soare PL: Intellectual development in shunted hydrocephalic children. Am J Dis Child 127:664–71, 1974.
Robinson JS, Kuwamura K, Raimondi AJ: Complications of ventriculoperitoneal shunting procedures. In Proceedings of the Symposium on Myelomeningocele, Cincinnati, Ohio. New York, Grune, Stratton, 1977.

"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

Chapter 16

Pediatric Neuroanesthesia

Badr A. Ishak

Several factors are responsible for the advances we witnessed over the last decade in the field of pediatric neurosurgery. Better understanding of the pathophysiology of increased intracranial pressure and its control, the pharmacological effects of anesthetic agents on the cerebral blood flow and metabolism, preoperative control of metabolic disturbances, specialized postoperative intensive care units, aggressive invasive monitoring, and newer diagnostic tools, such as computed transmission tomography and somatosensory evoked potentials, enabled the pediatric neurosurgeon to deal successfully with lesions considered inoperable a few years ago.

Physiologic Aspects

Cerebral Blood Flow

Measurement of Cerebral Blood Flow (CBF)

Kety and Schmidt[1] pioneered the quantitative measurement of CBF, using nitrous oxide as an inert tracer. The lower limit is 44 ml/100 g/min, which represents the global blood flow. With the introduction of radioisotopes as tracers (^{85}Kr, ^{133}Xe) and the use of multiple external scintillation counters, regional or focal blood flow was found to range between 20 ml/100 g/min in the white matter to 80 ml/100 g/min in the gray matter.[2]

Other methods for determination of CBF are the hydrogen clearance technique, where injected or inhaled hydrogen is measured by a small platinum electrode inserted in different areas of the brain,[3] and the injection of microspheres[4] and autoradiographic studies of diffusible tracers.[5] Because of their invasiveness, the use of these techniques is limited to animal studies.

Control of Cerebral Blood Flow (CBF)

Arterial Carbon Dioxide Tension ($PaCO_2$)

Respiratory acidosis due to increased $PaCO_2$ causes increased CBF. On the other hand, respiratory alkalosis due to passive or active hyperventilation causes arteriolar constriction and decreased CBF (Fig. 16.1). Carbon dioxide, being highly soluble, crosses the blood-brain barrier easily and increases the H^+ concentration in the perivascular and extracellular fluid, causing dilatation of smooth muscle in the arteriolar wall, thereby reducing vascular resistance and increasing CBF.[6] The clinical application of hyperventilation in neurosurgical patients is well recognized, although it was found that the cerebral vessels are maximally constricted at a $PaCO_2$ of 20 torr and that the brain may show signs of hypoxemia at extreme degrees of hyperventilation.[7] Readjustment of the cerebrospinal fluid bicarbonate during chronic hyperventilation limits the usefulness of long-term control of CBF by mechanical ventilation.[8]

There is also some evidence that this relationship between $PaCO_2$ and CBF may be different in premature and newborn infants, as it was shown that, in the sheep fetus and newborn lamb, an increase in $PaCO_2$ causes an increase in CBF, while a decrease in $PaCO_2$ is not associated with a drop in CBF.[9] Other investigators found a lack of reactivity of CBF to changes in $PaCO_2$ in the newborn monkey.[10]

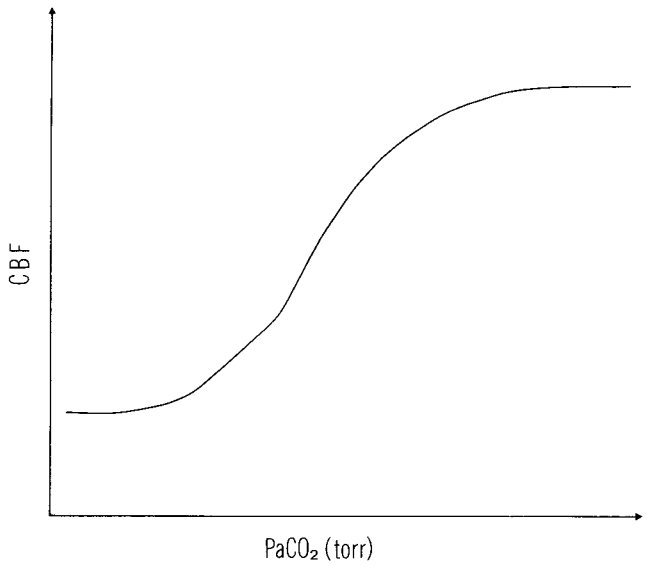


Figure 16.1. The effect of $PaCO_2$ on cerebral blood flow.

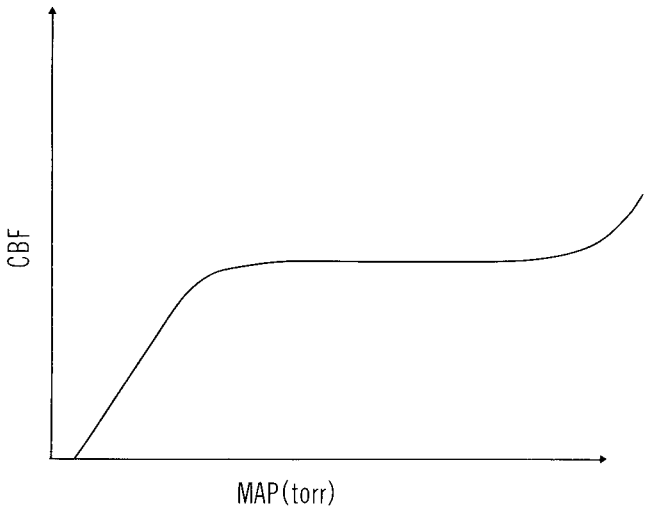


Figure 16.2. The effect of mean arterial pressure (MAP) on cerebral blood flow.

Cerebral Perfusion Pressure (CPP)

The CPP is defined as the difference between the mean arterial pressure (MAP) and the intracranial pressure (ICP). The ability of the cerebral blood vessels to maintain constant blood flow over a wide range of perfusion pressure exists also in other vital organs, such as the heart and kidney, and is known as autoregulation (Fig. 16.2).

The normal range of autoregulation in normotensive adults with a mean arterial pressure of 90 torr is between 50 and 150 torr.[11] In hypertensive patients with a mean arterial pressure of 125–180 torr, it was found that the lower limit of autoregulation is 90–125.[12] It is of interest to note that the breaking point of the lower limit of the autoregulatory mechanism in both normotensive and hypertensive adults is approximately two thirds the mean arterial pressure. Furthermore, there is evidence that the same relationship may exist in the healthy newborn infant whose normal blood pressure is reported to be 72/47,[13] with a mean arterial pressure less than that required for adequate cerebral perfusion in the adult, indicating that the breaking point is set at a lower level than that in adults. This was shown experimentally in newborn lambs with a mean arterial pressure of 60 that evidence of inadequate cerebral perfusion was apparent at a CPP of 40 torr.[9]

Other factors that affect the lower limit of autoregulation are shown in Figure 16.3. The autoregulatory mechanism is known to be lost in certain situations such as head trauma, hypoxia, tumors, exposure to inhalation anesthetics, and after the use of hypotensive durgs.[14–16] Thus it is important to control blood pressure in this group of patients, because any increase in blood pressure is associated with a passive increase in cerebral blood flow, resulting in an increase in intracranial pressure, which in turn lowers CPP.

Arterial Oxygen Tension (PaO_2)

The sensitivity of the brain to hypoxemia is explained by several factors: its high resting oxygen consumption, its dependence on oxidative metabolic pathways, and the lack of any appreciable oxygen stores. There is little change in cerebral blood flow during normoxia and hyperoxia, while there is marked increase of cerebral bloodflow in the presence of hypoxemia, at a PaO_2 of 50 torr or less[17] (Fig. 16.4); these data also correlate with the finding of tissue acidosis at this level,[18] thus suggesting the same mechanism of change in the prearteriolar $[H]^+$, as in the case of hypercapnia. The premature and newborn infants have a normal PaO_2 of 65–70 torr,[19] indicating their higher tolerance for hypoxemia than the adult, although it was found in animal studies[9] that a drop in PaO_2 of 7–11 torr caused an increase in cerebral blood flow in the lamb fetus, indicating a much narrower range of PaO_2 at which a normal cerebral perfusion pressure is maintained.

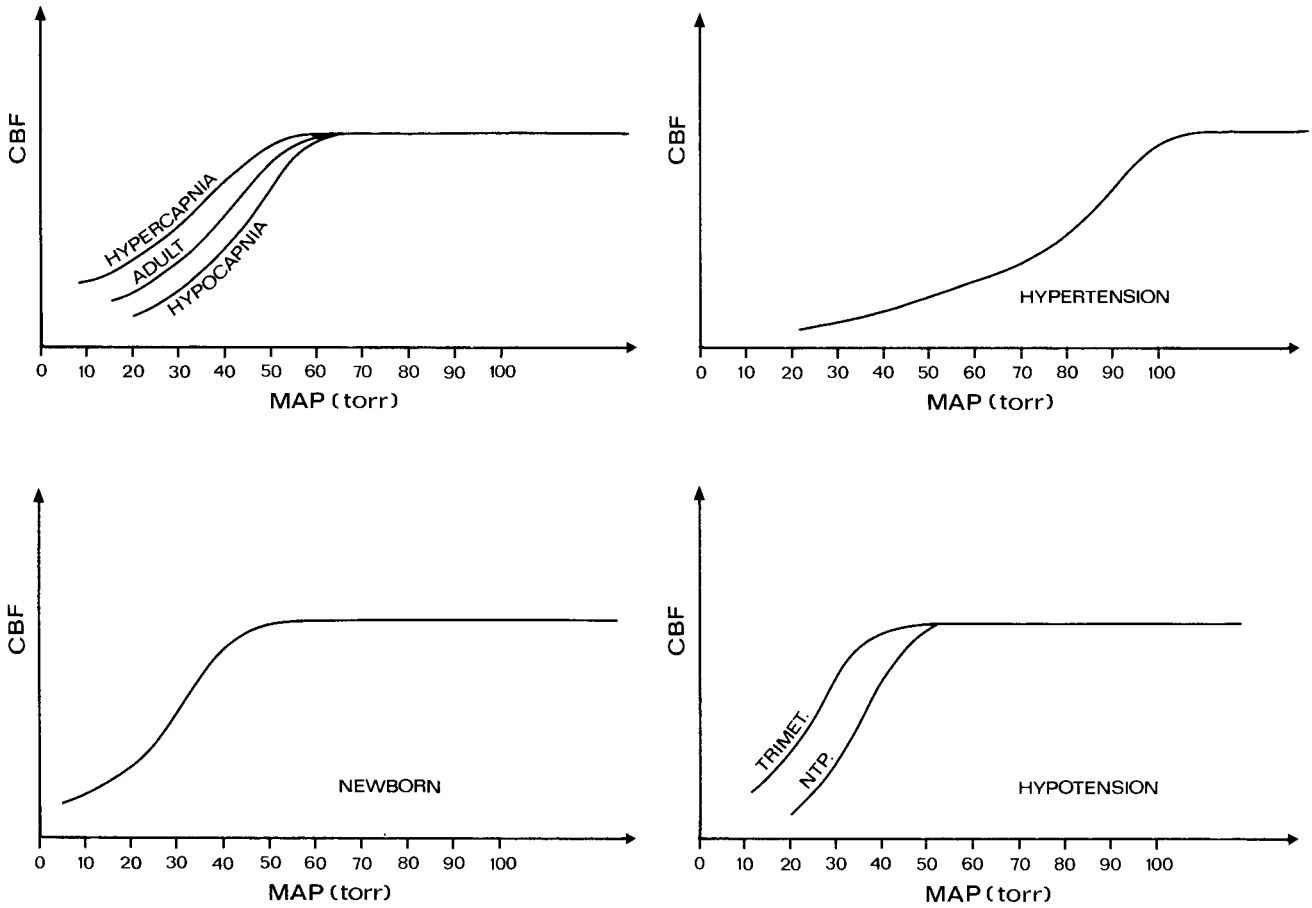


Figure 16.3. Factors which influence the lower limit of autoregulation (Ref. 9, 24, 12, 25).

Metabolic and Neurogenic Control

Marked changes in regional cerebral blood flow can occur with local activity of brain tissue,[20] which is coupled with an increase in cerebral oxygen consumption (CMRO2).[21] On the other hand, an increase in global blood flow is known to occur during grand mal seizures, and also secondary to pain or anxiety,[22,23] hence the importance of avoiding painful stimulation during neurological examination, even in comatose patients.

The autonomic nervous system does not contribute significantly to the regulation of cerebral blood flow, as stimulation of the cervical sympathetic chain causes a 15% decrease in cerebral blood flow[24-26]

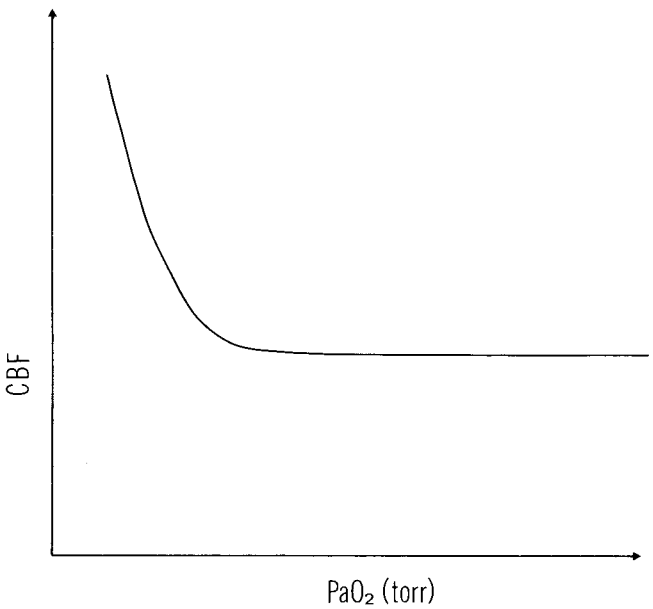


Figure 16.4. The effect of PaO_2 on cerebral blood flow.

Intracranial Pressure (ICP)

The importance of understanding the pathophysiology of increased ICP cannot be overemphasized for the practicing anesthesiologist, as many of the techniques and drugs routinely used have a profound effect on ICP. Monro[27] and Kellie[28] have postulated that, because the skull is indistensible, in the presence of constant craniospinal volume an increase in volume of any of its constituents will cause an increase in pressure. Although their hypothesis is not literally observed in

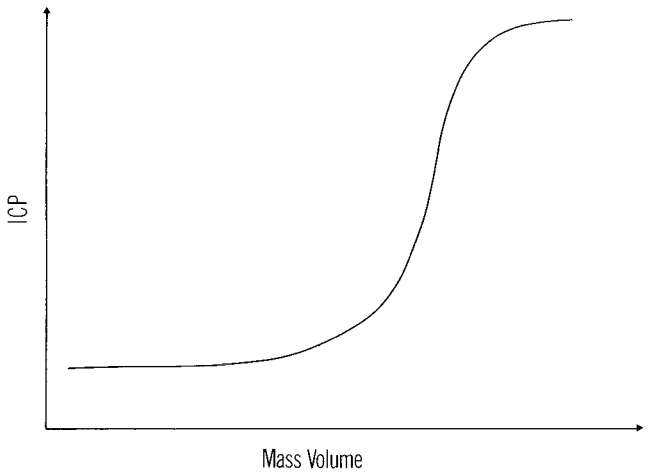


Figure 16.5. Intracranial volume-pressure relationship (Compliance curve).

patients, it forms the basis of the intracranial volume-pressure relationship (Fig. 16.5) in which a gradual increase in intracranial volume, such as in a growing mass lesion, is associated with an initial small rise in pressure, which is due to increased absorption of cerebrospinal fluid. This process is pressure-dependent.[29] Also, compression of the thin-walled cerebral vessels cause a reduction in cerebral blood volume, thus allowing the mass lesion to expand temporarily with minimum change in ICP. Exhaustion of these compensatory mechanisms results in a marked rise in ICP with any further, even slight, increase in volume to a plateau where ICP equals the mean arterial pressure, a level at which cerebral blood flow ceases. The progressive rise in ICP causes a reduction in cerebral perfusion pressure, which results eventually in lowered tissue perfusion and metabolic acidosis due to the accumulation of lactic acid, which in turn produces cerebrovascular paralysis.[30] Furthermore, the absence of vascular reactivity increases the cerebral blood volume in the presence of arterial hypertension, thus accentuating intracranial hypertension and also enhancing the formation of cerebral edema.[31]

Role of Blood Pressure in Intracranial Pressure (ICP)

In the presence of intact autoregulation, changes in arterial blood pressure have little effect on ICP except in extremes of blood pressure, that is, a mean arterial pressure of more than 150 torr or less than 50 torr. Loss of autoregulation due to intracranial pathology[15] or exposure to inhalation anesthetics[32] during and following deliberate hypotension[16] or hypovolemic hypotension,[33] can cause a passive change in ICP with changes in mean arterial pressure. Control of arterial pressure in neurosurgical patients during the recovery period, and also when moderate hypotension is present, may improve surgical conditions by reducing the brain bulk due to decreased flow to those areas of the brain with lost autoregulation.

Role of Arterial Blood Gases in Intracranial Pressure (ICP)

The influence of changes in arterial oxygen and carbon dioxide tension on cerebral brain flow has been discussed. Hypocapnia, by reducing cerebral brain flow, thereby causing a reduction in ICP, is an established technique in neuroanesthesia and in the management of nonsurgical patients with increased ICP. Extreme degrees of hypocapnia should be avoided because the cerebral blood vessels are maximally constricted at a PCO_2 of 20 torr, and further hyperventilation does not produce added reduction in cerebral blood flow. Also, signs of brain ischemia, as evidenced by increased tissue lactate levels, are present with excessive hyperventilation.[7] Finally, the large tidal volumes required cause a reduction in cerebral perfusion due to the accompanying increase in the mean intrathoracic pressure.[34] It should be noted that the effect of hyperventilation on reducing ICP is a temporary one because of decreased absorption of cerebrospinal fluid during the period of hyperventilation, and the readjustment of cerebrospinal fluid pH regains the original vascular tone.[8]

Hypercapnia, on the other hand, causes an increase in cerebral blood volume and a corresponding increase in ICP, which returns rapidly to normal with correction of the respiratory acidosis, unlike the increase in ICP that follows hypoxemia, persisting even after correcting the hypoxemia. Arterial hypertension in the presence of hypercapnia enhances cerebral edema[35] and increased ICP, which may persist after correcting the respiratory acidosis. A similar situation also exists in the presence of combined hypoxemia and hypercarbia (asphyxia).

Role of Anesthetic Agents in Intracranial Pressure (ICP)

All inhalation anesthetics used clinically were shown to increase ICP,[36-39] which is secondary to an increase in cerebral blood volume due to the vasodilatory effect of inhalation anesthetics on the cerebral arterioles, leading to distension of the cerebral veins due to the fixed drainage capacity of the venous sinuses.[40] In contrast, intravenous anesthetic drugs such as thiopental and althesin,[41,42] sedative drugs such as droperidol,[43] and potent analgesic drugs such as fentanyl, in the absence of respiratory acidosis,[42] will cause a decrease in cerebral blood flow and, therefore, intracranial pressure. Ketamine, on the other hand, is the only intravenous anesthetic agent that causes marked increase in cerebral flow and ICP,[44] thus limiting its use in neuroanesthesia.

The undesirable effect of inhalation anesthetics on ICP is well documented in animal models, and in human beings. Furthermore, this effect seems to be accen-

tuated in the presence of intracranial mass lesions,[45] although it was found that the increase in ICP due to halothane could be markedly obtunded by prior hyperventilation to an arterial carbon dioxide tension of less than 30 torr[46] before the introduction of halothane. This finding, by no means as universal as Gordon[47] has suggested, shows a marked rise in ICP when halothane was introduced after a period of hyperventilation in some patients with space-occupying lesions. Enflurane, apart from the fact that it increases ICP, which may not be abolished by hyperventilation,[48] also causes seizurelike EEG activity, which may be enhanced by the presence of hypocapnia, auditory stimulation, or a high inspired concentration of the anesthetic,[49] thus limiting its use in neuroanesthesia.

Role of Hypothermia in Intracranial Pressure (ICP)

Hypothermia causes a predictable and consistent fall in the cerebral metabolic rate and oxygen consumption, an effect which is traditionally expressed as Q10 which represents the factor by which the cerebral metabolic rate changes with a 10-degree I alteration in body temperature and is considered to be in the range of 2–3 in the human brain.[50] Hypothermia was also found to reduce cerebral blood flow[51] and ICP.[52] But the morbidity associated with the use of hypothermia and because it is a time-consuming procedure, especially in adult patients, the use of this technique was curtailed in clinical anesthesia. In our institution, we reserve the use of moderate hypothermia—30–32° C—to patients with arteriovenous malformations and aneurysms, where temporary clamping of vital cerebral vessels may be considered.[53]

The clinical implications of the above discussion may be summarized as follows:

1. Anesthetic agents that tend to increase intracranial pressure, such as inhalation anesthetics and ketamine, should be avoided in patients with signs of increased intracranial pressure.
2. Hyperventilation followed by the introduction of halothane is warranted in this group of patients.
3. The ideal anesthetics for neurosurgery include barbiturates as an induction agent and neuroleptanesthetic drugs for maintenance.
4. Because the effects of changes in the arterial blood gases, blood pressure, and the mean intrathoracic pressure are cumulative on intracranial pressure, hypoxemia, hypocapnia, and bucking should certainly be avoided.

Measurement and Interpretation of Intracranial Pressure (ICP)

The value of monitoring ICP in neurosurgical patients is well established,[54] since it provides an excellent diagnostic tool, particularly in those patients where clinical signs of increased intracranial pressure are lacking. Also, it may have a place in predicting the outcome in patients with severe head injuries.[55] Several methods have been used to monitor ICP insertion of an intraventricular catheter. This technique was described and used by early workers[56] and is still utilized in our institution, especially in hydrocephalic children on extraventricular drainage. This method has several advantages, as the fluid coupling allows repeated calibrations of the externally located transducer. Also, cerebrospinal fluid could be easily aspirated to control an acute rise in ICP. Ventriculitis is the main hazard associated with this monitoring technique,[57] while obstruction with debris may interfere with the accuracy of recording. Extradural bolts were used for ICP measurement to avoid infection, and this method was shown to compare favorably with intraventricular pressure,[58] although misplacement of the device could again result in faulty pressure recording. Recently, a subarachnoid bolt was described[59] in which the tip of the bolt is inserted through an opening in the dura.

However, cerebrospinal fluid drainage is not possible while the incidence of infections is reduced by inserting a Micropore filter on the distal end of the tubing (which does not dampen the pressure tracing[11]) and by the use of prophylactic antibiotics.[60]

Lundberg[61] considered ICP to be normal if it is between 5 and 10 torr, slightly increased between 11 and 20 torr, moderately increased between 21 and 40 torr, and severely increased if above 40 torr. The intracranial pressure waveform shows normal fluctuations with cardiac systole, and also slower fluctuations during respiration, whether spontaneous or controlled. This dynamic state of intracranial pressure waveform enhanced further studies to determine the usefulness of its analysis. Janny and coworkers[62] observed the increase in intracranial pulse pressure with increased ICP. Furthermore, Schettini and coworkers[63] calculated brain-relative stiffness from the slope of the dynamic pressure waveform and demonstrated the effect of hyposmotic edema, induced hypotension, hypovolemic hypotension, induced hypertension, hypercapnia and cerebral venous hypertension (Fig. 16.6). These and other studies[64,65] seem to indicate that computer analysis of ICP pressure waveform is a promising technique.

Preoperative Evaluation and Management of the Pediatric Neurosurgical Patient

The incidence of congenital anomalies of the central nervous system, such as meningomyelocele, may require surgical interference during the first few hours following birth, when the presence of other factors, such as prematurity or respiratory distress syndrome, may compli-

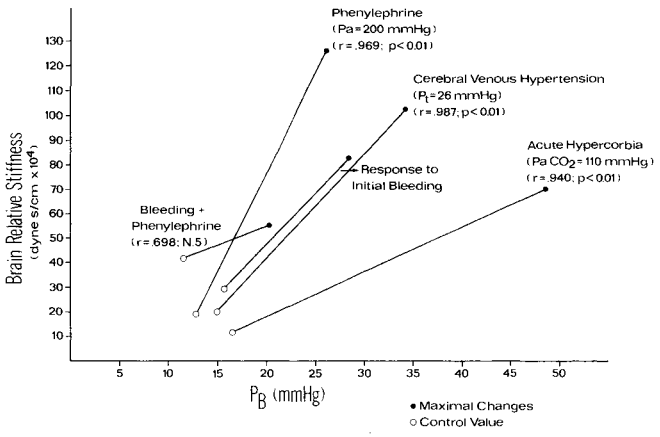


Figure 16.6. The influence of cerebral vascular factors on the brain relative stiffness (From Schettini, Malig, Moreshead. In Intracranial Pressure, Springer-Verlag, Berlin, 1972).

cate the anesthetic management. On the other hand, an adolescent with craniopharyngioma will require adequate preoperative control of intracranial pressure and any endocrine disturbances to minimize intra- and postoperative complications.

Preoperative Evaluation of the Newborn Infant. The indications of surgery in the newborn are few: repair of meningomyelocele (usually attempted during the first 24 hours), shunting procedures in congenital, postinfectious, or posthemorrhagic hydrocephalus, and, rarely, for elevation of depressed skull fractures, which are almost invariably due to birth trauma. The majority of newborns tolerate anesthesia well, with the exception of those recovering from meningitis or intraventricular hemorrhage, who may show other systemic and central nervous system dysfunctions or prematurity.

Respiratory Distress and Failure. The common causes of respiratory distress in the neonate are pulmonary atelectasis, especially in low birth weight infants, idiopathic respiratory distress syndrome, muconium aspiration, and spontaneous pneumothorax. These disorders are manifested clinically by intercostal and subcostal retractions, tachypnea, expiratory wheeze or grunt, cyanosis while breathing 40% oxygen, and deterioration in the level of consciousness. Physiological signs include arterial carbon dioxide tension greater than 75 torr and oxygen tension less than 100 torr while breathing 100% oxygen. The presence of 3 clinical and one physiological signs indicates acute respiratory failure.[66]

Preoperative management includes arterial blood gases and chest x-ray, and, because of the danger of brain damage from hypoxia or retrolental fibroplasia from hyperoxia, controlled oxygen therapy is indicated. In severe respiratory distress, an endotracheal tube is inserted and respiration is either assisted by continuous positive airway pressure (CPAP) or controlled by mechanical ventilator (Table 16.1).

Fluid and Electrolytes. Induction of anesthesia will invariably cause hypotension in dehydrated infants. Also, the presence of electrolyte disturbances could interfere with the action and duration of muscle relaxants. Clinically, dehydration may be classified as mild if the water deficit is 5% of the infant's body weight and manifested by dry skin, depressed fontanel in supine position; in moderate dehydration, the water loss is 5–10% of the body weight, and the infant shows mottled skin, tachycardia, oliguria, sunken eyeballs, and flaccidity; in severe dehydration of 15% or more of body weight, the infant is moribund and shows the classic signs of shock.[67] Hypovolemia is associated with arterial hypotension and low cardiac output, which predisposes to metabolic acidosis. This, in turn, leads to further depression of the myocardium, hence the importance of correcting any degree of metabolic acidosis (Table 16.2).

Table 16.1. Respiratory Distress Syndrome: Signs and Management

Respiratory Distress in Neonates
Pulmonary atalectasis
Idiopathic respiratory distress syndrome
Muconium aspiration
Spontaneous pneumothorax
Signs (Clinical)
Intercostal, subcostal retractions
Tachypnea
Expiratory wheeze or grunt
Cyanosis on FiO_2 0.4
Deterioration in level of consciousness
Signs (Physiological)
$PaCO_2 > 75$ torr
$PaO_2 < 100$ torr on FiO_2 1.0
Management
Arterial blood gases
Chest x-ray
Controlled O_2 therapy
Continuous positive airway pressure, controlled ventilation

Table 16.2. Common Metabolic and Fluid Disturbances in Neonates

Disorder	Management
Dehydration	
5%: Dry skin, slight depression of fontanel	Replace $^1/_2$ the estimated deficit with lactated Ringer's during the first hour and remainder over the next 4 hours
10%: Mottled, tachycardia, oliguria, sunken eyes, flaccid	
15%: Moribund, signs of shock	
Acidosis Arterial or capillary blood gases	Sodium bicarbonate 1 meq/ml required = base deficit × body weight (kg) × 0.3
Hypoglycemia Glucose level <30 mg% in premature <40 mg% in term infant	10% or 20% dextrose in water
Hypocalcemia Serum calcium < 5 mg%	Calcium gluconate 100 mg/kg

Table 16.3. Common Neurosurgical Procedures in Infants and Children

1. Craniosynostosis
2. Craniofacial synostosis (Crouzon's and Apert's syndromes)
3. Aneurysms and arteriovenous malformations
4. Hydrocephalus
5. Tumors
6. Intracranial hematomas
7. Skull fractures
8. Spinal cord injuries and tumors

The older child may require surgery for a variety of conditions (Table 16.3). It must be stressed that the presence of intracranial pathology is often the cause of other systemic disturbances that should be evaluated and adequately controlled before surgery.

Respiratory System

Disturbances of respiration often accompany intracranial pathology, and may present as periods of apnea in infants with intraventricular hemorrhage. Upper airway obstruction is also known to occur in the Arnold-Chiari malformation and hydrocephalus,[68] which may be due to impairment of blood flow to the brainstem as it herniates caudally, and the resultant ischemia causes bilateral abductor vocal cord paralysis evidenced clinically by stridor.[69] Increased intracranial pressure leading to mechanical stretching of the vagus nerve or its cervical rootlets may also be a factor.[70] Temporary relief of the laryngeal obstruction by the insertion of endotracheal tube may be required until surgical treatment as insertion of a shunt or decompression of the posterior fossa is undertaken, after which the respiratory obstruction generally improves. It should also be noted that some of these infants still develop periods of apnea even after securing an airway; thus the use of an apnea monitor is recommended.[68]

Aspiration pneumonitis is a well-documented complication in patients with head injuries, obtunded level of consciousness, also those with Chiari II malformation due to cricopharyngeal achalasia.[71] Preoperative management of these patients consists of protecting the airway from further aspiration by an endotracheal tube, the administration of prophylactic antibiotics and chest physiotherapy to minimize the high mortality associated with untreated aspiration.[72] Respiratory pattern disturbances occur secondary to head trauma, severe infections, and intracranial tumors,[73] while pulmonary edema may complicate the course of neurosurgical patients with severe head injuries, intracranial hemorrhage, tumors, and seizures.[74] Table 16.4 summarizes the indications of preoperative mechanical ventilation in neurosurgical patients.

Cardiovascular System

The occurrence of arterial hypertension in the presence of elevated ICP is part of the well-recognized Cushing triad. Electrocardiographic changes such as T-wave inversion, S-T segment depression, and even the appearance of Q waves were all described in patients with subarachnoid hemorrhage and space-occupying lesions.[76] Atrial fibrillation[77] and cardiac arrest were reported to follow head injuries,[78] although the exact mechanism of these dysrhythmias remains unclear. Congestive cardiac failure may be the only presenting sign in infants with large arteriovenous malformations of the brain,[79] and is evidenced by cardiac enlargement on x-ray film and signs of ventricular strain on the electrocardiogram. Adequate preoperative control with digitalis and diuretics is required, although rarely effective in the presence of a large fistula.

Table 16.4. Indications for Preoperative Intermittent Positive Pressure Breathing

1. Respiratory rate < 10/min in adult or > 70/min in neonate
2. Markedly irregular respiratory patterns
3. Combined chest and head injury with pulmonary involvement
4. Central hyperventilation with $PaCO_2 < 20$ torr
5. Head injury in coma with impending respiratory failure
6. Elevated ICP > 25 torr
7. Severe intrapulmonary shunts VD/VT > 50%
8. Upper airway obstruction

Modified from Miller JD, Sullivan HG: Management of acute intracranial disasters. IAC 17:59, 1979, with permission.

Table 16.5. Shapiro's Classification

Condition	Stimulus	Response
Alert	Normal visual, auditory Somatosensory, visceral	Immediate, complete Proper verbalization
Lethargic	Augmented, external	Delayed, incomplete Proper verbalization
Obtunded	Augmented, external	Difficult to arouse Obeys, no verbalization
Stuporous	Intense, noxious	Brisk defensive movements
Comatosed	Intense, noxious	Inappropriate or absent

Table 16.6. Preoperative Hormonal Abnormalities

Diabetes insipidus
- Polyuria
- Polydysia
- Inability to concentrate urine
- Urine/serum osmolality < 1.0
- Increased serum sodium and osmolality

Replacement:
- Vasopressin tannate 1–5 U q 24–48 hr
- DDAVP nasal spray 5–15 μg q 12–24 hr

Adenocorticotropic hormone (ACTH) deficiency
- Low 17-OH corticosteroids in 24-hr urine
- Low serum cortisol

Replacement:
- Cortisone acetate 20 mg/m^2/day × 6 day of surgery

Thyroid-stimulating hormone (TSH) deficiency
- Serum level of TSH < 10 mg/ml
- Serum level of T4 < 4 mg/ml
- Decreased radioactive iodine uptake

Growth hormone deficiency
- Insulin-induced hypoglycemia test

Gonadotropins deficiency
- Insignificant in children under 12 years

Central Nervous System

Preoperative evaluation of the central nervous system is prerequisite for safe and successful intraoperative management of neurosurgical patients. The primary interest of the anesthesiologist lies in evaluating the level of consciousness, as well as the presence of elevated intracranial pressure and the site of the lesion, as the latter will determine the position of the patient during surgery. Several scoring systems are used for evaluating the level of consciousness. Of these the Glasgow Coma Scale[80] is widely used in neurosurgical units, while, for rapid evaluation, Shapiro's classification[11] is both adequate and useful, as shown in Table 16.5. However, the CCS, as described in this text, is preferable.

The presence of elevated intracranial pressure is detected by the clinical and radiological signs, and it is usually managed preoperatively by one or more of the following regimen:

1. Dexamethasone 0.25–0.5 mg/kg/day
2. Glycerol 1.5–2.0 g/kg/single dose via nasogastric tube or maximum dose of 3–6 g/kg/day
3. Mannitol 1.5–2.0 g/kg/single dose iv or maximum dose of 4–8 g/kg/day
4. Mechanical hyperventilation to control sudden rise in intracranial pressure
5. Insertion of ventriculoperitoneal shunt is used routinely in our institution in the management of patients with intracranial lesions and elevated intracranial pressure a week prior to the planned craniotomy.

Endocrine Disturbances

Patients with parasellar and suprasellar tumors such as craniopharyngioma, optic nerve glioma, hypothalamic glioma, and also pituitary tumors, may present preoperatively with hormonal disturbances due to either direct pressure of the tumor mass on the pituitary gland or interference with the portal system of the pituitary stalk. The following table (Table 16.6) summarizes the different neuroendocrine disturbances that may accompany these tumors preoperatively.

The postoperative period requires careful management of the fluid and electrolyte balance, continued steroid therapy, particularly with altered demand due to fever, and sodium retention or even decreased water intake in the presence of obtunded consciousness.

Anesthetic Management

Preoperative Medication

The essential purpose of preoperative medication is twofold: to alleviate apprehension and fear, and to avoid excessive secretions and unwanted vagal reflexes such as bradycardia, which is common in the pediatric age group.

Preoperative Medication for Children Over Two Years Old

Patients over two years of age and with functioning ventriculoperitoneal shunts or without clinical or radiological signs of increased intracranial pressure may be sedated by pentazocine 1–1.5 mg/kg together with atropine sulfate 0.02 mg/kg intramuscularly 45 minutes before induction of anesthesia.

Preoperative Medication for Children Under Two Years Old

Patients less than two years old, or those with malfunctioning shunts or with clinical or radiological signs of increased intracranial pressure are premedicated with

atropine only because excessive sedation may cause an acute rise in intracranial pressure due to respiratory depression and retention of carbon dioxide, although, in agitated children, diazepam 0.1 mg/kg can be given orally the night before surgery and may be repeated two hours before the induction of anesthesia, as, in addition to its sedative effect, it causes reduction in intracranial pressure by reducing the cerebral blood flow.[81]

Induction of Anesthesia

Anesthesia in Infants

Infants with open fontanels or children with functioning ventriculoperitoneal shunts and children without clinical or radiological signs of increased intracranial pressure, inhalation induction with halothane is both rapid and pleasant and respiration is gradually assisted, then controlled as soon as the patient is adequately anesthetized. An intravenous route is then secured by using an indwelling cannula of appropriate size.

Endotracheal intubation is facilitated by the use of the short-acting depolarizing muscle relaxant succinylcholine in brief procedures such as shunt insertion. On the other hand, long-acting nondepolarizing relaxants such as pancuronium are used for longer operations. Intubation of the trachea is not attempted until full muscle paralysis has ensued, to avoid any increase in intracranial pressure that may accompany tracheal intubation.[82]

Anesthesia in Older Children and Adolescents

In older children and adolescents, patients with signs of increased intracranial pressure, patients with malfunctioning ventriculoperitoneal shunts, and those children with full stomach, a rapid sequence induction of anesthesia is preferred in which, after securing an intravenous route, a sleep dose of sodium pentothal (4–5 mg/kg) is injected; following a period of preoxygenation, endotracheal intubation is rapidly accomplished with the use of succinylcholine in a dose of 2 mg/kg. Manual hyperventilation is then started, and a long-acting muscle relaxant is used for the remainder of the procedure.

Maintenance of Anesthesia

Ventilation is routinely controlled in all neurosurgical patients. We utilize for that purpose the Elema servo 900B ventilator coupled with the Elema end tidal carbon dioxide monitor which provides an accurate, yet a noninvasive, method of adjusting ventilation, thus eliminating the need for repeated determination of arterial blood gases. Also, this ventilator proved to be extremely versatile, as it is easy to ventilate premature infants as well as adults without the need to modify its circuit. All inspired gases are humidified, and, in longer procedures, particularly in small children, they are also heated to 38° C. Anesthesia is maintained with a long-acting nondepolarizing muscle relaxant such as pancuronium together with 0.5–0.7 MAC of an inhalation agent as halothane or ethrane. The fluid deficit due to preoperative fasting and also for maintenance is replaced by the infusion of 5% dextrose in lactated Ringer's solution at the rate of 4 ml/kg/hr.

Intraoperative Monitoring

All patients are placed on heating/cooling mattresses thermostatically controlled to the desired temperature. Blood replacement is started if the blood loss exceeds 15% of the estimated blood volume or to maintain a minimum hematocrit value of 30% at all times. Hourly determination of arterial blood gases will check the adequacy of ventilation, and also will detect the onset of metabolic acidosis, which may complicate the use of sodium nitroprusside. Furthermore, serum electrolyte levels may be required to detect any drop that may result from excessive diuresis.

The inhalation agents are discontinued at the end of the procedure, when the muscle relaxant is reversed in the presence of adequate respiration, stable cardiovascular system, and intact protective reflexes, such as coughing and swallowing. The patient is then extubated and transferred to the recovery room where close hemodynamic and neurological monitoring is continued.

Special Considerations

Meningomyelocele Repair and Chiari Malformation

Because it may be difficult to place infants with large myelomeningoceles or encephaloceles supine during induction of anesthesia, we usually induce anesthesia in the lateral position. Also, as these patients are known to become hypothermic due to immaturity of the heat-regulating mechanisms, every effort should be made to keep their temperature within the normal thermal environment. Extreme care should be taken while positioning patients with Arnold-Chiari malformation and both respiratory arrest and cardiovascular instability are reported during manipulation of the cervical spine before and during surgery.[84,85]

Shunting Procedures

The long-term treatment of some types of hydrocephalus, such as those secondary to aqueductal stenosis, of the postinfectious and posthemorrhagic varieties, as well as the Arnold-Chiari malformation, may require shunting of the cerebrospinal fluid around the site of obstruction to different body cavities—commonly, the periotoneal cavity or the right atrium. Unfortunately, this procedure is compounded by a variety of complications, which requires repeated exposure to anesthetics. The common complications the anesthesiologist should be familiar with are infection, obstruction, and discon-

nection. Although the Uni-Shunt[86] used routinely in our institution, is devoid of this complication, shunt tube extrusion, the formation of localized peritoneal cysts and hernias,[87] catheter migration, and bowel perforation,[88] may occur. Ventriculoatrial shunts, on the other hand, are likely to have more serious complications, such as arrhythmias or pulmonary valvular dysfunctions due to migration of the distal end of the catheter, and also embolization of the right atrium and the pulmonary artery.[89] The anesthetic management of these children may cause different respiratory and cardiovascular changes, particularly bradycardia, which responds to small doses of atropine 0.1–0.2 mg. Insertion of the abdominal end of the catheter is usually facilitated by the use of a trocar, which requires the presence of some tone in the abdominal muscles to avoid injury to internal organs or perforation of the bowel, and for this reason we do not use muscle relaxants in those patients in whom the trocar will be used to insert the distal end of the shunt tubing into the peritoneal cavity. Ketamine should be avoided in hydrocephalic children, and also ethrane, particularly in patients with a history of seizures.

Posterior Fossa Craniotomy

Exploration of the posterior fossa in the pediatric patient is indicated for resection of a tumor, evacuation of an abscess or a hematoma, ligation of an aneurysm or arteriovenous malformation, and for implantation of electrodes for chronic cerebellar stimulation to control epilepsy[90] or to alleviate spasticity in cerebral palsy.[91] The presence of space-occupying lesions in the posterior fossa results in some unique problems that may affect the anesthetic management:

1. Increased intracranial pressure is liable to occur early because of rapid exhaustion of the compensatory mechanisms due to the small size of the posterior fossa.
2. Blockade of the narrow aqueduct results in obstructive hydrocephalus and ventricular enlargement.
3. The increased pressure in the posterior fossa eventually causes medullary coning evidenced clinically by stiffness of the neck, bradycardia, and periods of apnea. These patients require careful positioning, and marked flexion of the neck should be avoided.*
5. The rigid tentorium could cause a pressure gradient between the supratentorial compartment and the posterior fossa in the presence of space-occupying lesions, which may be accentuated by exposure to halothane[92] or by rapid reduction of the supratentorial pressure by hyperventilation, thus leading to upward or transtentorial herniation. The anesthetic implications of these facts require avoiding excessive hyperventilation, particularly if the patient is placed in the sitting position, as this may, in addition, lead to rupture of one of the bridging veins and the formation of subdural hematoma. A similar situation may also exist after the use of osmotic diurectis, such as mannitol; therefore such drugs should be avoided.

Access to the posterior fossa may be accomplished with the patient in the lateral or park bench position, which is ideal for insertion of cerebellar stimulator electrodes. The front of the chest should be prepped and draped to allow easy insertion of the stimulator mechanism.

The prone position is used in children under one year of age to prevent the excessive weight stress on the developing spine that the sitting position would cause. Although the prone position provides good access to the midline structures, lesions in the superior parts of the posterior fossa are difficult to deal with in this position. Further pooling of blood in the surgical field may hinder vision. From the anesthetic point of view, the prone position may present difficulties in airway management, and, for that reason, the use of extralong flexible endotracheal tubes is advised. Also, proper support of the pelvis and the chest by placing the child on lateral rolls or a similar device will allow adequate ventilation and prevent interference with venous return.

The sitting position provides an excellent visual and surgical approach to most posterior fossa lesions and is still preferred in our institution, yet it suffers two undesirable drawbacks. The first drawback is arterial hypotension caused by pooling of the circulating blood volume in the dependent lower limbs, and this is likely to happen in hypovolemic patients from decreased water intake or in whom osmotic diuretics were used preoperatively. This situation is easily corrected by rapid administration of lactated Ringer's solution to achieve a central venous pressure level of 2–4 torr, and by wrapping the legs with elastic bandages. It is also recommended that the patient be placed in a semireclining position with the knees supported in flexion at the level of the heart; this position will not cause a significant drop in cardiac output, in contrast to that which occurs in patients placed in the upright sitting position. Venous air embolism, the second complication of the sitting position, is due to entrainment of air from an open venous sinus at the surgical field in the presence of a vertical gradient between the open vessel and the level of the heart, and also because the venous sinuses are attached to the dura, which prevent them from collapsing, thus allowing a continuous flow of air to the heart. The importance of the vertical gradient as an essential factor for the occurrence of air embolism was shown in a clinical study by Albin and coworkers,[93] where 25% of patients in the sitting position, 8% in

* The reader interested in history, and the value of clinical examination, is referred by the Author to his reference to Vesalius' comments on hydrocephalus for comparison to these observations by W. Johak.

the lateral position, 18% in the supine position, and 10% in the prone position had clinical evidence of venous air embolism in the presence of a vertical gradient ranging between 5 and 65 cm. Venous air embolism is by no means a rare occurrence in pediatric neurosurgical patients. The incidence of air embolism in the sitting position is about 30% if sensitive methods of detecting air is used.[94] The entrance of air to the right atrium initiates a sequence of changes depending mainly on the rate of entainment. If the rate is slow, it produces mechanical obstruction of the distal pulmonary vessels, and also a reflex spasm of the unembolized pulmonary arterioles.[95] This leads to a rise in pulmonary artery pressure and an increase in ventilation perfusion mismatch, evidenced clinically by hypoxemia and a drop in end expiratory carbon dioxide concentration. Acute pulmonary edema is a documented complication of air embolism,[96-98] due possibly to the rise in pulmonary artery pressure or alveolar hypoxemia. Both the ventilation perfusion mismatch and the pulmonary edema lead to arterial hypoxemia, which, if sufficient, will cause myocardial failure, hypotension, and cardiovascular collapse. Hypotension, particularly in the sitting position, may also cause ischemia of the vasomotor centers and a drop in the peripheral vascular resistence,[99] thus accentuating the preexisting hypotension. The rise in pulmonary artery pressure eventually leads to a rise in right ventricular and right atrial pressures, which, in the presence of a patent foramen ovale, may cause paradoxical embolization of air to the systemic circulation.[100] This may explain unexpected neurological deficits following neurosurgical procedures complicated by air embolism. Rapid entrainment of air causes an air lock in the right ventricle and the pulmonary outflow track, thus leading to a rapid drop in cardiac output and cardiovascular collapse. Early detection of air embolism is critical if its life-threatening sequelae are to be avoided, and of the noninvasive devices used, the doppler remains by far the most sensitive,[101] as it is capable of detecting minute amounts of air—as small as 0.2 ml. Another noninvasive device is the end-tidal carbon dioxide analyzer, which we use as a backup for the doppler in all posterior fossa craniotomies in the sitting position.[102] The insertion of a Swan-Ganz catheter allows monitoring pulmonary artery pressure, which is another sensitive method of detecting air pressure,[103] although it is an invasive technique and has its own morbidity. The clinical signs of air embolism appear after a fairly large amount of air is embolized. These signs include cardiac dysrhythmias, particularly tachycardia and, less often, bradycardia; premature ventricular contractions; arterial hypotension together with hypoxemia; and the Mill-Wheel murmur over the heart. The treatment of venous air embolism should be initiated if air entry is suspected, particularly during dissection of the spinal muscles or the elevation of bone flaps, with the aim of stopping further air entrainment by identifying and coagulating the open venous channel. Lowering the head of the patient will decrease the vertical gradient, thus preventing further air entry. Then 100% oxygen should be administered because the presence of nitrous oxide in the inspired mixture of gases will increase the size of the air bubble.[104] The aspiration of air from an accurately placed central venous catheter may be extremely useful in removing the trapped air from the right side of the heart.[105,106] Vasopressors are used to correct the hypotension.

Anesthetic Adjuncts

Deliberate Hypotension

Pharmacologically induced hypotension in pediatric neurosurgery is becoming an important part of the anesthetic management of not only patients with arteriovenous malformation of the brain, aneurysms, and vascular tumors, but also in long reconstructive procedures to minimize the blood loss and to improve the surgical field. Deliberate hypotension is used also to control, the intraoperative hypertension that may accompany light levels of anesthesia when inhalation agents such as halothane are avoided. Arterial hypotension can be induced by several methods, some of which, although effective, are not recommended because of their long duration of action or unwanted metabolic effects.

Inhalation Agents

Deep anesthesia with inhalation agents such as halothane produces hypotension by their depression of the myocardium, while they have no effect on the peripheral vascular resistance.[107] Thus these reduce agents tissue perfusion and predispose to metabolic acidosis.

Vasodilators and Sympathetic Ganglion Blockers

Presently the popular drugs used for deliberate hypotension include sodium nitroprusside (SNP), which is a direct vasodilator agent that acts mainly on the resistance vessels,[108] and glyceryl trinitrate (GTN), a direct vasodilator that acts on the capacitance vessels.[109] Another useful group of drugs are the sympathetic ganglion blockers such as trimethaphan camsylate and pentolinium tartrate, both of which produce arterial hypotension by arteriolar and venular dilation.[110] Table 16.7 summarizes the differences among the three most commonly used hypotensive drugs.

Close monitoring of the arterial blood pressure is essential, preferably by an indwelling arterial cannula, while catheterization of the urinary bladder will allow monitoring of urinary output, which is an indicator of adequacy of cardiac output. It should be noted that resistance to NIP is an early sign of toxicity, together with the development of metabolic acidosis and the

Table 16.7. Most Commonly Used Hypotensive Drugs

	SNP	GTN	Trimethaphan
Onset	Rapid	Rapid	Rapid
Preparation	0.01%	0.04–0.1%	0.2%
Initial dose mcg/kg/min	0.2–1.0	0.2–1.0	20–50
Maximum dose (Ref: 111)	5 mg/kg	Unknown	Unknown
Histamine release	Possible	None	None
Metabolism	Cyanide thiothianate	Denitration in the liver	Inactivated by plasma cholinesterase

SNP = Sodium nitroprusside.
GTN = glyceryl trinitrate.

presence of low arterial to mixed venous oxygen content difference, all of which are indications for discontinuation of the drug.

Hypothermia

At present there are few indications for the clinical use of hypothermia in neurosurgery. It is used mainly during the surgical clipping of cerebral aneurysms when temporary clamping of a major cerebral vessel is planned, or during surgical excision of a large arteriovenous malformation. Hypothermia may be used when hypotension is contraindicated, for examples, in vertebral and basilar artery aneurysms with signs of arterial steal from the brainstem. Moderate hypothermia, to a core temperature of 28–32° C, is most commonly used because profound hypothermia requires the use of cardiopulmonary bypass.[112] The protective effect of hypothermia on the brain and other body organs is well documented,[113] and several methods have been described for induction of hypothermia in neurosurgical procedures, such as the use of cardiopulmonary bypass[114] through a thoracotomy or with a closed chest,[115] and selective cooling of the brain by diverting the blood from the carotid arteries through a heat exchanger[116] then back to cool the brain. Nevertheless, these methods require heparinization of the patient, with its inherent morbidity and complications; for that reason, only surface cooling is usually adopted, by placing the patient between two cooling blankets. Hypothermia has a profound metabolic effect, not only on the brain, but also on the cardiovascular system, by causing a predictable drop in the pulse rate and electrocardiographic changes such as prolonged PR and QT intervals, widening of the WRS complex, and changes in the S-T segment and inverted T waves, which may indicate myocardial insufficiency. Premature ventricular contractions and atrial and ventricular fibrillation occur if the temperature drops below 28° C. Respiration has to be adjusted with the induction of hypothermia due to the reduced carbon dioxide output with depressed metabolism. Also, shivering should be prevented by the use of muscle relaxants or phenothiazines. Finally, the effect of muscle relaxants is potentiated in the presence of hypothermia due mainly to depressed renal function. Rewarming is a slower process, and it may be extended to the immediate recovery period. Care should be taken to guard against thermal burns that may occur in the presence of vasoconstriction.

References

1. Kety SS, Schmidt CF: Determiniation of cerebral blood flow in man by the use of nitrous oxide in low concentrations. Am J Physiol 43:53–65, 1945.
2. Hoedt-Rasmussen K, Sveindottir E, Lassen NA: Regional cerebral blood flow in man determined by intracranial injection of radioactive inert gas. Circ Res 18:237, 1966.
3. Pasztor E, Symon L, Dorsch NWC, Branston NM: The hydrogen clearance method in assessment of blood flow in cortex, white matter and deep nuclei of baboons. Stroke, 4:556, 1973.
4. Pamier JL, Weyne J, Demeester G, Leusen I: Influence of changes in acid-base composition of the ventricular system on cerebral blood flow in cats. Arch 333:337, 1972.
5. Reivich M, Jehle J, Sokoloff L, Ketty SS: Measurement of regional cerebral blood flow with antipyrine-14C in awake cats. J Appl Physiol 27:296, 1969.
6. Severinhaus JW, Chiodi H, Egar EA et al.: Cerebral blood flow in man at high altitude: The role of cerebrospinal fluid pH in normalization of flow in chronic hypocapnia. Circ Res 19:274, 1966.
7. Harp JR, Woolman H: Cerebral metabolic effects of hyperventilation and deliberate hypotension. Br J Anaesth 45:256, 1973.
8. Rosmoff HL: Distribution of intracranial contents with controlled hyperventilation: Implications for neuroanesthesia. Anesthesiology 24:640, 1963.
9. Purves MJ, James IM: Observations on the control of cerebral blood flow in sheep fetus and newborn lamb. Circ Res 25:651, 1969.
10. Reivich M, Brann AW, Shapiro H: Reactivity of cerebral vessels to CO_2 in the newborn rhesus monkey. Eur Neurol 6:132, 1971.
11. Shapiro HM: Intracranial hypertension: Therapeutic and anesthetic considerations. Anesthesiology 43:445–71, 1975.
12. Strandgaard S, Olsen J, Skinhoj E, et al.: Autoregulation of brain circulation in severe arterial hypertension. Br Med J 159:507–10, 1973.

13. Moss AJ, Leibling W, Adams FH: The flush method for determining blood pressure in the first year of life. Pediatrics 21:950, 1958.
14. Brock M, et al.: Regional cerebral blood flow and vascular reactivity in cases of brain tumor. In Ross Russel RW (ed): Brain and Blood Flow. London, Pitman Medical, 1971, p 281.
15. Palvolgyi R: Regional cerebral blood flow in patients with intracranial tumors. J Neurosurg 31:149, 1969.
16. Crockard HA, Brown FD, Mullen JA: The effect of trimethaphan and sodium nitroprusside on cerebral blood flow in the rhesus monkey. Acta Neurochir 35:85, 1976.
17. Kogure K, Schienberg P, Rienmuth OM et al.: Mechanisms of cerebral vasodilatation in hypoxia. J Appl Physiol 29:223, 1970.
18. Siesjo BK, Nielsen L: Influence of arterial hypoxemia upon labile phosphates and upon extracellular and intracellular lactate and pyruvate concentration in the rat brain. Scand J Clin Lab Invest 27:83, 1971.
19. Duc G: Assessment of hypoxia in the newborn, suggestions for a practical approach. Pediatrics 48:469, 1971.
20. Olesen J: Contralateral focal increase in cerebral blood flow in man during arm work. Brain 94:635, 1971.
21. Raichle, M.: Sensori-motor area increase of oxygen uptake and blood flow in the human brain during contralateral hand exercise. Preliminary observations by the 0–15 method. In Ingvar DH, Lassen NA (ed): Brain Work, the Coupling of Function. Metabolism and Blood Flow in the Brain. Munksgaard, Copenhagen, 1975, p 372.
22. Ingvar DH: Patterns of brain activity revealed by measurement of regional cerebral blood flow. In Ingvar DH, Lassen NA (ed): Brain Work, the Coupling of Function. Metabolism and Blood Flow in the Brain. Copenhagen, Munksgaard, 1975, p 397.
23. Kety SS: Discussion on psychoactive drugs and anxiety, their influence on cerebral circulation and metabolism. In Ingvar DH, Lassen NA (ed): Brain Work the Coupling of Function, Metabolism and Blood Flow in the Brain. Copenhagen, Munksgaard, 1975, p 472.
24. Michenfelder JD, Theye RA: Canine systemic and cerebral effects of hypotension induced by hemorrhage, trimethaphan, halothane and nitroprusside, Anesthesiology 46:188, 1977.
25. Fitch W, MacKenzie A, et al.: The effects of decreasing arterial blood pressure on cerebral blood flow in the baboon, influence of sympathatic nervous system. Circ Res 37:550, 1975.
26. Mayer MW, Klassen AC: Regional brain blood flow during sympathetic stimulation. Stroke 4:370, 1973.
27. Monro A: Observations on the structure and function of the nervous system. Edinburgh, Creech and Johnston, cited by Mc Lone DG, Bondaceff W and Raimondi AS. J. of Neureopath and Experimental Neurol. 30:627, 1971.
28. Kellie G: An account of the appearances observed in the dissection of two or three individuals presumed to have perished in the storm of third and whose bodies were discovered in the vicinity of Leith on the morning of the 4th of November, 1821, with some reflections on the pathology of the brain. Transaction of the Medico-Chirurgical Society of Edinburgh 1:84, 1824.
29. Sahar A, Hochwald GM, Ranshoff J: Cerebrospinal fluid and cranial sinus pressure. Arch Neurol (Chicago) 23:413, 1970.
30. Langfitt TW, Weinstein JD, Kassel NF: Cerebral vasomotor paralysis produced by intracranial hypertension. Neurology 15:622–41, 1965.
31. Schutta HE, Kassel NF, Langfitt TW: Brain swelling produced by injury and aggravated by arterial hypertension. Brain 91:281–94, 1968.
32. Keaney NP, Pickerodt VWA, McDowal DG, et al.: Cerebral circulatory and metabolic effects of hypotension produced by deep halothane anesthesia. J Neurol Neurosurg Psychiatry 36:898, 1973.
33. Freeman J, Ingvar DH: Elimination of hypoxia of cerebral blood flow autoregulation and EEG relationship. Exp Brain Res 5:61, 1968.
34. Schettini A, McKay L, Mahig J, Modell JH: The response of brain surface pressure to hypercapnia, hypoxia and hyperventilation. Anesthesiology 36:4, 1972.
35. Marshall WJS, Jackson JLF, Langfitt TW: Brain swelling caused by trauma and arterial hypertension. Arch Neurol (Clin) 21:545, 1969.
36. Wollman H, Alexander SC, Cohen PJ et al.: Cerebral circulation of man during halothane anesthesia. Anesthesiology 25:180, 1970.
37. Michenfelder JD, Theye RA: Effect of methoxyflurane on canine cerebral blood flow. Anesthesiology 38:123, 1973.
38. Jorgensen PB, Hendriksen HT: The effect of fluorxene on intracranial pressure in patients with intracranial space-occupying lesions. Br J Anaesth 45:599, 1973.
39. Theye RA, Michenfelder JD: The effect of nitrous oxide on canine cerebral metabolism. Anesthesiology 29:1119, 1968.
40. McDowall DG, Barker J, Jennett WB: Cerebrospinal fluid pressure measurement during anesthesia. Anaesthesia 21:189, 1966.
41. Shapiro HM, Galindo A, Wyte SR, et al.: Rapid intraoperative reduction of intracranial pressure with thiopentone. Br J Anaesth 45:1057, 1973.
42. Takahaski T, Takasaki M, Namiki A, Dohi S: Effects of althesin on cerebrospinal fluid pressure. Br J Anaesth 45:179, 1973.
43. Fitch W, Barker J, Jennett WB, et al.: The influence of neuroleptanalgesic drugs on cerebrospinal fluid pressure. Br J Anaesth 41:800, 1969.
44. List WF, Crumrine RS, Cascorbi HF, Weiss WH: Increased cerebospinal pressure after ketamine. Anesthesiology 36:98, 1972.
45. Jennett WB, Barker J, Fitch W, McDowall DG: Effect of anaesthesia on intracranial pressure in patients with space-occupying lesions. Lancet 1:61, 1969.
46. Adams RW, Gronert GA, Stundt TM, Michenfelder JD: Halothane hypocapnia cerebrospinal fluid pressure in neurosurgery. Anesthesiology 37:510, 1972.
47. Gordon E: The action of drugs on the intracranial contents. In Boulton TB, et al. (eds): Progress in Anesthesiology. Amsterdam, Excerpta Medica, 1970, p 60.
48. Zatoni J, Siani C, Rivano C: In Lavin P, Beer R, Weithoff E (eds): Proceedings of the First Symposium on Modern Anaesthetic Agents. Berlin, Springer-Verlag, 1975, p 272.
49. Michenfelder JD, Cucchiara RF: Canine cerebral oxygen

consumption during enflurane anesthesia and its modification during induced seizures. Anesthesiology 40:575, 1974.
50. Smith AL, Wollman H: Cerebral blood flow and metabolism, effect of anesthetic drugs and techniques. Anesthesiology, 36:378, 1972.
51. Michenfelder JD, Theye RA: Hypothermia: Effect on canine brain and whole body metabolism. Anesthesiology 29:1107, 1968.
52. McQueen JD, Jeanes LD: Influence of hypothermia on intracranial hypertension. J Neurosurg 19:277, 1962.
53. Ishak B, Gutierrez F, Seleny FL: Anesthetic management of arteriovenous malformations of the brain in infants and children. Anesthesiol Rev 12:23, 1978.
54. Johnston IH, Jennett B: The place of continuous intracranial pressure monitoring in neurosurgical practice. Acta Neurochir 29:53, 1973.
55. Troupp H: Ventricular pressure recording after severe brain injuries. Eur Neurol 11:227, 1974.
56. Lundberg N: Continuous recording and control of ventricular fluid pressure in neurosurgical practice. Acta Psychiatry Neurol Scand Suppl 149, 1960.
57. Miller JD: Intracranial pressure monitoring. Br J Hosp Med 19(5):497, 1978.
58. Croneos NJ, Turner JM, Givson RM, et al.: Comparison of extradural with intraventricular pressure in patients after head injury. In Brock M, Dietz H (eds): Intracranial Pressure II. Berlin, Springer-Verlag, 1976, p 51.
59. Vries JK, Becker DP, Young HF: A subarachnoid screw for monitoring intracranial pressure (technical note). J Neurosurg 39:416, 1973.
60. Wyler AR, Kelly WA: Use of antibiotics with external ventriculostomies. J Neurosurg 37:185, 1972.
61. Lundberg N: Monitoring of the intracranial pressure. In Critchley M, O'Leary JL, Jennet B (eds): Scientific Foundations of Neurology. Philadelphia Davis FA, 1972, p 356–71.
62. Janny P, Jouan JP, Janny M et al: A statistical approach to long term monitoring of intracranial pressure. In Brock M, Dietz H (eds): Intracranial Pressure. Berlin, Springer-Verlag, 1972.
63. Schettini A, Malig J, Moreshead G: Influence of cerebral vascular factors on brain relative stiffness. In Brock M, Dietz H (eds): Intracranial Pressure. Berlin, Springer-Verlag, 1972, p 27–32.
64. Szewczykowsli J, Dytko P, Junicki A, et al.: A method of estimating intracranial decompression in man. J Neurosurg 45:155, 1976.
65. Avezaat CJJ, van Eijindhoven JHM, Moolenaar WCJ: A new method of monitoring intracranial volume pressure relationship. In Becks JWF, Bosch DA, Brock M (eds): Intracranial Pressure III. Berlin, Heidelberg, New York, Springer-Verlag, 1976.
66. Downes JJ, Raphealy RC: Pediatric intensive care. Anesthesiology 43:238, 1975.
67. Gregory G: Anesthetic management of specific surgical problems: Premature and term neonate. The Annual Refresher Course Lectures, ASA, 1979, p 102.
68. Holinger PC, Holinger LD, Reichert TJ: Respiratory obstruction and apnea in infants with bilateral abductor vocal cord paralysis: Meningomyelocele, hydrocephalus and Arnold-Chiari malformation. J Pediatr 92:368, 1978.
69. Emery JL, Levic R: Movement of the brain stem vessels around the brain stem in children with hydrocephalus and Arnold-Chiari deformity. Dev Med Child Neurol (suppl) 11:49, 1966.
70. Krisch WM, Duncan BR, Black FO, et al.: Laryngeal palsy in association with meningomyelocele, hydrocephalus and the Arnold-Chiari malformation. J Neurosurg 28:207, 1969.
71. Gendell HM, McCallum JE, Reigel DH: Cricopharyngeal achalasia associated with Arnold-Chiari malformation in childhood. Child's Brain 4:65, 1978.
72. Cameron JL, Mitchell WH, Zuidema GD: Aspiration pneumonia, clinical outcome following documented aspiration. Arch Surg 106:49, 1973.
73. North JB, Jennett S: Abnormal breathing patterns associated with acute brain damage. Arch Neurol 31:338, 1974.
74. Ducker TB: Increased intracranial pressure and pulmonary edema. Part I. Clinical study of 11 patients. J Neurosurg 28:112, 1968.
75. Miller JD, Sullivan HG: Management of acute intracranial disasters. IAC 17:59, 1979.
76. Hersch C: Electrocardiographic changes in subarachnoid hemorrhage, meningitis and intracranial space-occupying lesions. Br Heart J 26:785, 1964.
77. Marshall AJ: Transient atrial fibrillation after minor head injury. Br Heart J 38:948, 1976.
78. McLaurin RL, King LR: Recognition and treatment of metabolic disorders after head injury. Clin Neurosurg 19:281, 1972.
79. Glatt BS, Rowe RD: Cerebral arteriovenous fistula associated with congestive heart failure in the newborn. Report of two cases. Pediatrics 26:596, 1960.
80. Jennett B, Teasdale G: Aspects of coma after severe head injuries. Lancet, 1:878, 1977.
81. Cotev S, Shalit MW: Effects of diazepam on cerebral blood flow and oxygen uptake after head injury. Anesthesiology 43:117, 1975.
82. Burney RG, Winn R: Increased cerebrospinal fluid pressure during laryngoscopy and intubation for induction of anesthesia. Anesth Analg (Cleve) 54:687, 1975.
83. Smith C: An endo-oesophageal stethoscope. Anesthesiology 15:566, 1954.
84. Mullan S, Raimondi AJ: Respiratory hazards in surgical treatment of Arnold-Chiari malformation. J Neurosurg 19:675, 1962.
85. Ishak B, McLone D, Seleny FL: Intra-operative autonomic dysfunction associated with Arnold-Chiari malformation. Child's Brain 7:146, 1980.
86. Raimondi AJ, Matsumoto S: A simplified technique for performing the ventriculo-peritoneal shunt. J Neurosurg 26:357, 1967.
87. Grosfeld JL, Cooney DR, Smith J, Campbell RL: Intraabdominal complications following ventriculoperitoneal shunt procedures. Pediatrics 54:791, 1974.
88. Wilson CB, Bertan V: Perforation of the bowel complicating peritoneal shunts for hydrocephalus. Am Surg 32:60, 1966.
89. Hougen TJ, Emmanoulides GC, Moss AJ: Pulmonary

valvular dysfunction in children ventriculovenous shunts for hydrocephalus: A previously unreported condition. Pediatrics 55:836, 1975.
90. Cooper IS, Amin I, Upton A, et al.: Chronic cerebellar stimulation in epilepsy: Clinical and anatomical studies. Arch Neurol 33:559, 1976.
91. Cooper IS, Ricklan M, Amin I, et al.: Chronic cerebellar stimulation in cerebral palsy. Neurology 26:744, 1976.
92. Fitch W, McDowall DG: The effect of halothane on the intracranial pressure gradients in the presence of intracranial space-occupying lesions. Br J Anaesth 43:904, 1971.
93. Albin MS, Chang JL, Babinski M, et al.: Intracardiac catheters in neurosurgical anesthesia. Letter to the editor. Anesthesiology 50:67, 1979.
94. Michenfelder JD, Miller RH, Gronert GA: Evaluation of an ultrasonic device (doppler) for diagnosis of venous air embolism. Anesthesiology 36:164, 1972.
95. Josephson S: Pulmonary hemodynamics during experimental air embolism. Evidence of vasoconstriction. Scand J Clin Lab Insert 26 (Suppl) 115:113, 1970.
96. Chander WF, Dimchoff DJ, Taren JA: Acute pulmonary edema following venous air embolism during a neurosurgical procedure. J Neurosurg 40:400, 1974.
97. Ishak BA, Seleny FL, Noah Z: Venous air embolism, a possible cause of acute pulmonary edema. Anesthesiology 45:453, 1976.
98. Perschau RA, Munson ES, Chapin JC: Pulmonary interstitial edema after multiple venous air embolism. Anesthesiology 45:364, 1976.
99. Adornato DC, Gildenberg PL, Ferario CM, et al: Pathophysiology of intravenous air embolism in dogs. Anesthesiology 49:120, 1978.
100. Gronert GA, Messick JM, Cucchiara RF, Michenfelder JD: Paradoxical air embolism for a patent foramen ovale. Anesthesiology 50:548, 1979.
101. Gildenberg PL, O'Brien RP, Britt WJ, Frost EAM: The efficacy of doppler monitoring for the detection of venous air embolism. J Neurosurg 54:75, 1981.
102. Chang JL, Albin MS, Hung TK: Analysis and comparison of venous air embolism detection methods. Neurosurgery 7:135, 1980.
103. Munson ES, Pual WL, Perry JC, et al: Early detection of venous air embolism using a Swan-Ganz catheter. Anesthesiology 42:223, 1975.
104. Steffey EP, Johns BH, Egar EI: Nitrous oxide intensifies the pulmonary arterial pressure response to venous injection of carbon dioxide in the dog. Anesthesiology 52:52, 1980.
105. Michenfelder JD, Terry HR, Daw EG, Miller RH: Air embolism during neurosugery: A new method of treatment. Anesth Analg (Cleve) 45:390, 1966.
106. Tateishi H: Prespective study of air embolism. Br J Anaesth 44:1306, 1972.
107. Jordan WS, Graves CL, Boyd WA, et al: Cardiovascular effects of three techniques for inducing hypotension during anesthesia. Anesth Anal (Cleve) 50:1059, 1971.
108. Franciosa JA, Buiha NH, Limas CJ, et al.: Improved left ventricular function during nitroprusside infusion in acute myocardial infarction. Lancet 1:650, 1972.
109. Fahmy NR: Nitroglycerin as a hypotensive drug during general anesthesia. Anesthesiology 49:17, 1978.
110. Wang HH, Liu MP, Katz RL: A comparison of the cardiovascular effects of sodium nitroprusside and trimethaphan. Anesthesiology 46:40, 1977.
111. Michenfelder JD, Tinker JH: Cyanide toxicity and thiosulfate protection during chronic administration of sodium nitroprusside in the dog. Anesthesiology 47:441, 1977.
112. Sellick BA: Induced hypothermia. In Hewer CL, (ed): Recent Advances in Anaesthesia and Analgesia. London, Churchill, 1969.
113. Michenfelder JD, Vihlein A, Dawe EF, Theye RA: Moderate hypothermia in man, hemodynamic and metabolic effects. Br J Anaesth 37:938, 1965.
114. Drew CE: Profound hypothermia in cardiac surgery. Report of three cases. Lancet, 1:478, 1959.
115. Michenfelder JD, Kirklin JW, Vihlein A, et al.: Clinical experience with a closed chest method of producing profound hypothermia and total circulatory arrest in neurosurgery. Ann Surg 159:125, 1964.
116. Kristiansen K, Krog J, Lund I: Experience with selective cooling of the brain. Acta Chir Scand Supl 253:151, 1960.

"Crito, We owe a cock to Aesculapius."

SOCRATES
(dying words)

Chapter 17

Neurologic Intensive Care

Badr A. Ishak
Zehava L. Noah

Intensive care units represent a natural and expected extension in the care of critically ill patients who require close medical and nursing observation because of rapidly changing physiological variables or are in need of advanced monitoring. At present, neurologic intensive care units (NICU) are limited to medical centers with adequate radiological support, including computed transmission tomography, and neurophysiological monitoring facilities such as EEG and somatosensory evoked potentials (SSEP).

Criteria for Admission to the Neurologic Intensive Care Unit

Severe Head Injuries

The management of severe head injuries undoubtedly starts at the site of the accident or trauma, as some of the associated temporary conditions like airway obstruction or apnea may worsen the initial intracranial insult, and are probably a reason for the high mortality during the first two hours after severe head injuries.[1] Different studies[2,3] have shown a significantly better outcome after severe head injuries in the pediatric age group, and for that reason all patients with head injuries are routinely addmitted to NICU even in the presence of an unfavorable prognostic score.[4,5] The initial neurological assessment[6] is done in the emergency room, and if the descision is made to secure an airway that is compromised due to obstruction, hypoventilation, apnea, or even in the presence of marked spontaneous hyperventilation, which may be due to central lactic acidosis from localized tissue hypoxemia following the cerebral insult.[7,8] Insertion of the endotracheal tube should only be performed after adequate anesthesia and muscular relaxation to avoid the increase in intracranial pressure that accompanies laryngoscopy or bucking during intubation. A CT scan or occasionally an angiogram may be required to exclude surgically amenable lesions before transferring the patient to NICU where treatment is continued. Pulmonary involvement is common with head injury. It may be due to concomitant lung contusion, or hypoxemic insult during a period of arterial hypotension, manifested clinically as adult respiratory distress syndrome,[10] where progressive arterial hypoxemia, decreased lung compliance with reduced functional residual capacity, and the appearance of the characteristic lung infiltrates of shock lung on chest x-ray film are the clues for clinical diagnosis.[11-13] Aspiration of vomitus shortly after the head injury may also lead to severe aspiration pneumonitis. Early treatment of the pulmonary complications with controlled ventilation, positive end expiratory pressure (PEEP) reverse these changes in the majority of cases,[14] although high levels of PEEP, by increasing the mean intrathoracic pressure transmitted to the right atrium may lead to a drop in the cerebral perfusion pressure (CPP).[15] Hence it is advisable not to exceed the optimum PEEP required for maximum therapeutic effect as evidenced by improved oxygenation. Also the patient should be nursed in a 30-degree head-up tilt, so as to counteract the hydrostatic effect of PEEP on the intracranial pressure.[16] Acute neurogenic pulmonary edema (NPE) is a well-documented serious complication of head inju-

ry.[17] It also occurs with other intracranial pathology and peripheral nervous disorders such as poliomyelitis.[18] The occurrence of NPE in patients with head injuries, even in the absence of previous pulmonary disease, may be due to an increase in ICP, which produces intense sympathetic stimulation in an effort to perfuse the ischemic medullary centers,[19] causing marked systemic hypertension leading eventually to left ventricular failure and the development of pulmonary edema. The management of NPE consists of controlled ventilation, PEEP, diuretics, and cardiac support.

States of Altered Consciousness and Coma

When a comatosed child is admitted to NICU, he or she will require a rapid assessment of, not only central nervous system deficits, but other systemic dysfunctions that may present with coma or altered consciousness. The presence or absence of focal neurologic signs will place the patient in one of four categories[20]:

1. No focal signs (normal ICP)
   a. Some metabolic encephalopathies (renal, hepatic)
   b. Drug overdose
   c. CNS infections
   d. Trauma (concussion)
   e. Epilepsy (postictal state)
2. No focal signs (elevated ICP)
   a. Metabolic encephalopathies (Reye's syndrome, lead poisoning, posthypoxic encephalopathy)
   b. CNS infections
   c. Trauma (subdural, subarachnoid hematomas)
   d. Brain tumors (midline)
   e. Hydrocephalus
3. Focal signs (normal ICP)
   a. Vascular occlusions
   b. CNS infections (encephalitis)
   c. Trauma (contusion)
   d. Epilepsy (Todd's paralysis)
4. Focal signs (elevated pressure)
   a. Trauma (subdural, epidural hematomas)
   b. Tumors
   c. CNS infections (abscess)
   d. Vascular (AVM)

Focal neurologic signs always indicate structural brain disease, which can often be detected by a CTT scan, EEG, or cerebral angiography.

Severe Intracranial Infections

Severe septic meningitis due to pneumococcal or *Hemophilus* infections and encephalitis are the usual causes for admission to NICU in the pediatric age group, particularly if rapidly progressive as evidenced by a deteriorating level of consciousness or signs of brainstem impaction, and also the presence of other superimposed systemic infections.[21] Intensive treatment is directed toward management of the elevated ICP, avoiding the development of acute cerebral edema by fluid restriction and diuretics, and proper antibiotic therapy.

Postcirculatory Arrest Encephalopathy

The presence of coma or other sings of CNS damage following hypoxic cerebral insult after successful resuscitation from cardiac arrest, especially if witnessed, require intensive management directed toward neuronal conservation. The importance of such treatment after global brain insult was shown to be particularly effective in dealing with children who suffered near-drowning, where complete recovery was achieved even after apparently established brain death.[22] In one study, aggressive CNS-oriented therapy resulted in complete recovery in 77% of comatosed children who suffered near-drowning,[23] which is significantly higher than in a similar study where only routine therapeutic measures were taken,[24] especially in decorticate and decerebrate patients.

Intractable Convulsions

Recurrent convulsions are commonly due to epilepsy, whether idiopathic or organic, which follow trauma, intracranial hemorrhage, and infections, or it may be secondary to different metabolic derangements such as hypoglycemia from hyperinsulinism and adrenocortical insufficiency, as well as hepatic and renal failure. The main objective of the intensive treatment is to control the acute convulsive phase and also to prevent recurrence by recognizing the cause if permanent damage to the CNS is to be avoided.[25]

Postoperative care

NICU is often used for the postoperative monitoring and care of patients recovering from major neurosurgical procedures. During this critical period, a marked rise in ICP may occur following craniotomies where minimal or no decompression of the intracranial contents was accomplished, such as biopsy of a tumor.[26,27] The availability of trained respiratory therapy personnel helps to provide optimum oxygen therapy and mechanical support to ventilation and airway protection if required.

Monitoring in the Neurologic Intensive Care Unit

Guidelines for Monitoring

One of the main functions of specialized intensive care units is to provide system-oriented monitoring, and this is no exception to NICU. To provide a rational and practical approach to the problem of monitoring, a patient scoring system to determine the level of monitoring is proposed, derived from the American Society of Anesthesiologists' (ASA) physical status classification[28] in

combination with Shapiro's CNS evaluation system,[29] as the former alone does not provide an adequate assessment of the CNS.

The ASA physical status classification:

1. Normal healthy patient	one point
2. Mild systemic disease	two point
3. Severe systemic disease, limited activity	three points
4. Incapacitating systemic disease	four points
5. Moribund, not expected to survive 24 hours	five points

Shapiro's CNS evaluation classification:

1. Alert, with immediate appropriate response	one point
2. Lethargic, with delayed incomplete response and approporiate verbalization	two points
3. Obtunded, who is difficult to arouse but obeys with no verbalization	three points
4. Stuporous, obeys but without verbalization and responds in defensive movements	four points
5. Comatose, with no verbal and absent or inappropriate motor response	five points

A score of 2–3 requires minimum or basic monitoring; a score of 4–6 requires enhanced monitoring; specialized monitoring is reserved for patients with scores of 6 or more.

Levels of Monitoring

Basic Monitoring

Patients recovering from minor surgery and who had stable vital signs before and after surgery only require routine nursing observation on a more frequent basis, including the respiratory and pulse rates, temperature, and noninvasive measurement of the blood pressure by auscultation,[30] oscillometry,[31] distal flow detection,[32] or ultrasonic devices.[33]

Enhanced Monitoring

For those patients with scores of 4–6, a more invasive monitoring approach may be required, which often includes cannulation of a peripheral artery for continuous pressure and intermittent blood gases monitoring. An Allen test[34] should be routinely performed before cannulation of the radial artery at the wrist to avoid occluding the arterial supply to the hand in the presence of a dominent radial artery. Central venous catheters, are also useful in fluid management, as they reflect the right atrial pressure, which represents the state of the cardiac preload, yet they are inadequate indicators of left ventricular function in the presence of multiple trauma, acute and chronic respiratory failure.[35]

Specialized Monitoring

This level of monitoring is recommended for patients with clinical scores of 6 or more, as it provides maximal amounts of data, which help in the management of these critically ill patients and may include intracranial pressure monitoring, as well as pulmonary artery and pulmonary wedge pressure monitoring.

System Monitoring in the Neurologic Intensive Care Unit

Respiratory System

1. Respiratory rate (B)
2. Tidal volume (E)
3. Minute volume (E)
4. Vital capacity (E)
5. End tidal carbon dioxide concentration (E)
6. Inspired oxygen concentration (B)
7. Arterial blood gases (E)
8. Chest x-ray (B)
9. Shunt fraction (Qs/Qt.) (S)
10. Alveolar arterial oxygen difference (S)

(B) Basic (E) Enhanced (S) Specialized

From the abovementioned parameters, the rate, tidal volume, and minute volume are checked periodicaly in mechanically ventilated patients. Meanwhile an increasing respiratory rate in a spontaneously breathing patient may indicate the onset of respiratory failure or may be due to a hypermetabolic state as fever. Regular estimation of vital capacity is a helpful guide for the feasibility of extubation. Calculation of the shunt fraction or the alveolar arterial oxygen difference is reserved for patients with severe pulmonary involvement.

Cardiovascular System

1. Heart rate (B)
2. Blood pressure (B)
3. ECG (E)
4. Arterial cannulation (E)
5. Central venous pressure (E)
6. Pulmonary artery wedge pressure (S)

(B) Basic (E) Enhanced (S) Specialized

The use of pulmonary artery catheters may be required as part of specialized monitoring of patients in barbiturates coma in whom measurement of cardiac output and cardiac index provides a handle on the fluid management and consequently hemodynamic stability.

Oxygen Transport

1. Arterial oxygen tension (E)
2. Arterial oxygen content (E)
3. Arterial oxygen saturation (E)
4. Mixed venous oxygen content (S)
5. Arterial, mixed venous oxygen content difference(S)

6. Oxygen consumption (S)
7. Oxygen flux (S)
8. P_{50} (S)

(E) Enhanced (S) Specialized

Oxygen availability is dependent on three variables: the cardiac output, arterial oxygen saturation, and the hemoglobin content. Those variables are evaluated collectively by determination of the oxygen flux,[36] while the P_{50} reflects the availability of oxygen at the tissue level,[37] which is affected by different factors, such as the level of 2,3-DPG, temperature, and pH. Excessive desaturation of hemoglobin at the tissue level due to sluggish peripheral circulation is evidenced by increased arterial, mixed venous oxygen content difference, and may be an early sign of low cardiac output.

Fluid and Electrolytes Balance

1. Serum electrolytes: Na, K, Cl, Ca, PO_4, Mg (B)
2. 24-hour fluid balance chart (B)
3. Serum glucose (B)
4. Serum osmolality (E)

(B) Basic (E) Enhanced

Disturbances in water and electrolytes may occur in critically ill neurosurgical patients for a variety of reasons. Excessive hydration may result from the use of hypotonic solutions or 5% dextrose in water, or secondary to the use of ultrasonic nebulizers to humidify the inspired gases. Furthermore, water retention is known to complicate prolonged mechanical ventilation.[38,39] The syndrome of inapropriate antidiuretic hormone (SIADH) may complicate the course of patients with head trauma,[40,41] meningitis,[42] encephalitis,[43] brain tumors,[40] and subarachnoid hemorrhage,[44] and is manifested by lowered serum osmolality and hyponatremia, which may cause personality changes if slight, or vomiting and convulsions if severe. These patients respond well to water restriction, diuretics, and occasionally the use of hyperosmolar saline solution (3%). On the other hand, dehydration may result from severe water restriction, chronic use of osmotic diuretics, or in the presence of diabetes insipidus. Daily estimation of serum glucose will detect intolerance or latent diabetis mellitus, which may be a late complication of head injuries.[45,46]

Hematology and Hemostasis

1. Hemoglobin estimation (B)
2. Hematocrit (B)
3. Red cell count (B)
4. White cell count and differential (B)
5. Prothrombin time (PT) (E)
6. Partial thromboplastin time (PTT) (E)
7. Platelet count (E)
8. Blood cultures (E)

(B) Basic (E) Enhanced

Hematological parameters provide information regarding the capacity of oxygen transport and the adequacy of the defence mechanisms, while coagulation screening will detect any clotting abnormalities known to be associated with severe head trauma,[47,50] massive blood transfusions, or hemolytic reactions to incompatible blood, shock, and severe infections,[51] usually in the form of hypercoagulability and increased fibrinolytic activity,[48] which is thought to be due to increased sympathetic discharge.[49] In this syndrome of disseminated intravascular coagulation (DIC), the formation of fibrin thrombi and consumption of factors V and VII, together with loss of platelets and activation of the fibrinolytic system suggest the presence of thrombin in the systemic circulation.[52] Clinically, patients with DIC may show thrombotic, hemorrhagic, or mixed signs. Occasionally, laboratory signs are the only evidence of the presence of DIC. The presence of prothrombin time (PT) of more than 15 seconds, fibrinogen level of less than 160 mg/100 ml, and platelet count of less than 150,000 confirms the diagnosis of DIC.[52] The therapeutic aim is directed to the cause, also maintenance of normovolemia and careful replacement therapy of the deficient clotting factors by fresh frozen plasma and platelet concentrate, and rarely heparin may be used to help prevent microthrombi formation,[53] although its use is controversial and may even be dangerous in patients with head injuries, as it may cause subarachnoid hemorrhage.

Metabolic

1. Body temperature (B)
2. Serum albumin (E)
3. Serum lactate (E)
4. BUN and creatinine (E)
5. Urine osmolality (E)
6. Serum ammonia and SGPT/SGOT (S)

(B) Basic (E) Enhanced (S) Specialized.

Metabolic derangements may complicate acute trauma, major neurosurgical procedures, and prolonged coma.[54,55] The presence of serum albumin of less than 3.5 g/100 ml is a an indication of a severe degree of negative nitrogen balance and reflects the increased caloric demands associated with the aforementioned conditions, although chronic administration of steroids may also cause low serum albumin.[56] Adequate caloric intake should be provided via a nasogastric tube in patients incapable of oral feeding, nevertheless in a minority of patients who develop gastrointestinal complications, such as bleeding, may require total parenteral

nutrition (TPN) through a centrally placed indwelling catheter. The basic considerations for TPN are as follows[57]:

1. Nitrogen 0.2–0.4 g/kg/day
2. Energy 40–45 cal/kg/day
3. Nitrogen: energy ratio 1:200
4. Glucose: fat emulsions energy provision ratio 1:1
5. Potassium (mmol): nitrogen (g) ratio 6.0:1.
6. Magnesium (mmol): nitrogen (g) ratio 1.0:1.0.
7. Phosphate 0.5–0.75 mmol/kg/day
8. Adequate electrolytes and water-soluble vitamins
9. Essential fatty acids
10. Even distribution over 24 hours

Central Nervous System Monitoring

1. Clinical assessment (B)
2. EEG (E)
3. CTT scan (E)
4. Ultrasound scan (E)
5. Evoked potentials (S)
6. CSF measurement (S)
7. Cerebral venous blood measurement (S)

(B) Basic (E) Enhanced (S) Specialized

Despite the recent development of various sophisticated monitoring techniques for the functional evaluation of the CNS, clinical assessment still plays an essential role in evaluating patient's response to treatment or the development of further complications. The Glasgow Coma Scale[6] provides a practical approach to clinical monitoring of patients with head injuries as well as other neurosurgical conditions in NICU, particularly if other variables such as the vital signs, lateralization, and signs of brainstem functions are also incorporated as suggested by Marsh[59] (Fig. 17.1). The CCS is preferable for children. Furthermore, there is evidence that such clinical scoring systems may be of value in predicting the outcome in patients with sever head injuries, also in nontraumatic coma.[60,61] One of the enhanced functional monitoring tools of the CNS is the EEG, which is recognized as a sensitive indicator of brain hypoxia and hypoperfusion,[62] nevertheless, it was shown that the presence of severe EEG abnormality, which appears in younger patients undergoing cardiopulmonary bypass, does not reflect the presence of irreversible brain damage.[63] It has been also recommended that EEG should not be used as the only method of confirming brain death.[64] Continuous EEG monitoring may be accomplished by either the cerebral function monitor (CFM)[65] in which both the frequency and amplitude of the EEG are integrated and displayed as a single line, although specific information regarding dominant frequencies is unobtainable. The other method used for continuous EEG monitoring is the compressed spectral array (CSA), which provides information about frequency and amplitude through Fourier analysis of the EEG spectrum[66] and more recently the introduction of the density-modulated spectral array (DSA),[67] which allows a three-dimensional display of the EEG.

The determination of CSF acid-base balance and gas tensions helps to detect the changes in cerebral perfusion,[8] although they reflect only global rather than regional changes. Nevertheless, the severity of the metabolic acidosis due to accumulation of lactic acid in the hypoxic or injured tissue is related to the extent of the cerebral insult.[46] Serial evoked potentials and CTT scans are useful prognostic tools and also for the assessment of progress of the disease.

Guidelines for Management of Patients with Intracranial Pathology

Control of Blood Pressure

The loss of cerebrovascular autoregulation in patients with intracranial pathology leads to an increase in cerebral blood flow in the presence of arterial hypertension, which in turn promotes the development of cerebral edema by translocating water to the extracellular space, thus increasing tissue hydrostatic pressure, which leads eventually to regional reduction in cerebral perfusion.[68,69] Hypotension, on the other hand, causes a reduction in CBF, which, if excessive, may result in brain ischemia.[70] Hence it is essential to maintain the mean arterial pressure within normal limits by the infusion of 5% albumin or the use of vasoactive infusions such as dopamine if fluid intake is restricted. While arterial hypertension may require the use of vasodilator drugs such as trimethaphan or sodium nitroprusside, yet their use in patients with intracranial mass lesion may lead to an initial rise in the intracranial pressure, and for that reason, they should be started gradually in patients with monitored ICP.[71] It should also be noted that the reduction in mean arterial pressure in patients with critical cerebral blood flow may cause further reduction in CBF. Thus the advantage of controlling arterial hypertension must be assessed in individual patients.

Control of ventilation

The indications for controlling ventilation in neurosurgical patients were discussed in Chapter 16, Table 16.4. In addition to controlled respiration, hyperventilation has long been the recognized method of reducing intracranial pressure during neurosurgical procedures[72] and also results in clinical improvement in a variety of intracranial catastrophes,[73,74] which is probably due to normalization of CSF pH. Nevertheless hyperventilation may have an unpredictable effect on regional cerebral blood flow in areas of the brain with lost autoregula-

tion.[75] Also there is a potential for the development of cerebral hypoxemia due to the extreme vasoconstriction that accompanies hyperventilation, yet moderate degrees of hyperventilation caused improvement in the outcome of head-injured patients with elevated intracranial pressure.[76]

Sedation and Neuromuscular Blockade

The undesirable effect of painful stimuli, even in comatose neurosurgical patients, was recently stressed.[77] Also the presence of an endotracheal tube may promote bucking, which in turn decreases the cerebral perfusion pressure by its effect on the intrathoracic pressure. For these reasons, mild sedation is always advisable. Among the commonly used drugs 15 chlorpromazine in small, repeated doses intravenously with careful watch on the arterial blood pressure, as it may cause hypotension, particularly in hypovolemic patients; long-acting barbiturates, diazepam, and droperidol may also be used for sedation with an added beneficial effect of reducing the cerebral metabolic rate and cerebral blood flow.[78,79]

Muscular relaxation with long-acting nondepolarizers such as pancuronium helps to control the respiration, and by reducing the central venous pressure, improves cerebral perfusion. The absence of afferent impulses from proprioceptors in muscles and joints in a paralyzed patient decreases the general neuronal traffic in the brain and may explain the beneficial effect of immobilization on postischemic brain damage.[80]

Control of Temperature

Hypothermia produces a predictable drop in cerebral metbolic rate and oxygen consumption, which is frequently used during surgical correction of complex congenital cardiac defects in infants and children, and also during clamping of arteriovenous malformations and vascular tumors of the brain. In contrast hyperpyrexia causes an increase in cerebral oxygen consumption and may increase the severity of experimentally induced cerebral injury.[81] The antimetabolic and protective effects of hypothermia was suggested by different authors for the management of global hypoxic, and metabolic encephalopathies such as in drowning, Reye's syndrome, and even spinal cord injuries.[23,82,83] Nevertheless, because hypothermia adds a tremendous burden on the nursing staff in NICU, together with the high morbidity due to lowered defense mechanisms against infections, increased blood coagulability and viscosity, and cardiac irritability and shivering, which may increase the total oxygen consumption severalfold, limit the routine use of hypothermia. The tendency at the present time is to maintain normothermia and particularly to avoid hyperthermia in all neurosurgical patients.

Hematological and Metabolic Homeostasis

Normalization of acid-base balance in patients with intracranial pathology prevents central hyperventilation, which results from metabolic or respiratory acidosis, and central hypoventilation from metabolic alkalosis seen frequently after blood transfusion due to metabolic breakdown of citrates to bicarbonates. This pharmacological control of acid-base status is usually supplemented with mechanical support of respiration, as the response of the respiratory centers may lag after the corrected acid-base balance. Maintainence of a hematocrit value of not less than 30% will insure an optimum oxygen-carrying capacity, as well as improved perfusion due to decreased viscosity. Water and electrolyte requirements are given via an intravenous route during the first few days following admission to NICU. Balanced salt solutions with added dextrose are preferred to dextrose in water. It must be also noted that these patients will be in a negative nitrogen balance after the second or third day, so adequate caloric intake should be provided by a nasogastric feeding tube or by the institution of TPN.

Drug Therapy

Steroids

Steroids cause clinical improvement in some focal neurologic lesions associated with cerebral edema, particularly some brain tumors and abscesses, while they are less effective in generalized brain injuries, as in head trauma. Neurological improvement often precedes any reduction in intracranial pressure,[84] which may be attributed to the edema-reducing effect of steroids,[85] and also by stabilization of cellular membrane,[86] and possibly by reduction in CSF formation.[87] The recommended daily dose is 0.25–0.5 mg/kg up to 40 kg.[88]

Osmotherapy

Hyperosmolar agents reduces intracranial pressure by withdrawing water from normal rather than edematous tissues.[89] Urea suffers the disadvantage of rebound increase in the water content of the brain after the initial diuresis as it slowly crosses the blood-brain barrier, causing an increase in intracellular osmotic pressure leading to intracellular translocation of water. Mannitol, a carbohydrate with a molecular weight three times larger than that of urea, causes minimal rebound and is widely used. For a rapid effect, mannitol 0.5–2.0 g/kg is given intravenously as 20% solution. A transient hypervolemia and hypertension invariably follow the administration of mannitol, which usually disappear after the start of diuresis. Occasionally, mannitol may be administered chronically to control elevated intracranial pressure, where the dose is limited to 4–8 g/kg/day.[88] Monitoring of plasma osmolality is essential so as to prevent renal failure, which is liable to occur if plasma osmolality is allowed to exceed 330 mosml/l. It should be also noted that the range of dosage of mannitol may widely vary, and may be minimized if combined with steroids[84] or hyperventilation.[90] The use of glycerol as an osmotic dehydrating agent is also described[91]

in a dose of 1.5–2.0 g/kg orally or 3–6 g/kg/day via a nasogastric tube.[88] However, it tends to cause gastric irritation and possible aspiration, as well as intravascular hemolysis and hemoglobinuria.[92] Chemical or loop diuretics such as furosemide may also be used as a dehydrating agent. In addition, it inhibits cerebrospinal fluid production[93] and may be preferred in patients with cardiac or renal disease.[94]

Barbiturates

The pharmacological and metabolic effects of barbiturates on te brain and their potential for protection against hypoxic insults are well documented.[95–97] Although in sedative doses barbiturates have no significant effect on CBF and CMRO2,[98] yet higher doses produce progressive reduction of both CBF and CMRO2, and will also cause a drop in intracranial pressure, possibly by its effect on CBF.[99] These potentially advantageous effects of barbiturates led to extensive clinical applications of barbiturates in different situations, such as severe head trauma,[100] prophylaxis against stroke following aneurysm surgery,[101] postcardiac arrest encephalopathy,[23,97] and also in metabolic encephalopathies such as Reye's syndrome.[102] Nevertheless, other studies showed that the use of barbiturates does not significantly improve the outcome.[103,104] At present we limit the use of barbiturates to those patients with markedly increased intracranial pressure resistant to conventional treatment with hyperventilation and osmotic dehydration. The recommended dose to initiate barbiturate coma is 3–5 mg/kg of pentobarbital intravenously, followed by 1–2 mg/kg/h so as to attain a blood level of 2.5–4 mg/dl.[105]

References

1. Sevit S: Fatal road accidents in Birmingham and their cause. Injury 4:281, 1973.
2. Bruce DA, Raphaely RC, Goldberg AI, Zimmerman RA, et al: The pathophysiology, treatment and outcome following severe head injuries in children. Child Brain 5:174, 1979.
3. Judson JA: Nonoperative treatment of severe head injuries: The Aukland Experience. In Turbuhovich RV (ed): Management of Acute Intracranial Disasters. Boston, Little, Brown, 1979, p 174.
4. Jennet B, Teasdale G, Braakman R, et al: Predicting the outcome in individual patients after severe head injury. Lancet 1:1031, 1976.
5. Overgaard J, Christensen S, Hvid-Hansen O, et al: Prognosis after head injury based on early clinical examination. Lancet 2:631, 1973.
6. Teasdale G, Jennett B: Assessment of coma and impaired consciousness—A practical guide. Lancet 2:18, 1974.
7. Froman MB, Crampton-Smith AC: Hyperventilation associated with low pH of the cerebrospinal fluid after intracranial hemorrhage. Lancet 1:780, 1966.
8. Gordon E, Rossanda M: The importance of cerebrospinal acid-base status in the treatment of unconscious patients with brain lesions. Acta Anaesth Scand 12:51, 1968.
9. Burney RG, Winn R: Increased cerebrospinal fluid pressure during laryngoscopy and intubation for induction of anesthesia. Anesth Analg (Cleve) 54:687, 1975.
10. Laver MB, Loenstein E: Lung function following trauma in man. Clin Neurosurg 19:133, 1972.
11. Markello R, Schuder R, Border J: Arterial-alveolar N2 differences documenting ventilation perfusion mismatching following trauma. J Trauma 14:423, 1974.
12. Brueggemann MW, Loudon RG, Mc-Laurin RL: Pulmonary compliance changes after experimental head injury. J Trauma 16:16, 1976.
13. Ramachandran PR, Fairly HB: Changes in functional residual capacity during respiratory failure. Can Anaesth Soc J 17:359, 1970.
14. Powers SR, Mannal R, Neclerio M, et al: Physiologic consequences of positive end-expiratory pressure (PEEP) on ventilation. Ann Surg 178:265, 1973.
15. Aidinis SJ, Lafferty J, Shapiro HM: Intracranial response to PEEP. Anesthesiology 45:275, 1976.
16. Frost EAM: Effects of positive end expiratory pressure on intracranial pressure and compliance in brain-injured patients. J Neurosurgery 47:195, 1977.
17. Ducker TB, Simmons RL, Martin AM: Pulmonary edema as a complication of intracranial disease. Am J Dis Child 118:638, 1969.
18. Baker AB: Poliomylitis 16: A study of pulmonary edema. Neurology 7:743, 1957.
19. Ducker TB, Simmons RL: Increased intracranial pressure and pulmonary edema. II. The hemodynamic response of dogs and monkeys to increased intracranial pressure. J Neurosurgery 28:118, 1968.
20. Huttenlocher PR: The comatose child. In Vaughan VC III, McKay RJ (eds): Nelson's Textbook of Pediatrics. Philadelphia, Saunders, 1975, p 1410.
21. Baird DR, Whittle HC, Greenwood BM: Mortality from pneumococcal meningitis. Lancet 2:1344, 1976.
22. Montes JE, Conn AW: Near drowning: An unusual case. Can Anaesth Soc J 27:172, 1980.
23. Conn AW, Montes JE, Barker JE, Edmonds JF: Cerebral salvage in near drowning following neurological classification by triage. Can Anaesth Soc J 27:201, 1980.
24. Modell JH, Graves SA, Kuck EJ: "Near drowning." Correlation of level of consciousness with survival. Can Anaesth Soc J 27:211, 1980.
25. Aicardi J Chevril JJ: Convulsive status epilepticus in infants and children. A study of 239 cases. Epilepsia 11:187, 1970.
26. Leech P, Barker J, Fitch W: Changes in intracranial pressure during the termination of anaesthesia. Br J Anaesth 46:316, 1974.
27. Jorgensen PB, Misfeldt BB: Intracranial pressure during recovery from nitrous oxide and halothane anaesthesia in neurosurgical patients. Br J Anaesth 47:977, 1975.
28. The American Society of Anesthesiologists classification

of physical status. Amendment by the house of delegates of the American Society of Anesthesiologists. Anesthesiology 24:111, 1963.
29. Shapiro HM: Intracranial hypertension: Therapeutic and anesthetic considerations. Anesthesiology 43:445, 1975.
30. Kirkendall WM, Burton AC, Epstein FH, et al: Recommendations for human blood pressure determination by sphygmomanometers. American Heart Association, New York, 1967.
31. Cliffe P: Transducers for measurement of pressure. In Scurr C, Feldman S (eds): Scientific foundations of anaesthesia. London, William Heinemann Medical Books, 1970.
32. Moss AJ, Leibling W, Adams FH: Flush method for determining blood pressure in the first year of life. Pediatrics 21:950, 1958.
33. Hernandez A, Goldring D, Hostmann AF: Measurement of blood pressure in infants and children by the doppler ultrasonic technique. Pediatrics 48:788, 1971.
34. Allen EV: Throboangitis oblitrans: Methods of diagnosis of occlusive arterial lesions distal to the wrist with illustrative cases. Am J Med Sci 178, 1929.
35. Civetta JM, Gabel JC, Laver MB: Disparate ventricular function in surgical patients. Surg Forum 22:136, 1971.
36. Nunn JF, Freeman J: Problems of oxygenation and oxygen transport during haemorrhage. Anaesthesia 19:206, 1964.
37. McConn R: The oxyhemoglobin dissociation curve in acute disease. Surg Clin NA 55:627, 1975.
38. Quist J, Pontoppidan H, Wilson RS et al: Hemodynamic response to mechanical ventilation with PEEP, the effect of hypervolemia. Anesthesiology 42:45, 1975.
39. Kumar A, Pontoppidan H, Baratz RA et al: Inappropriate response to increased plasma ADH during mechanical ventilation in acute respiratory failure. Anesthesiology 40:215, 1974.
40. Carter NW, Rector FC, Seldin DW: Hyponatremia in cerebral disease resulting from inappropriate secretion of antidiuretic hormone. N Engl J Med 264:67, 1961.
41. Becker RM, Daniel RK: Increased antidiuretic hormone production after trauma to the craniofacial complex. J Trauma 13:112, 1973.
42. Mangos JA, Lobeck CC: Studies of sustained hyponatremia due to central nervous system infection. Pediatrics 34:503, 1964.
43. Rivot RL, Sigler MH: Hyponatremia with herpes simplex encephalitis: Possible relationship of limbic lesions and ADH secretion. Arch Neurol 10:595, 1964.
44. Joynt RL, Afifi A, Harison J: Hyponatremia in subarachnoid hemorrhage. Arch Neurol 13:633, 1965.
45. King LR, Knowles HC, McLaurin RL, Lewis HP: Glucose tolerance and plasma insulin in cranial trauma. Ann Surg 137:337, 1971.
46. McLaurin RL, King LR: Recognition and treatment of metabolic disorders after head injuries. Clin Neurosurg 19:281, 1972.
47. Attar S, Boyd D, Layne E, et al.: Alteration in coagulation and fibrinolytic mechanisms in acute trauma. J Trauma 9:39, 1969.
48. Innes D, Sevitt S: Coagulation and fibrinolysis in injured patients. J Clin Pathol 17:1, 1964.
49. Sevitt S: The boundaries between physiology, pathology and irreversibility after injury. Lancet 2:1203, 1968.
50. Keimowitz RM, Annis BL: Disseminated intravascular coagulation associated with massive brain injury. J Neurosurg 39:178, 1973.
51. Minna JD, Robboy SJ, Colman RW: Disseminated Intravascular Coagulation in Man. Charles C Thomas, Springfield Il, 1974.
52. Colman RW, Robboy SJ: Postoperative disseminated intravascular coagulation and fibrinolysis. Urol Clin North Am 3:379, 1976.
53. Pitney WR: Disseminated intravascular coagulation. Sem Haematol 8:65, 1971.
54. Bryan-Brown CW, Savitz MH, Elwyn DH, et al: Cerebral edema nonresponsive to conventional therapy in neurosurgical patients with unsuspected nutritional failure. Crit Care Med 1:1125, 1973.
55. Cuthbertson DP, Tilstone WJ: Metabolism in the post injury period In Bodansky O, Stewart CP (eds): Advances in Clinical Chemistry. Academic Press, New York, 1969.
56. Owen JA: Role of endocrine changes in the plasma protein response to injury. In Bodansky O, Stewart CP (eds): Advances in Clinical Chemistry Academic Press, New York, 1969.
57. Lee HA: Fluid balance and parenteral feeding. In Gray C, Nunn, Utting JE (eds): General Anaesthesia. London, Butterworths, 1980.
58. Caronna JJ, Leigh J, Show D, et al: The outcome of medical coma: Prediction by bedside assessment of physical signs. Am Neurol Ass 32:349, 1975.
59. Marsh ML, Marshall LF, Shapiro HM: Neurosurgical intensive care. Anesthesiology 47:149, 1977.
60. Jennett B, Teasdale G, Galbsaith S, et al.: Severe head injury in three countries. J Neurol Neurosurg Psychiatry 40:291, 1977.
61. Bates D, Caronna JJ, Cartildge NEF et al: A prospective study of nontraumatic coma: Methods and results in 310 patients. Ann Neurol 2:211, 1977.
62. Trojaborg W, Boysen G: Relation between EEG, regionalcerebral blood flow and internal carotid artery pressure during carotid endarterectomy. Electroencephal Clin Neurophysiol 34:61, 1973.
63. Coons RE, Keats AS, Cooley DA: Significance of electroencephalographic changes occurring during cardiopulmonary by-pass. Anesthesiology 20:804, 1959.
64. Hughes J: Limitations of the EEG in the diagnosis of brain death. In Korein J (ed): Brain death, interrelated medical and social issues. Ann NY Acad Sci 315:121, 1978.
65. Prior PF: the EEG in acute cerebral anoxia. Excerpta Medica, Amsterdam, 1973.
66. Bickford RG, Billinger TW, Fleming RA, et al: The compressed spectral array (CSA): A pictorial EEG. Proc San Diego Biomed Suppl 11:365, 1972.
67. Fleming RA, Smith N Ty: On line representation of three variable data. Proc San Diego Biomed Suppl 13:199, 1974.
68. Bleyaert A, Safar P, Nemoto RM, Stezoski SW: Blood pressure control after cardiac arrest. Crit Care Med 6:92, 1978.

69. Katzman R, Clasen R, Klatzo I, Meyer JS, et al: Brain edema in stroke. Stroke 8:512, 1977.
70. Kovach AG, Sandor P: Cerebral blood flow and brain function during hypotension and shock. Ann Rev Physiol 38:571, 1976.
71. Cotrell JE, Patel K, Tundorf H, Ransohoff J: Intracranial pressure changes induced by sodium nitroprusside in patients with intracranial mass lesions. J Neurosurg 48:329, 1978
72. Lundberg N, Kjallquist A, Bien C: Reduction of increased intracranial pressure by hyperventilation. A therapeutic aid in neurological surgery. Acta Psychiatr Scand (suppl) 139:1, 1959.
73. Katsurada K, Sugimoto T, Ouji Y: Significance of cerebrospinal fluid bicarbonate ions in the management of patients with cerebral injury. J Trauma 9:799, 1969.
74. Gordon E, Rossanda M: Further studies on cerebrospinal fluid acid-base status in patients with brain lesions. Acta Anaesthesiol Scand 14:97, 1970.
75. Brock M, Hadjidimos AA, Schurmann K: Possible adverse effect of hyperventilation on rCBF during the acute phase of total proximal occlusion. In Brock C, Fieschhi, Ingvar DH (eds): Cerebral Blood Flow. Berlin, Springer, 1969, p 254.
76. Jannes HE, Langfitt TW, Kumar VS: Analysis of the response of therapeutic measures to reduce intracranial pressure in head injured patients. J Trauma 16:437, 1976.
77. Lassen NA, Christansen MS: Physiology of cerebral blood flow. Br J Anaesth 48:719, 1976.
78. Escuret E, Roquefeuil B, Frerebeu P et al: Effect of hyperventilation associated with administration of central nervous depressants in brain injuries. In Ingvar DH, Lassen NA (eds): Cerebral Function, Metabolism and Circulation. Copenhagen, Munksgaard, 1977, p 154.
79. Carlesson A, Lindqvist M: Effect of chlorpromazine or haloperidol on formation of 3-methoxytyramine and normetanephrine in mouse brain. Acta Pharmacol Toxicol 20:140, 1963.
80. Bleyeart A, Safar P, Stezoski W, et al: Amelioration of post ischemic brain damage in the monkey by immobilization and controlled ventilation. Crit Care Med 6:112, 1978.
81. Classen RA, Pandolfi S, Laring J, et al: Experimental study of relation of fever to cerebral edema. J Neurosurg 41:567, 1974.
82. Siebke H, Rod T, Breivik H, et al: Survival after 40 minutes submersion without cerebral sequelae. Lancet 1:1275, 1975.
83. Albin MS: Resuscitation of the spinal cord. Crit Care Med 6:270, 1978.
84. Miller HD, Leech P: Effects of mannitol and steroid therapy on intracranial volume-pressure relationship in patients with cerebral edema. J Neurosurg 42:274, 1975.
85. Leiberman A, Brun VL, Glass P, et al: Use of high dose corticosteroids in patients with inoperable brain tumors. J Neurol Neurosurg Psychiatry 40:678, 1977.
86. Demopoulos HB, Milvy P, Kakari S, Ransohoff J: Molecular aspects of membrane structure in cerebral edema. In Reulen HJ, Schurmann K (eds): Steroids and Brain Edema. New York, Springer-Verlag, 1972.
87. Martins AN, Ramirez A, Soloman LS, Wiese GM: The effect of dexamethasone on the rate of formation of cerebrospinal fluid in the monkey. J Neurosurg 41:550, 1974.
88. Bell WE, McCormick WF: Increased intracranial pressure in children. In Schaffer AJ, Markowitz M (eds): Major Problems in Clinical Pediatrics. Philadelphia, Saunders, 1978, p 23.
89. Pappius HM, Dayes LA: Hypertonic urea: Its effects on the distribution of water and electrolytes in normal and edematous tissue. Arch Neurol (Chicago) 13:395, 1965.
90. Marshall LF, Smith RW, Rauscher LA, Shapiro HM: Mannitol dose requirement in brain injured patients. J Neurosurg 48:169, 1978.
91. Cantore G, Guidetti B, Virno M: Oral glycerol for the reduction of intracranial pressure. J Neurosurg 21:278, 1965.
92. Hagnvik K, Gordon E, Lins L, et al: Glycerol—induced hemolysis with hemoglobinuria and acute renal failure. Lancet 1:75, 1974.
93. Clasen RA, Pandofli S, Casey D: Furosemide pentobarbital in cryogenic cerebral injury and edema. Neurology (Minneap) 24:642, 1974.
94. Cotrell JE, Robustelli A, Post K, Turndorf H: Furosemide and mannitol—induced changes in serum osmolality and electrolytes. Anesthesiology 47:28, 1977.
95. Goldstein A, Wells BA, Keats AS: Increased tolerance to cerebral anoxia by pentobarbital. Arch Int Pharmacodyn Ther 161:138, 1966.
96. Yatsu FM, Diamond I, Gwazina C, et al: Experimental brain ischemia, protection from irreversible damage with rapid-acting barbiturate (methohexital). Stroke 3:726, 1972.
97. Bleyaert AL, Nemoto EM, Safar P, et al: Thiopental amelioration of brain damage after global ischemia in monkeys. Anesthesiology 49:390, 1978.
98. Kety SS, Woodford RB, Harmel MH, et al: Cerebral blood flow and metabolism in scizophrenia, the effect of barbiturates seminarcosis, insulin coma, and electroshock. Am J Psychiatry 104:765, 1948.
99. Shapiro HM, Galindo A, Wyte SR, Harris AB: Rapid intraoperative reduction of ICP with thiopentone. Br J Anesth 45:1057, 1973.
100. Marshall LF, Smith RW, Shapiro HM: The outcome with aggressive treatment in severe head injuries. I. Significance of intracranial pressure monitoring. II. Acute and chronic barbiturate administration in the management of head injury. J Neurosurg 50:20, 1979.
101. Haff JT, Pitts LH, Spetzler K, Wilson CB: Barbiturates for protection from cerebral ischemia in aneurysm surgery. In Ingvar DH, Lassen NA (eds): Cerebral Function, Metabolism and Circulation. Copenhagen, Munksgaard, 1977, p 158.
102. Marshall LF, Shapiro HM, Rauscher LA, Kaufman NM: Pentobarbital therapy for intracranial hypertension in metabolic coma. Crit Care Med 6:1, 1978.
103. Miller JD: Barbiturates and raised intracranial pressure. Ann Neurol 6:189, 1979.
104. Trauner DA: Treatment of Reye syndrome. Ann Neurol 7:2, 1980.
105. Crocker JFS, Bagnell PC: Reye's syndrome: A clinical review. Can Med Assoc J 124:375, 1981.

Index