N. R. Galloway

Common Eye Diseases and their Management

Springer-Verlag Berlin Heidelberg GmbH

N.R. Galloway

Common Eye Diseases and their Management

With 119 Figures

Springer-Verlag Berlin Heidelberg GmbH
1985

N.R. Galloway, BA, MB, ChB, DO, FRCS, MD
Consultant Ophthalmologist and Clinical Teacher in Ophthalmology, Nottingham University Medical School, Nottingham

ISBN 978-3-540-13659-0 ISBN 978-1-4471-3521-0 (eBook)
DOI 10.1007/978-1-4471-3521-0

Library of Congress Cataloging in Publication Data
Galloway, N.R.
Common eye diseases and their management.
Includes bibliographies and index. 1. Eye – Diseases and defects. I. Title.
[DNLM: 1. Eye Diseases. WW 140 G174c] RE46.G34 1984 617.7 84-13944

Originally published by Springer-Verlag Berlin Heidelberg New York in 1985

Filmset by Herts Typesetting, 84 Fore Street, Hertford.
Printed by Page Bros (Norwich) Limited, Mile Cross Lane, Norwich.

2128/3916-543210

Preface

This book is intended for the medical student and non-specialist postgraduate. It is being published at a time when hospital waiting lists for eye care in Great Britain have reached record levels and the need for more teaching at medical student level is becoming imperative. In many medical schools the students have very little exposure to the specialty; some students none at all. This may be because in past years the emphasis has been on teaching ophthalmology at a postgraduate level and a considerable development of postgraduate teaching has occurred. It had been inferred that more exposure of qualified general practitioners to the subject would reduce the hospital waiting lists since more eye care would take place in general practice. It would appear that the reverse has taken place: general practitioners, being now more aware of eye problems, send even more cases to the hospitals. In the absence of an expansion of the hospital service there is an urgent need to improve primary care ophthalmology and this can best be done by placing more emphasis on the teaching of medical students.

It is against this background that the need to provide a new basic textbook in ophthalmology arises. The book is, therefore, aimed primarily at the medical student. It does, however, aim to make preliminary reading for the postgraduate student. The approach has been essentially clinical, with the aim of convincing the student of an important truth: ophthalmic practice is highly effective in the curing of eye diseases and the restoration of eyesight. Some revision of the anatomy and physiology of the eye has been included but only in relation to disease states. This applies particularly to the optics, a branch of physics sometimes shunned by the medical student, and here only the necessary details which will further an understanding of the relevant associated diseases are included. Although the book is intended to be clinical and hence practical, it is in no way a surgical manual, but in so far as some of the simpler techniques could quite easily be performed in general practice, they are described in more detail.

The selection of material for the book has been influenced by my own experience of common misunderstandings of medical students, and some of the chapters have been adapted fairly closely from my own regular lectures. These have been modified from time to time according to 'feedback' from the students themselves.

Ophthalmology was at one time practised exclusively in isolated specialised hospitals, but the scene is now being shifted to the new style district general hospitals where the practice is more in association with other disciplines. The need to see the eye as part of a human being rather than an isolated organ is emphasised, and attention is drawn to the links between ophthalmology and other specialties.

It has been a deliberate policy to write this book in a repetitive manner so that previously mentioned facts keep reappearing in the text. At the same time the need for brevity in a book of this kind has led to the omission of some rarer conditions, a knowledge of which has little practical value. To balance this, the more common and important eye conditions have been explained at some length.

1984 N.R. Galloway

Acknowledgements

I would like to thank my secretary, Mrs A. Padgett, for all the hard work that she has contributed in assembling the manuscript. My thanks are also due to Mr Michael English, Senior Registrar, Eye Department, University Hospital, Nottingham, for his reading of the text and his helpful suggestions. Professor A.T. Birmingham has also helped with reading and elimination of errors, and I am grateful to Dr J.F. Conway, General Practitioner, for useful discussions. My special thanks are due to Mr Geoffrey Lyth of the Audio-Visual Department for his drawings, but also to the rest of the staff of the department for their help with photographs. I would like to thank many generations of medical students at Nottingham University who have, by their questions and comments, helped to mould the shape of the book. Finally I must acknowledge the unfailing patience and constant encouragement of Mr Michael Jackson of Springer.

Contents

Section I

Introducing the Eye

1 The Scope of Ophthalmology

Those who regard science as the study of more and more about less and less will find that medicine is no exception to this process of specialisation. It is mainly by the intensive study of minutiae that studies can become effective and lead to the prevention and cure of disease. The eye and its surrounding structures provide us with an ideal terrain for this type of specialisation. The importance of the eye and its function in our daily lives is sometimes underrated, but a consideration of the part played by vision in our consciousness soon makes us realise the value of vision. If we think of dreams, memories, of photographs and of almost anything in our daily existence, it is difficult to express them without visual references. After a little careful consideration of the meaning of blindness, it is easy to sense the rational and irrational fears that our patients present in daily life. In a modern European community the effects of blindness are not so apparent as in former years, and blind people tapping their way around streets or begging for food are less in evidence to remind us of the deprivation which they suffer. This is due to the effective application of preventive medicine and to the efficacy of modern surgical techniques.

The broad and detailed scientific interest in the eye and vision is witnessed by the vast number of journals which are available, possibly more than in any other specialty. There are several hundreds of ophthalmological journals all contributing to the scientific literature of the subject. As an organ of specialisation, the eye has another advantage; it can be seen. Using the slit-lamp microscope it is possible to examine living nerves, including central nervous system tissues and blood vessels, in a manner which is not possible in other parts of the body without biopsy. So much are the component parts of the eye on display to the clinician that when a patient presents to an eye casualty department with symptoms, the explanation of the symptoms should be made evident by careful examination. Compare this with the vague aches and pains which present to the gastroenterologist or the neurologist, symptoms which ultimately resolve without any cause being found for them. The student or newly qualified doctor must be warned that if the patient presents with eye symptoms and no abnormality can be found after examination, then he must look again as it is likely that something has been missed. If this warning could be heeded, then undoubtedly much more eye disease and visual disability could be prevented.

Most of the work of the ophthalmologist is necessarily centred around the globe of the eye itself and there are a number of diseases which are limited to this region, sometimes without there being any apparent involvement of the rest of the body. Ophthalmology is usually classed as a surgical specialty but it provides a bridge between surgery and medicine. Most of the surgery is performed under the microscope and there is overlap with the fields of the plastic surgeons and the neurosurgeons. On the medical side, the ophthalmologist has links with the general physicians, especially when concerned with vascular problems in the eye or with diabetes.

Historical Background

Modern clinical ophthalmology can be dated back to the invention of the ophthalmoscope in 1851 by Von Helmholtz. Prior to this, a proper examination of the living organ was impossible and our knowledge of the eye was based on post-mortem studies which could at times be misleading. Even nowadays, the lay idea of an eye is that of a flat unidimensional part of our facial expression; the expressions 'twinkling in an eye' and 'smiling eyes' witness this. Von Helmholtz must have seen the black central pupil of the eye as a hole to inspect rather than as the facial feature which dilates in anger or pain. Today his task seems simple to us: that of peering into a small unilluminated hole. We are all aware of the occasional glimpse of unfocused detail at the back of an eye obtained by viewing a cat in the headlights of a car or of the red reflex seen from the back of an eye in an ill-positioned family photograph. The ophthalmoscope provides a means of illuminating the eye and viewing the contents of the globe along the path of the illuminating light beam. Following the introduction of the ophthalmoscope, large numbers of previously unknown diseases were described; the changes in the eye seen in association with systemic diseases such as hypertension and anaemia became recognised, as were also several blinding conditions limited to the eyes themselves such as glaucoma and macular degeneration.

But we must not belittle developments which had occurred before the invention of the ophthalmoscope. In the 18th century considerable advances had been made in the technique and instrumentation of cataract surgery, and the science of optics was being developed to enable the correction of refractive errors. The development of ophthalmology in the 17th century is revealed in the writings of the famous diarist, Samuel Pepys. Although we have no record of his eye condition other than his own, he did consult an oculist at the time and unfortunately received little comfort or effective treatment. His failing eyesight brought his diary to an abrupt end in spite of the use of 'special glasses' and the medicaments which caused him great pain. Once the ophthalmoscope had been invented, a further series of advances in general medicine were to give a great boost to ophthalmology. The use of cocaine allowed pain-free eye surgery, and more extensive surgery became possible with the introduction of general anaesthesia towards the end of the 19th

century. The use of eserine eyedrops to reduce the intraocular pressure in glaucoma was introduced at the same time, and the sight-saving value of this drug has probably been greatly underestimated. The preventable toll of visual handicap resulting from glaucoma in the elderly is one of the major problems of ophthalmology today. Cataract surgery saw great advances at the beginning of this century with the introduction of the intracapsular cataract extraction. In the 1920s, successful attempts were being made to replace the detached retina, which had previously been an irreversible cause of blindness. Such early surgical techniques have now been developed to produce some of the most dramatic means of restoring sight. As a spin-off from the last war came a revolutionary idea of 'spare parts' surgery in the eye. The observation that crashed fighter pilots were able to tolerate small pieces of perspex in their eyes led to the use of acrylic intraocular implants, the lens of the eye being replaced by an artificial one. Such spare-part surgery has now become commonplace in the elderly, as will be seen in Chapter 10. The operating microscope was introduced in the 1960s, and with it came the development of fine suture materials and the use of instruments too small for manipulation with the naked eye. Until recently the vitreous has been a no-man's land for the ophthalmic surgeon because of the technical difficulties of carrying out procedures within the eye without causing damage. Instruments have recently been developed which can cut, aspirate and inject fluid simultaneously, all these procedures being carried out through a fine-bore needle. This has led to great changes, particularly in the management of diabetic retinopathy.

In the early days of the development of the specialty, a number of specialised eye hospitals were built throughout Britain. The first of these was Moorfields Eye Hospital, founded largely to combat the epidemic of trachoma which was prevalent in London at the time. Subsequently other eye hospitals appeared in the main cities of England, often the result of pressures of local needs such as, for example, the treatment of industrial accidents. In recent years, there has been a tendency for eye departments to become incorporated within the larger general hospital. In theory at least, the system makes better economic sense from the point of view of cost of services to patients and also provides better liaison with other specialties.

2 The Human Eye

The Basic Structure of the Eye (Figs. 2.1 and 2.2)

Anatomy of the Globe

The globe can be considered to be arranged in three layers.

Outer layer

The outer layer is a tough fibrous envelope which consists anteriorly of the cornea and posteriorly of the sclera. The cornea is transparent whereas the sclera, which is continuous with it, is white. The junction of cornea and sclera is known as the limbus. In this region the epithelium on the outer surface of the cornea becomes continuous with the conjunctiva, a thin, loose skin which covers the anterior part of the sclera, from which it is separated by loose connective tissue. Above and below, the conjunctiva is reflected onto the inner surface of the upper and lower lids. This mucous membrane therefore lines the posterior surface of the eyelids and there is a mucocutaneous junction on the lid margin.

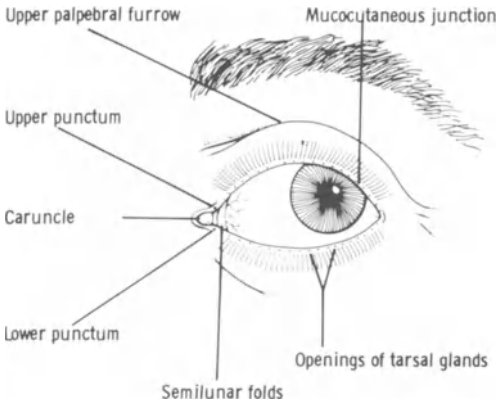

Fig. 2.1. Surface anatomy.

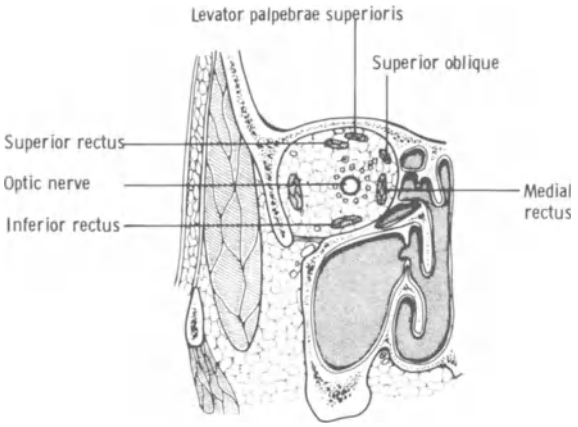

Fig. 2.2. Anatomy of the orbit.

Middle layer

The middle layer is highly vascular. If one were to peel the sclera away from this layer (not an easy task), the remaining structure would resemble a grape since this middle layer, which is called the uvea, is heavily pigmented as well as being very vascular. The anterior part of the uvea forms the bulk of the iris and the ciliary body and hence inflammation of the iris is called either anterior uveitis or iritis. The posterior part of the uvea is called the choroid.

Inner layer

The inner layer of the eye, which lines the vascular uvea, is the neurosensory layer. This layer forms the retina posteriorly, but anteriorly it comes to line the inner surface of the ciliary body and iris as a two-layered endothelium. These same two layers can be traced into the retina, which is composed of an outer

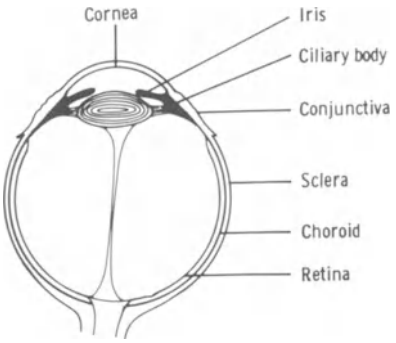

Fig. 2.3. The layers of the globe.

pigment epithelium and an inner sensory part which contains the rods and cones, bipolar cells and ganglion cells (Fig. 2.3).

Blood supply

The blood supply of the globe is derived from three sources: the central retinal artery, the anterior ciliary arteries, and the posterior ciliary arteries. All these are derived from the ophthalmic artery which is a branch of the internal carotid. The central retinal artery runs in the optic nerve to reach the interior of the eye and its branches spread out over the inner surface of the retina supplying its inner half. The anterior ciliary arteries emerge from the insertion of the recti muscles and perforate the globe near the iris root to join an arterial circle in the ciliary body. The posterior ciliary arteries are the fine branches of the ophthalmic artery which penetrate the posterior pole of the eye. Some of these supply the choroid and two or more larger vessels run anteriorly to reach the arterial circle in the ciliary body. The larger vessels are known as the long posterior ciliary arteries, and those supplying the choroid are known as the short posterior ciliary arteries. The branches of the central retinal artery are accompanied by an equivalent vein but the choroid, ciliary body and iris are drained by approximately four vortex veins. These leave the posterior four quadrants of the globe and are familiar landmarks for the retina surgeon (Fig. 2.4).

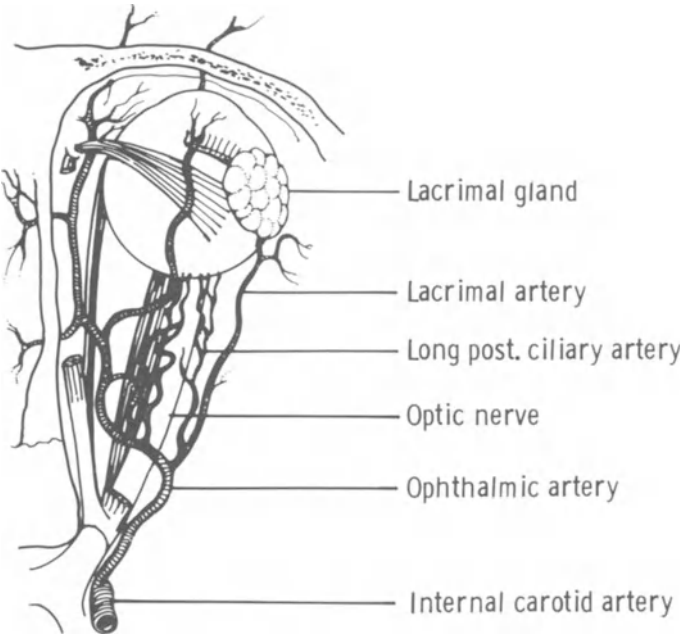

Fig. 2.4. Blood supply of the globe.

Optic nerve

The optic nerve meets the posterior part of the globe slightly nasal to the posterior pole and very slightly above the horizontal meridian. Inside the eye this point is seen as the optic disc. There are no light-sensitive cells on the optic disc — and hence the blind spot which anyone can find in their field of vision. The optic nerve contains about one million nerve fibres, each of which has a cell body in the ganglion cell layer of the retina (Fig. 2.5). Nerve fibres sweep across the innermost part of the retina to reach the optic disc. They can be seen with the ophthalmoscope by carefully observing the way light is reflected off the inner surface of the retina (Fig. 2.6). The retinal vessels are also embedded on the inner surface of the retina. There is therefore a gap, which is the thickness of the transparent retina, between the retinal vessels and the stippled pigment epithelium. Apart from the optic nerve, the posterior pole of the

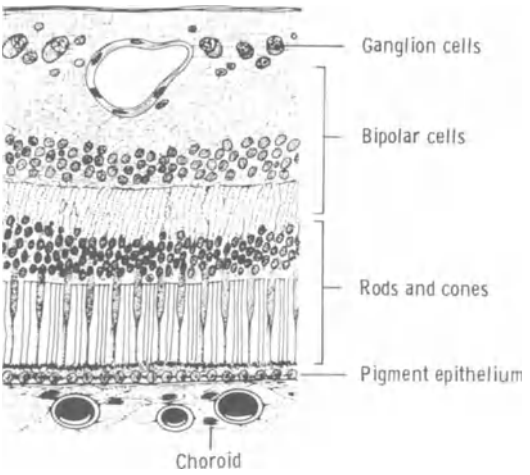

Fig. 2.5. The layers of the retina.

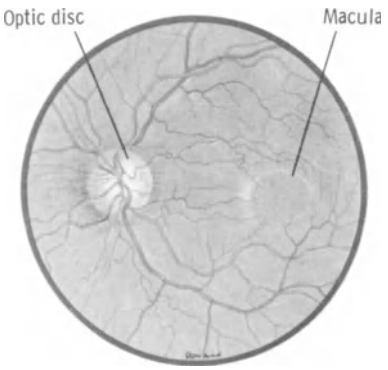

Fig. 2.6. The optic fundus. (With acknowledgements to I.C. Michaelson, 'Textbook of the Fundus of the Eye', Churchill Livingstone, Edinburgh.)

globe is also perforated by several long and short ciliary nerves. These contain parasympathetic, sympathetic and sensory fibres, which mainly supply muscles of the iris (dilator and sphincter) and ciliary body (ciliary muscle). Patients can experience pain when the iris is handled under inadequate local anaesthesia, and pain is also sometimes experienced during laser coagulation treatment of the retina — this would seem to prove the existence of sensory fibres in the iris and retina. The cornea is extremely sensitive, but again the only sensory endings are those for pain.

How to Find Out What a Patient Can See

One obvious way to measure sight is to ask the patient to identify letters which are graded in size. This is the basis of the standard Snellen test for visual acuity (Fig. 2.7). This test only measures the function of a small area of retina at the posterior pole of the eye called the macula. If we stare fixedly at an object, for example a picture on the wall, and attempt to keep our eyes as still as possible,

Fig. 2.7. Snellen test type.

it soon becomes apparent that we can only appreciate detail in a small part of the centre of the field of vision. Everything around us is ill-defined and yet we can detect the slightest twitch of a finger from the corner of our eyes. The macula region is specialised to detect fine detail, and the whole peripheral retina is concerned with the detection of shape and movement. In order to see, we use the peripheral retina to help us scan the field of view. The peripheral retina may be considered as equivalent to the TV cameraman who moves the camera around to the relevant views and allows the camera (or macula) to make sense of the scene. If the macula area is damaged by, for example, senile degeneration, then the patient may be unable to see even the largest print on the test type and yet have no difficulty in walking about the room. Navigational vision is largely dependent on the peripheral field of vision. On the other side of the coin, the patient with marked constriction of the peripheral field of vision but preservation of the central field may behave as though blind. The same patient could read the test chart down to the bottom *once he has found it.* This situation sometimes arises in patients with advanced chronic simple glaucoma.

It should be becoming clear that measuring the visual acuity alone, although very useful, is not an adequate measure of vision. For a proper clinical examination we need to assess the visual acuity, the visual fields and the colour vision. A number of other facets of visual function can also be measured, such as dark adaptation or the perception of flicker.

Visual Acuity

The familiar Snellen chart has one large letter at the top which is designed to be just visible to a normal-sighted person at 60 m. The chart is viewed from a

Fig. 2.8. The Stycar test.

distance of 6 m. If a patient is just able to see this large letter, the vision is recorded as 6/60. Below the large letter are rows of smaller letters decreasing in size down to the bottom. The size of letter normally visible to a normal-sighted person at 6 m is usually on the second-to-bottom line. Patients reading this line are said to have a vision of 6/6. If a patient cannot read the top letter, he is taken nearer to the chart. If the top letter becomes visible at 3 m, the acuity is recorded as 3/60. If the letter is still not visible, the patient is asked if he can count fingers (recorded as 'CF') and, failing this, if he can see hand movements ('HM'). Finally, if even hand movements are not seen, the ability to see a light is tested ('P of L').

Young children and illiterates can be asked to do the 'E' test in which they must orient a large wooden letter E so that it is the same way up as an indicated letter E on a chart. Perhaps better than this is the Stycar test (Fig. 2.8) in which the child is asked to point at the letter on a card which is the same as the one held up at 6 m. Other ways of measuring visual acuity are discussed in Chapter 16.

Visual Field

Some measurements of the visual field can be made by sitting facing the patient and asking if the movement of one's fingers can be discerned. The patient is instructed to cover one eye with a hand and the observer also covers

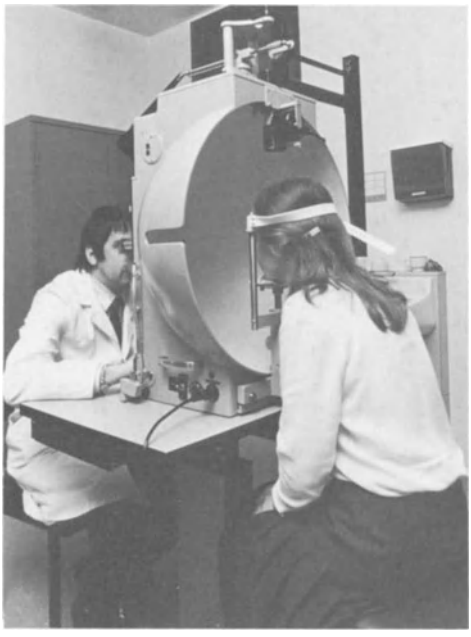

Fig. 2.9. The Goldmann perimeter.

one of his eyes so that he can check the patient's field against his own. The test can be made more accurate by using a pin with a red head on it as a target.

None of these confrontation methods can match the accuracy of formal perimetry. A number of specialised instruments of varying complexity are available. Using such equipment, the patient is presented with a number of different-sized targets in different parts of the visual field, and a map of the field of vision is charted. An accurate map of the visual field is often of great diagnostic importance. In the past it has been customary to map out the central part of the visual field using the Bjerrum screen, and the peripheral field using a perimeter. The Goldmann perimeter is now widely used; this instrument allows both central and peripheral fields to be plotted out on one chart (Fig. 2.9).

Colour Vision

The Ishihara plates provide a popular and effective method for screening for colour vision defects (Fig. 2.10). The patient is presented with a series of plates on which are printed numerous coloured dots. The normally sighted subject will see numbers on the majority of the plates, whereas the colour-defective patient will fail to see many of the numbers. The test is easy to do and will effectively screen out the more common red–green deficiency found in 8% of the male population. Other tests, such as the Farnsworth 100 Hue test, are available for the more detailed analysis of colour vision defects.

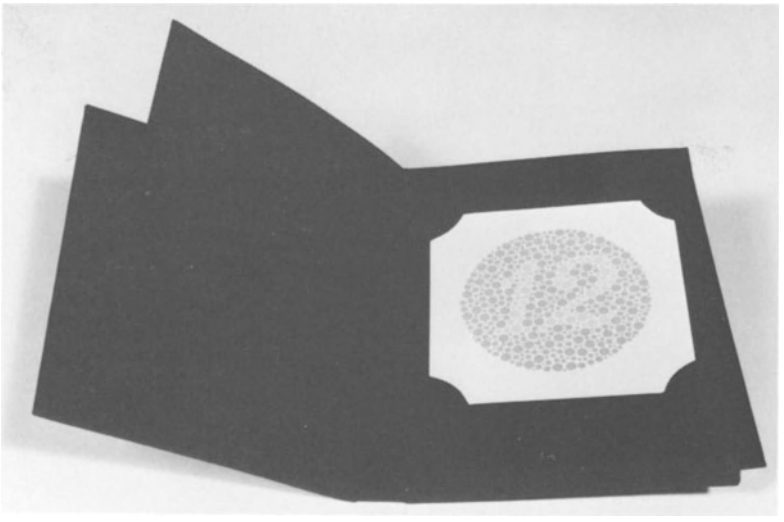

Fig. 2.10. Ishihara test for colour vision.

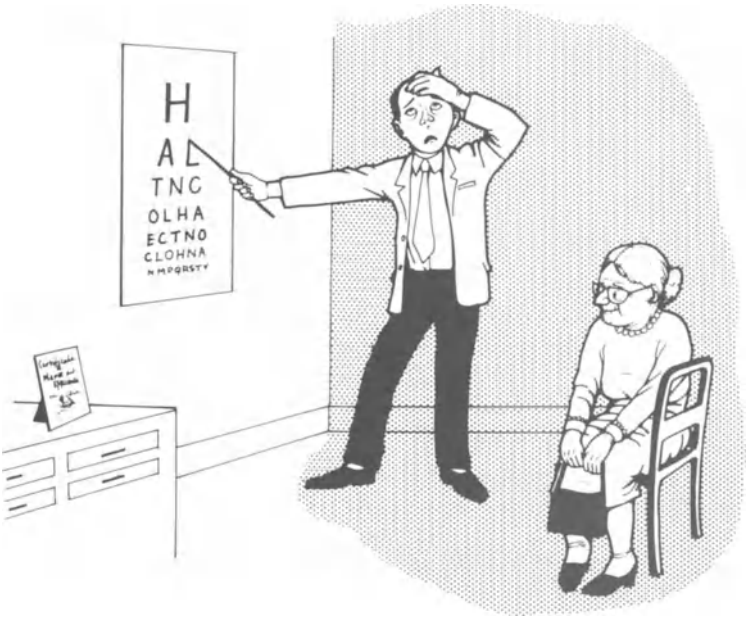

Fig. 2.11. "I borrowed my husband's glasses...."

Spectacles

Measurement of the visual acuity may not be valid unless the patient is wearing the correct spectacles. Some patients when asked to *read* a Snellen chart will put on their reading glasses. Since these glasses are designed for close work, the chart may be largely obscured and the uninitiated doctor might be surprised at the poor level of visual acuity (Fig. 2.11). If the glasses have been left at home, long sight or short sight can be largely overcome by asking the patient to view the chart through a pinhole. In an ophthalmic department a check of the spectacle prescription is a routine part of the initial examination. Figure 2.12 shows how the converging power of the optical media and the length of the eye are mismatched to produce the need to wear spectacles.

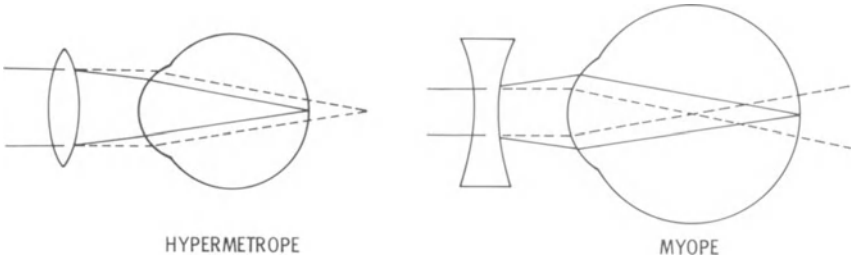

Fig. 2.12. Optical defects of the eye.

How to Start Examining an Eye

External Eye and Lids

The eyelids should be inspected to make sure that the lid margins and puncta are correctly apposed to the globe and that there are no ingrowing lashes. Rodent ulcers on the skin of the lids can easily be missed, especially if obscured by cosmetics. The presence of ptosis should be noted and the ocular movements assessed by asking the patient to follow a finger upwards, downwards and to each side. Palpation of the skin around the eyes may reveal an orbital tumour or a swollen lacrimal sac. Palpation with the end of a glass rod is sometimes useful to find points of tenderness when the lid is diffusely swollen. Such tenderness can indicate a primary infection of a lash root or the lacrimal sac. Both surfaces of the eyelids should be examined. The inside of the lower lid can easily be inspected by pulling down the skin of the lid with the index finger. The upper lid can be everted by asking the patient to look down, grasping the lashes gently between finger and thumb, and rolling the lid margins upwards and forwards over a cotton-wool bud or glass rod. The lid will usually remain in this everted position until the patient is asked to look up. Foreign bodies quite often lodge themselves under the upper lid and they can only be removed by this means. As a general rule, if a patient complains that there is something in his eye, there usually is, and if you find nothing, it is necessary to look again more closely or refer the patient for microscopic examination. A feeling of grittiness may be due to inflammation of the conjunctiva and this may be accompanied by evidence of purulent discharge in the lashes. The presence of tear overflow and excoriation of the skin at the outer canthus should also be noted.

The Globe

Much ophthalmic disease has been described and classified using the microscope. In spite of this, many of the important eye diseases can be

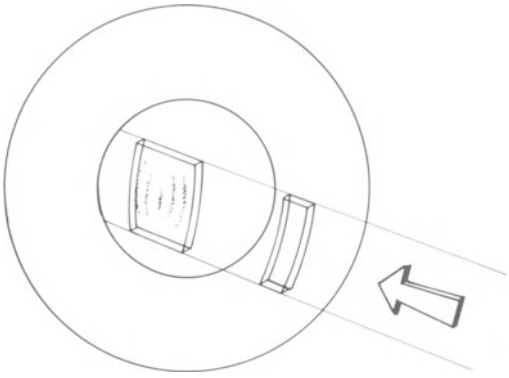

Fig. 2.13. Focal illumination.

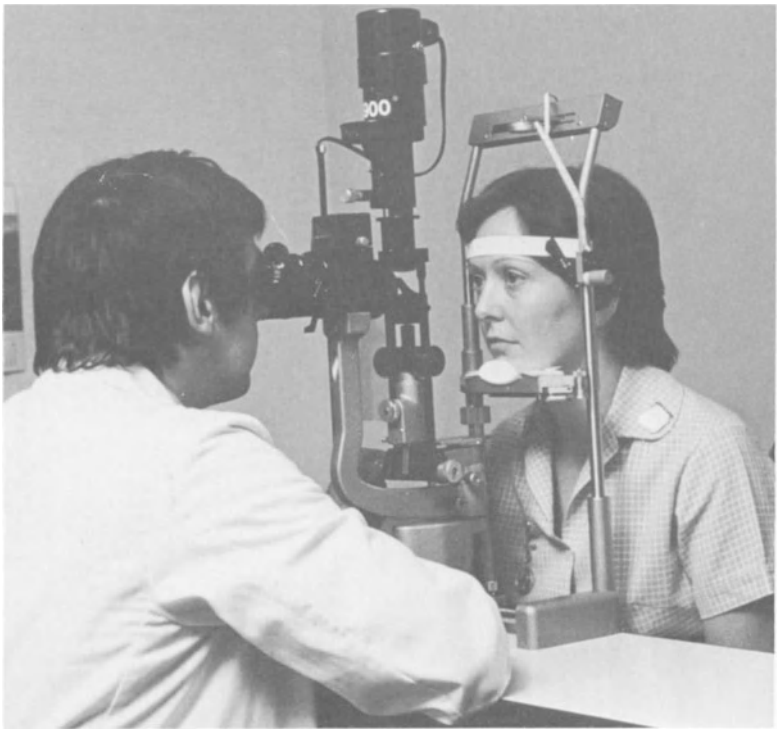

Fig. 2.14. The slit-lamp microscope.

diagnosed using a hand magnifier and an ophthalmoscope. At this point it is important to understand the principle of examining the eye with a focused beam of light. If a pencil of light is directed obliquely through the cornea and anterior chamber, it can be made to illuminate structures or abnormalities otherwise invisible. One might inspect the glass sides and water of a fish tank using a strong, focused torch in the same manner (Fig. 2.13). Many ophthalmoscopes incorporate a focused beam of light which can be used for this purpose. The principle has been developed to a high degree in the slit lamp (Fig. 2.14). This instrument allows a focused slit of light to be shone through the eye, which can then be examined by a binocular microscope. By this means an optical section of the eye can be created. The method can be compared with making a histological section where the slice of tissue is made with a knife rather than a beam of light. The slit lamp is sometimes called the biomicroscope. By means of such optical aids, the cornea must be carefully inspected for scars or foreign bodies. The presence of vascular congestion around the corneal margin may be of significance. A routine should be developed for examining the pupil reactions. The patient is asked to look at a distant object and the size of each pupil noted. The light of the torch is then presented to each eye, approaching from the side, and the direct reaction to

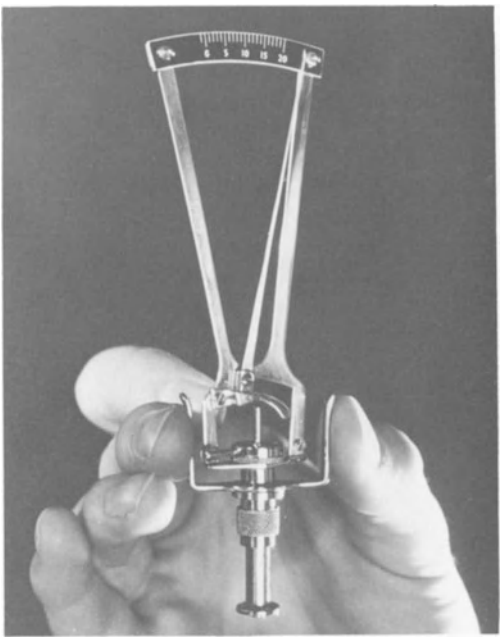

Fig. 2.15. The Schiøtz tonometer.

light noted. The procedure is repeated, but this time the reaction of the opposite pupil — the consensual reaction — is noted. Finally the patient is asked to look at one's finger to obtain the reaction to convergence. Closer inspection of the iris may show that it is atrophic or fixed by adhesions. Turbidity or cells in the aqueous may be seen in the beam of the inspection light. The lens and anterior parts of the vitreous can be examined by the same means.

Once the anterior segment of the eye has been examined, the intraocular pressure is measured. The cheapest way to do this outside an eye department is to use the Schiøtz tonometer (Fig. 2.15); (for the method see Chapter 11). Applanation tonometry is more accurate and although this instrument is normally attached to the slit lamp, a hand-held instrument is now available (Fig. 2.16).

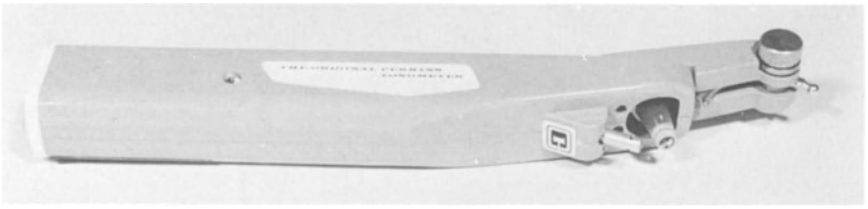

Fig. 2.16. The Perkins' applanation tonometer.

At this stage the pupil may be dilated for better examination of the fundi and optical media. A short-acting mydriatic is preferable, e.g. tropicamide 1% (Mydriacyl). These particular drops take effect after 10 minutes and take 4 or 5 hours to wear off. Patients should be warned that their vision will be blurred and that they will be more sensitive to light over this period. Most people find that their ability to drive a car is unimpaired, but there is a potential medicolegal risk if the patient subsequently has a car accident. Once the pupils have been dilated, the eye can then be examined with the ophthalmoscope.

How to Use the Ophthalmoscope

Before the middle of the last century, nobody had seen the inside of a living eye and much of the science of medical ophthalmology was unknown. In 1851 Hermann von Helmholtz introduced his ophthalmoscope and it rapidly became used in clinics dealing with ophthalmological problems. The task of Helmholtz was to devise a way of looking through the black pupil and at the same time illuminate the interior of the globe. He solved the problem by arranging to view the fundus of the eye through an angled piece of glass. A light projected from the side was reflected into the eye by total internal reflection. Most modern ophthalmoscopes employ an angled mirror with a small hole in it to achieve the same end. They also incorporate a series of lenses which can be interposed between the eye of the patient and that of the observer, thereby overcoming any refractive problems which may defocus the view. These lenses are positioned by rotating a knurled wheel on the side of the ophthalmoscope. A number on the face of the instrument indicates the

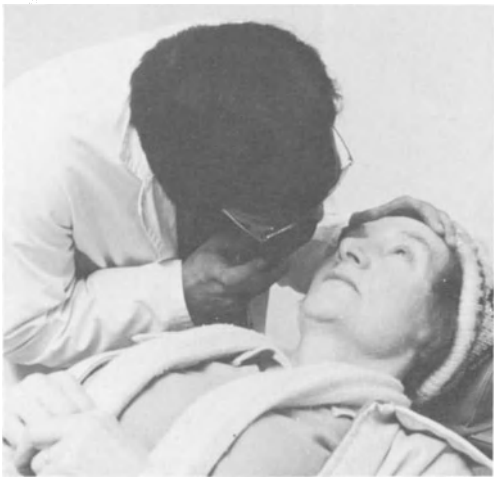

Fig. 2.17. Direct ophthalmoscopy.

strength of the lens. When choosing an ophthalmoscope, it is worth remembering that large ones take larger batteries which last longer (or, better still, they may have rechargeable batteries); small ophthalmoscopes are handy for the pocket. Some ophthalmoscopes have a wider field of view than others and this is an advantage when learning to use the instrument.

If examining the patient's right eye, it is best to hold the ophthalmoscope in the right hand and view through one's own right eye. A left eye should be viewed through the left eye using the left hand (Fig. 2.17). It is best if the patient is seated and the doctor is standing. The first thing to observe is the red reflex, which simply refers to the general reddish colouring seen through the pupil. If viewed from about 30 cm away from the eye, very slight and subtle opacities or defects in the optical media may be seen, against the background of the red reflex. The patient's eye must always be brought into focus by rotating the lens wheel on the ophthalmoscope.

Having observed the red reflex, the eye can be approached closely and the focus of the ophthalmoscope adjusted so that fundus detail becomes visible. It is best to look for the optic disc first, remembering its position nasal to the posterior pole and slightly above the horizontal meridian. The patient should be asked to look straight ahead at this point. The important points to note about the disc are the clarity of the margins, the colour, the nature of the central cup, the vessel entry and the presence or absence of haemorrhages. Once the disc has been examined carefully, the vessels from the disc can be followed. For example, the upper temporal branch vessels can be followed out to the periphery and back, then the lower temporal branch vessels, then the upper nasal and finally the lower nasal. Having examined the vessels, ask the patient to look directly at the ophthalmoscope light and the macula region should come into view. At first this may look unremarkable, like a minute dot of light which follows our own light. More careful examination will reveal that it has a yellowish colour. To obtain a better view of the macula it is usually

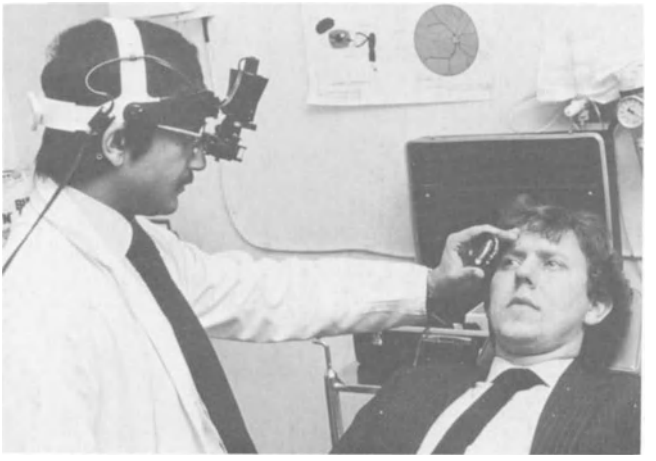

Fig. 2.18. Indirect ophthalmoscopy.

necessary to examine it with a special attachment on the slit-lamp microscope, the Hruby lens. A fundus photograph is also very helpful. After viewing the macula the general fundus background should be observed. The appearance here depends upon the complexion of the patient: in a lightly pigmented subject it is possible to see through the stippled pigment epithelium and obtain an indefinite view of the choroidal vasculature; in heavily pigmented subjects the pigment epithelium is uniformly black and prevents any view of the choroid which lies behind it. Finally, the peripheral fundus can be inspected by asking the patient to look to the extremes of gaze and by refocusing the ophthalmoscope. Examining the peripheral fundus demands some special skill, even with the ordinary ophthalmoscope, but it is best seen using the triple mirror gonioscope. This is a modified contact lens which has an angled mirror attached to it. A view through this mirror is obtained using the slit-lamp microscope.

There are a number of other methods of examining the fundus. The ophthalmoscope described above is known as the direct ophthalmoscope. Shortly after the introduction of direct ophthalmoscopy, the indirect ophthalmoscope was introduced. If one examines an eye with the pupil dilated through a mirror with a hole in it, the patient being at arm's length from the observer and the mirror being held close to the observer's eye, then the red reflex is seen. If a convex lens is placed in the line of sight about 8 cm from the patient's eye, then, rather surprisingly, a clear wide field inverted view of the fundus is obtained. The view can be made binocular, and the binocular indirect ophthalmoscope is an essential tool of the retina surgeon (Fig. 2.18). If we want a really magnified view of the fundus, then the slit-lamp microscope can be used. However, a special lens must be placed in front of the patient's eye, either in the form of the triple mirror contact lens or the Hruby lens (Fig. 2.19). Another useful way of examining the fundus is by means of

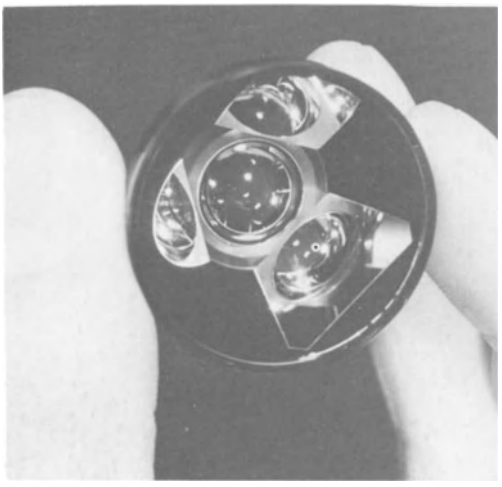

Fig. 2.19. The triple mirror gonioscope.

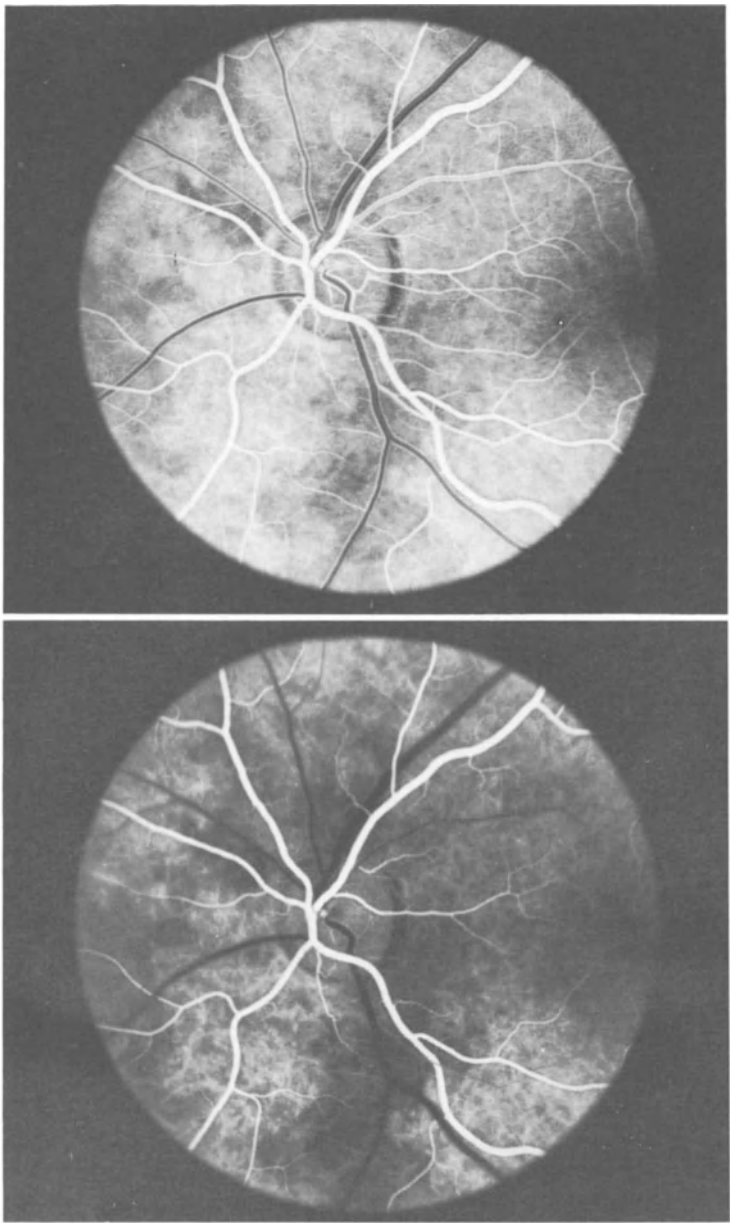

Fig. 2.20. Fluorescein angiogram of normal fundus. Two successive views to show transit of dye through retinal vessels. (With acknowledgement to Mr M.D. Sanders.)

fundus photography. A special type of fundus photograph known as a fluorescein angiogram shows up the retinal vessels, including the capillaries, in great detail. The technique involves taking repeated photographs in rapid succession after the injection of the dye fluorescein into the antecubital vein. The dye in the vessels is selectively photographed by using filters in the camera (Fig. 2.20). Finally, video filming is becoming an important method for observing changing events in the fundus.

Other Tests Available in an Eye Department

Several special tests are available to measure the ability of the eyes to work together. A department, known as the orthoptic department, is usually set aside within the eye clinic for making these tests. When there is a defect of the ocular movements this can be monitored by means of the Hess chart (see chapter on squint, Chapter 13). The ability to use the eyes together is measured on the synoptophore, and any tendency of one eye to turn out or in can be measured with the Maddox rod and Maddox wing test (Fig. 2.21). The use of contact lenses and also of intraocular implants has demanded more accurate measurements of the cornea and of the length of the eye. A keratometer is an instrument for measuring the curvature of the cornea, and the length of the eye can now be accurately measured by ultrasound. If one eye appears to protrude forwards and one wishes to monitor the position of the globes relative to the orbital margin, then an exophthalmometer is used (Fig. 2.22). X-rays of the eye and orbit are still often used. An x-ray is essential if an intraocular foreign body is suspected and it is useful for detecting bony abnormalities in the walls

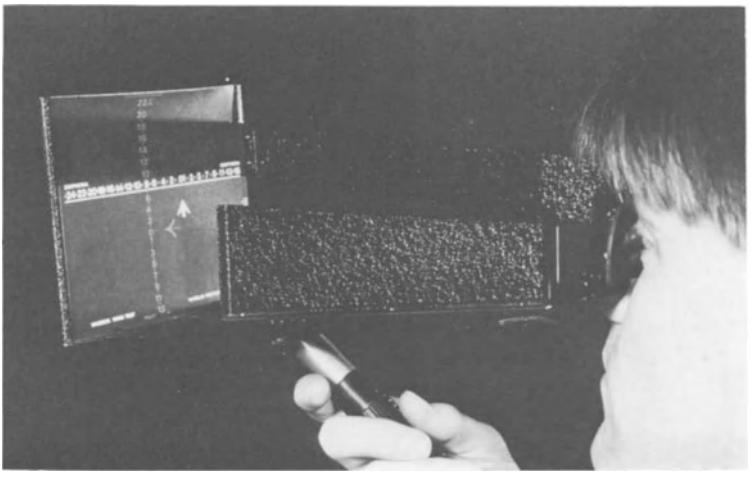

Fig. 2.21. The Maddox wing test.

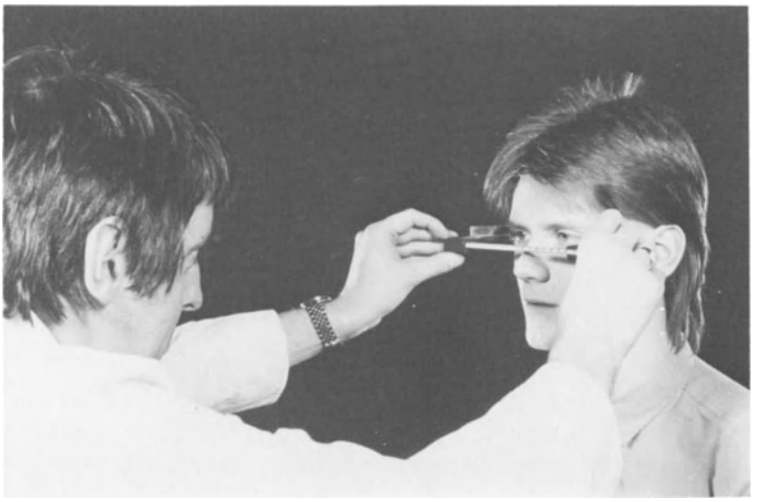

Fig. 2.22. The exophthalmometer.

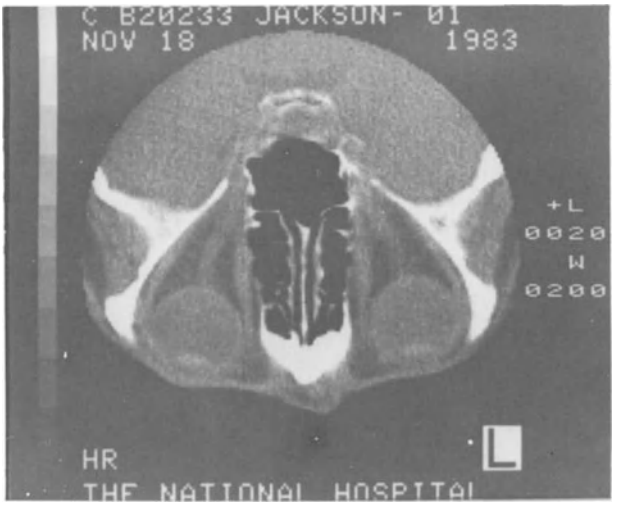

Fig. 2.23. CAT scan of normal human orbits. (With acknowledgement to Mr M.D. Sanders.)

of the orbit due to tumours. Ultrasonography has been mentioned as a technique for measuring the length of the eye; it may also be used to depict tissue planes within the eye, showing, for example, the extent of a retinal detachment or the presence of vitreous membranes. Computer-assisted tomography is becoming an important diagnostic technique, especially for lesions in the orbit (Fig. 2.23). Electroretinography provides a measure of the electrical changes which take place in the retina when the eye is exposed to light. It can indicate retinal function in the same way that the

electrocardiogram indicates cardiac function. The visually evoked potential (VEP) is a measure of minute electrical changes over the back of the scalp which occur when the eyes are stimulated with a flashing light. This test has been shown to be useful in detecting previous damage to the optic nerve in patients with suspected multiple sclerosis.

The eye is probably the most measured organ in the body. There are many other ingenious instruments that are not in regular clinical use but which may be found in the laboratory.

Further Reading

Chawla HB (1981) Essential ophthalmology. Churchill Livingstone, Edinburgh
Glasspool M (1982) Problems in ophthalmology. MTP Press, Lancaster
Harrington DO (1981) The visual fields: a textbook and atlas of clinical perimetry. CV Mosby, St Louis
Miller SJH, ed. (1978) Parsons' diseases of the eye. Churchill Livingstone, Edinburgh
Newell FW (1982) Ophthalmology: principles and concepts, 5th edn. CV Mosby, St Louis

Section II

Primary Care Eye Problems

The aim of this section is to present some of the more commonly occurring eye conditions which are likely to confront a general practitioner in his surgery or a casualty officer in the general or ophthalmic casualty department. Those conditions which would be normally referred for more extensive investigations or treatment are dealt with in subsequent chapters, although their differential diagnoses are considered here.

3 Long Sight, Short Sight

When the patient with an ophthalmological problem first enters the doctor's surgery, it is useful to notice whether he or she is long sighted or short sighted. The long-sighted person tends to have smaller eyes than normal, whereas the short-sighted person tends to have larger eyes than normal. Sometimes this is evident on inspection of the eyes and eyelids, but the long-sighted person will usually be wearing convex or converging lenses and these tend to make the eyes look bigger, whereas the short-sighted person will be wearing concave lenses which make the eyes look smaller (Fig. 3.1). Therefore, glasses which magnify

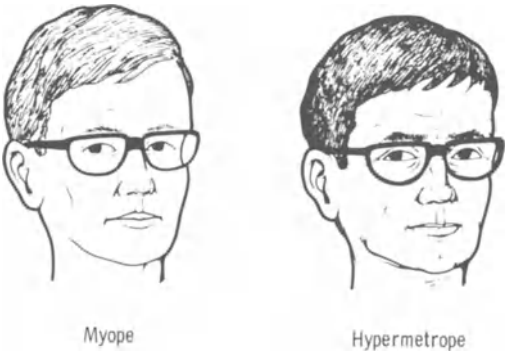

Fig. 3.1. Long sight and short sight. Note line of cheek behind lens.

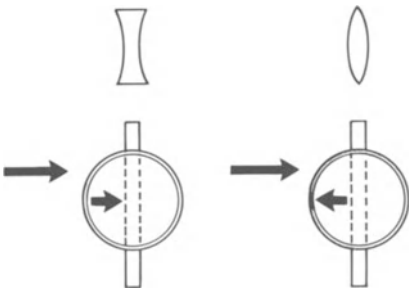

Fig. 3.2. Concave lens 'with'; convex lens 'against'. Try this for yourself in the clinic.

Fig. 3.3. A long-sighted person.

Fig. 3.4. A short-sighted person.

the eyes belong to the hypermetrope. The nature of the spectacle lens can be verified by moving it backwards and forwards in front of one's hand. If the hand appears to move in the opposite direction to that of the movement of the spectacle lens, it is convex (Fig. 3.2). The spectacles of the myopic (or short-sighted) patient contain diverging concave lenses and if these are held in front of one's hand and moved to and fro, the hand appears to move with the direction of the movement of the glasses. After a little practice it is possible to recognise long-sighted or short-sighted patients simply by looking at them, providing the glasses that they are wearing are strong enough. Apart from providing a knowledge of the patient's visual world, this information may be helpful when making a diagnosis.

The long-sighted person is more susceptible to narrow angle glaucoma, especially in late middle age. A number of elderly people are long sighted simply because they have undergone cataract surgery. The optic disc of the long-sighted person tends to be pink with an ill-defined margin and may be smaller than the short-sighted disc. Sometimes long-sighted people have one eye which is weaker than the other, especially when the hypermetropia is unequal on the two sides. The weakness is due to amblyopia of disuse, a common condition which may confuse the diagnosis of other diseases (Fig. 3.3).

Short-sighted patients are more susceptible to retinal detachment, especially in late middle age; they are also more likely to develop cataracts in later years than are people with normal sight (Fig 3.4). Retinal degenerative changes at the posterior pole are another complication of high myopia. The optic disc of the short-sighted person is slightly larger than normal and rather more pale. Its margins are usually well defined. Table 3.1 gives those conditions more commonly associated with either short sight or long sight.

Having observed the nature of the spectacle lenses we have now made a step towards diagnosing the eye condition: if the patient is middle aged and complaining of evening headaches and seeing haloes round street lights, he

Table 3.1. Eye disease and refractive error

Myopia ('short sight')	Hypermetropia ('long sight')
Conditions associated with myopia	*Conditions associated with hypermetropia*
Retinal detachment	Narrow angle glaucoma
Macula haemorrhages	Concomitant squint
Cataract	Amblyopia of disuse
Myopic chorioretinal degeneration	
Mongolism	
Keratoconus (conical cornea)	
Conditions causing myopia	*Conditions causing hypermetropia*
Cataract	Cataract surgery
Diabetes mellitus	Dislocated lens
Congential glaucoma	Macula oedema
Accommodation spasm, or 'pseudomyopia'	Intraocular or orbital tumours
	Retinal detachment

may have subacute glaucoma. If he is elderly, he may have had cataract surgery. Figure 3.4 shows a short-sighted person. If he is complaining of seeing the sudden appearance of numerous black spots combined with flashes of light, he may be about to have a retinal detachment. If he is simply complaining of visual deterioration, it may be the result of cataract formation. The short-sighted person who experiences a sudden loss of reading vision in one eye may have suffered a macula haemorrhage, but he could also have a detached retina.

If we take note of whether a patient is long sighted or short sighted at an early stage, then this information can influence the type of questions that are best asked when taking a history. Furthermore, such information is essential when we look at the fundus. The paler disc of the myope can be mistaken for optic atrophy by the uninitiated and likewise the pink disc with indefinite margins seen in hypermetropes may be mistaken for papilloedema.

4 Common Diseases of the Eyelids

The Watering Eye

Quite often patients present at the clinic or surgery complaining of watering eyes. It may be the golfer whose glasses keep misting up on the fairways or the housewife who is embarrassed by tears dropping onto the food when cooking, or it may be the 6-month-old baby whose eyes have watered and discharged since birth. Sometimes an elderly patient may complain of watering eyes when on examination there is no evidence of tear overflow but the vision has been made blurred by cataracts. Some degree of tear overflow is quite normal in windy weather, and the anxious patient may over-emphasise this; it is important to assess the actual amount of overflow by asking the patient whether it occurs all the time both in and out of doors.

An eye may water because the tears cannot drain away adequately or because there is excessive secretion of tears.

Impaired Drainage

Normally the tears drain through two minute openings at the inner end of the lid margins known as the upper and lower puncta respectively. Most of the

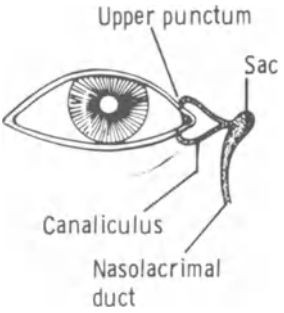

Fig. 4.1. The lacrimal passageway.

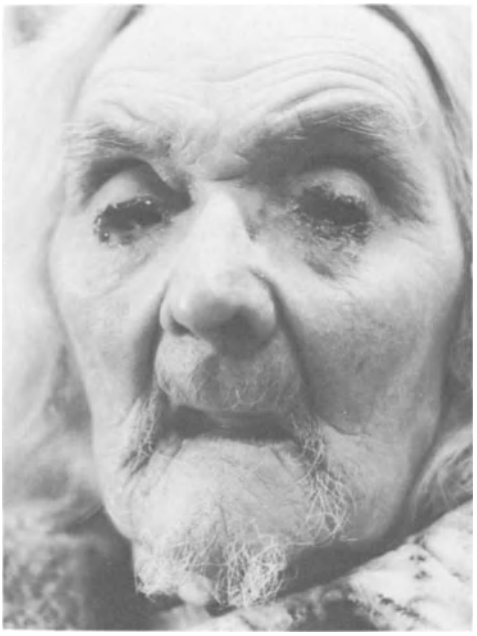

Fig. 4.2. Bilateral entropion. The inturned lower lids are largely obscured by purulent conjunctival discharge.

tears drain through the lower punctum. The puncta mark the opening of the lacrimal canaliculi and these small tubes conduct tears medially to the common canaliculus and thence into the tear sac (Fig. 4.1). The tear sac is connected directly to the nasolacrimal duct which opens into the inferior meatus of the nose below the inferior turbinate bone. The lacrimal puncta are easily visible to the naked eye and in the elderly the opening may be on top of a little mound like a miniature volcano. Inadequate drainage of tears may be due to misplacement of the punctum; the lower lid in elderly patients sometimes becomes turned inwards (entropion) due to the fact that the whole tarsal plate rotates on a horizontal axis. This in turn is related to slackening of the fascial attachments of the lower margin of the tarsal plate. Whenever these patients screw up their eyes the eyelids turn in and the lashes rub on the cornea. Such patients complain of watering sore eyes and the matter can be corrected very effectively by lid surgery (Fig. 4.2). Entropion may also result from scarring and contracture of the conjunctiva.

Not only may the punctum become turned inwards, but it may also be turned outwards. Sometimes the eversion may be very slight but enough to cause problems. Sometimes the patient may have been treating himself with proprietary drops or ointment which have given rise to a skin reaction on the lower lid. This reaction may slightly contract the skin and cause punctal eversion. Further rubbing of the eyelids and application of drops exacerbate the condition and ectropion (or eversion of the lower lid) may ensue (Fig. 4.3). Ectropion may also be corrected very effectively by surgery and it may also be caused by scarring and contracture of the skin of the lid following injury (cicatricial ectropion).

Fig. 4.3. Ectropion.

Drainage of tears along the lacrimal canaliculi depends to some extent on the muscular action of certain fibres of the orbicularis oculi muscle. This band of fibres encloses the lacrimal sac and it is thought that the walls of the sac are thereby stretched, producing slight suction along the canaliculi. Whatever the exact mechanism, when the orbicularis muscle is paralysed the tear flow is impaired, even if the position of the punctum is normal. Sometimes patients who have suffered a Bell's palsy may complain of a watering eye even though they appear to have otherwise made a complete recovery.

Misplacement of the drainage channels, particularly of the punctum and canaliculus, may thus affect the outflow of tears, but perhaps more commonly the drainage channel becomes blocked. In young infants with lacrimal obstruction the blockage is usually at the lower end of the nasolacrimal duct and takes the form of a plug of mucus or a residual embryological septum which has failed to become naturally perforated. In these cases there is nearly always some purulent discharge which may be expressed from the tear sac by gentle pressure with the index finger over the medial palpebral ligament. The mother is shown how to express this material twice daily and is instructed to instil antibiotic drops three or four times daily. This treatment alone may resolve the problem but it sometimes becomes necessary to syringe and probe the tear duct under a short anaesthetic. Usually one waits until the child is about 6 months old before probing. In adults the obstruction is more often in the lower canaliculus or the common canaliculus and in these cases syringing may be performed after the instillation of local anaesthetic drops. Unfortunately, syringing alone is not always effective and at this point the patient must decide whether to accept what may be a minor disability or

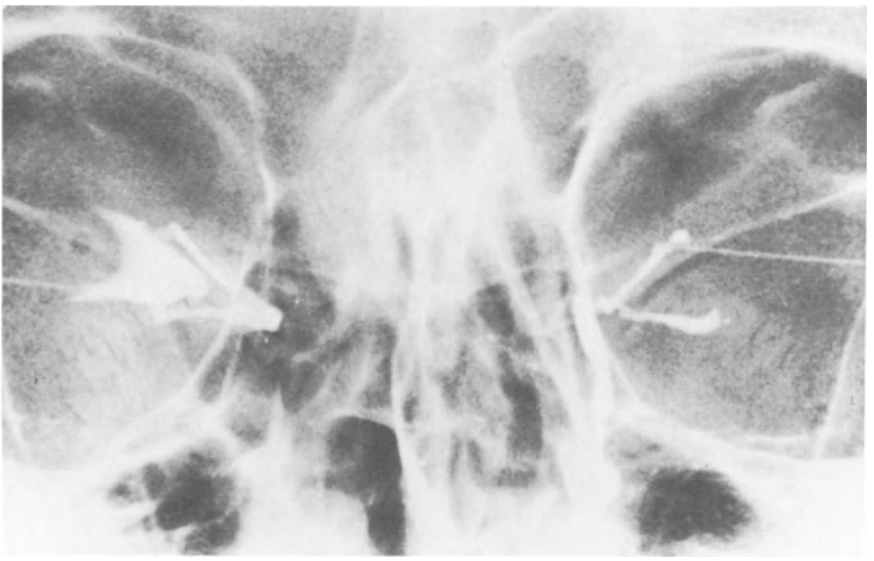

Fig. 4.4. Dacryocystogram. (With acknowledgement to Mr R. Welham.)

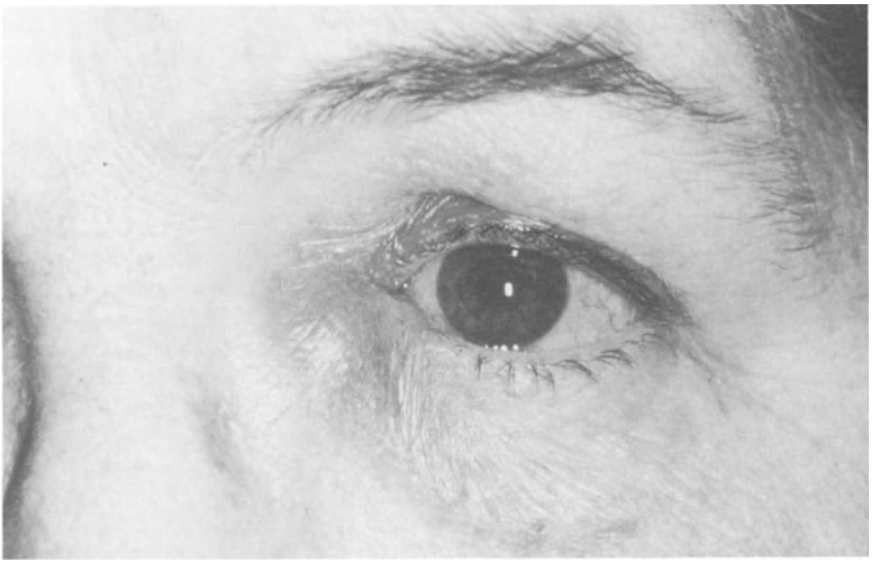

Fig. 4.5. Acute dacryocystitis. (With acknowledgement to Mr R. Welham.)

proceed to undergo more radical surgery of the tear passageway. Sometimes, in adults, the obstruction is in the nasolacrimal duct and in these cases an effective cure can usually be achieved by performing an operation to connect the lacrimal sac directly with the nasal cavity (dacryocystorhinostomy). The initial investigation of lacrimal obstruction entails syringing and, if this does not give the information required, it is possible to display the tear duct by x-ray using a radio-opaque contrast medium which is injected into the lower canaliculus with a lacrimal syringe (Fig. 4.4). The technique is known as dacryocystography.

Sometimes the lacrimal sac may become infected. This may occur in either children or adults but is more common in adult females. Acute dacryocsytitis can present initially as a watering eye and, in its early stages, the diagnosis may be missed if the tear sac is not gently palpated and found to be tender. Subsequently there is marked swelling and tenderness at the inner canthus and eventually the abscess may point and burst. In its early stages the condition can be aborted by the use of local and systemic antibiotics, but once the abscess has formed it is probably better to incise and drain it under general anaesthesia and, if necessary, insert a rubber or plastic drain. Perhaps surprisingly, the condition usually heals without any visible scar although it may recur in the future (Fig. 4.5).

Very rarely, the lacrimal canaliculi may become infected by the fungus *Actinomycoses,* and a small telltale bead of pus can usually be expressed from the punctum. The condition is very resistant to ordinary antibiotic treatment, and is best treated by opening up the punctum with a fine knife specially designed for the purpose — the procedure being called canaliculotomy — and then irrigating the canaliculi and tear sac with a suitable antibiotic.

The diagnosis of lacrimal obstruction therefore depends firstly on an examination of the eyelids, secondly on syringing the tear ducts, and if necessary on dacryocystography. Figure 4.6 illustrates the diagnostic use of lacrimal syringing.

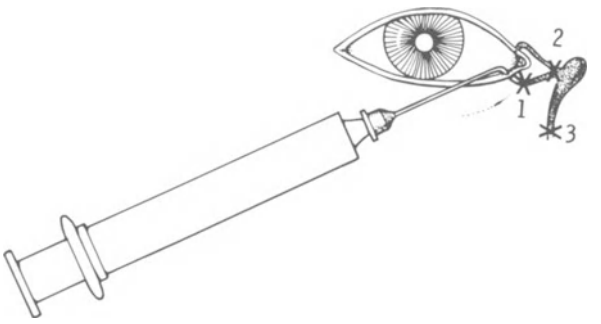

Fig. 4.6. Diagnostic use of lacrimal syringing. **1.** Obstruction in canaliculus shown by regurgitation of saline back through punctum. **2.** Common canaliculus obstruction shown by return of saline through upper punctum. **3.** Obstruction in nasolacrimal duct shown by filling of sac.

Excessive Secretion of Tears

A wide range of conditions affecting the eye may cause an excessive production of tears, from acute glaucoma to a corneal abscess, but these do not usually present as a watering eye since the other symptoms such as pain or visual loss are more evident to the patient. Occasionally the unwary doctor may be caught out by an irritative lesion on the cornea which mimics the more commonplace lacrimal obstruction. For example a small corneal foreign body or an ingrowing eyelash may present in this way. Not uncommonly, a lash may become lodged in the lower canaliculus and its removal, sometimes after weeks of discomfort, produces instant relief and gratitude.

The Dry Eye

The normal tear film is in three layers and the integrity of this film is essential for comfort and, more important, for good vision. The anterior or outermost layer is formed by the oily secretion of the meibomian glands, the middle layer consists of watery tears from the lacrimal and accessory lacrimal glands, and the layer next to the cornea is mucinous to allow proper wetting by the watery component of the tears. This three-layered film is constantly maintained by the act of blinking.

A patient may complain of dryness of the eyes simply because the conjunctiva is inflamed, but when the tear film really is defective the complaint is often soreness and irritation of the eyes and not necessarily dryness. The diagnosis of a dry eye depends on a careful examination and it is quite erroneous to assume that the tear film is inadequate simply because the patient appears to be improved by the instillation of artificial tears.

Causes

Systemic disease with lacrimal gland involvement
  Sarcoidosis
  Rheumatoid arthritis (Sjögren's syndrome)
Trachoma
Neuroparalytic keratitis
Exposure keratitis
Stevens–Johnson syndrome and pemphigoid
Old age

Signs

Slit-Lamp Examination

The tear film can normally be seen, especially where it is slightly thicker along

the margin of the lower lid, and the dryness of the eye may be evident by direct examination. Prolonged deficiency of tears may be associated with the presence of filaments — microscopic strands of mucus and epithelial cells which stain with rose bengal. Punctate staining of the corneal epithelium is also seen after applying a drop of fluorescein. In some dry eye syndromes, for example Stevens–Johnson syndrome or ocular pemphigoid, keratinisation of the cornea and conjunctiva with the formation of contracting adhesions between opposed surfaces of the conjunctiva is seen. A similar change is apparent following chemical burns of the eye.

Schirmer's Test

One end of a special filter-paper strip is placed between the globe and the lower lid. The other end projects forward and the time taken for the tears to wet the projecting strip is measured. The test is not a very accurate measure of tear secretion but it provides a useful screening method (Fig. 4.7).

Tear Film Break-Up Time

Using the slit lamp, the time for the tear film to break up when the patient stops blinking is measured. This test is sometimes used as an index of mucin deficiency.

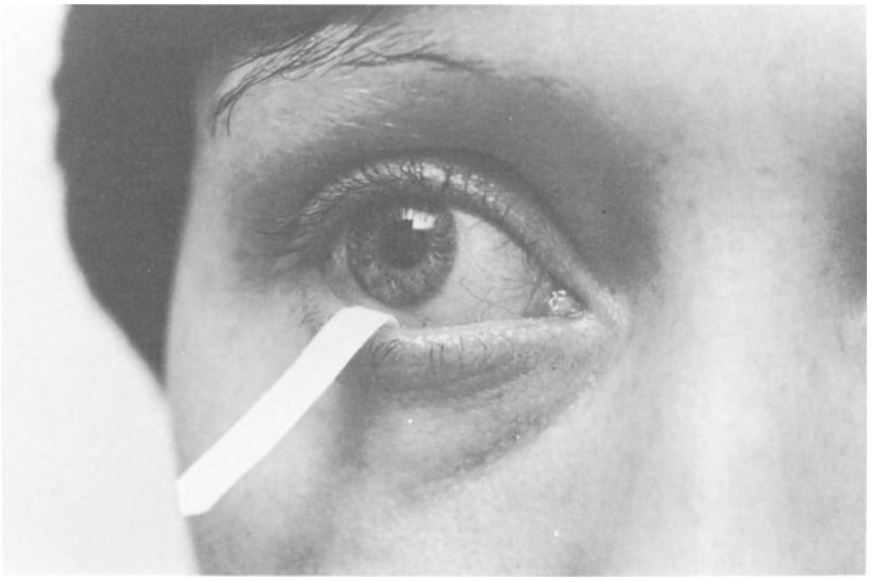

Fig. 4.7. Schirmer's test.

Management

This, of course, depends on the cause of the dry eye, and the underlying systemic cause may require treatment in the first place. Artificial tear drops are a mainstay in treatment and may be prescribed hourly or more frequently if necessary. A local antibiotic may also be required, and lid surgery may sometimes be indicated.

Deformities of the Eyelids

The Normal Eyelid

Figure 4.8 is a diagram of the normal eyelids in cross-section. The lids contain two antagonistic voluntary muscles: the more superficial orbicularis oculi,

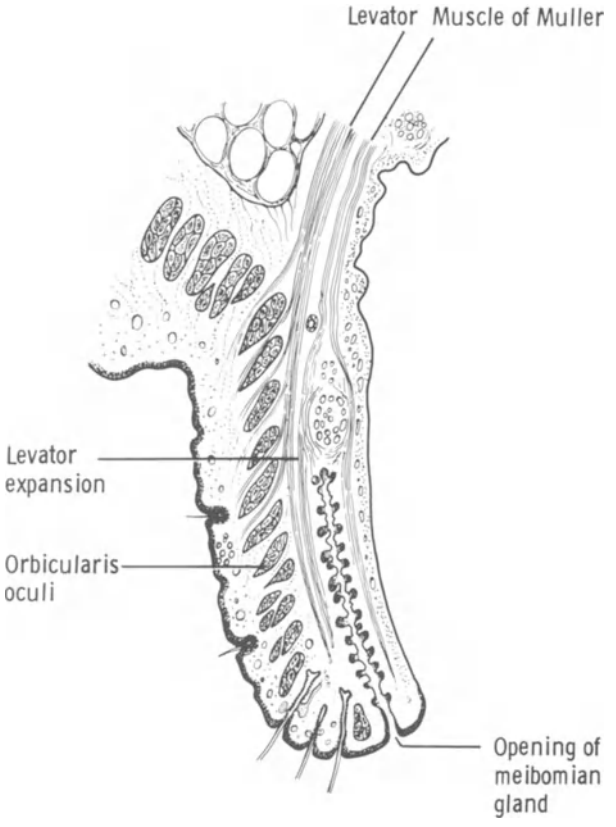

Fig. 4.8. Cross-section of normal eyelid.

supplied by the seventh cranial nerve, which closes the eyes, and the levator palpebrae superioris. Strictly speaking, it is the tendon of the levator which is present in the eyelid and this muscle opens the eyes and is supplied by the third cranial nerve. We must not forget that there is also some smooth muscle in both upper and lower eyelids which has clinical importance apart from its influence on facial expression when the subject is under stress. Loss of tone in this muscle accounts for the slight ptosis in Horner's syndrome; increased tone is seen in thyrotoxic eye disease. These muscles are attached to the skeleton of the lid which is the tarsal plate, a plate of fibrous tissue (not cartilage) which contains the meibomian glands.

Epicanthus

Figure 4.9 shows that this is characterised by vertical folds of skin at the inner canthus. These folds are seen quite commonly in otherwise normal infants and they gradually disappear as the facial bones develop. Children with epicanthus may appear to the uninitiated to be squinting and this can cause considerable parental anxiety. It is important to explain that the squint is simply an optical illusion rather than just tell the parents that there is nothing the matter. Epicanthus persists into adult life in Mongolian races, and is occasionally seen in European adults. It may also be associated with other deformities of the eyelids.

Fig. 4.9. Epicanthus.

Entropion

This usually affects the lower eyelid and generally occurs over the age of 50. It is a rare deformity in newborn infants but nearly always resolves spontaneously during the first few months. Entropion in late middle age is common and the patient presents with a sore and irritable eye and possibly associated purulent conjunctivitis. Closer inspection reveals the inturned eyelid, which can be restored to its normal position by slight downwards pressure on the lower lid, only to turn in again when the patient forcibly closes the eyes. The condition is often associated with muscular eyelids and sometimes seems to be precipitated by repeatedly screwing up the eyes. Slackening of the fascial sling of the lower lid with ageing combined with the action of the orbicularis muscle allows this to happen. This common type of entropion is called spastic entropion and it can be promptly cured without leaving a visible scar by minor lid surgery. Other types of entropion are seen following scarring of the conjunctival surface of the eyelids and one must mention the entropion of the upper lids caused by trachoma which is very rare in Britain but all too common in the Middle East and countries where trachoma is still rife.

Ectropion

This commonly seen outward turning of the lower lid in the elderly is also eminently treatable and responds well to minor surgery. Senile ectropion may begin with slight separation of the lower lid from the globe leading to overflow of tears and infection of the conjunctiva. The resulting irritation leads to eye rubbing and further downward pulling of the eyelids and a viscious circle develops. Like entropion, ectropion may be cicatricial and result from scarring of the skin of the eyelids, but it may also occur following a seventh cranial nerve palsy due to the complete inaction of the orbicularis oculi; this is called paralytic ectropion.

Lagophthalmos

This is the term used to denote failure of closure of the eyelids during sleep or inadequate blinking due to lid deformity. In either case the cornea is not adequately lubricated and exposure keratitis develops. If untreated this can lead to a serious situation: initially the cornea shows punctate staining when a drop of fluorescein is placed in the conjunctival sac, and subsequently a corneal ulcer with possible penetration of infection into the eye may occur. Pus is seen in the anterior chamber (hypopion keratitis) and in the absence of prompt and intensive treatment with antibiotics the eye may be lost.

As a general principle it is important to realise that the sight may be lost simply because the eyes cannot blink. The principle applies especially to the unconscious or the anaesthetised patient where a disaster may be avoided by taping or padding the lids and applying an antibiotic ointment.

Blepharospasm

Slight involuntary twitching of the eyelids is very common and not usually considered to be of any pathological significance other than being a symptom of fatigue or sometimes of an anxiety state. The condition is termed 'myokymia'. True blepharospasm is rare. It may be unilateral or bilateral and may cause great inconvenience and worry to the patient. Many types of treatment have often been tried but the most effective seems to be surgical division of the affected peripheral branches of the facial nerve, a rather lengthy procedure which is sometimes justified.

Redundant Lid Skin

Excessive skin on the eyelids is commonly seen in elderly people, often as a family characteristic. It may result from chronic oedema of the eyelids due, for example, to thyrotoxic eye disease or to renal disease. The problem is made worse in some cases by herniation of orbital fat through the orbital septum, and excision of the redundant skin and orbital fat may sometimes be necessary.

Ptosis

Drooping of one upper lid is an important clinical sign. In ophthalmic practice, ptosis in children is usually congenital and in adults is either congenital or due to a third cranial nerve palsy. These more common causes must always be kept in mind but there are a large number of other possible ones. When confronted with a patient whose upper lid appears to droop, the first thing to decide is whether the lid really is drooping or whether the other lid is retracted. The upper lid may droop because the eye is small and hypermetropic or shrunken from disease. Having eliminated the possibility of such 'pseudoptosis', the various other causes can be considered, beginning on the skin of the eyelid — styes, meibomian cysts — and advancing centrally through muscle — myasthenia gravis — along nerves — oculomotor palsy, Horner's syndrome — to the brainstem. Marked ptosis with the eye turned down and out is an oculomotor palsy, whereas very slight ptosis, often not noticed by the patient nor sometimes by the doctor, is more likely to mean Horner's syndrome. This syndrome is due to damage to the sympathetic nervous supply to either upper or lower lids or both and is characterised by slight ptosis, small pupil, loss of sweating on the affected side of the face and slight enophthalmos (posterior displacement of the globe).

The management of ptosis depends on the cause and thus on accurate diagnosis. Surgical shortening of the levator palpebrae superioris muscle is effective in some cases of congenital ptosis and sometimes in long-standing third cranial nerve palsies. Such a measure would be disastrous if applied to a patient with myasthenia gravis or with corneal anaesthesia. Children with congenital ptosis need to be assessed very carefully before considering surgery.

Normally surgery is reserved for those cases where the lid threatens to droop across the visual axis and thereby interfere with the sight or where there is an unacceptable head tilt. In one rather strange type of congenital ptosis, the problem disappears when the mouth is opened and the patient may literally wink unavoidably when chewing. Under these circumstances the surgeon must again be very wary, especially against a background of pressure from anxious parents.

Causes of Ptosis

Pseudoptosis:	small eye, atrophic eye, lid retraction on other side
Mechanical ptosis:	inflammation, tumour, excess skin
Myogenic ptosis:	myasthenia gravis
Neurogenic ptosis:	sympathetic — Horner's syndrome, IIIrd palsy, any lesion in the pathway of these nerves, carcinoma of the lung may cause Horner's syndrome
Drugs:	guanethidine eyedrops cause ptosis
Congenital	

Meibomian Cysts and Styes

These two conditions are very common and few of us escape suffering from one or the other at some time. They can both be disabling and sufferers from them very often seek medical attention.

Meibomian Infection

The openings of the meibomian glands become infected at any age resulting in a meibomitis, seen initially as redness along the line of a gland when the lid is everted. A small abscess may then form with swelling and redness of the whole eyelid and this may point and burst either through the conjunctiva or, less often, through the skin. The orifice of a gland may become occluded and the gland becomes distended and cystic. The retained secretions of the gland set up a granulating reaction and the cyst itself may become infected. The patient may complain of soreness and swelling of the eyelid which subsides leaving a pea-sized swelling which remains for many months and sometimes swells up again. During the stage of acute infection the best treatment is local heat, preferably in the form of steam. This produces considerable relief and is preferable to the use of either local or systemic antibiotics. Antibiotics are not required unless the patient shows signs of septicaemia, or has troublesome recurrences. Once a pea-sized cyst remains in the tarsal plate, this can be promptly removed under local anaesthetic unless the patient is a child. The method of removal involves everting the eyelid and incising the cyst through

Fig. 4.10. Meibomian cyst.

the conjunctiva and then curetting out the contents. Postoperatively, local antibiotic drops or ointment are prescribed (Fig. 4.10).

Styes

These are distinct from meibomian infections, being the result of infection of the lash root. The eyelid may swell up and become painful and at this stage the site of the infection may be uncertain. However, a small yellow pointing area is eventually seen around the base of an eyelash. Hot steaming is the most effective treatment, and once the pus is seen the eyelash can be gently epilated with resulting discharge and subsequent resolution of the infection.

Children aged 6–10 years sometimes seem to go through periods of their lives when they may be dogged by recurrent styes and meibomian infections, much to the distress of their parents. Under these conditions very frequent baths and hairwashing are advised and sometimes a long-term systemic antibiotic may be considered. Recurrent lid infections may raise the suspicion of diabetes mellitus but in practice this is rarely found to be an underlying cause.

Eyelid infections such as these very rarely cause any serious problems other than a few days off work, and it is extremely unusual for the infection to spread and lead to orbital cellulitis. Recurrent swelling of the eyelid in spite of incision and curettage may indicate the need for a lid biopsy, but in practice tumours of the eyelid such as carcinoma of the meibomian gland are extremely rare.

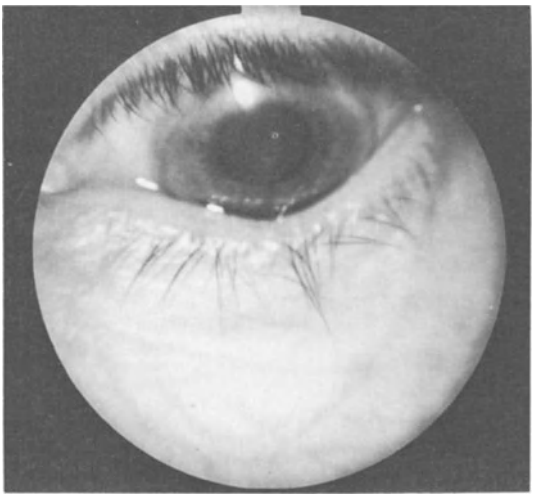

Fig. 4.11. Trichiasis. Two ingrowing lashes on the lower lid had been causing a sore eye for 3 months.

Ingrowing Lashes (Trichiasis)

The lashes may grow in an aberrant manner even though the lids themselves are in good position. This may be the result of chronic infection of the lid margins or may follow trauma. Sometimes one or two aberrant lashes appear for no apparent reason (Fig. 4.11). The lashes tend to rub on the cornea producing irritation and secondary infection. The condition is referred to as 'trichiasis'. When one or two lashes are found to be the cause of the patient's discomfort, it is common practice simply to epilate them with epilating forceps. This produces instant relief, but often the relief is short lived because the lashes regrow in the same way. At this stage the best treatment is to destroy the lash root by electrolysis prior to epilation. Needless to say, before removing lashes it is essential to be familiar with the normal position of the lash line and to realise, for example, that hairs are normally present on the caruncle. As stated above, occasionally a lash is found projecting from the lacrimal punctum having been washed there like a stick in a drain. This is seen surprisingly often in ophthalmic practice and the simple removal of this lash may end weeks or even months of irritation and recurrent infection in the conjunctiva. When the lash line is grossly distorted by injury or disease, the rubbing of the lashes on the cornea can be prevented by fitting a protective contact lens or, if this measure proves impractical, it may be necessary to transpose or excise the lashes and their roots.

Blepharitis

This refers to a chronic inflammation of the lid margins due to staphylococcal infection. The eyes become red rimmed and there is usually an accumulation

of scales giving the appearance of fine dandruff on the lid margins. The condition is often associated with seborrhoea of the scalp. Sometimes it becomes complicated by recurrent styes or chronic infection of the meibomian glands. The eye itself is not usually involved although there may be a mild superficial punctate keratitis as evidenced by fine staining of the lower part of the cornea with fluorescein. In more sensitive patients the unsightly appearance may cause difficulties, but in more severe cases the discomfort and irritation may interfere with work. Severe recurrent infection may lead to irregular growth of the lashes and trichiasis.

In the management of these patients it is important to explain the chronic nature of the condition and the fact that certain individuals seem to be prone to it. Attention should be given to keeping the hair, face and hands as clean as possible and to avoid rubbing the eyes. When the scales are copious they can be gently removed with cotton wool moistened in sodium bicarbonate lotion twice daily. Dandruff of the scalp should also be treated with a suitable shampoo. A local antibiotic may be applied to the lid margins twice daily with good effect in many but not all cases, and in severe cases with ulceration of the lid margin it may be necessary to consider a systemic antibiotic, preferably after culturing the causative organism. Local steroids when combined with an antibiotic are very effective, but the prescriber must be aware of the dangers of using steroids on the eye and long-term treatment with steroids should be avoided. Steroids should not be used without monitoring the intraocular pressure.

Lid Tumours

Benign Tumours

Papilloma. Commonly seen on lids near or on margin, may be sessile or pedunculated, sometimes keratinised. Caused by a virus. Easily excised but care must be taken if excision involves the lid margin (Fig. 4.12).

Fig. 4.12. Lid margin papilloma.

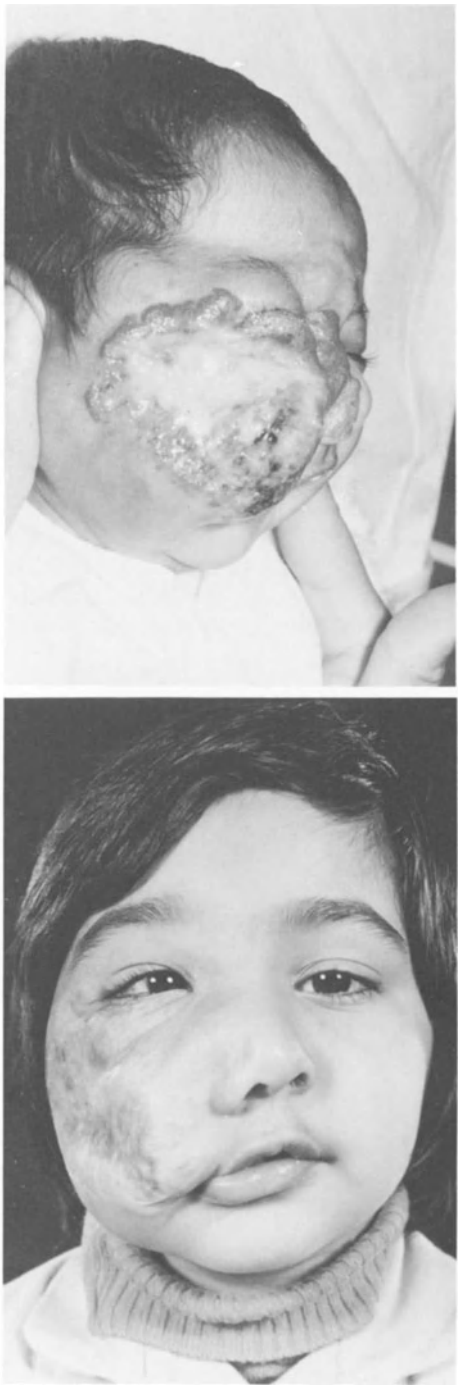

Fig. 4.13a. Large disfiguring haemangioma in infancy. **b.** The same lesion in later childhood showing spontaneous regression.

Naevus. Flat brown spot on skin, may have hairs, very rarely becomes malignant.

Haemangioma. When seen as red 'strawberry mark' on the eyelids at or shortly after birth, may regress completely during first years of life. Cavernous haemangioma is raised and may be very disfiguring. This also may regress spontaneously (Fig. 4.13). 'Port wine stain' is name applied to capillary haemangioma. This is usually unilateral and when the eyelids are involved there is risk of association with haemangioma of the choroid, congenital glaucoma and haemangioma of the meninges on the ipsilateral side (Sturge–Weber syndrome). Children with port wine stains involving the lids need full ophthalmological and neurological examinations.

Dermoid Cyst. Quite commonly seen lump in or just below the eyebrow. Feels cystic and sometimes attached to bone. Present in children as minor cosmetic problem. The cysts are lined by keratinised epithelium and contain dermal appendages and cholesterol. X-ray before removal since it may extend deep into skull.

Xanthelasma. Seen as yellowish plaques in skin; usually begin at medial end of lids. Associated rarely with diabetes, hypercholesterolaemia and histiocytosis. Usually no associated systemic disease.

Malignant Tumours

Basal Cell Carcinoma. The most common malignant tumour of the lids, usually on lower lid. Small lump, tends to bleed forming central crust with slightly raised hard surround. Tumour is locally invasive only but should be excised to avoid spread into bone. Radiotherapy is effective where local excision is impractical (Fig. 4.14).

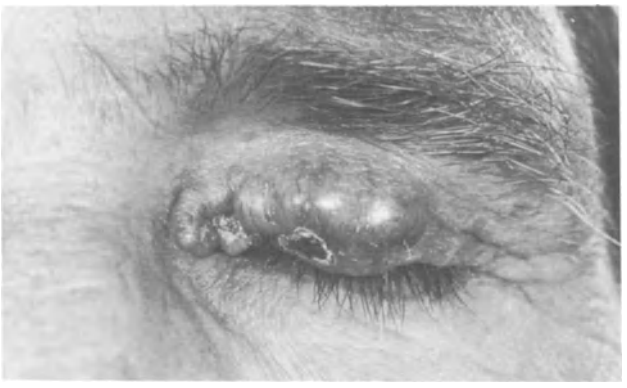

Fig. 4.14. Cystic basal cell carcinoma which has extended to involve most of the upper lid.

Squamous Cell Carcinoma. Tends to resemble basal cell carcinoma and biopsy is needed to differentiate. May also be mimicked by a benign self-healing lesion known as keratoacanthoma.

Malignant Melanoma. A raised black-pigmented lesion. Highly malignant; rare.

Lid Injuries

Probably the commonest injury to the eyelids is due to the presence of a foreign body under the lid — a subtarsal foreign body. A small particle of grit lodges near the lower margin of the lid, but to see it the lid must be everted. Every student should be familiar with the simple technique of lid eversion. This is performed by gently grasping the lashes of the upper lid between finger and thumb and placing a glass rod horizontally across the lid at the same time. The eyelid is then gently everted by drawing the lid margin upwards and forwards. The manoeuvre is only achieved if the patient is asked to look down beforehand, and the everted lid is replaced by asking the patient to look upwards. If a small foreign body is seen, it is usually a simple matter of removing it using a bit of a cotton-wool bud or even the edge of a clean handkerchief if the former is not available (Fig. 4.15).

Cuts on the eyelids may be the result of a car accident involving a broken windscreen but the lids may also be injured by chemical burns or flash burns. Exposure to ultra-violet light, as from a welders arc or in snow blindness, may cause oedema and erythema of the eyelids which appears after a delay of some hours but resolves spontaneously after about 2 days. When the eyelids have been cut right through, suturing is more simple if the cut is parallel to the lid margin. When the cut is full thickness and involving the lid margin, it must be sewn up in three layers taking especial care to oppose the wound accurately in order to prevent notching of the lid as healing takes place.

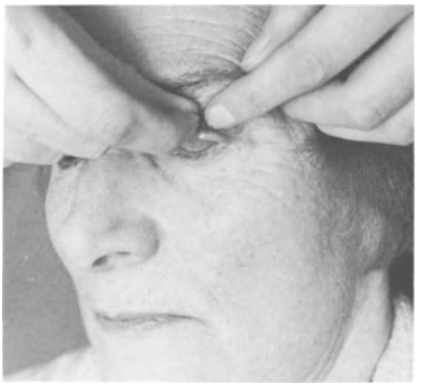

Fig. 4.15. Everting the upper lid.

5 Common Diseases of the Conjunctiva and Cornea

Subconjunctival Haemorrhage

This is common and tends to occur spontaneously or sometimes after straining or, especially, vomiting. The eye becomes suddenly red and although the patient may experience a slight pricking, the condition is usually first noticed in the mirror or by a friend. The haemorrhage gradually absorbs in about ten days and investigations usually fail to reveal any underlying cause. Very rarely, it is necessary to cauterise the site of bleeding if the haemorrhage is repeated so often that it becomes a nuisance to the patient (Fig. 5.1).

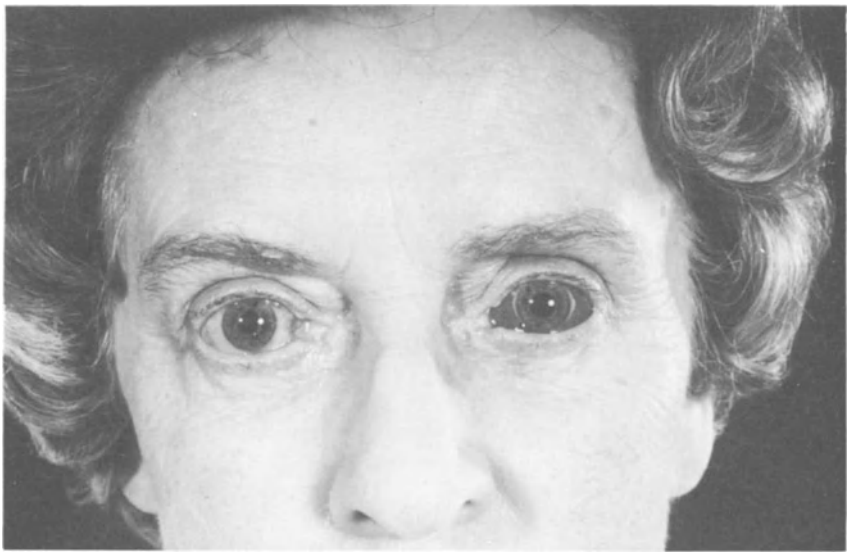

Fig. 5.1. Subconjunctival haemorrhage.

Conjunctivitis

Inflammation of the conjunctiva is extremely common in the general population and the general practitioner is often expected to find out the cause and treat this condition. If we consider that the conjunctiva is a mucous membrane which is exposed during the waking hours to wind and weather more or less continuously, year in year out, then it is not surprising that this membrane is rather susceptible to inflammation. Furthermore, the conjunctiva can be compared with the lining of a joint, the eye being considered as an unusual type of ball and socket joint. The analogy takes on more meaning when the relation between conjunctivitis and some joint disease is seen.

There are a large number of different specific causes of conjunctivitis. Some of these are interesting but rare and it is important that the student obtains an idea of the relative importance and frequency of the different aetiological factors. For this reason, in this chapter a more or less categorical list is given of the different causes, but in the chapter on the red eye (Chapter 6) you will find a plan of approach to the red eye which deals with the important and more common causes of conjunctivitis seen in day-to-day practice.

Although the conjunctiva is continuously exposed to infection, it has special protection from the tears which contain immunoglobulins and lysozyme. The tears also help to wash away debris and foreign bodies and this protective action can explain the self-limiting nature of most types of conjunctivitis.

Symptoms

In all types of conjunctivitis the eye becomes red and feels irritable and gritty as if there were a foreign body under the lid. There is usually some discharge and if marked this may make the eyelids stick together in the mornings. Itchiness may also be present, especially in cases of allergic conjunctivitis. The discharge around the eyelids tends to make vision intermittently blurred and the patient may volunteer that the sight is cleared by blinking.

Signs

The conjunctiva appears hyperaemic and there may be evidence of purulent discharge on the lid margins causing matting together of the eyelashes. The redness of the conjunctiva extends to the conjunctival fornices and is usually less marked at the limbus. When a rim of dilated vessels is seen around the cornea, the examiner must suspect a more serious inflammatory reaction within the eye. Apart from being red to a greater or lesser degree, the eyes also tend to water, but a dry eye may lead one to suspect conjunctivitis due to inadequate tear secretion. Drooping of one or both upper lids is a feature of some types of virus conjunctivitis and this may be accompanied by enlargement of the preauricular lymph nodes. The ophthalmologist should

train himself to feel for the preauricular node as a routine part of the examination of such a case. Closer inspection of the conjunctiva may reveal numerous small papillae giving the surface a velvety look, or the papillae may be quite large. Giant papillae under the upper lids are a feature of spring catarrh, a form of allergic conjunctivitis. Close inspection of the conjunctiva may also reveal follicles or focal lymphoid hyperplasia. Being deep to the epithelium, they are small, pale, raised nodules and although they are not specific for any particular type of inflammation they are commonly seen in viral conjunctivitis. Follicles under the upper lids are especially characteristic of trachoma.

Microscopy

The examination of a severe case of conjunctivitis of unknown cause is not complete until conjunctival scrapings have been taken. A drop of local anaesthetic is placed in the conjunctival sac and the surface of the conjunctiva gently scraped with the blade of a sharp knife. The material obtained is placed on a slide and stained with Gram's stain and Giemsa's stain. The infecting organism may thus be revealed or the cell type in the exudate may indicate the underlying cause.

Conjunctival Culture

In most cases of conjunctivitis it may be good medical practice to take a culture from the conjunctival sac, but such a measure may not always be possible if a microbiological service is not near at hand.

Causes

1. Bacterial
2. Chlamydial
3. Viral
4. Other infective agents
5. Allergic
6. Secondary to
   - lacrimal obstruction
   - corneal disease
   - lid deformities
   - degenerations
   - systemic disease
7. Unknown cause

Bacterial Conjunctivitis

In Britain the commonest organisms to cause conjunctivitis are the *Pneumococcus, Haemophilus influenzae* and *Staphylococcus aureus.* The last-mentioned is normally associated with chronic lid infections, and the acute purulent conjunctivitis, known more familiarly as 'pink eye', is usually caused by the *Pneumococcus.* Chronic conjunctivitis may also be caused by

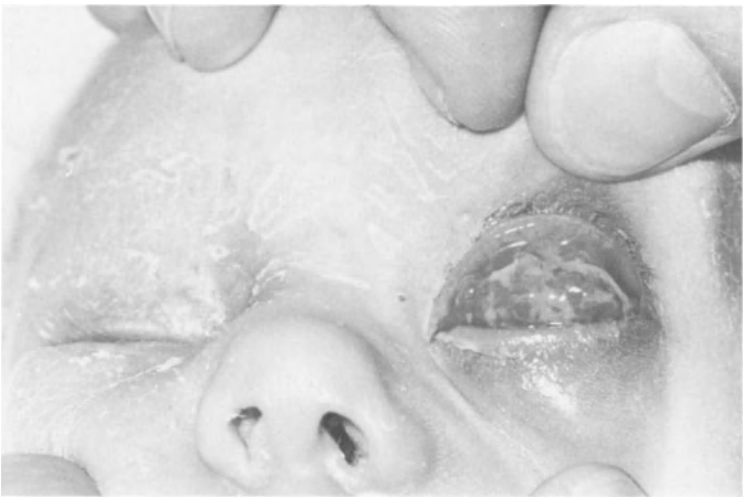

Fig. 5.2. Ophthalmia neonatorum. (With acknowledgement to Mr A. Fielder.)

Moraxella lacunata but this organism is rarely isolated from cases nowadays. An important but rare form of purulent conjunctivitis is that due to *Neisseria gonorrhoeae;* this is still an occasional cause of a severe type of conjunctivitis seen in the newborn babies of infected mothers. Untreated, the cornea also becomes infected, leading to perforation of the globe and permanent loss of vision. Purulent discharge, redness and severe oedema of the eyelids are features of the condition, which is generally known as ophthalmia neonatorum. Ophthalmia neonatorum may also be caused by *Staphylococcus* and the chlamydial virus (see Inclusion Conjunctivitis of the Newborn, p. 55). The disease is notifiable and any infant with purulent discharge from the eyes, particularly between the 2nd and 12th day post partum, should be suspect. At one time the special blind schools were filled with children who had suffered ophthalmia neonatorum, but an active campaign against this cause of blindness began at the end of the last century when Carl Crede introduced the principle of careful cleansing of the infant's eyes and the instillation of silver nitrate drops. Blindness from this cause has now disappeared in Britain but there is still a low incidence of ophthalmia neonatorum (Fig. 5.2).

Pink eye is the name given to the type of acute purulent conjunctivitis which tends to spread rapidly through families or around schools. The eyes begin to itch and within an hour or two produce a sticky discharge which causes the eyelids to stick together in the mornings. If the disease is mild, it can be treated by cleaning away the discharge with cotton wool, and it does not usually last longer than 2 or 3 days. More severe cases may warrant the prescription of antibiotic drops instilled four times daily for 5 days. A conjunctival culture should be taken prior to starting treatment. Common-sense precautions against spread of the infection should also be advised, although they are not always successful.

Attempts to culture bacteria from the conjunctival sac of cases of chronic conjunctivitis do not yield much more than commensal organisms. One particular kind of chronic conjunctivitis in which the inflammation is sited mainly near the inner and outer canthi is known as angular conjunctivitis. Another feature of this is the excoriation of the skin at the outer canthi from the overflow of infected tears. The clinical picture has been recognised in association with infection by the bacillus *Moraxella lacunata.* Often zinc sulphate drops and the application of zinc cream to the skin at the outer canthus are sufficient treatment in such cases.

Chlamydia Conjunctivitis

The *Chlamydia* comprise a group of large viruses which are sensitive to tetracycline and erythromycin and which cause relatively minor disability to the eyes in northern Europe and the United States when compared to the severe and widespread eye infection seen especially in Africa and the Middle East. Inclusion conjunctivitis ('inclusion blenorrhoea') is the milder form of chlamydial infection. The condition tends to be transmitted venereally and it may cause a more severe type of conjunctivitis in the newborn child, which can also involve the cornea. The infection is usually self-limiting, lasting several months, but it responds to treatment with tetracycline. In children and adults the oily drops of tetracycline should be used at least four times daily. In adults the treatment can be supplemented with systemic tetracycline, but the drug should not be used systemically in pregnant mothers or children under 7 years old. The diagnosis depends on the results of conjunctival culture and examination of scrapings and the association of a follicular conjunctivitis with cervicitis or urethritis.

Trachoma. Although a doctor practising in Great Britain may rarely see a case of trachoma, and even then only in immigrants, it is the commonest cause of

Fig. 5.3. Trachoma. Both corneae were severely involved.

blindness in the world and, furthermore, the disease affects about 15% of the world's population. It is spread by direct contact and perpetuated by poverty and unhygienic conditions. There is evidence that the virus is the same one which causes inclusion blenhorroea. The disease begins with conjunctivitis which, instead of resolving, becomes persistent, especially under the upper lid where scarring and distortion of the lid may result. The inflammatory reaction spreads to infiltrate the cornea from above and ultimately the cornea itself may become scarred and opaque. At one time trachoma was common in Britain, especially after the Napoleonic wars at the end of the 18th century. It had been eliminated by improved hygienic conditions long before the introduction of antibiotics (Fig. 5.3).

Adenoviral Conjunctivitis

Acute viral conjunctivitis is common. Several of the adenoviruses may cause it. Usually the eye symptoms follow an upper respiratory tract infection and, although nearly always bilateral, one eye may be infected before the other. The affected eye becomes red and discharges; characteristically the eyelids become thickened and the upper lid may droop. The ophthalmologist's finger should feel for the telltale tender enlarged preauricular lymph node. In some cases the cornea becomes involved and subepithelial corneal opacities may appear and persist for several months. If such opacities are situated in the line of sight, the vision may be impaired. There is no known effective treatment but it is usual to treat with an antibiotic drop to prevent secondary infection.

From time to time, epidemics of viral conjunctivitis occur and it is well recognised that spread may result from the use of improperly sterilised ophthalmic instruments or even contaminated solutions of eyedrops.

Other Infective Agents

The conjunctiva may be affected by a wide variety of organisms, some of which are too rare to be considered here, and sometimes the infected conjunctiva is of secondary importance to more severe disease elsewhere in the eye or in the rest of the body. Molluscum contagiosum is a virus infection which causes small umbilicated nodules to appear on the skin of the lids and elsewhere on the body, especially the hands. It may be accompanied by conjunctivitis when there are lesions on the lid margin. The infection is usually easily eliminated by curetting each of the lesions. Infection from ***Phthirus pubis*** (the pubic louse) involving the lashes and lid margins may initially present as conjunctivitis but observation of nits on the lashes should give away the diagnosis.

Allergic Conjunctivitis

Several types of allergic reaction are seen on the conjunctiva and some of these also involve the cornea. They may be listed as follows.

Hay Fever Conjunctivitis. This is simply the commonly experienced red and watering eye that accompanies the sneezing bouts of the hay fever sufferer. The eyes are itchy and mildly injected and there may be conjunctival oedema. If treatment is needed, adrenaline drops may be helpful or sodium chromoglycate eyedrops may be used on a more long-term basis.

Atopic Conjunctivitis. Unfortunately, patients with asthma and eczema may experience recurrent itching and irritation of the conjunctiva. This is relieved by applying local steroid drops, but in view of the long-term nature of the condition, these should be avoided if possible because of their side-effects. (Local steroids can cause glaucoma in predisposed individuals.) Although atopic conjunctivitis tends to improve over a period of many years, it may result in repeated discomfort and anxiety for the patient, especially as the cornea may become involved, showing a superficial punctate keratitis or, in the worst cases, abscess formation and scarring. The diagnosis is usually evident from the history but conjunctival scrapings show the presence of eosinophils. Patients with atopic keratoconjunctivitis have a higher risk than normal for the development of herpes simplex keratitis and the condition is also associated with the rare corneal dystrophy known as keratoconus or conical cornea.

Vernal Conjunctivitis (spring catarrh). Some children with an atopic history may develop a specific type of conjunctivitis characterised by the presence of giant papillae under the upper lid. The child tends to develop severely watering and itchy eyes in the early spring, which may interfere with schooling. Eversion of the upper lid reveals the raised papillae which have been likened to cobblestones. Occasionally the cornea is also involved initially by punctate keratitis but sometimes it may become vascularised. It is often necessary to treat these cases with local steroids, for example prednisolone drops applied if needed every 2 hours for a few days to enable the child to return to school, then reducing this dose as much as possible to a maintenance dose over the worst part of the season. Less severe cases may respond well to sodium chromoglycate drops and these may be useful as a long-term measure (Fig. 5.4).

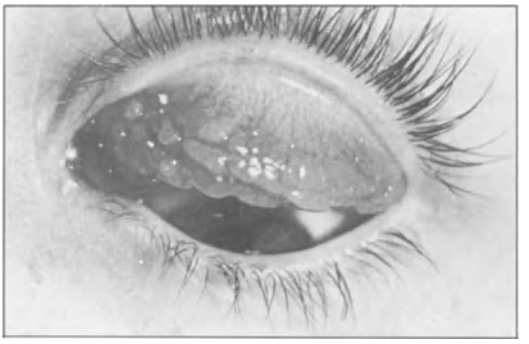

Fig. 5.4. Spring catarrh. Large follicles seen under the upper lid. (With acknowledgement to Institute of Ophthalmology, London, and Churchill Livingstone, Edinburgh: Perkins E.S., Hansell P. (1971) Atlas of diseases of the eye.)

Secondary Conjunctivitis

Inflammation of the conjunctiva may often be secondary to more important primary pathology. The following are some of the underlying possible causes of this type of conjunctivitis.

1. Lacrimal obstruction
2. Corneal disease
3. Lid deformities
4. Degenerations
5. Systemic disease

Lacrimal obstruction may cause recurrent unilateral purulent conjunctivitis and it is important to consider this possibility in recalcitrant cases because early resolution may be achieved simply by syringing the tear ducts. *Corneal ulceration* from a variety of causes is often associated with conjunctivitis and here the treatment is aimed primarily at the cornea. Occasionally the presence of one of the two common acquired *lid deformities,* entropion and ectropion, may be the underlying cause. Sometimes the diagnosis may be missed, especially in the case of entropion when the deformity is not present all the time. Other lid deformities may also have the same effect. A special type of *degenerative change* is seen in the conjunctiva which is more marked in hot, dry, dusty climates. It appears that the combination of lid movement in blinking, dryness and dustiness of the atmosphere and perhaps some abnormal factor in the patient's tears or tear production may lead to the heaping up of subconjunctival yellow elastic tissue which is often infiltrated with lymphocytes. The lesion is seen as a yellow plaque on the conjunctiva in the

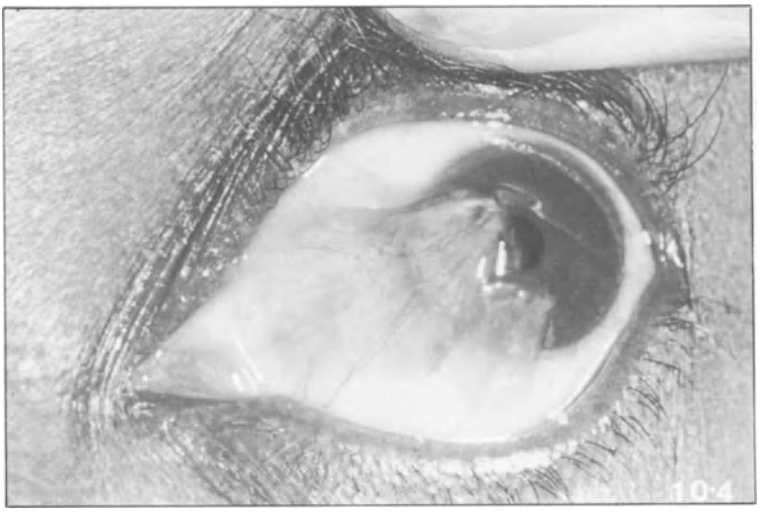

Fig. 5.5. Pterygium. (With acknowledgement to Institute of Ophthalmology, London, and Churchill Livingstone, Edinburgh: Perkins E.S., Hansell P. (1971) Atlas of diseases of the eye.)

exposed area of the bulbar conjunctiva and usually on the nasal side. Such early degenerative changes are extremely common in all climates as a natural ageing phenomenon, but under suitable conditions the heaped-up tissue spreads onto the cornea, drawing a triangular band of conjunctiva with it. The eye becomes irritable due to associated conjunctivitis and in worst cases the degenerative plaque extends across the cornea and affects the vision. The early stage of the condition, which is common and limited to a small area of the conjunctiva, is termed a pingueculum and the more advanced lesion spreading onto the cornea is known as a pterygium. Pterygium is more common in India and China as well as the Middle East than in Europe and it is rarely seen in white races living in temperate climates. Treatment is by surgical excision if the cornea is significantly affected; antibiotic drops may be required if the conjunctiva is infected (Fig. 5.5).

Finally, when considering secondary causes of conjunctivitis one must be aware that redness and congestion of the conjunctiva with secondary infection may be an indicator of *systemic disease.* Examples of this are the red eye of renal failure and gout and also polycythaemia rubra. The association of conjunctivitis, arthritis and non-specific urethritis makes up the triad of Reiter's syndrome. Some diseases cause abnormality of the tears and these have already been discussed with dry eye syndromes, the most common being

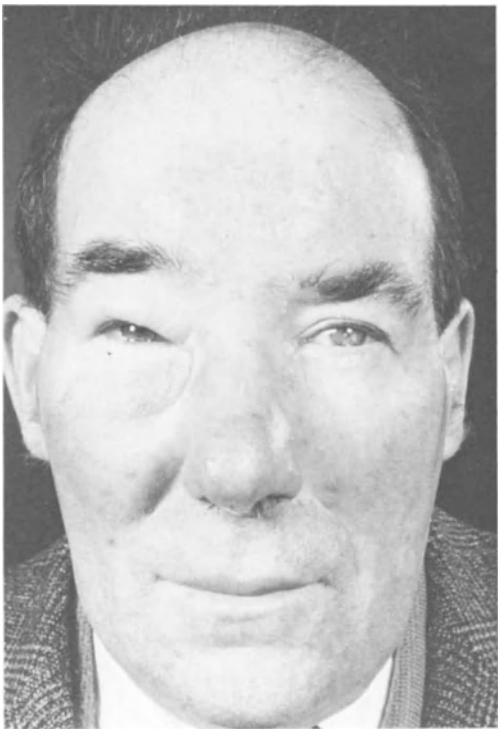

Fig. 5.6. Acne rosacea.

rheumatoid arthritis, but there are other rarer diseases which upset the quality or production of tears such as sarcoidosis, pemphigus and Stevens–Johnson syndrome. Thyrotoxicosis is a more common systemic disease which is associated with conjunctivitis, but the other eye signs such as lid retraction, conjunctival oedema and proptosis are usually more evident. A rather persistent type of conjunctivitis is seen in patients with acne rosacea. Here, the diagnosis is usually, but not always, made evident by the appearance of the skin of the nose, cheeks and forehead, but the corneal lesions of rosacea are also quite characteristic. The cornea becomes invaded from the periphery by wedge-shaped tongues of blood vessels associated with recurrent corneal ulceration. Rosacea keratoconjunctivitis is seen less commonly now, perhaps because it responds well to treatment with local and systemic tetracycline, a treatment which has now largely replaced the use of local steroids (Fig. 5.6).

Corneal Foreign Body

Small particles of grit or dust very commonly become embedded in the cornea and every casualty officer is aware of the increasing incidence of this occurrence on windy dry days. Small foreign bodies also become embedded as the result of using high-speed grinding tools without adequate protection of the eyes. The dentist's drill may also be a source of foreign bodies, but the most troublesome are those particles that have been heated by grinding or chiselling. It is important to have some understanding of the anatomy of the cornea if one is attempting to remove a corneal foreign body. One must realise, for example, that the surface epithelium can be stripped off from the underlying layer and can regrow and fill raw areas with extreme rapidity. Under suitable conditions the whole surface epithelium can reform in about 48 hours. The layer underlying or posterior to the surface epithelium is known as Bowman's membrane and if this layer is damaged by the injury or cut into

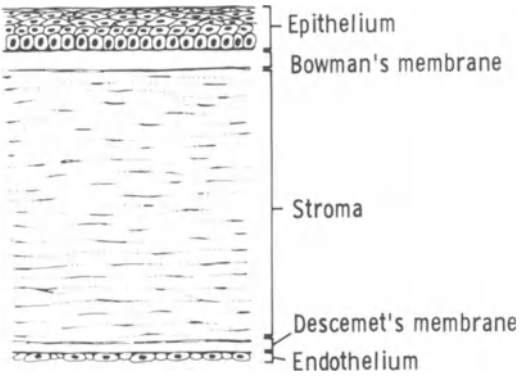

Fig. 5.7. The cornea.

unnecessarily by overzealous use of surgical instruments, then a permanent scar may be left in the cornea. When the epithelium alone is involved there is usually no scar, and healing results in perfect restoration of the optical properties of the surface.

Figure 5.7 shows the normal anatomy of the cornea. Remember that the epithelium is covered by the tear film. This is actually in three layers: a mucous layer immediately against the epithelium, a watery layer, and an outermost oily layer. The integrity of these three components of the tear film is essential for seeing, and small irregularities on the surface of the cornea may upset the tear film as well as the basic optical properties of the tissue itself. The stroma of the cornea is surprisingly tough, permitting some degree of boldness when removing deeply embedded foreign bodies, but it should be remembered that if the cornea has been perforated, then the risk of intraocular infection or loss of aqueous dictates that the wound should be repaired under full sterile conditions in the operating theatre.

Signs and Symptoms

Patients usually know when a foreign body has gone into their eye and the history is clear cut — but not always. Occasionally the complaint is simply a red sore eye which may have been present for some time. Spotting these corneal foreign bodies is really lesson number one in ocular examination. It involves employing the important basic principles of examining the anterior segment of the eye. Most foreign bodies can be seen without the use of the slit-lamp microscope if the eye is examined carefully and with a focused beam of light. Figure 5.8 demonstrates the great advantage of the focused beam, and in fact this very principle is used in slit-lamp microscopy. If the foreign body has been present for any length of time, there will be a ring of ciliary injection around the cornea due to the dilatation of the deeper episcleral capillaries which lie near the corneal margin. Ciliary injection is a sure warning sign of corneal or intraocular pathology.

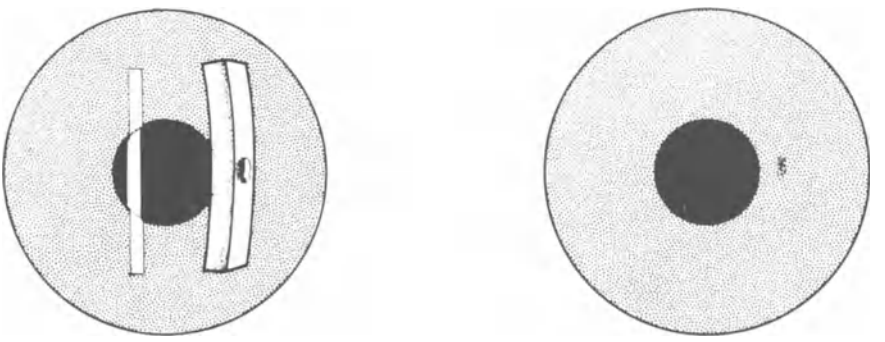

Fig. 5.8. Focal illumination of corneal foreign body.

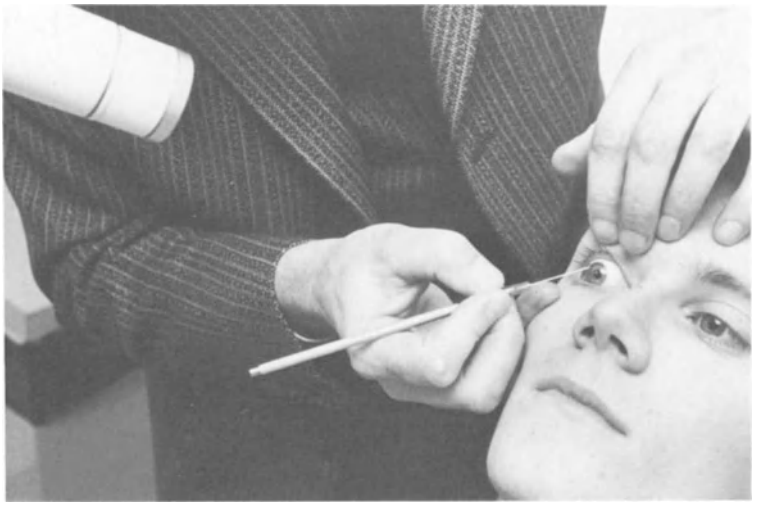

Fig. 5.9. Removing a corneal foreign body.

Treatment

The aim of treatment is, of course, to remove the foreign body completely. Sometimes this is not as easy as it may seem, especially when a hot metal particle lies embedded in a 'rust ring'. In instances when it is clear that much digging is going to be needed, it may be prudent to leave the rust ring for 24 hours, after which it becomes more easy to remove. The procedure for removing a foreign body should be as follows: the patient lies down on a couch or dental chair and one or two drops of proparacaine hydrochloride 0.5% (Ophthaine) or a similar local anaesthetic are instilled onto the affected eye. A good light on a stand is needed, preferably one with a focused beam, and the eyelids are held open with a speculum (Fig. 5.9). The doctor will also usually require some form of optical aid in the form of special magnifying spectacles, for example 'Bishop Harman's glasses'. Many foreign bodies can be easily removed with a cotton-wool bud and this should always be tried first. If this fails, a blunt spud may succeed. When the foreign body is more deeply embedded, it will be necessary to pick it off with a sharp pointed instrument such as a scalpel blade. There are a variety of instruments specially designed for this purpose. Once the foreign body has been removed, an antibiotic drop is placed in the eye and the speculum is withdrawn. The lids are then splinted together by means of a firm pad and bandage. (A light pad under which the eyelids can open and shut freely is of no benefit and simply a nuisance to the patient.) There is no doubt that the corneal epithelium heals more quickly if the eyelids are splinted in this way. It is usually advisable to see the patient the following day if possible to make sure that all is well, and if the damaged spot on the cornea is no longer staining with fluorescein, the pad can be left off. Antibiotic drops should be continued at least three times daily for a few days

after the cornea has healed, and the visual acuity of the patient should always be checked before he is finally discharged.

There are one or two factors that should always be borne in mind when treating patients with corneal foreign bodies: in most instances healing takes place without any problem but, very rarely, the vision may be permanently impaired by scarring. Also on very rare occasions, the site of corneal damage becomes infected and if neglected the infection may enter the eye and cause endophthalmitis with total blindness of the affected eye. This is a well-recognised tragedy which should never happen in an age of antibiotics. Of course, if the eye has been perforated, endophthalmitis is a more or less inevitable sequel in the absence of antibiotic treatment. One only has to examine old hospital case notes from the pre-antibiotic era to obtain proof of this.

Let us remember that **a perforating injury of the eye is a surgical emergency.** Any doubt about the possibility of a perforating injury of the cornea can usually be resolved by examining it carefully with the slit-lamp microscope. One other factor to bear in mind is the possibility of a retained intraocular foreign body. Sometimes the patient may be quite unaware of such an injury and this may mislead the doctor into underestimating the serious nature of the problem. The answer for the doctor is 'when in doubt, x-ray', especially when a hammer and chisel or high-speed drill have been used. A retained intraocular foreign body may not set up an inflammatory reaction until several weeks or even months have elapsed (Fig. 5.10).

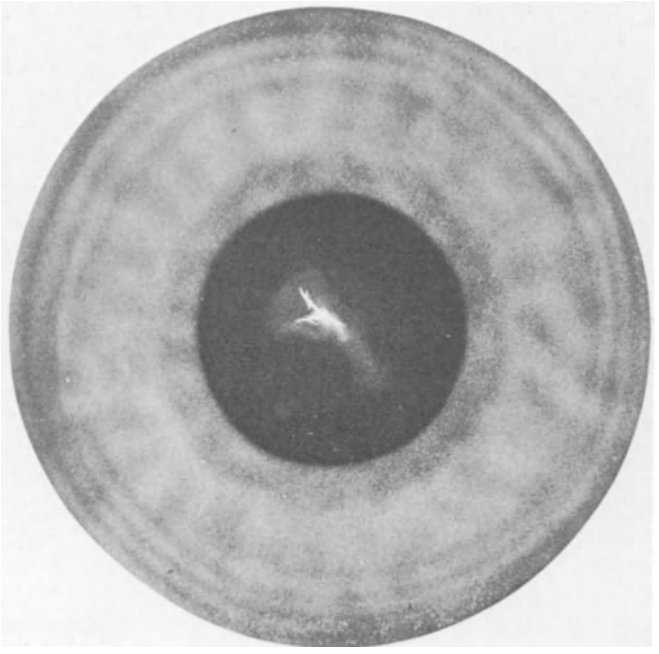

Fig. 5.10. Beware the full-thickness corneal scar. When in doubt, x-ray.

Corneal Ulceration

Corneal ulcers may arise spontaneously (primary) or they may result from some defect in the normal protective mechanism, or sometimes they are part of a more generalised susceptibility to infection (secondary). The nerve endings in the cornea are pain-sensitive endings and a light touch is felt as a sharp pain. Furthermore, stimulation of these nerves causes a vigorous blink reflex and the eye begins to water excessively. A very effective protective mechanism is therefore brought into action which tends to clear away infection or foreign bodies and warns the patient of trouble. In most instances of corneal ulceration the eye is painful, photophobic and waters. The conjunctiva is usually injected and there may be ciliary injection.

Types of Corneal Ulcer

Due to Direct Trauma

The corneal epithelium becomes disrupted and abraded by certain characteristic injuries. It is surprising how the same old story keeps repeating itself: the mother caught in the eye by the child's fingernail, the edge of a newspaper, the backlash from the branch of a tree. The injury is excruciatingly painful and the symptoms are often made much worse by the rapid eye movements of an anxious patient and sometimes by vigorous rubbing of the eye. The patient complains that there is something in the eye and once the diagnosis has been made it may be difficult to persuade the patient that there is no foreign body. A denuded area of cornea is seen which stains with fluorescein. It may not be possible to examine the patient until a drop of local anaesthetic has been instilled into the eye, but, as a general rule, **local anaesthetic drops should not be used to treat 'sore eyes'.** This is because healing is impaired and serious damage to the eye may result. Anaesthetic drops should only be used as a single-dose diagnostic measure in such cases. Treatment involves the instillation of a mydriatic such as homatropine 1% or an antibiotic such as chloramphenicol 0.5%, after which especial care is needed to fix the eyelids. This is probably best achieved by directly sticking the eyelids together with two vertically placed short strips of micropore surgical tape. Over the closed eyelids is then placed a square of Vaseline gauze, and over this a pad and firm bandage. The patient is then given some analgesic tablets to take home and is advised to rest quietly until the eye is inspected the following day. The pad can be left off once the epithelium has healed over, but even then the patient should continue to instil an antibiotic ointment in the eye at night for several weeks. The reason for taking a little trouble over the management of a patient with a corneal abrasion is the recurrent nature of the condition. All too often after some months or even a few years, the patient begins to experience a sharp pain in the injured eye on waking in the morning. It is as if the cornea, or the weak part of the cornea, becomes stuck to the

posterior surface of the upper lid during the night. It has been claimed that recurrent corneal abrasion is related to failure to close the eyes completely during sleep. The pain wears off after an hour or two and when the patient presents to the doctor there may be no obvious cause for the symptoms. In actual fact careful examination with the slit lamp reveals very minute cysts or white specks at the site of the original abrasion, indicating a weak area of attachment of the corneal epithelium. Severe recurrent corneal abrasion is best dealt with in an eye department where slit-lamp control is available.

Due to Bacteria

The commonest ulcer of this type is known as a 'marginal ulcer'. The patient complains of a persistently red eye which is moderately sore. Examination reveals conjunctival congestion which is often mainly localised to an area adjacent to the corneal ulcer. The ulcer is seen as a white spot near the corneal margin but there is usually a small gap of clear cornea between it and the limbus (the corneoscleral junction). Such marginal ulcers are thought to be due to *Staphylococcus aureus*, mainly because they are often associated with *Staphylococcus aureus* blepharitis. On the other hand, it is not possible to grow the organism from the corneal lesion, and for this reason it is said that the infiltrated area is some form of allergic response to the infecting organism. Furthermore, these marginal ulcers respond very rapidly to treatment with a steroid–antibiotic mixture. It is essential that the usual precautions before applying local steroids to the eye are taken, that is to say, the possibility of herpes simplex infection should be excluded and the intraocular pressure should be monitored if the treatment is to continue on a more long-term basis. A wide range of other bacteria are known to cause corneal ulceration, but, by and large, infections only occur as a secondary problem when the defences of the cornea are impaired.

Due to Viruses

Apart from other rare types of virus infection, there is one outstanding example of this — herpes simplex keratitis. The condition seems to be more common than it used to be, perhaps because the incidence of other types of corneal ulcer has become less with the more liberal use of local antibiotics on the eye. Every eye casualty department has one or two patients with the debilitating condition which may put a patient off work for many months. Fortunately it is only certain cases that cause such a problem, and most instances of this common condition give rise to a week or 10 days of incapacity. Herpes simplex is thought to produce a primary infection in infants which is transferred from the lips of the mother and may be subclinical. Sometimes a vesicular rash develops around the eyelids accompanied by fever and enlargement of the preauricular lymph nodes. Whatever the initial manifestation of primary infection, it is thought that many members of the population harbour the virus in a latent form so that overt infection in an adult

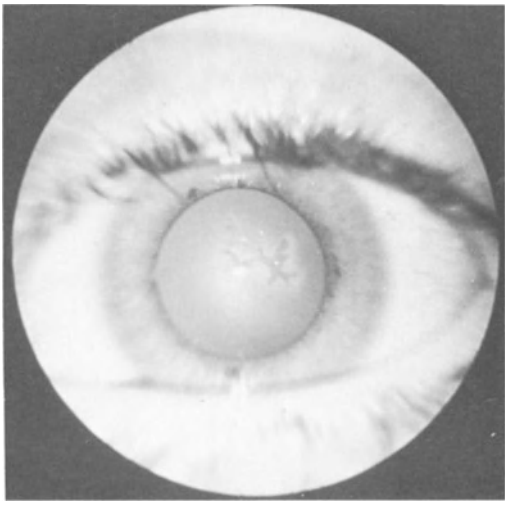

Fig. 5.11a. Dendritic ulcer of cornea.

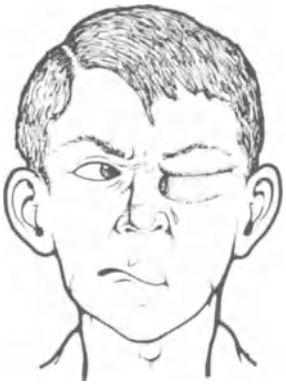

Fig. 5.11b. "My eye seemed much better at first on those steroid drops."

tends to appear in association with other illnesses. Most people are familiar with the cold sores that appear on the lips due to herpes simplex. Sometimes, after a cold, one eye becomes sore and irritable and inspection of the cornea shows the very characteristic corneal changes of herpes simplex infection. A slightly raised granular, star-shaped or dendriform lesion is seen which takes up fluorescein (Fig. 5.11). The virus can be cultivated from this lesion and the size of the dendriform figure is some guide to prognosis. A large lesion extending across the cornea, especially across the optical axis (i.e. the centre of the cornea), is likely to be the one which is going to give trouble and it is better that the patient should be warned about it at this stage. After a few days, or sometimes weeks, the epithelial lesion heals and at this point complete resolution may occur or an inflammatory reaction may appear in the stroma deep to the infected epithelium. The eye remains red and irritable to an

incapacitating degree and further dendritic ulcers may subsequently appear. In worse cases the cornea may become anaesthetic so that although the eye may be more comfortable, the problems of a numb cornea are added to the original condition. Healing tends to occur with a vascular scar.

Treatment of Herpes Simplex Keratitis. The initial management depends on the size of the lesion and the age of the patient. Small dendritic ulcers in young people where the lesion is away from the optical axis may be treated by cauterising the area of damaged epithelium. After instilling local anaesthetic drops, the lids are held apart with a speculum and the diseased epithelium is removed with a cotton-wool bud. The area is then touched with iodine, ether, or copper sulphate, an antibiotic drop is instilled and a firm pad and bandage applied. Following this procedure the eye may become very sore and the patient is given an analgesic and advised to remain in bed. Often the corneal epithelium will heal after 48 hours and the condition will be cured. Larger ulcers may not respond very satisfactorily to this treatment, and it is dangerous to cauterise the elderly cornea. Where cautery is not indicated or where there has been a recurrence in spite of cautery, an antiviral agent together with an antibiotic should be instilled locally. Examples of currently used antiviral agents are idoxuridine, trifluorothymidine, cytarabine, acyclovir and human interferon inducers. Unfortunately none of these agents is curative, but they are thought to have some effect on acute rather than chronic cases. Early diagnosis and treatment seem to give the best chance of avoiding recurrences. **Steroids should not be used in the treatment of dendritic ulcers of the cornea** (Fig. 5.11a). It is well recognised that steroid drops enhance the replication of the herpes simplex virus. They reduce the local inflammatory reaction and may give the false impression that the eye is improving. However, persistent use of local steroids in such cases may result in corneal thinning and even corneal perforation. Once the dendritic ulcer has healed, residual stromal infiltration is then sometimes treated by carefully gauged doses of steroids, but this should be under strict ophthalmological supervision. In more severe cases, secondary iritis or secondary glaucoma may complicate the picture and require special treatment. The decision whether or not to apply a pad to the eye depends on the state of the corneal epithelium and also on the patient's response. In the worst cases it may be advisable to perform a tarsorrhaphy, that is to say, the lids are stitched together in such a way that they remain closed when the stitches are removed. Surprisingly, the keratitis seems to heal usually in 1 to 2 weeks when this is done and the patient may be able to return to work providing the work does not require the use of both eyes. It is unwise to open the tarsorrhaphy for many months, preferably leaving it for at least a year. When herpetic keratitis has taken its toll leaving a scarred cornea, the sight may eventually be restored again by a corneal graft. Unfortunately, recurrences still often occur and dendritic ulcers may appear on the graft.

Due to Damage to the Corneal Nerve Supply

When the ophthalmic division of the trigeminal nerve is damaged by disease or

injury, the cornea may become numb and there is a high risk of corneal ulceration. Such neurotrophic ulcers are characteristically painless and easily become infected, with possible disastrous results. A tarsorrhaphy may be needed to save the eye but sometimes a soft contact lens may suffice. Before embarking on the treatment of an anaesthetic cornea, the cause should be established and this may involve a full neurological investigation.

Due to Exposure

When the normal 'windscreen wiper' mechanism of the lids is faulty, as, for example, when the eyelids have been injured or in a case of facial palsy, then the surface of the cornea may dry and become ulcerated. The same problem occurs in the unconscious patient unless great care is taken to keep the eyelids closed. Most cases of Bell's palsy recover sufficiently quickly to prevent exposure keratitis, but when severe and when recovery is poor, a tarsorrhaphy, or at least treatment with an eyepad and local anaesthetic ointment at night, may be needed. It is important to bear in mind that the same risk is evident in patients with severe thyrotoxic exophthalmos.

Corneal Degeneration

There are a number of specific corneal degenerations, some of which are inherited and most of which cannot be diagnosed without the aid of the slit-lamp microscope. For this reason they will not be dealt with in any detail here. Keratoconus (or conical cornea) is perhaps the commonest. It is still rare in the general population but is familiar to general practitioners looking after student populations because it tends to appear in this age group. The condition is bilateral and may be inherited as an autosomal recessive trait. It should be suspected in patients who show a rapid change of refractive error, particularly if a large amount of myopic astigmatism suddenly appears. Often, but not always, there is an associated history of asthma and hay fever. The cornea shows central thinning and protrudes anteriorly. This may be observed with the naked eye by asking the patient to sit down and then standing behind him so that one can look down on his downturned eye. By holding up the upper lids one can make an estimate of the abnormal shape of the cornea by noting how the cornea shapes the lower lid. Alternatively, the patient's cornea can be observed using Placido's disc. This ingenious instrument is simply a disc with a hole in the centre of it through which one observes the patient's cornea. On the patient's side of the disc is a series of concentric circles which can be seen by the observer reflected on the patient's cornea (Fig. 5.12). Distortion of these circles indicates the abnormal shape of the cornea. Of course, more accurate assessment of the cornea can be made by observing it with the slit-lamp microscope and still more information can be obtained by keratometry, that is, using an instrument to measure the curvature of the cornea in different meridians. Keratoconus tends to progress slowly and contact lenses may be very helpful. Sometimes a corneal graft is required.

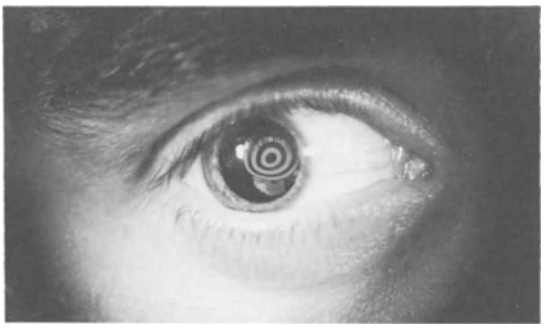

Fig. 5.12. Placido's disc in the diagnosis of keratoconus. The image of the disc can be seen reflected from the cornea.

Apart from the inherited corneal degenerations, certain changes are often seen in the cornea with ageing such as arcus senilis and endothelial pigmentation. Band degeneration refers to a deposition of calcium salts in the anterior layers of the cornea. The calcification is first seen at the margin of the cornea in the 'nine o'clock and three o'clock' area, but it may gradually extend across the normally exposed part of the cornea. It is seen in cases of chronic iridocyclitis, in particular in patients with juvenile rheumatoid arthritis and also in those with sarcoidosis. In fact, band degeneration is seen in any eye which has become degenerate or in cases of long-standing corneal disease (Fig. 5.13). Although band degeneration and keratoconus can, if sufficiently advanced, be diagnosed quite easily with the naked eye, most degenerative conditions of the cornea can only be diagnosed and classified under the microscope. A rare but dramatic example is the Kayser–Fleischer ring. This is a ring of brown pigment which is seen lying deeply in the cornea around the corneal margin but just inside the limbus (the corneoscleral junction). The

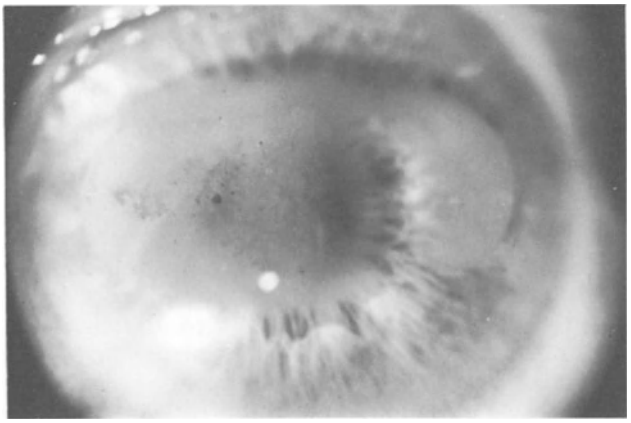

Fig. 5.13. Band degeneration of the cornea.

appearance is pathognomonic of hepatolenticular degeneration. A diffuse pigment stippling of the inner surface of the cornea, which again is only visible with the microscope, may be part of the much more common clinical picture known as Fuch's dystrophy. The observation of such changes takes on a special importance when cataract surgery is being considered and they will be discussed in more detail in a later chapter (see Chapter 10).

Corneal Oedema

To the naked eye, corneal oedema may not be very obvious but careful inspection will reveal a lack of lustre when the affected cornea is compared with that on the other side. The normal sparkle of the eye is no longer evident and the iris has become less well defined. Microscopically a bedewed appearance is seen, minute droplets being evident in the epithelium. When the stroma is also involved this may seem misty and may be also infiltrated with inflammatory cells which are seen as powdery white dots. When the oedema is long standing the droplets in the epithelium coalesce to produce blisters or bullae.

The more important causes of corneal oedema are as follows.

1. Acute narrow angle glaucoma
2. Virus keratitis
3. Trauma
4. Contact lenses
5. Postoperative
6. Fuch's endothelial dystrophy

When the intraocular pressure is suddenly raised from any cause, the cornea becomes oedematous. The normal cornea requires to be relatively dehydrated in order to maintain its transparency, and the necessary level of dehydration seems to depend on active removal of water by the corneal endothelium as well as an adequate oxygen supply from the tears. This mechanism is impaired not only by raising the intraocular pressure, but also by infection or trauma. Senile degenerative changes may also be the sole underlying cause due to failure of the endothelial pumping mechanisms. Contact lenses, if ill-fitting and worn for too long a period, may prevent adequate oxygen reaching the cornea, with resulting oedema.

The management of corneal oedema depends on the management of the underlying cause. Oedema due to endothelial damage may respond in its early stages to local steroids and sometimes a clear cornea may be maintained by the use of osmotic agents such as hypertonic saline or glycerol. Chronic corneal oedema tends to be painful and often acute episodes of pain occur when bullae rupture leaving exposed corneal nerves. In such cases it may be necessary to consider a tarsorrhaphy, or in some instances a corneal graft may prove beneficial. The pain of corneal oedema is a late symptom and in its early stages

oedema simply causes blurring of the vision and the appearance of coloured haloes around light bulbs. This is simply a 'bathroom window' effect. Haloes are also seen by patients with cataracts, so that defects in other parts of the optical media of the eye may give a similar effect.

Absent Corneal Sensation

Corneal sensation is supplied by the fifth nerve. About 70 nerve fibres are present in the superficial layers of the cornea and they can often be seen when the cornea is examined with the slit-lamp microscope. They appear as white threads running mainly radially. Corneal anaesthesia can be assessed by asking the patient to gaze straight ahead and then lightly touching the cornea with a fine wisp of cotton wool. Care must be taken not to touch the lid margins when doing this. The blink reflex is then noted and it is also important to ask the patient what has been felt. In the case of elderly people the blink reflex may be reduced, but a slight prick should be evident when the cornea is touched. Attempts to quantify corneal anaesthesia have led to the development of graded strengths of bristle which can be applied to the cornea instead of cotton wool.

Corneal anaesthesia may result from a lesion at any point in the fifth cranial nerve from the cornea to the brainstem. In the cornea itself, herpes simplex infection may ultimately result in anaesthesia. Herpes zoster is especially liable to lead to this problem and, because this condition may often be treated at home rather than in the ward, it will be considered in more detail here.

Herpes Zoster Ophthalmicus

This is due to the varicella–zoster virus, the same virus that causes chickenpox. It is thought that the initial infection with the virus occurs with an attack of childhood chickenpox and that the virus remains in the body in a latent form, subsequently to manifest itself as herpes zoster in some individuals. The virus appears to lodge in the Gasserian ganglion. The onset of the condition is heralded by headache and the appearance of one or two vesicles on the forehead. Over the next 3 or 4 days the vesicles multiply and appear in the distribution of one or all of the branches of the fifth cranial nerve. The patient may develop a temperature and usually experiences malaise and considerable pain. Sometimes a chickenpox-like rash appears over the rest of the body. The eye itself is most at risk when the upper division of the fifth nerve is involved; there may be vesicles on the lids and conjunctiva, and when the cornea is affected punctate-staining areas are seen which become minute subepithelial opacities. After 4 days to a week, the infection reaches its peak; the eyelids on the affected side may be closed by swelling, and oedema of the lids may spread across to the other eye (Fig. 5.14). The vesicles become pustular and then form crusts which are then shed over a period of 2 or 3 weeks. In most cases

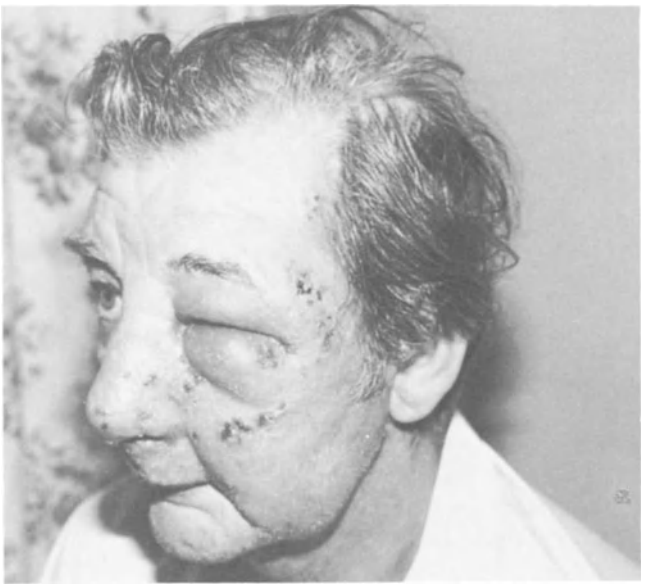

Fig. 5.14. Herpes zoster of the eye.

complete resolution occurs with remarkably little scarring of the skin considering the appearance in the acute stage. However, the cornea may be rendered permanently anaesthetic and the affected area of skin produces annoying paraesthesiae, amounting quite often to persistent rather severe neuralgia which may dog the patient for many years. Other complications include extraocular muscle palsies or, rarely, encephalitis. Iridocyclitis is fairly common and glaucoma may develop and lead to blindness if untreated. At present there is no known effective treatment other than the use of local steroids for the uveitis and acetazolamide for the glaucoma. The disease has to run its course and the patient, who is usually elderly, may require much support and advice, especially when post-herpetic neuralgia is severe. It is accepted practice to treat the eye at risk with antibiotic drops and a weak mydriatic. Analgesics are, of course, also usually needed, often on a long-term basis.

Other causes of corneal anaesthesia include surgical division of the fifth cranial nerve for trigeminal neuralgia or any space-occupying lesion along the nerve pathway. The possibility of exposure and drying of the cornea must always be borne in mind in the unconscious or the anaesthetised patient since corneal ulceration and infection will soon result if this is neglected.

Corneal anaesthesia due to nerve damage is nearly always permanent and, if it is complete, it may often be necessary to protect the eye by means of a tarsorrhaphy. Lesser degrees of corneal anaesthesia may be treated by instilling an antibiotic ointment at night and, if a more severe punctate keratitis develops, by padding the eye.

6 The Red Eye

Redness of the eye is one of the commonest signs in ophthalmology, being a feature of a wide range of ophthalmological conditions, some of which are severe and sight threatening whereas some are mild and of little consequence. Occasionally the red eye may be the first sign of important systemic disease. It is important that any practising doctor has an understanding of the differential diagnosis of this common sign, and a categorisation of the signs, symptoms and management of the red eye will now be made from the standpoint of the non-specialist general practitioner.

First of all, let me point out that the simplest way of categorising these patients is in terms of their visual acuity. As a general rule, if the sight, as measured on the Snellen test chart, is impaired, then the cause may be more serious. The presence or absence of pain is also of significance but as this depends in part on the pain threshold of the subject, it may be a misleading symptom. Disease of the conjunctiva alone is not usually painful whereas disease of the cornea or iris is generally very painful.

The red eye will therefore be considered under two headings: the red eye which sees well and is not painful, and the red eye which does not see well and is acutely painful.

Red Eye Which Is Not Painful and Sees Normally

Subconjunctival Haemorrhage

Careful examination of the eye will easily confirm that its redness is due to blood rather than dilated blood vessels, and the redness may be noticed by someone other than the patient. The condition is common and resolves in about 10 days. It is extremely unusual for a blood dyscrasia to present with subconjunctival haemorrhages, and although vomiting or a bleeding tendency may also be rare causes, the normal practice is to reassure the patient rather than embark on extensive investigations, because the majority of cases are due to spontaneous bleeding from a conjunctival capillary.

Conjunctivitis

Examination of the eye will reveal inflammation, that is, dilatation of the conjunctival capillaries and larger blood vessels, associated with more or less discharge from the eye. The exact site of the inflammation should be noted and it is especially useful to note whether the deeper capillaries around the margin of the cornea are involved. The resulting pink flush encircling the cornea is called 'ciliary injection' and is a warning of corneal or intraocular inflammation. For clinical purposes it is useful to divide conjunctivitis into acute and chronic types.

Acute

This is usually infective and due to a bacterium; it is more common in young people. It may spread rapidly through families or schools without serious consequence other than a few days' incapacity. When adults develop acute conjunctivitis it is worth searching for a possible underlying cause, especially a blocked tear duct when the condition is unilateral. Sometimes an ingrowing lash may be the cause or occasionally a free-floating eyelash lodges in the lacrimal punctum. The important symptoms of acute conjunctivitis are redness, irritation and sticking together of the eyelids in the mornings. Management entails finding the cause and using antibiotic drops if the symptoms are severe enough to warrant this. However, it must be remembered that the inadequate and intermittent use of antibiotic eyedrops may simply encourage growth of resistant organisms.

Chronic

This is a very common cause of the red eye and almost a daily problem in non-specialised ophthalmic practice. If we consider that the conjunctiva is a mucous membrane which is daily exposed to the elements, it is perhaps not surprising that after many years it tends to become chronically inflamed and irritable. The frequency and nuisance value of the symptoms are reflected in the large across-the-counter sales of various eyewashes and solutions aimed at relieving 'eyestrain' or 'tired eyes'. The symptoms of chronic conjunctivitis are therefore redness and irritation of the eyes with a minimal degree of discharge and sticking of the lids. If there is an allergic background, itching may also be a main feature. The chronically inflamed conjunctiva accumulates minute particles of calcium salts within the mucous glands. These conjunctival concretions are shed from time to time producing a feeling of grittiness. When confronted with such a patient there are a number of key symptoms to be elicited and these can be related to a check list of causes mentioned below.

Key Symptoms
Environmental factors, especially eyedrops, make-up or foreign bodies
Lids stick in mornings?
Do the eyes itch?
Emotional stress or psychiatric illness?

Check List of Causes

Eyelids
Deformities such as entropion or ectropion
Displaced eyelashes
Chronic blepharitis

Refractive error
A proportion of patients who have never worn glasses and need them or who are wearing incorrectly prescribed or out-of-date glasses present with the features of chronic conjunctivitis, the symptoms being relieved by the proper use of spectacles. The cause is not clear but possibly related to rubbing the eyes.

Dry eye syndrome
The possibility of a defect in the secretion of tears or mucus can only be confirmed by more elaborate tests, but this should be suspected in patients with rheumatoid arthritis or sarcoidosis.

Foreign body
Contact lenses and mascara particles are the commonest foreign bodies to cause chronic conjunctivitis.

Stress
Often a period of stress seems to be closely related to the symptoms and perhaps eye rubbing is also the cause in these patients.

Allergy
It is very unusual to be able to incriminate a specific allergen for chronic conjunctivitis, unlike allergic blepharitis. On the other hand, hay fever and asthma may be the background cause.

Infection
Chronic conjunctivitis may begin as an acute infection, usually viral and usually following an upper respiratory tract infection.

Drugs
The long-term use of adrenaline drops may cause dilatation of the conjunctival vessels and irritation in the eye. In 1974 it was shown that the ß-blocking drug practolol (since withdrawn from the market) could cause a severe dry eye syndrome in rare instances. Since then there have been several reports of mild reactions to other available ß-blockers, although such reactions are difficult to distinguish from chronic conjunctivitis from other causes.

Systemic causes
Congestive cardiac failure, renal failure, Reiter's disease, polycythaemia, gout, rosacea as well as other causes of orbital venous congestion such as orbital tumours may all cause vascular congestion and irritation of the conjunctiva. Migraine may also be associated with redness of the eye on one side and chronic alcoholism is a cause of bilateral conjunctival congestion.

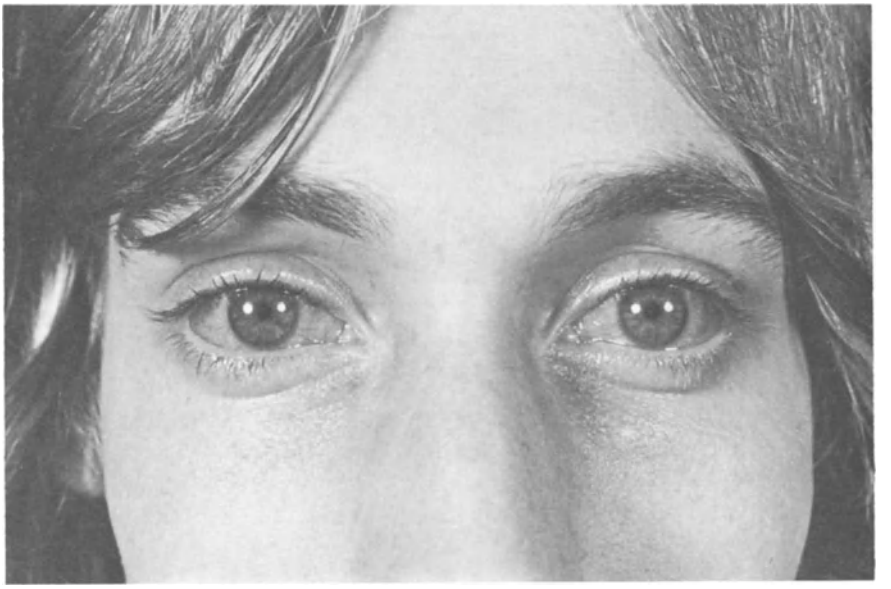

Fig. 6.1. Episcleritis. The condition is usually unilateral; this case was unusual.

Episcleritis

Sometimes the eye becomes red due to inflammation of the connective tissue underlying the conjunctiva, that is, the episclera. The condition may be localised or diffuse. There is no discharge and the eye is uncomfortable although not usually painful. The condition responds to sodium salicylate given systemically and to the administration of local steroids, and the underlying cause is often never discovered, although there is a well-recognised link with the collagen diseases, especially rheumatoid arthritis. Episcleritis tends to recur and may persist for several weeks producing a worrying cosmetic blemish in a young person (Fig. 6.1).

Scleritis

Inflammation of the sclera is a less common cause of the red eye; the aetiology is similar to that of episcleritis. It is most often seen in association with rheumatoid arthritis and sometimes may become severe and progressive to the extent of causing perforation of the globe. For this reason steroids must be administered with extreme care.

Red Painful Eye Which Cannot See

It is worth emphasising again that the red painful eye with poor vision is likely to be a serious problem, often requiring urgent admission to hospital or at least intensive out-patient treatment as a sight-saving measure. The following are the principal causes.

Acute Glaucoma

The important feature here is that acute glaucoma occurs in long-sighted people and there is usually a previous history of headaches and seeing haloes round lights in the evenings. The iris sphincter is damaged by the raised intraocular pressure and for this reason the pupil is semidilated. Oedema of the cornea causes the eye to lose its lustre and gives the iris a hazy appearance. The eye is extremely tender and painful and the patient may be nauseated and vomiting. Immediate admission to hospital is essential where the intraocular pressure is first controlled medically and then surgery is carried out to prevent recurrence, both on the affected eye and then, usually 1 week later, on the unaffected eye (Fig. 6.2). **Mydriatics should never be given to patients with suspected narrow angle glaucoma.**

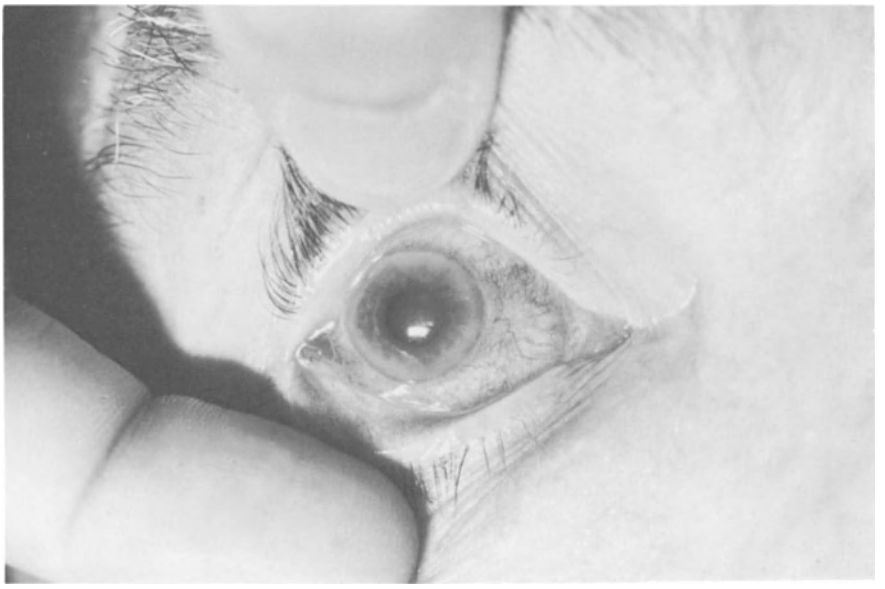

Fig. 6.2. Acute glaucoma. The patient had been vomiting. The cornea looked hazy and the pupil was semidilated.

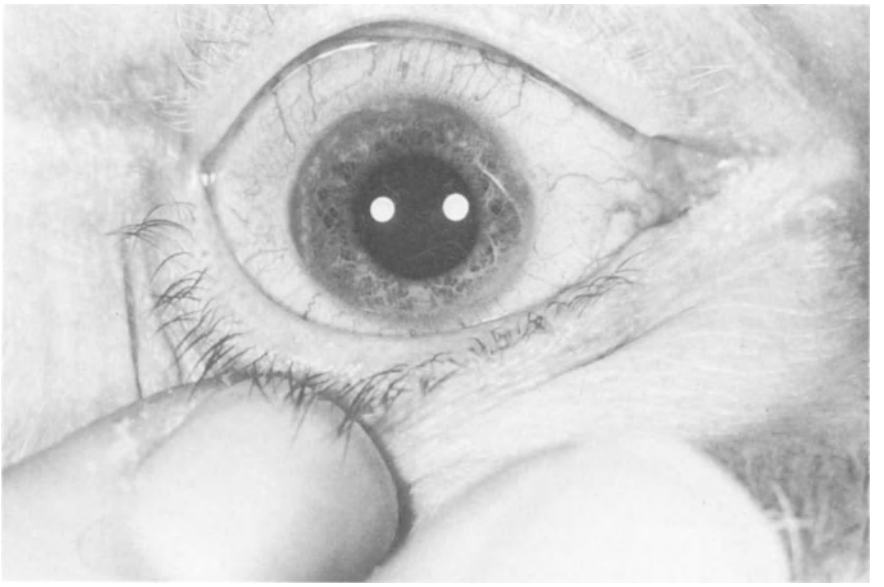

Fig. 6.3. Acute iritis. The cornea is bright; the pupil was originally constricted but has been dilated with mydriatic drops.

Acute Iritis

The eye is painful, especially when attempting to view near objects, but the pain is never so severe as to cause vomiting. The cornea remains bright and the pupil tends to go into spasm and is smaller than on the normal side. Acute iritis is seen from time to time mainly in the 20–40 age group, whereas acute glaucoma is extremely rare at these ages. Unless severe and bilateral, acute iritis is treated on an out-patient basis with local steroids and mydriatic drops (Fig. 6.3). Some expertise is needed in the use of the correct mydriatic, and systemic steroids should be avoided unless the sight is in jeopardy. Because the iris forms part of the uvea, acute iritis is the same as acute anterior uveitis. In many cases no systemic cause can be found but it is important to exclude the possibility of sarcoidosis or ankylosing spondylitis. It is also advisable to treat any concomitant infection of the teeth or sinuses. The condition lasts for about 2 weeks but tends to recur over a period of years. After two or three recurrences there is a high risk of the development of cataract, although this may form slowly.

Acute Keratitis

The characteristic features are sharp pain, often described as a foreign body in the eye, marked watering of the eye, photophobia and difficulty in opening the

affected eye. The clinical picture is very different from those of the above two conditions and the commonest causes are the herpes simplex virus or trauma. The possibility of a perforating injury must always be borne in mind. Sometimes children are reticent about any history of injury for fear of incriminating a friend, and sometimes a small perforating injury is surprisingly painless. The treatment of acute keratitis has already been discussed in this chapter and the management of corneal injuries will be considered in Chapter 15.

Thrombotic Glaucoma

The elderly patient who presents with a blind and painful eye and who may also be diabetic should be suspected of having thrombotic glaucoma. A fairly well-defined sequence of events enables the diagnosis to be inferred from the history. The first event is occlusion of the central retinal vein on one side and the patient notices that the vision of one eye has become blurred. Some elderly patients do not seek attention at this stage and some degree of spontaneous recovery may seem to occur before the onset of secondary glaucoma. Fortunately, only a small proportion of cases develop this severe complication, which usually occurs, surprisingly enough, after 100 days, hence the term 'a hundred day glaucoma'. Once the intraocular pressure rises, the eye tends to become painful and eventually degenerates in the absence of treatment, and sometimes even in spite of treatment, and thrombotic glaucoma remains as one of the few indications for surgical removal of the eye.

7 Failing Vision

Failing Vision in an Eye Which Looks Normal

Very often a patient will present with reduction of vision in one or both eyes and the eyes themselves may look normal. In the case of a child, the parents may have noticed an apparent difficulty in reading or the vision may have been noticed to be poor at routine school testing. If ophthalmoscopy reveals normal fundi, the diagnostic field is considerably narrowed but it is vital to check the pupil reaction before instilling mydriatic drops to examine the fundus. At any age, the possibility of refractive error must first be excluded by testing the patient for spectacles. When the fundus is normal, the likely diagnosis depends largely on the age of the patient. Infants with visual deterioration usually require an examination under anaesthesia to exclude the possibility of an inherited retinal degeneration, and electroretinography must be performed at the same time. Other children, particularly those in the age group from 9 to 12 years, must first be suspected of some emotional upset, perhaps due to domestic upheaval or stress at school, which may make them reluctant to read the test chart. Sometimes such children discover that exercising their own power of accommodation produces blurring of vision and they may present with accommodation spasm. The commonest cause of unilateral visual loss in children is amblyopia of disuse. This important cause of visual loss with a normal fundus is considered in more detail in the chapter on squint (Chapter 13). When, for whatever cause, one retina fails to receive a clear and correctly orientated image for a period of months or years during the time of visual development, then the sight of the eye remains impaired. The condition is treatable if caught before the visual reflexes are fully developed, that is, before the age of 8 years. Young adults who present with unilateral visual loss and normal fundi may, of course, also have untreated or inadequately treated amblyopia of disuse and the diagnosis may be confirmed by looking for a squint or a refractive error more marked on the affected side. We must also remember that retrobulbar neuritis presents in young people as sudden loss of vision on one side with aching behind the eye and a reduced pupil reaction on the affected side. This contrasts with amblyopia of disuse in which the pupil is normal. Migraine is another possibility to be considered in such patients.

Elderly patients who present with visual loss from normal fundi usually give the history of a stroke and are found to have a homonymous hemianopic field defect due to an embolus or thrombosis in the area of distribution of the posterior cerebral artery. Hysteria and malingering are also causes of unexplained visual loss, but these are extremely rare and it is important that the patient is investigated very carefully before such a diagnosis is made.

Quite a proportion of patients who complain of loss of vision with eyes that look normal on superficial inspection show changes on ophthalmoscopy. The three important potentially blinding but eminently treatable ophthalmological conditions must be borne in mind: cataract, chronic simple glaucoma, and retinal detachment. These diseases are limited to the eye itself, but disease elsewhere in the body can often first present as a visual problem and in this context we must remember what has been the commonest cause of blindness in young people — diabetic retinopathy, as well as the occasional case of severe hypertension. Intracranial causes of visual loss are perhaps less common in general practice and for this reason easily missed. Intracranial tumours may present in an insidious manner, in particular the pituitary adenoma, and the diagnosis may be first suspected by careful plotting of the visual fields. In the case of the elderly patient who complains of visual deterioration in one eye, the ophthalmoscope all too commonly reveals senile macular degeneration which is worse on one side. However, it is also common to find that the patient has suffered a thrombosis of the central retinal vein or one of its branches. Unlike the situation with a central retinal artery occlusion, which is less common, some vision is preserved when the central retinal vein is thrombosed, and indeed the dramatic fundus appearance may be at variance with the symptoms. Temporal arteritis is another important vascular cause of visual failure in the elderly.

Finally, there are a large number of less common conditions, only one or two of which will be mentioned at this point. At any age the ingestion of drugs may affect the sight, but there are very few proved oculotoxic drugs still on the

Table 7.1 Failing vision in a normal-looking eye

	Fundus normal	Fundus abnormal
Child	Refractive error Disuse amblyopia Inherited retinal degeneration Emotional stress	Cataract Macular degeneration Posterior uveitis
Young adult	Refractive error Retrobulbar neuritis Intracranial space-occupying lesion Drug toxicity	Diabetic retinopathy Retinal detachment Macular disease Hypertension Posterior uveitis
Elderly	Homonymous hemianopia	Temporal arteritis Macular degeneration Central vein thrombosis Chronic glaucoma Cataract Vitreous haemorrhage

market. One important example is chloroquine. When a dose of 100 g in 1 year is exceeded, there is a high risk of retinotoxicity which may not be reversible. Although senile macular degeneration is normally recognised as a disease of the elderly, the same problem may occur in younger people and, rarely, in children. In young adults a particular type of macular problem known as central serous retinopathy may occur. The condition tends to resolve spontaneously after a few weeks, although treatment by laser coagulation is occasionally needed. Unilateral progressive visual loss in young people may also be due to posterior uveitis, which is the same thing as choroiditis. The known causes and management of this condition will be discussed in a later chapter (see Chapter 17).

The more common causes of failing vision in a normal-looking eye may be summarised as shown in Table 7.1.

Treatable Causes

Nobody can deny that the practice of ophthalmology is highly effective. There are many eye diseases which can be cured or arrested and it is possible to restore the sight fully from total blindness. It is a tragedy of modern times that the commonest cause of blindness in the world — trachoma — is eminently treatable. The most important treatable cause of visual failure in Britain is cataract, and of course no patient should be allowed to go blind from this condition, although this does occasionally happen (Fig. 7.1). Retinal

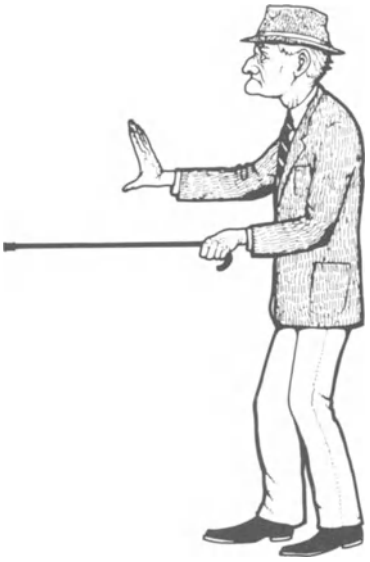

Fig. 7.1. "The family thought it was just old age."

detachment is less common than cataract but it provides a situation where the sight may be lost completely and then be fully restored. For the best results, surgery must be carried out as soon as possible, before the retina becomes degenerate, whereas delay prior to cataract surgery does not usually affect the outcome of the operation. Acute glaucoma is another instance where the sight is lost and restored by surgery. The treatment of chronic glaucoma has less impression on the patient because it is aimed at preventing visual deterioration, although in sight-saving terms it is equally effective.

It is very easy to overlook the value of antibiotics in saving sight. Prior to their introduction, many more eyes had to be removed following injury or infection. Systemic and locally applied steroids also play a sight-saving role in the management of temporal arteritis in the elderly and in the treatment of uveitis. In recent years considerable hope has appeared for patients with severe diabetic retinopathy. The combined effect of laser coagulation and scrupulous control of the diabetes is now reducing the number of cases of proliferative retinopathy who go blind each year. In the past, about half of the cases with this particular kind of retinopathy would be expected to go blind in 5 years and many of these patients were young and at the height of their careers. The proper management of ocular trauma often has a great influence on the visual result, and the rare but dreaded complication of ocular perforating injuries — sympathetic ophthalmia — can now be effectively treated with systemic steroids. Amblyopia of disuse has already been mentioned in this chapter. The treatment is undoubtedly highly effective in some cases but the results are disappointing when the diagnosis is made when the child is too old or when there is poor patient cooperation.

Untreatable Causes

Ophthalmologists are sometimes asked if the sight can be restored to a blind eye and, as a general rule, one can say that if there is no perception of light in the eye, then it is unlikely that the sight can be improved irrespective of the cause. There are several ophthalmological conditions for which there is no known effective treatment and it is sometimes important that the patient is made aware of this at an early stage in order to avoid unnecessary anxiety, and perhaps unnecessary visits to the doctor. Most degenerative diseases of the retina fail to respond to treatment. If the retina is out of place, it can be replaced, but old retinae cannot be replaced with new. So far, there has been no firm evidence that any drug can alter the course of the inherited retinal degenerations such as retinitis pigmentosa, although useful information is beginning to appear about the biochemistry of these conditions. Macular degeneration tends to run a progressive course, in spite of claims for effective treatment which have appeared from time to time over the years, and although most cases do not become completely blind, it accounts for the loss of reading vision in many elderly people. Some myopic patients are susceptible to degeneration of the retina in later years; known as myopic chorioretinal

degeneration, it may account for visual deterioration in myopes who have otherwise undergone successful cataract or retinal detachment surgery.

Scarring of the retina following trauma is another cause of permanent and untreatable visual loss, but the most dramatic and irrevocable loss of vision occurs following traumatic section of the optic nerve. However, one must be careful here before dismissing the patient as untreatable because on rare occasions a contusion injury to the eye or orbit may result in a haemorrhage into the sheath of the optic nerve. Some degree of visual recovery may occur in these patients and it has been claimed that recovery may be helped by surgically opening the nerve sheath. It is hardly necessary to say that any neurological damage proximal to the optic nerve tends to produce permanent and untreatable visual loss, as exemplified by the homonymous hemianopic field defect which may follow a cerebrovascular accident. There is one odd exception: visual loss due to optic neuritis. Patients with retrobulbar neuritis (optic neuritis) nearly always recover their vision again whether they receive treatment or not, the explanation being that the visual loss is due to pressure from oedema rather than to damage to the nerve fibres themselves.

Malignant tumours of the eye come into the category of untreatable causes of visual failure because the eye may require to be removed with the idea of avoiding metastatic spread. In fact serious attempts are now being made at removing tumours locally or treating by radiotherapy if at all possible, and this applies to the important malignant intraocular tumours, the malignant melanoma of the choroid and the retinoblastoma.

8 Headache

Headache must be one of the commonest symptoms, and few specialties escape from the diagnostic problems that it may present. We must begin with the realisation that more or less everyone suffers from headache at some time or another. In fact, the majority of headaches that present have no detectable cause and are often labelled psychogenic if there seems to be a background of stress. The implication is that the sufferer is perhaps exaggerating mild symptoms in order to gain sympathy from his or her spouse, or even perhaps the doctor (Fig. 8.1). One must, of course, be extremely cautious about not accepting symptoms at their face value, and certainly cerebral tumours have been overlooked for this very reason. If the psychogenic headache is the commonest, then headache due to raised intracranial pressure and an intracranial space-occupying lesion must be the most important, and between these two a whole spectrum of causes must be considered. It is essential, therefore, to memorise a permanent check list in order that obvious causes are not omitted.

Fig. 8.1. "It's like a massive pin sticking through my head, doctor."

History

Often the history is the total disease in the absence of any physical signs and it is therefore important to note the nature of the pain, the total duration and frequency of the pain, the time of day it occurs and its relation to other events or the taking of analgesics. Headaches that are present 'all the time' and are described in fanciful terms tend not to have an organic basis; the patient with an organic headache is not usually smiling. The time of day may be important: raised intracranial pressure has the reputation for causing an early morning headache which is described as bursting or throbbing and which may be made worse by straining or coughing. We must always remember the triad of headache, vomiting and papilloedema in this respect, especially as the vomiting may not be accompanied by nausea and is not necessarily mentioned by the patient. The family history should also be noted, especially where there is a history of migraine.

Classification

When considering the different common causes of headache, an anatomical classification is a useful way of providing a reference list. The following should be considered by the examining doctor.

Cerebrospinal Fluid

A rise or fall from normal of the cerebrospinal fluid pressure is associated with headache. When the CSF pressure is raised the patient usually experiences a bursting pain which may interrupt sleep or appear in the early morning. It tends to be intermittent and made worse by coughing or lying down. It may also, of course, be accompanied by papilloedema and vomiting, and another important symptom is transient blurring or obscuration of vision. The situation of the pain is usually diffuse rather than focal, but we must remember that a bursting headache made worse by coughing is sometimes described by otherwise healthy individuals. When the rise of intracranial pressure is due to a space-occupying lesion, signs of focal brain damage may also be present.

Blood Vessels

A variety of diseases involving the blood vessels may cause headache. The commonest is probably migraine. Classical migraine is thought to be due to an initial spasm followed by dilatation of the meningeal arteries. There is usually

a family history of the same problem showing dominant inheritance, and attacks may sometimes be precipitated by stress or taking certain foods such as cheese. Before the headache begins there is usually a visual aura characterised by a shimmering effect before one or both eyes which spreads across the vision, or the appearance of zig-zag lines known as fortifications because of their resemblance to the silhouette of a fortress. The visual disturbances may take the form of a hemianopic scotoma or, very rarely, of a formed hallucination but, whatever their nature, they tend to last for about 10–20 minutes and are followed by a headache which is centred above one eye and often described as a boring pain. The headache lasts for anything between 1 hour and 24 hours and then disperses. The patient may experience nausea and vomiting as the attack ends. Migraine may begin quite early in childhood and continue at regular intervals for many years. Migraines are more common in women and tend to improve at the time of the menopause. Atypical migraine can sometimes pose a diagnostic problem. The visual aura may appear by itself or the migraine attack may be accompanied by gastrointestinal symptoms or by ophthalmoplegia. The attack may be preceded by oliguria and fluid retention and be followed by a diuresis. Very rarely, a permanent hemianopic scotoma or ophthalmoplegia may result from an attack of migraine, but in these circumstances the original diagnosis must be reviewed very carefully.

There is some doubt as to whether essential hypertension causes headaches but there is no doubt that when the blood pressure becomes acutely raised a severe headache may ensue, accompanied often by blurring of the vision. Any adults with headaches should have their blood pressure measured. Another form of headache associated with abnormality of the blood vessels is that due to an intracranial aneurysm of the internal carotid artery or one of its branches. The pain in this case is usually throbbing in nature and there may be other signs of a space-occupying lesion at the apex of the orbit such as cranial nerve palsies or a bruit heard with the stethoscope. In the case of elderly patients, the possibility of temporal arteritis must always be kept in mind. This is an inflammation of the walls of many of the medium-sized arteries in the body but it tends to affect the temporal arteries preferentially. The walls of the vessels become thickened by inflammatory cells and giant cells mainly in the media and there is fibrosis of the intima. The lumen of the affected vessels becomes occluded. Affected patients are usually over the age of 70 and complain of tenderness of the scalp, especially over the temporal arteries which may be seen and felt to be inflamed and no pulse can be felt in them. The headache is made particularly bad by attempting to brush the hair. The importance of this type of headache rests on the fact that the eye is involved in about 60% of cases and the patient may suddenly go blind in one eye and then, a short time later, blind in the other. The diagnosis depends on finding a markedly raised ESR (above 70 mm/h) and, if necessary, performing a temporal artery biopsy. Whenever an elderly patient presents with occlusive vascular disease in the eye and a high ESR, this diagnosis should be considered because it is possible to treat the condition by means of systemic steroids. Once steroids have been instituted, the dose should be very carefully monitored, preferably in cooperation with a general physician, and titrated against the value of the ESR.

Other less common causes of vascular headaches include intracranial angioma and subarachnoid or subdural haemorrhage.

Blood

Changes in the blood itself may also be associated with headache. It is easy to forget that anaemic patients often have headaches which can be cured by treating the anaemia. Likewise, patients with polycythaemia may also complain of headache. Hypoglycaemia is another recognised cause; here, the symptoms occur after strenuous exercise or may be due to an overdose of insulin in a diabetic.

Nerves

In some respects this type of headache resembles migraine, although it is more common in men in the third or fourth decade. The word cluster refers to the timing of the attacks, which may be repeated several times over a few weeks followed by a period of remission for several months. The pain is described as being very severe and unilateral. There is conjunctival congestion and constriction of the pupil on the affected side, and the attack may last from minutes to hours. Tenderness over the side of the face and nasal discharge are also features. Raeder's paratrigeminal neuralgia probably merges with cluster headache, being described as severe ocular pain associated with miosis and ptosis but preservation of ipsilateral sweating. Trigeminal neuralgia can be easily distinguished from these other forms of headache by its distribution over one or all of the terminal branches of the trigeminal nerve and the fact that the very severe pain is triggered by touching a part of the cheek or by chewing or swallowing. The pain is so severe that the patient may become suicidal, and surgical division of the trigeminal nerve at the level of the Gasserian ganglion may be necessary. It is important to be sure that the severity of the symptoms justifies this measure in view of the difficult postoperative problems that may result such as corneal anaesthesia or weakness of the orbicularis muscle.

Post-herpetic neuralgia is an extremely debilitating form of headache experienced by elderly people. The pain seems to be more severe in the elderly and it may persist for many years. The cause of the headache is usually evident when one inspects the skin of the forehead which is slightly whitened and scarred from the previous attack of herpes zoster. Apart from the use of analgesics, antidepressant drugs may also be of help, together with the application of local heat or vibration.

Bones

In this condition the bones of the head enlarge and grow abnormally, the abnormal growth being associated with headache and incidentally an increase

in hat size. The eyes themselves may show optic atrophy, and close inspection of the fundi may reveal the curious appearance of wavy lines known as angioid streaks. These are due to cracks in Bruch's membrane, the membrane which separates the pigment epithelium from the choroid. Oxycephaly is a congenital defect of the skull due to premature closure of the sutures; patients sometimes complain of headaches as well as visual loss due to optic nerve compression. Multiple myeloma is the name given to a malignant proliferation of plasma cells within the bone marrow. There is also an excessive production of immunoglobulins. Osteolytic bone lesions occur especially in the skull, and headache may be an accompaniment. The disease is more common in the elderly and is accompanied by a high ESR. Diagnosis is made by examining the urine for Bence–Jones protein and the serum for abnormal immunoglobulins. Disease of the cervical vertebrae is another cause of headache, due to the effect of spasm of the neck muscle. Relief of the pain has been claimed to have been effected by manipulation of the neck, but the exact diagnosis must be made before embarking on such treatment.

Meninges

It is presumed that the pain and headache which accompany meningitis or encephalitis are mediated through the sensory nerve supply to the meninges. The pain-sensitive structures in the middle and anterior cranial fossa are supplied by the fifth cranial nerve, and inflammation may produce referred pain to the region of the eye.

The Eyes

The classical eye headache is that of subacute narrow angle glaucoma. Here the headache is an evening one, tends to be over one eye and is nearly always accompanied by blurring of vision and seeing coloured haloes around street lights. If the intraocular pressure is measured when the headache is present and is found to be normal, then it is unlikely that narrow angle glaucoma is the correct diagnosis. On the other hand, the diagnosis cannot be so easily excluded if the headache is absent. Patients with narrow angle glaucoma are long sighted — therefore beware the middle-aged, long-sighted patient with evening headaches and blurring of vision. Chronic simple glaucoma very rarely causes headache because the rise of intraocular pressure is too gradual and not great enough. The possibility should be borne in mind when a patient experiences headache following ocular trauma or eye surgery, that there may be secondary glaucoma. This type of glaucoma often responds well to treatment but if ignored may lead to rapid blindness. Acute iritis is associated with headache but in practice rarely presents as such because the other ocular symptoms override this. Patients developing endophthalmitis complain of severe pain in the eye and headache but this is a rare cause.

It has been argued that refractive error does not cause headache, but nothing could be further from the truth. Refractive headache is most

commonly seen in uncorrected hypermetropes, sometimes in children, but more commonly in adults aged 30–40 years who are beginning to have difficulty in accommodating through their long sightedness. For reasons of vanity, patients may have been deliberately avoiding the use of glasses and it may have to be explained to them that they have the choice of having headaches or wearing glasses. In the patient with no refractive error, the onset of presbyopia may be accompanied by headache which is sometimes delayed until the morning after prolonged reading. An otherwise normal person aged 45 should be suspected of having presbyopic headaches. Uncorrected myopes do not usually complain of headaches. If the spectacle prescription is incorrect for any reason, then a sensitive person may experience headache, but it is surprising how some people may tolerate an incorrect spectacle lens without complaint. Ocular muscle imbalance is an unusual cause of headache but it is an important one because it can be corrected with dramatic relief to the patient. Usually the patient has a considerable difference in refractive error between the two eyes and when the eyes are dissociated by such means as the cover test or the Maddox wing test, one eye tends to drift upwards or downwards. Relief of symptoms may be achieved by incorporating a prism in the spectacle lens or, if the deviation is marked, by ocular muscle surgery. Horizontal imbalance of the ocular movements is less closely linked with headache, although there is a group of patients, usually young adults under stress, who seem unable to converge their eyes on near objects; instead they allow one eye to drift outwards when reading. Some elderly patients have the same problem but do not so often have associated headache. This so-called convergence insufficiency can be greatly improved by a course of convergence exercises and provides one of the few instances where exercises of the eye muscles have any therapeutic value. However, the exercises are of doubtful value in the elderly.

Pain Referred From Other Sites

Sinusitis is well recognised as a common cause of headache and the patient with headache should be questioned about recent upper respiratory tract infections or previous history of sinus disease. Tenderness over the affected sinus is an important sign. The headache tends to begin after rising in the morning and reaches a peak later in the morning. Pain from an infected tooth may be referred over the side of the face and cause some diagnostic confusion but it is usually worse when chewing or biting. Pain from a middle-ear infection may cause similar problems. The temporomandibular joint is a recognised source of referred pain over the side of the face, and malfunctioning of the joint may result from incorrect jaw alignment or poorly fitting dentures.

Drugs

Over-indulgence in alcohol is one of the three causes of morning headache, the

other two being raised intracranial pressure and acute sinusitis. The diagnostic difficulty with alcoholism tends to be failure of the patient to admit to drinking. It may seem strange that such a patient should ever seek a doctor's opinion about headache, but chronic alcoholics do occasionally seek medical attention for their symptoms without relating them to alcohol intake and perhaps urged on by an anxious relative or friend. Chronic poisoning by other drugs is too rare a cause of headache in ophthalmic practice to be considered here, but it may have to be borne in mind.

Post-Traumatic Headache

Nearly all patients who have suffered a significant head injury complain of headaches. The pain may remain severe for many months and in the worst cases may last a year or two. Usually no obvious explanation can be found, apart from the original injury, and suspicion about the nature of the pain may be aroused if it is relieved once adequate financial compensation for the injury has been obtained. Usually this aspect of the symptom can be assessed by considering the personality of the patient together with the severity of the injury.

9 Contact Lenses

The widespread use of contact lenses means that the general practitioner and the ophthalmic casualty department find themselves confronted with more and more patients who have run into wearing problems of one kind or another, and for this reason some of the likely emergency requirements are considered here.

Types

As long ago as 1912 a glass contact lens was being produced, but because of the manufacturing difficulties and wearing problems, the widespread use of this type of optical aid was delayed until the introduction of plastic scleral lenses in 1937. The obvious advantage of placing a lens directly on the cornea over the wearing of spectacles is the cosmetic one, but the system also has optical advantages. Because the lens moves with the eye there are none of the problems associated with looking through the edge of the lens experienced by the wearer of spectacles. In addition a more subtle effect is the more accurate representation of image size on the retina in subjects with high degrees of refractive error. Although the moulded scleral contact lenses are still occasionally used, they have been largely replaced by the modern 'hard lens' which is much smaller and thinner and hence causes less interference with corneal physiology. In 1960 the hydrophilic contact lens was introduced. This had the great advantage of being soft and malleable and hence more comfortable to wear, but optically it has never been quite so good as the hard lens, especially when the patient has high degrees of astigmatism. Several different materials have now been used in the production of soft lenses and these differ in their ability to take up water or oxygen. Lenses are now being made which can be worn for long periods without needing to be removed and cleaned.

Side-Effects

The commonest complication of wearing modern contact lenses is losing them. Patients are well advised to have a pair of glasses at hand in case they have contact-lens-wearing problems or a lens is lost. More serious trouble may result from clumsy handling of the lens or leaving a hard lens in the eye for too long a period. Such patients quite often present with severe pain in the eye, and examination reveals a partially healed corneal abrasion. This must be treated in the usual manner and the patient advised against wearing the lens again for several weeks, depending on the extent of the abrasion. The contact lenses themselves should also be examined by the patient's fitter to make sure that they are not faulty. Bearing in mind the troubles which may ensue when an abrasion becomes recurrent, the indications for wearing the lenses in the first place should be reconsidered. Another sequel to wearing contact lenses, either hard or soft, is the appearance of chronic inflammatory changes in the conjunctiva, often characterised by a follicular conjunctivitis. The resulting irritation and redness of the eyes may persist for some weeks after the wearing of the contact lenses ceases. Unfortunately these symptoms may appear after wearing lenses successfully for some years and they may tend to recur in spite of renewing the lenses. Some patients who tolerate contact lenses very well may develop corneal changes after some years. Peripheral vascularisation may become evident and in neglected cases there may be band degeneration of the cornea. Some contact-lens wearers complain of recurrent blurring of their vision and this may be due to an ill-fitting lens producing corneal epithelial oedema or simply to the excessive accumulation of mucus on the lens (Fig. 9.1).

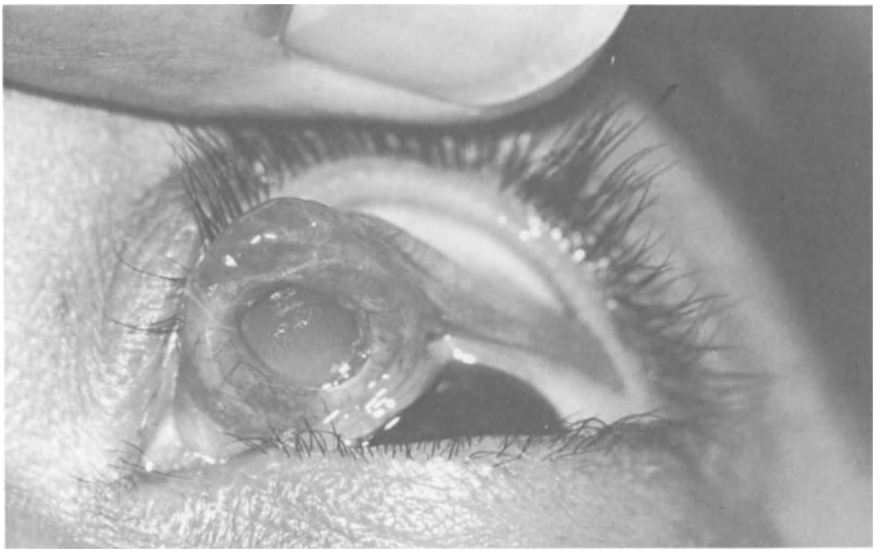

Fig. 9.1. Hard contact lens left for many weeks in the superior conjunctival fornix.

Indications

These may be considered as either cosmetic or therapeutic.

Cosmetic

There are obvious cosmetic advantages for the wearer of contact lenses, especially the teenager, but the potential wearer should realise the possible difficulties involved: the need to clean and sterilise the lenses and the need for some degree of finger dexterity when they are inserted and removed. They may be required for certain pursuits such as golf or athletics where the spectacle wearer is handicapped by misting up of the glasses in wet weather. Patients over the age of 45 or 50 will find that they require reading glasses as well and these, of course, must be worn over the contact lenses, thereby somewhat reducing the cosmetic value of the latter.

Therapeutic

There are instances when the contact lens may result in much better vision than spectacles, for example in patients with high degrees of corneal astigmatism which is not fully correctable with glasses. This accounts for the benefit of contact lenses in patients with keratoconus. Soft contact lenses are sometimes used as 'bandage lenses' to protect the cornea after corneal burns or in patients with bullous keratopathy. The contact lens has a special importance in the correction of unilateral aphakia (see Chapter 10) by reducing the image size on the retina to such an extent that the two eyes can once again be used together. If eyedrops are being regularly instilled into the eyes, soft contact lenses may absorb the drug being used or the preservative in the drops — in fact, attempts have been made to use soft contact lenses as a slow-release system by impregnating them with the drug before fitting.

Further Reading

Beard C (1981) Ptosis. CV Mosby, St Louis
Davson H (1980) The physiology of the eye. Churchill Livingstone, Edinburgh
Duke-Elder Sir Stewart (1965) System of ophthalmology, Vol. VIII: Diseases of the outer eye, Parts 1 and 2. Kimpton, London
Duke-Elder Sir Stewart (1974) System of ophthalmology, Vol. XIII: The ocular adnexa (2 vols). Kimpton, London
Duke-Elder Sir Stewart, Abrams D (1978) The practice of refraction. Churchill Livingstone, New York
Peyman GA, Sander DR, Goldberg MF (1980) Principles and practice of ophthalmology, Vol. I. Saunders, Philadelphia
Roskin NH, Appenzeller O (1980) Headache, monograph in major problems in international medicine, No. 19. Saunders, Philadelphia

Section III

Problems of the Eye Surgeon

The eye surgeon is confronted by problems which have been selected to a greater or lesser extent by the general practitioner or the optician. In the larger teaching centres he may be in a position to see patients selected in turn by other ophthalmologists and thus he may be able to gain a very detailed experience of relatively few aspects of the subject. In this section the cardinal eye problems which confront any eye surgeon are described. Being surgical problems, they are all fairly rapidly responsive to treatment and often dramatically so, involving the restoration of sight to the blind patient. In other cases the treatment may simply arrest the progress of visual failure or relieve the patient of pain or discomfort.

10 Cataract

Cataract means an opacity of the lens and it is the commonest potentially blinding condition which confronts the eye surgeon. Fortunately the results of surgery are very good, a satisfactory improvement of vision being achieved in about 90% of cases. It is usually possible to forewarn the patient when there is an extra element of doubt about the outcome. To the uninformed patient the word cataract strikes a note of fear and it may be necessary to explain that opacities in the lens are extremely common in elderly people. It is only when the opaque lens fibres reach the stage of significantly interfering with the vision that the name 'cataract' is applied. Many patients have a slight degree of cataract which advances so slowly that they die before any visual problems arise. Nobody need now go blind from cataract; however, one still encounters elderly people who, from ignorance or neglect, are left immobilised by this form of blindness, and it is especially important that the general practitioner is able to recognise the condition.

The Lens

The human lens is a surprising structure. It is avascular and yet it is actively growing throughout life, albeit extremely slowly. It receives its nourishment from the aqueous which bathes it. The lens is enclosed in an elastic capsule and for this reason tends to assume a spherical shape, or would do if the moulding of the lens fibres allowed. In situ the shape of the lens is retained by a series of taut fibres known as the zonule. The fibres exert radial tension on the lens but the tension is reduced when the circular part of the ciliary muscle contracts. The reduced tension of the zonule allows the lens to assume a more spherical shape and hence the anterior–posterior diameter of the lens increases. As a result, the refracting power of the lens increases, that is to say, light rays are more bent and the eye becomes focused on near objects. This process of accommodation, which is affected by relaxation of the lens but contraction of the ciliary muscle, gradually becomes less and less effective as we grow older, probably because the lens becomes less malleable rather than because the ciliary muscle is becoming weaker. This reduction in the range of

accommodation explains why a little child will present an object close to an adult's eyes and expect him or her to see it clearly. It also explains why, in the mid-forties, it becomes necessary to hold a book further from the eyes if it is to be read easily, and also the subsequent inability to read without the assistance of a spectacle lens which provides additional converging power. The need for reading glasses occurs in people with normal eyes at about the age of 45 (presbyopia) but this is only a milestone in a slowly progressive path of deterioration which begins at birth.

Histological section of the lens reveals that beneath the capsule there is an anterior epithelium with a single layer of cells, but no such layer is evident beneath the posterior capsule. Furthermore, if one follows the single-layered anterior epithelium to the equator of the lens, then the epithelial cells can be seen to elongate progressively and lose their nuclei as they are traced into the interior of the lens. Thus one can deduce from histological sections that the lens fibres are being continuously laid down from the equator. The actual arrangement of the lens fibres is quite complex, each fibre being made up of a prismatic six-sided band bound to its fellow by an amorphous cement substance.

Slit-lamp examination of the lens reveals the presence of the lens sutures which mark the points of junction of the ends of lens fibres. Two such sutures are usually seen, both often taking the form of the letter 'Y', the posterior suture being inverted. The lens fibres contain proteins known as 'crystallins' which have the property of setting up an antigen–antibody reaction if they are released into the eye from the lens capsule. One other feature of the lens which can usually be seen with the slit-lamp microscope is an object looking like a pig's tail which hangs from the posterior capsule. This is the remains of the hyaloid artery, a vessel which runs in the embryonic eye from the optic disc to the vascular tunic of the lens which is present at that stage (Fig. 10.1).

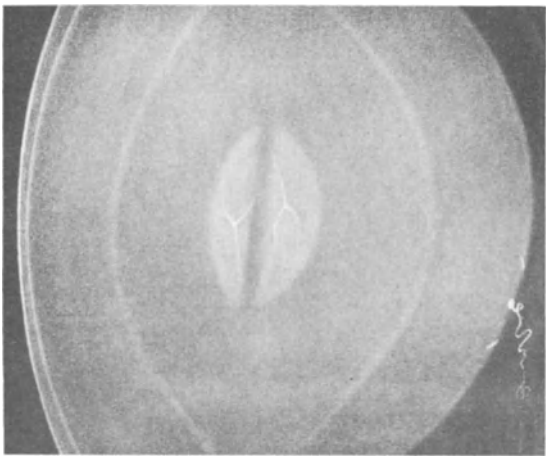

Fig. 10.1. Cross-section of child's lens: aqueous on left, vitreous on right. (With acknowledgement to M.L. Berliner, 1949.)

Aetiology

Having learned of the complex structure of the lens, perhaps one should be more surprised that the lens retains its transparency throughout life than that some of the lens fibres may become opaque. There are a number of reasons why lens fibres become opaque but the commonest and most important is ageing. The various causes will now be considered.

Senility

Although the majority of cataracts are associated with senility, the exact biochemical cause in these patients is not yet understood. We know that certain families are more susceptible to senile cataract, but a degree of opacification of the lens is commonplace in the elderly. Often the opacity is limited to the peripheral part of the lens and the patient may be unaware of any problem. It is usual to limit the term 'cataract' to the situation where the opacities are causing some degree of visual incapacity. Elderly patients are often reassured to learn that their eye condition is part of the general ageing process and that only in certain instances does the opacification progress to the point where surgery is required.

Diabetes

The new house surgeon working in an eye ward must be impressed by the number of diabetics with cataracts who pass through his hands, and might be forgiven for deducing that diabetes is a common cause of cataract. To see the situation in perspective he must also realise that both cataract and diabetes are common diseases of the elderly and one would expect the conditions to coincide quite often. Of course, the matter has been investigated from the statistical point of view and it has been shown that there is a somewhat higher incidence of cataract in diabetics, mainly because they tend to develop senile cataracts at a slightly earlier age. A special type of cataract is seen in young diabetic patients and in these cases the lens may become rapidly opaque in a matter of months. Fortunately this is not very common, usually occurring in insulin-dependent patients who may have had difficulty with the control of their diabetes. It is claimed that in its early stages this type of cataract can be reversible, but such an occurrence is so rare that it has not presented much opportunity for study.

Secondary

Cataract may be secondary to disease in the eye or elsewhere in the body.

Secondary to Disease in the Eye. More or less any terminal event in the eye tends to be associated with cataract. Advanced uncontrolled glaucoma is often associated with an opaque lens, as are chronic iridocyclitis and intraocular tumours. Certain specific eye diseases are accompanied by cataract; for example, patients who suffer from the inherited retinal degeneration, retinitis pigmentosa, sometimes develop a particular type of opacity in the posterior part of the lens. The removal of such a cataract may sometimes restore a considerable amount of vision, at least for a time.

Secondary to Disease Elsewhere. It may be recalled that the lens is ectodermal, being developed as an invagination of the overlying surface ectoderm. It is not surprising, therefore, that some skin diseases are associated with cataract. In particular, patients suffering from asthma and eczema may present to the eye surgeon in their late fifties. Dysfunction of the parathyroid glands is a very rare cause of cataract, mongolism another more common association.

Trauma

Contusion. A direct blow on the eye, if it is severe enough, may cause the lens to become opaque. A squash ball injury is a typical example of the type of force required. Sometimes the appearance of the cataract may be delayed, even for several years. The onset of unilateral cataract must always make one suspect the possibility of previous injury, but a cause-and-effect relationship may be difficult to prove in the absence of any other signs of previous contusion. It seems unlikely that a cataract will form unless there has been a direct blow on the eye itself, although occasionally medicolegal claims are made for compensation when a cataract has developed following a blow on the side of the head.

Perforation. A perforating injury of the eye bears a much higher risk of cataract formation. At present one of the commonest forms of perforating injury is that due to car windscreen glass. If the intruding piece of glass perforates the cornea without reaching the lens, usually the lens is spared and, in the absence of significant contusion, a cataract does not form. This, of course, also depends on careful management of the corneal wound and the prevention of infection. Unfortunately such perforating injuries may also involve splitting of the lens capsule with spilling out of the lens fibres into the anterior chamber. The series of events which follow such an injury is dependent on the age of the individual. When the lens capsule of a child is ruptured, a vigorous inflammatory reaction is set up in the anterior chamber and the lens matter is nearly always gradually absorbed over a period of about 1 month. This leaves behind the lens capsule and often a clear pupil; however, the refractive power of the lens is lost and, as will be seen later, this produces serious optical problems in the eye. When the lens capsule of an adult is ruptured, a similar inflammatory reaction ensues, but here there tends to be more fibrosis and a white fibrous plaque may remain to obstruct the pupil. Very rarely, it is possible for a lens to be perforated with subsequent opacity

limited to the site of perforation — indeed, one occasionally sees a foreign body within the lens surrounded by opaque fibres but limited to a small part of the lens.

Radiation. Visible light does not seem to cause cataract, although claims have been made that individuals from white races living for long periods in the Tropics may show a high incidence of cataracts. In practice this is not easy to confirm. In spite of public misapprehension, ultraviolet light probably does not cause cataract either, since the shorter wavelengths fail to penetrate the globe. Although these shorter wavelengths beyond the blue end of the visible spectrum do not penetrate the globe, they can produce a dramatic superficial burn of the cornea which usually heals in about 48 hours. This injury, which is typified by 'snow blindness' and 'welder's flash', will be discussed in Chapter 15. Prolonged doses of infra-red rays can produce cataract; this used to be seen occasionally in glass blowers and steel workers, but the wearing of goggles has now more or less eliminated this. X-rays and gamma-rays may also produce cataracts, as was witnessed by the mass of reports which followed the explosion of the atomic bombs at Nagasaki and Hiroshima. X-ray cataracts were recognised long before this; the threshold dose is in the region of 1000 rad but this varies with the exposure time. In practice the risk is only present when therapeutic x-rays are applied to lesions in the vicinity of the eye.

Congenital Factors

Many of the cases of congenital cataract seen in ophthalmic practice are inherited. Sometimes there is a dominant family history and there are many other possible associated defects, some of which fit into named syndromes. Acquired congenital cataract may result from maternal rubella infection during the first trimester of pregnancy. The association of deafness, congenital heart lesion and cataract must always be borne in mind. The ophthalmic house surgeon must take special care when examining the congenital cataract case preoperatively and likewise the paediatric house surgeon must bear in mind that congenital cataracts may be overlooked, especially if they are not very severe. Sometimes the cataracts may be slight at birth and gradually progress subsequently, or sometimes they may remain stationary until later years.

Toxicity

Toxic cataracts are probably rare, although several currently used drugs have been incriminated, the most notable being systemic steroids. Chlorpromazine has also been shown to cause lens opacities in large doses, and so has the use of certain miotics, including pilocarpine. Most of our knowledge of drug-induced cataracts is based on animal experiments, but the potential danger of new drugs causing cataract was emphasised before the last war after the introduction of dinitrophenol as a slimming agent. This produced a large number of lens opacities before it was eventually withdrawn.

Symptoms

Many patients complain of blurred vision which is usually worse when viewing distant objects. If the patient is unable to read small print, the surgeon may suspect that other pathology, such as macular degeneration, may be present. One must bear in mind that some elderly patients say that they cannot read when it is found that they *can* read small print when carefully tested. It is a curious fact that when the cataract is unilateral, the patient may claim that the loss of vision has been quite sudden. Elucidation of the history in these cases sometimes reveals that the visual loss was noted when washing and observing the face in the mirror. When one hand is lowered before the other, the unilateral visual loss is noted and occasionally interpreted as a sudden event. The history in cataract cases may be further confused by a natural tendency for patients to project their symptoms into the spectacles, and several pairs may be obtained before the true cause of the problem is found. In order to understand the symptoms of cataract it is essential to understand what is meant by index myopia. This simply refers to the change in refractive power of the lens which occurs as a preliminary to cataract formation. Index myopia may also result from uncontrolled diabetes. If we imagine an elderly patient who requires reading glasses in the normal way (for presbyopia) but no glasses for viewing distant objects, then the onset of index myopia will produce blurring of distance vision but also the patient will discover to his or her amazement that it is possible to read without glasses. In the same way the hypermetropic patient will become less hypermetropic and find that it is possible to see in the distance without glasses. The ageing fibres in the precataractous lens become more effective at converging light rays, so that parallel rays of light are brought to a focus more anteriorly in the eye.

Apart from blurring of vision, the cataract patient often complains of monocular diplopia. Sometimes even a very slight and subtle opacity in the posterior part of the lens can cause the patient to notice, for example, that car rear lights appear doubled, and this can usually be reproduced with the ophthalmoscope light. Monocular diplopia is sometimes regarded as a rather suspect symptom, the suggestion being that if the patient continues to see double even when one eye is closed, then he or she may not be giving a very accurate history. In actual practice nothing could be further from the truth and this is quite a common presenting feature of cataract.

Glare is another common presenting symptom. The patient complains that he or she cannot see so well in bright light and may even be wearing a pair of dark glasses. Glare is a photographic term but here it refers to a significant reduction in visual acuity when an extraneous light source is introduced. Light shining from the side is scattered in the cataractous lens and reduces the quality of the image on the retina. Glare becomes an important consideration when advising an elderly cataractous patient on fitness to drive. The visual acuity may be within the requirements laid down by law but only when the patient is tested in the absence of any sources of glare.

A consideration of all these factors makes it relatively easy to diagnose cataract even before examining the patient. To summarise, a typical patient

may complain that the glasses have been inaccurately prescribed, that the vision is much worse in bright sunlight, that sometimes things look double, and that there is difficulty in recognising people's faces in the street rather than difficulty in reading. Patients with cataracts alone do not usually complain that things look distorted or that straight lines look bent, nor do they experience pain in the eye.

Very rarely, cataracts become hypermature; that is to say, the lens enlarges in the eye and this in turn may lead to secondary glaucoma and pain in the eye.

In its late stages, a cataract matures and becomes white, so that exceptionally a patient may complain of a white spot in the middle of the pupil.

Signs

Reduced Visual Acuity

A reduction in visual acuity may, of course, be an early sign of cataract formation but this is not always the case; some patients see surprisingly well through quite marked lens opacities, and the effect on visual acuity as measured by the Snellen test type depends more on the position of the opacities in the lens than on the density of the opacity.

Findings of Ophthalmoscopy

The best way of picking up a cataract in its early stages is to view the pupil through the ophthalmoscope from a distance of about 50 cm. In this way the red reflex is clearly seen. The red reflex is simply the reflection of light from the fundus and it is viewed in exactly the same manner that one might view a cat's eyes in the headlamps of one's car or the eyes of one's friends in an ill-judged flash photograph. In fact, such a flash photo could well show up an early cataract if an elderly relative were included in the photograph. When using the ophthalmoscope, the opacities in the lens are seen often as black spokes against the red reflex. It is important to focus one's eyes onto the plane of the patient's pupil if the cataract is to be well seen, and it is preferable to dilate the pupil beforehand or at least examine in a darkened room. Typical senile lens opacities are wedge shaped, pointing towards the centre of the pupil. At the same time the central nucleus of the lens may take on a yellowish-brown colour, the appearance being termed 'lens sclerosis', and ultimately the lens may become nearly black in rare instances.

After inspecting a cataract with the ophthalmoscope held at a distance from the eye, one must then approach closer and attempt to examine the fundus. Further useful information about the density of the cataract can be obtained in this way. It is generally true that if the observer can see in, then the patient can see out. If there is an obvious discrepancy between the clarity of the fundus

and the visual acuity of the patient, then some other pathology must be sought. Sometimes the patient may not have performed too well on subjective testing if the test was not carried out with sufficient zeal; such an error should become evident when the fundus is viewed. Some types of cataract can be misleading in this respect — in particular those seen in highly myopic patients. Here there is sometimes a preponderance of nuclear sclerosis, which simply causes a distortion of the fundus while the disc and macula may still be seen fairly well.

Findings of Slit-Lamp Microscopy

A really detailed view of any cataract can be obtained with the slit lamp. By adjusting the angle and size of the slit beam, various optical sections of the lens can be examined, revealing the exact morphology of the cataract. The presence of small vesicles under the anterior lens capsule may be seen as an early sign of senile cataract. Cataracts secondary to uveitis or to drugs may first appear as an opacity in the posterior subcapsular region. For optical reasons, an opacity in this region tends to interfere with reading vision at an early stage. Opacities in the lens may appear in a wide range of curious shapes and sizes, and earlier in the century there was a vogue for classifying them with Latin names which are now largely forgotten. Such a classification is of some help in deciding the cause of the cataract, although it may sometimes be misleading. Congenital cataracts are usually quite easily identified from their morphology, as also are traumatic cataracts which follow soon after injury. When a unilateral cataract appears many years after a mild contusion injury, it may be difficult to distinguish this from senile cataract.

Other Important Signs

Certain other important signs need to be carefully elicited in a patient with cataracts. The pupil reaction is a particularly useful index of retinal function and it is not impaired by the densest of cataracts. A poor reaction may lead one to suspect senile macula degeneration or chronic glaucoma, but a brisk pupil with a mature cataract might be described as a 'surgeon's delight' because it indicates the likelihood of restoring good vision to a blind eye. The function of the peripheral retina can be usefully assessed by performing the light projection test. This entails seating the patient in a darkened room, covering one eye, and asking him to indicate, by pointing, the source of light from a torch positioned at different points in the peripheral field. Checking the pupil and the light projection test take a brief moment to perform and are by far the most important tests of retinal function when the retina cannot be directly seen. A number of other more sophisticated tests are now available; electroretinography, ultrasonography and measurement of the visually evoked potential to name some. Sometimes at least an area of the peripheral retina may be seen when the pupils have been dilated, and all cataract patients should be examined in this way before one embarks on more complex tests. A search

for the signs of cataract thus involves a full routine eye examination including a measurement of the best spectacle correction.

Management

There is no known medical treatment which may influence the progress of cataracts. Occasionally patients claim that their cataracts seem to have cleared, but such fluctuation in the density of lens opacities has not been demonstrated in a scientific manner, except possibly in the case of certain metabolic cataracts. In particular, there have been claims that galactosaemic cataracts may clear under the influence of treatment in their early stages. If one considers that a cataract is the result of the coagulation of lens protein, then it is perhaps not surprising that the opacity is irreversible.

Cataract is therefore a surgical problem, and the management of a patient with cataract depends on deciding at what point the visual impairment of the patient justifies undergoing the risks of surgery. The cataract operation itself has been practised since pre-Christian times, and developments in recent years have made it safe and effective in a large proportion of cases. The operation entails the removal of the lens either in toto or leaving the posterior part of the capsule behind. More and more often nowadays, the removed lens is replaced by an artificial one and it is important to have an understanding of the pros and cons of this if the patient is to be advised satisfactorily even at the primary care level.

In the earlier part of the century the technical side of cataract surgery necessitated waiting for the cataract to become 'ripe'. Nowadays no such limitations exist and it is theoretically possible to remove a clear lens; indeed, the decision regarding surgery depends upon whether the patient has a high chance of being better off afterwards. Such a decision is not always as simple as it may sound, even though nowadays it does not depend on purely technical considerations. In order to be able to advise patients on their cataracts, it is essential to understand the full significance of aphakia.

Aphakia

'Aphakia' means the absence of a lens. All cataract surgery entails the removal of the opaque lens by one means or another and we must now consider the way in which the eye is altered by such a deprivation.

Loss of Accommodation. This might seem to be the most obvious loss once the lens has been removed. In fact most patients undergoing cataract surgery have very little power of accommodation, being elderly; thus although they have no accommodation once the lens has been removed, this makes very little difference to them.

Loss of Refracting Power. The normal eye has four refracting surfaces: the

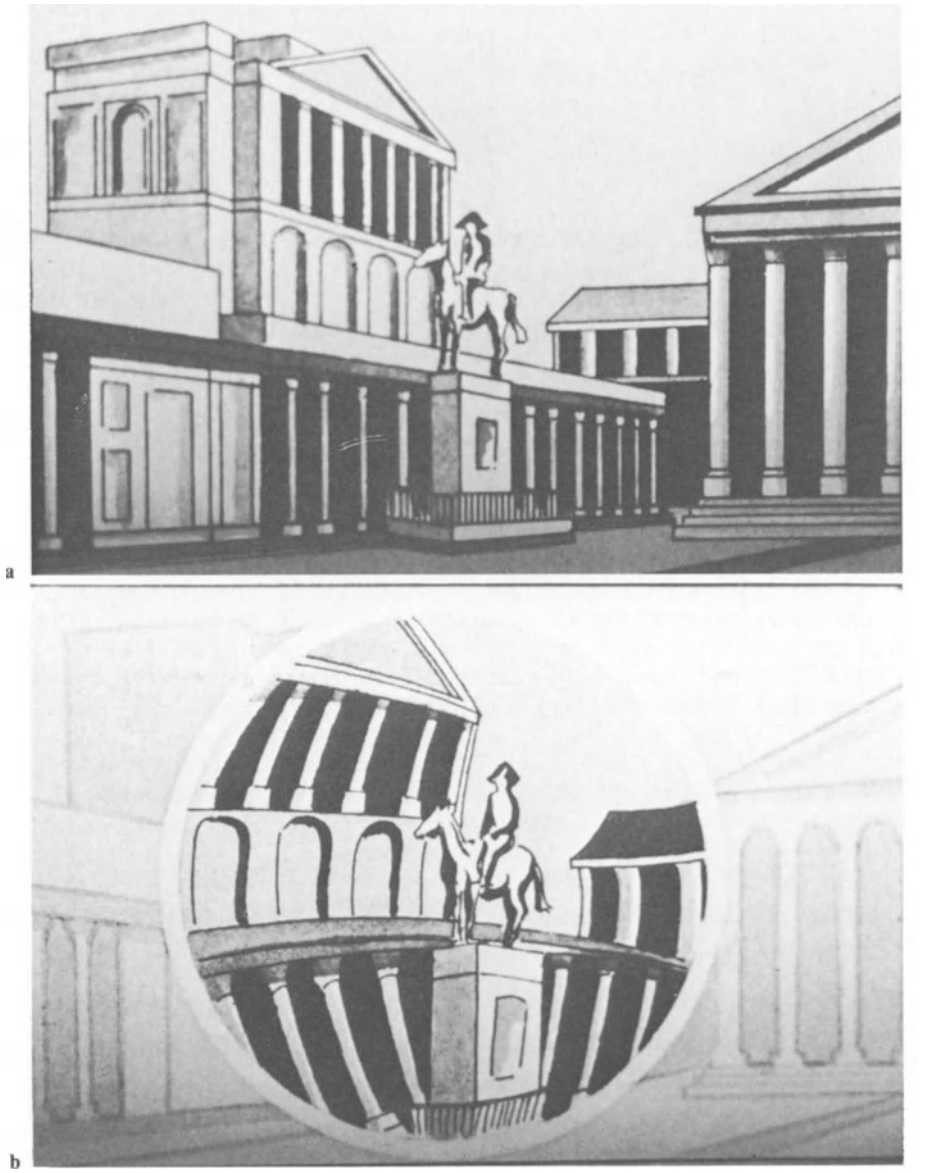

Fig. 10.2. The same scene as viewed **a** by a normal person and **b** by an aphakic patient.

anterior and posterior surfaces of the cornea, and the anterior and posterior surfaces of the lens. At each of these surfaces light rays are converged to the extent that parallel rays meeting the cornea are brought to a point focus on the retina. Removing the lens eliminates two of these surfaces, replacing them with an aqueous–vitreous interface across which there is negligible bending of light rays. This means that parallel light rays reaching the cornea are now brought to a focus somewhere behind the eye; that is to say, the eye becomes very long sighted. In actual fact the patient becomes hypermetropic to the degree of about 10 dioptres. Remember that a dioptre is the unit of measure of the converging power of a lens, being the reciprocal of the focal length in metres. A 10-dioptre hypermetrope is a very long-sighted person. It is somebody whose first act of the day is to find their spectacles because without them life is impossible. The 10-dioptre hypermetrope has to wear thick spectacles which make the eyes look larger. Fortunately this problem can now often be overcome, as will be described shortly.

Enlarged Image Size. If one looks at an object through a magnifying glass, it is common experience that when the magnifying glass is moved away slightly then the object looks larger. Let us now for a moment let our imagination run wild and consider being able to grasp the lens inside our own eye and to move it forward to the position of a spectacle lens. By the same principle it can be imagined that everything would look slightly larger. This is exactly the situation in the aphakic patient where the opaque lens is replaced by a spectacle lens. Aphakic patients very soon notice that the world looks rather a different place; some accept this rapidly and without question, whereas others find difficulty in coming to terms with their larger than life surroundings and the problems of distance judgement that this may entail. The need to wear strong glasses introduces certain other difficulties which may be rather vexing to the elderly patient who is unaccustomed to the problem. Not only do things look larger, but distortion is introduced so that straight lines in the peripheral field look curved and one has the impression of doorways looming up before one and people swooping past the peripheral field when walking in crowds. Fortunately there are some neat solutions to these difficulties which will be described in the next section (Fig. 10.2).

Changes in Colour Perception. Most patients comment on the amazing brightness of colours after cataract surgery. Comments such as 'I haven't seen colours like this since I was young' are quite frequent and presumably reflect the fact that the often rather brown opaque lens adds a dull sepia tone to the surroundings. Sometimes after cataract surgery, patients may complain that everything looks pink or sometimes, to confuse the issue, they say everything looks blue. These interesting effects have not been adequately explained.

Unilateral Aphakia

Unilateral aphakia presents a special problem owing to the fact there is a discrepancy between the image size on each retina. Our brains are not made to

accept different image sizes from each eye. When there is a difference in refractive error amounting to more than 4 dioptres, then the image difference is such that vision is seriously disturbed and the subject experiences double vision, headache and even nausea. In children this situation is resolved by suppressing the image from one eye, but in the elderly this is not possible and it is necessary to prescribe a spectacle lens for one eye only with a 'balancing lens' or even a frosted glass on the other side.

Contact Lenses. By placing the converging lens closer to the original site of the lens that has been removed, the image size on the retina can be reduced to an acceptable level so that the two eyes can be used together again. This is achieved by placing the lens actually on the cornea in the form of a contact lens. Furthermore, with this system the problems of peripheral distortion caused by looking through the edge of a thick spectacle lens are also eliminated because the contact lens moves with the eye. The snag here is that elderly people often have the greatest difficulty in handling contact lenses and although they are sometimes very successful this is not always the case. The younger unilateral aphake, following traumatic cataract for example, tends to be a better candidate for contact lenses, although even here problems of tolerance frequently arise after a time.

Intraocular Acrylic Implants. These represent the ideal theoretical solution to the problems of aphakia because the cataractous lens is replaced by a clear acrylic one. As might be expected, this introduces new risks into cataract surgery, risks which at worst can mean the loss of the eye. However, in recent years there have been important advances in our understanding of these risks and in the development and design of these implants, and the insertion of implants is now being practised on a wider scale by surgeons throughout the world.

When to Operate

Even though the decision to operate on a cataract must be made by the ophthalmic surgeon, the non-specialist general practitioner must understand the reasoning behind such a decision. In a National Health Service clinic the initial interview may necessarily be too brief to explain the problem fully to a patient with a limited understanding of biological matters. Some patients, having understood that they have a cataract, assume that it must be removed and may be vexed by the fact that 'nothing is being done'. The informed general practitioner can provide essential reassurance in these circumstances. At the other end of the scale there are patients who are reluctant to admit that they have a problem and who may refuse to seek specialist opinion, often because of some hidden fear. The general practitioner should be able to decide whether they are being reasonable to themselves and how much pressure and persuasion should be applied. It is important to realise that the decision to operate depends largely on the type of patient. The active businessman with a visual acuity of 6/12 may find that his work is seriously hampered and may

greatly appreciate the results of surgery. The elderly lady who, for other reasons, is confined to sitting and reading or watching television in one room may resent the fact that surgery changes her from someone who can read without glasses to an aphake with excellent vision but thick spectacles. In such a case it might be more prudent to wait until the vision has reached 6/24 or worse. Early surgery may be needed to keep a joiner in work for whom good binocular vision is essential. The patient with only one eye is another special case because here the surgeon runs the remote but possible risk of inflicting blindness on the patient. For this reason many surgeons avoid operating on an only eye unless the visual acuity is worse than 6/60, and the patient may find difficulty in understanding why his vision is allowed to become so bad before having his operation.

Time Spent in Hospital

Nowadays it is common practice for the cataract patient to be admitted on one day, have surgery the next and then remain in hospital for a further 3 or 4 days. Attempts have been made to perform day-cases cataract surgery, but this is not yet widely practised.

Convalescence

It is a fair generalisation to say that an eye requires 3 months for full healing to take place following a cataract operation. On the other hand, most of the healing takes place during the first postoperative fortnight and it is usual for the younger patient to return to work after about 1 month. In former days it was necessary for the patient to remain in bed and be completely restrained for some days, but the advent of more effective suturing has enabled the patient to be mobilised on the day after the operation. Usually some restraint, such as the avoidance of lifting heavy weights or bending, is advisable during the first fortnight after the operation.

The older patient who has just had a cataract operation is not in a position to look after him or herself. Food should be provided, and often someone is needed to instil drops two or three times a day. For these reasons a careful enquiry into the patient's domestic circumstances is needed before embarking on surgery, and the timing of the operation may depend upon this.

Age of the Patient

By itself, the age of the patient need have little influence on the decision to operate. Many people over the age of 100 years have had their cataracts successfully removed, and technically speaking cataract surgery becomes easier the older the patient. The general health of the patient must be taken into account, and indeed this may influence one's decision in unexpected ways.

Occasionally one is presented with someone who has difficulty in balancing,

perhaps as a result of Meniere's disease or some other cause, and who asks for cataract surgery with the idea that an improvement of their sight would benefit this. To render such a patient aphakic could prove disastrous, and the feasibility of an intraocular lens must first be considered. Sometimes cataract surgery is requested in a nearly blind demented patient on the grounds that the dementia may be improved. Although this occasionally happens, often the patient's mental state is made worse even though the sight is made better and this raises some interesting ethical problems for the surgeon.

Children

In the case of the child with congenital cataracts, the indications for surgery depend largely on the degree of opacification of the lens. An incomplete cataract may permit a visual acuity of 6/12 or 6/18 and yet the child may be able to read very small print by exercising the large amount of accommodation which children have available. Such a child could undergo normal schooling, and cataract surgery may never be required. A complete cataract demands early surgery and this may be undertaken during the first few months of life. There is a high incidence of retinal detachment in patients who have had surgery for congenital cataracts but this does not usually occur until the third or fourth decade. As might be expected, children adapt easily to aphakia and, furthermore, they often wear contact lenses very successfully.

Traumatic Cataract

This is usually a unilateral problem that presents all the difficulties of unilateral aphakia which have already been described. At the present time, the restoration of binocular vision is achieved by means of a contact lens, but the development of the intraocular lens may replace this in future years.

The Cataract Operation

Every medical student should witness at least one cataract operation during the period of training. It is an example of a classical procedure which has been practised for 3000 years. The earliest method for dealing with cataract was known as couching. This entailed pushing the lens back into the vitreous face, where it was allowed to sink back into the fundus of the eye. Although this undoubtedly proved a simple and satisfactory procedure in many instances, there is a tendency for the lens to set up a vigorous inflammatory reaction within the eye, with subsequent loss of sight.

Modern cataract surgery was founded by the French surgeon Jacques Daviel in the 18th century. The operation that he devised involved seating the patient in a chair and making an incision around the lower half of the cornea. The lens was then removed through the opening. The results he obtained were remarkable considering the technical difficulties that he must have

encountered. Subsequently the procedure was facilitated by lying the patient down and making the incision around the upper part of the cornea, where, in the postoperative period, it was protected by the upper lid. The use of local anaesthesia was introduced at the end of the last century and at the same time attempts were being made to suture the cornea back into position. By the beginning of this century two methods had evolved for the actual removal of the lens. The safest way was to incise the anterior lens capsule and then wash out or express the opaque nucleus, preserving the posterior lens capsule as a protective wall against the bulging vitreous face. This is known as the extracapsular technique and it is still employed in younger eyes and when certain types of intraocular artificial lens are being used. The intracapsular cataract extraction became the standard operation of choice in most patients over the age of 50 years during the early part of the century. It entailed removing the complete lens within its capsule and by this means avoided subsequent operations to open up residual opaque posterior capsule. Cataract surgery is now routinely performed under the microscope and several very fine sutures are used to secure the corneoscleral wound. The use of such sutures has reduced the risk of postoperative gaping of the wound and also the risk of bleeding into the eye from the wound edges.

Intraocular Acrylic Implants

Perhaps the most dramatic change in cataract surgery over the past few years has been the increasing use of the acrylic implant. The first implants were used after the Second World War. In this country they were pioneered by Harold Ridley, who had noticed the inert nature of perspex when imbedded in the eyes of fighter pilots (Fig. 10.3). Although these early acrylic lenses enabled many

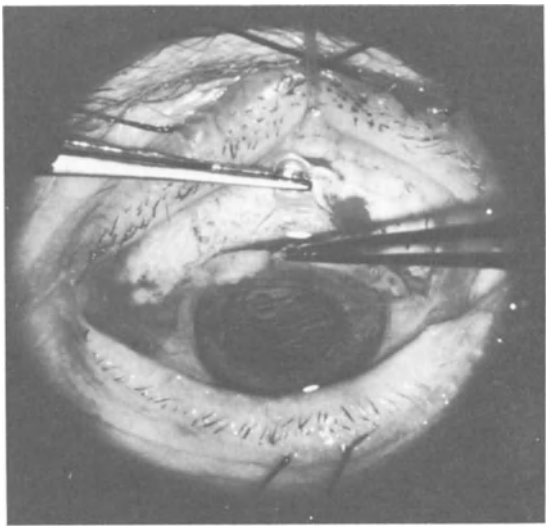

Fig. 10.3. Inserting an implant during a cataract operation.

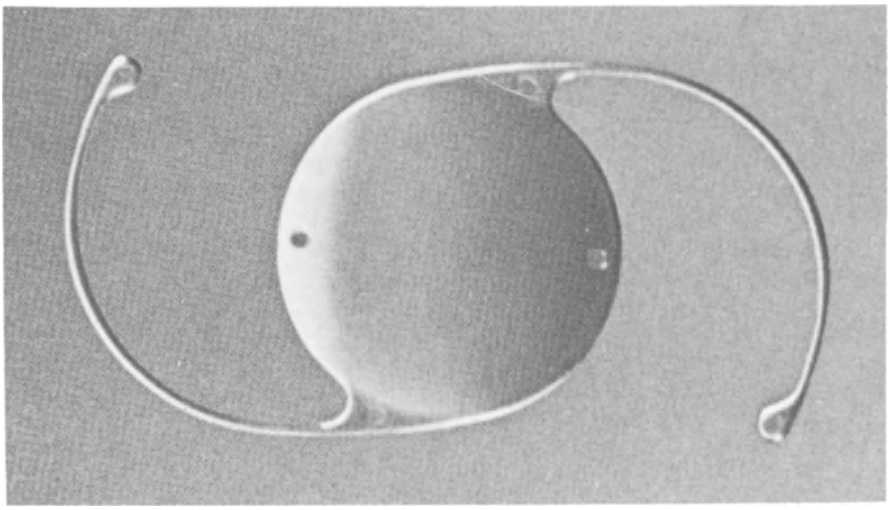

Fig. 10.4. An acrylic intraocular implant.

people to see without the disadvantages of aphakia, there was a significant morbidity which initially prevented their more widespread use. However, the wheels of progress had started turning and soon different types of lens were being designed. At the present time a wide range of lenses are being used and some of these are undoubtedly relatively safe and very effective. The various types of intraocular lens can be classified into those fitting into the anterior chamber and being supported at the angle, those which are fixed to the iris by means of loops, and those which fit into the area behind the iris. This last type of lens is now being inserted after extracapsular surgery and the results seem especially promising (Fig. 10.4).

From the patient's point of view, the results of successful cataract surgery with insertion of an acrylic implant seem almost miraculous. Often the sight is very clear quite soon after the operation and a visual acuity of 6/6, sometimes without glasses, is not unusual. Of course, the optical value of the lens which is implanted may be important and attempts are now being made to assess this requirement preoperatively. Sometimes there may be technical reasons for not inserting an implant and these may not become apparent until the middle of the operation. It is therefore necessary to explain this to the patient beforehand.

Drug Treatment During the Postoperative Period

The uncomplicated cataract patient is usually treated with antibiotic drops alone, perhaps chloramphenicol drops three times daily. If no implant has been inserted, many surgeons instil a mydriatic drop once or twice a day to dilate the pupil. When the eye is more inflamed than usual, or sometimes

routinely, local steroids are instilled together with the antibiotic. This treatment may need to be maintained for a month or more after the operation.

Summary

It is important to understand the benefits and risks of cataract surgery in order to be able to give the patient some advice as to when the cataract is bad enough to have an operation. An understanding of the features of aphakia is an essential background to any such advice.

Most patients who present with cataracts are diagnosed as having senile cataracts, and investigations as to the cause are limited to tests to exclude diabetes and to confirm that the patient is fit for surgery. An understanding of the signs of cataract depends to a large extent on the understanding of the meaning of index myopia.

Fig. 10.5. An elderly person cannot read without glasses unless she is myopic. Myopia in the elderly may be due to cataract. ('Rembrandt's mother', with acknowledgement to Rijksmuseum-Stichting.)

The following is a revision list of the features of aphakia.

1. Loss of accommodation
2. +10 dioptres hypermetropia
3. Enlarged image size
4. Changes in colour perception

The following is a list of the important aetiological factors.

1. Senility
2. Diabetes
3. Secondary to (a) disease in the eye — iridocyclitis, tumours, detachment, retinitis pigmentosa, myopia, terminal glaucoma
   (b) disease elsewhere — atopy, mongolism, myotonia congenita, galactosaemia, hypoparathyroidism
4. Trauma
5. Congenital factors
6. Toxicity

Figure 10.5 is a final reminder of the signs and symptoms of cataract. An elderly woman would not normally be able to read small print without glasses and her eyes must therefore be abnormal. She may have inherited myopia enabling her to focus on near objects without the need for presbyopic lenses, but the myopia may also be index myopia. This in turn could be due to early cataract formation, but it could also be due to untreated diabetes.

11 Glaucoma

The word 'glaucoma' refers to the apparent grey-green colour of the eye suffering from an attack of acute narrow angle glaucoma. Nowadays the term has come to cover a group of eye diseases characterised by raised intraocular pressure. These diseases are quite distinct and the treatment in each case quite different. Glaucoma might be defined as a 'pathological rise in the intraocular pressure'. This is to distinguish the normal elevation of intraocular pressure seen in otherwise normal individuals. Here we must consider what is meant by the 'normal intraocular pressure'.

Normal Intraocular Pressure

Measurement of the intraocular pressure in a large number of normal subjects reveals a normal distribution extending from pressures of 10–12 mmHg to pressures of 25–28 mmHg. The pattern of distribution fits a Gaussian curve, so that the majority of subjects have a pressue of about 16 mmHg. For clinical purposes it is necessary to set an arbitrary upper limit of normal. By and large, the eye can stand very low pressures remarkably well, but when the pressure is abnormally high, the circulation of blood through the eye becomes jeopardised and serious damage may ensue. For clinical purposes, an upper level of 22 mmHg is often accepted. Above this level, suspicions are raised and further investigations undertaken.

Maintenance of Intraocular Pressure

If the eye is to function as an effective optical instrument, it is clear that the intraocular pressure must be maintained at a constant level. At the same time, an active circulation of fluid through the globe is essential if the structures within it are to receive adequate nourishment. The cornea and sclera form a

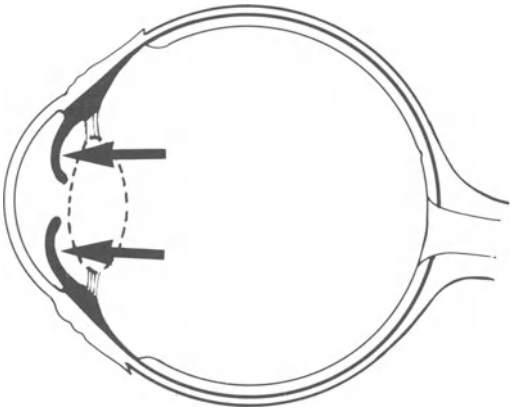

Fig. 11.1. Iris bombé.

tough fibrous and unyielding envelope and within this an even pressure is maintained by a balance between the production and drainage of aqueous fluid.

Aqueous is produced by the ciliary epithelium and a continuous flow is maintained through the pupil, whence it reaches the angle of the anterior chamber. A pressure gradient is constantly present and the iris, riding on this, is normally slightly bowed forward, as shown in Fig. 11.1. This phenomenon may become exaggerated in disease states, in which case it is known as 'iris bombé'.

On reaching the angle of the anterior chamber, aqueous passes through a grill known as the trabecular meshwork and thence reaches a circular canal embedded in the sclera known as Schlemm's canal. This canal runs in a ring around the limbus (corneoscleral junction) and from it minute channels radiate outwards through the sclera to reach the episcleral circulation. These channels are known as aqueous veins and they transmit clear aqueous to the episcleral veins which lie in the connective tissue underlying the conjunctiva. In actual fact, the proof of the site of drainage of aqueous can be verified by any medical student — it simply entails examining the white of the eye around the cornea with extreme care, using the high power of the slit-lamp microscope. After a time one can sometimes detect that some of the deeper veins convey parallel halves of blood and aqueous in the region beyond the junction of aqueous and episcleral vein.

The relative parts played by ciliary epithelium and trabecular meshwork in maintaining what is a remarkably constant intraocular pressure throughout life is not fully understood. It would appear that the production of aqueous is an active secretion whereas the drainage is more passive, although the rate of drainage can be altered by changing the tone of the ciliary muscle. In normal subjects the intraocular pressure does not differ in the two eyes by more than about 3 mmHg. Wider differences may lead one to suspect early glaucoma, especially if there is a family history of the disease. The normal intraocular pressure undergoes a diurnal variation, being highest in the early morning and

gradually falling during the first half of the day. This diurnal change may become exaggerated as the first sign of glaucoma.

Measurement of Intraocular Pressure

The Schiøtz tonometer is the instrument which provides the simplest and cheapest way of measuring the intraocular pressure. It requires a certain expertise in its use but any training doctor with reasonable dexterity should become proficient with practice. It is perhaps unfortunate that the instrument is rarely used by non-specialists, because the routine measurement of intraocular pressure is undoubtedly a sight-saving procedure. The instrument consists of a central plunger which moves freely within a hollow cylinder connected to a footpiece. The plunger is connected by a lever to a pointer on a dial (see Chapter 11).

To measure the pressure, the patient is asked to lie on a couch and a drop of local anaesthetic is instilled into the eyes. The eyelids of the left eye are held open gently, and without exerting any pressure on the eye, by the index finger and thumb of the left hand while the tonometer is lowered until the footpiece rides on the centre of the cornea. The plunger within the footpiece then impresses the cornea to a variable degree depending on the intraocular pressure. The amount of impression is indicated on the dial. The procedure is then repeated but this time the right finger and thumb keep open the right eyelids and the instrument is held in the left hand.

In ophthalmological clinics the Schiøtz tonometer has been superseded by a more rapid and more accurate technique known as applanation tonometry. The applanation tonometer is supplied as an accessory to the slit-lamp microscope. The principle of applanation is as follows. When two balloons are pushed together so that the interface is a flat surface, then the pressure within the two balloons must be equal. By the same argument, when a fixed flat surface is pressed against a spherical surface such as the cornea, then at the point at which the spherical surface is exactly flattened, the intraocular pressure is equal to the pressure being applied. The applanation head is a small perspex rod with a flattened end which is fitted to a moveable arm. The tension applied to the moveable arm can be measured directly from a dial on the side of the instrument. The observer looks through the rod using the microscope of the slit lamp, and the point at which exact flattening occurs can thus be gauged. For applanation tonometry, the patient is seated at the slit lamp and not lying down but it is still necessary to instil a drop of local anaesthetic beforehand. Because the measurement of the intraocular pressure is such a basic requirement in any eye clinic, attempts have been made to introduce even more rapid and efficient devices. Perhaps the most ingenious to date is the tonometer which measures the indentation of the cornea in response to a puff of air by a photo-electric method. A hand-held applanation tonometer is currently also used and undoubtedly this is now the ideal domiciliary instrument, although it is more expensive than the Schiøtz tonometer.

Clinical Types of Glaucoma

It has been mentioned that the word 'glaucoma' refers to a group of diseases and for clinical purposes these may be subdivided into four:

1. Chronic open angle glaucoma
2. Acute narrow angle glaucoma
3. Secondary glaucoma
4. Congenital glaucoma

Chronic Open Angle Glaucoma

The first important point to note about this disease is that it is very common, occurring in about 1% of the population over the age of 50 years. The second point is that the disease is inherited, and whereas the practice of screening the whole population for the disease is problematic in terms of finance, it is well worth screening the families of patients with the disease if those over the age of 40 are selected. This leads to the third point: that the incidence increases with age, being very rare under the age of 40. Chronic open angle glaucoma is also termed chronic simple glaucoma, or even 'CSG'. It is sad that this insidious, potentially blinding disease should affect those who are least likely to notice its onset, and elderly patients with advanced chronic open angle glaucoma are still seen from time to time in eye clinics.

Pathogenesis and Natural History

Histologically there are remarkably few changes to account for the raised intraocular pressure, at least in the early stages of the disease. Subsequently, degenerative changes have been described in the trabecular meshwork, with endothelial proliferation and oedema. It has been shown that in the majority of cases the problem is one of inadequate drainage rather than excessive secretion of aqueous. In the untreated patient the chronically raised pressure leads to progressive damage to the eye and eventual blindness. The rate of progress of the disease varies greatly from individual to individual. It is possible for gross visual loss to occur within months, but usually the process takes 5 years. Younger eyes survive a raised pressure rather better than older eyes, which may already have circulatory problems. Very few eyes can withstand a pressure of 40 mmHg for more than a week or two, or a pressure of 35 mmHg for more than a few months.

Chronic open angle glaucoma is nearly always bilateral, but often the disease begins in one eye, the other eye not becoming involved immediately. It is important to realise that the progress of chronic glaucoma can be arrested by treatment, but unfortunately many ophthalmologists experience the natural history of the disease by seeing neglected cases.

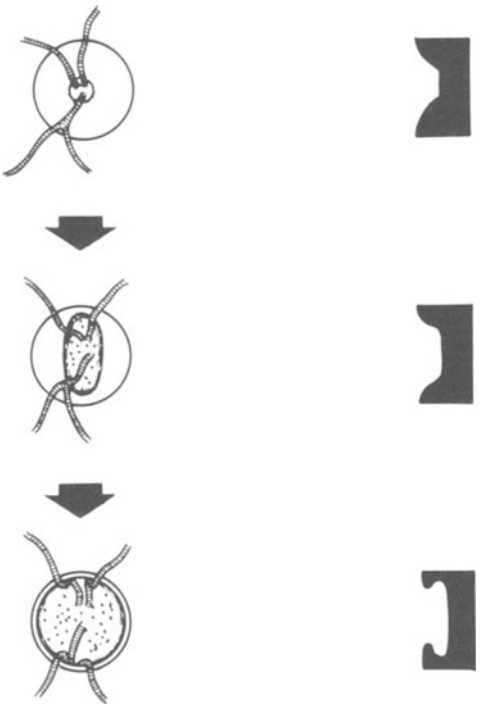

Fig. 11.2. The effect of glaucoma on the optic disc.

Symptoms

Most patients with chronic glaucoma have no symptoms. That is to say, the disease is detected before they have noticed any visual loss. Occasionally younger patients notice a defect in their visual field but this is unusual. Unfortunately the peripheral loss of visual field may pass unnoticed until it has reached an advanced stage.

Signs

The three cardinal signs are:

1. Raised intraocular pressure
2. Cupping of the optic disc
3. Visual field loss

The intraocular pressure creeps up gradually to 30–35 mmHg, and it is this gradual rise which accounts for the lack of symptoms. Such a rise in intraocular pressure impairs the circulation of the optic disc, and the nerve

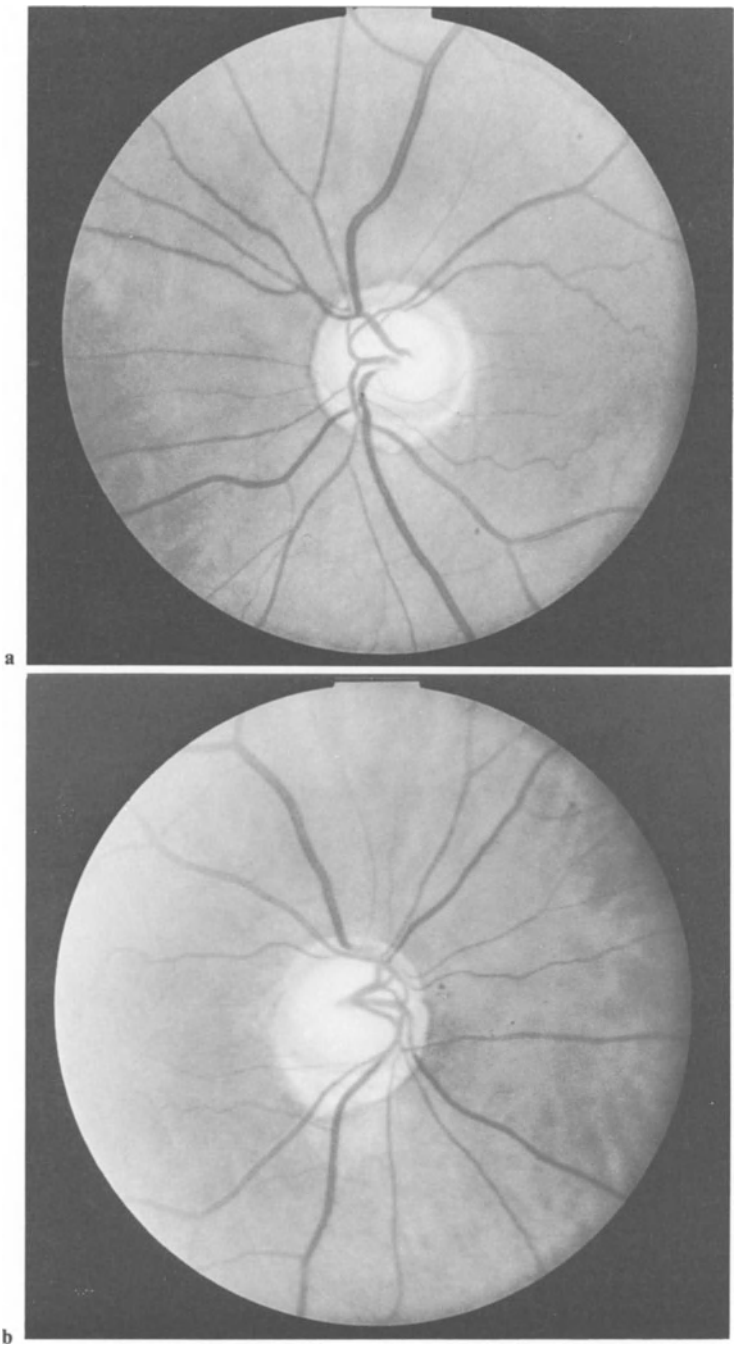

Fig. 11.3a. The left eye of a patient showing early glaucomatous cupping. **b.** The right eye of the same patient showing more advanced cupping.

fibres in this region become ischaemic. The combined effect of raised intraocular pressure and atrophy of nerve fibres results in gradual excavation of the physiological cup, and it is extremely useful to be able to identify this effect of raised intraocular pressure at an early stage. Figure 11.2 shows an optic disc undergoing various stages of pathological cupping. In the first instance the central physiological cup becomes enlarged, with its long axis arranged vertically. Loss of nerve tissue lays bare the minute openings in the sclera through which the optic nerve fibres pass, and the now pale centre of the disc is marked with small black dots. This prominence of the cribriform markings is seen in other types of optic atrophy. The central retinal vessels which formerly sloped gradually forwards and laterally from the centre of the disc now are seen to run along the floor of the disc before curving forwards around its eroded margin. It is particularly useful to observe the way in which the vessels enter and leave the nerve head (Fig. 11.3).

The changes in the visual field can be deduced from observing the disc and from considering the arrangement of the nerve fibres in the eye. If we gaze fixedly with one eye at a spot on the wall and then move a small piece of paper on the end of a paper clip, or even the end of our index finger, in such a manner as to explore our peripheral field, it is soon possible to locate the blind spot. In the case of the right eye, this is found slightly to the right of the point of fixation because it represents the projected position of the optic nerve head in the right eye. The blind spot is rounded and about 8–12° lateral to and slightly below the level of fixation. It has already been mentioned that the glaucomatous disc is initially excavated above and below so that the patient with early glaucoma has a blank area in the visual field extending in an arcuate manner from the blind spot above and below fixation. This typical pattern of field loss is known as the arcuate scotoma. If the glaucoma remains uncontrolled, this scotoma extends peripherally and centrally. It can be seen that even at this stage the central part of the field may be well preserved and the patient may still be able to read the smallest letters on the Snellen test chart. If the field loss is allowed to progress further, the patient becomes blind.

Treatment

For many years the mainstay in the treatment of chronic open angle glaucoma has been the use of miotic drops. The miotic of choice was pilocarpine, starting with the 1% solution and increasing to 4% if needed. Recently the treatment has undergone a minor revolution as the result of the introduction of the ß-blocker timolol. Pilocarpine itself is very effective in reducing the intraocular pressure. After about half an hour from the moment of instillation, the pupil becomes small and the patient experiences dimming of the vision, aching over the eyebrow and a spasm of accommodation which blurs the distance vision. At the same time the intraocular pressure in the majority of fresh cases of chronic glaucoma falls to within the normal range. After about 4 hours the intraocular pressure begins to rise again and the side-effects wear off. This, of course, means that a further drop of pilocarpine must be instilled if good control is to be continued. It is here that we find the

most difficult problem of treatment. Human nature is such that drops are rarely instilled four times daily on a regular basis, even though patients are genuinely anxious to preserve their eyesight. It is fortunate that timolol is effective over a 12-hour period and need be instilled only twice daily. As an ocular hypotensive agent it is probably not quite as effective as pilocarpine, but many cases of chronic glaucoma are now satisfactorily controlled by it and, furthermore, the drug may be used in combination with pilocarpine. Timolol has the further advantage that it does not cause any miosis and has no appreciable side-effects in most cases.

Anticholinesterase drugs such as echothiophate (Phospholine iodide) and demecarium bromide (Humorsol) are also very effective ocular hypotensive agents when administered in drop form. However, their use tends to be reserved for aphakic glaucoma because they are inclined to increase the rate of formation of cataracts. Systematic effects have been noted in children undergoing general anaesthesia combined with the use of succinylcholine.

The cholinergic drugs such as pilocarpine and the anti-cholinesterase drugs such as echothiophate iodide probably act by increasing the rate of outflow of aqueous, whereas timolol is thought to inhibit the production of aqueous. Adrenaline drops also have the effect of reducing aqueous production and they have been in use for some years as a supplement to pilocarpine. However, their effect is not very powerful and they tend to cause chronic dilatation of the conjunctival vessels in some patients as well as the deposition of pigment in the conjunctiva.

Oral acetazolamide is only occasionally used in chronic glaucoma because of its long-term side-effects. Acetazolamide (Diamox) is a carbonic anhydrase inhibitor which was introduced many years ago as a diuretic. Its diuretic action is not very well sustained, but it is a potent drug for reducing intraocular pressure. If a normal subject takes a 500-mg tablet of the drug, the eye becomes very soft after about an hour. Every patient taking acetazolamide experiences paraesthesiae of the hands and feet and some complain of gastric symptoms. Occasionally patients become lethargic or even confused. Young patients, particularly young males, may suffer haematuria or sometimes renal colic, and cases of renal failure have been recorded. It should be pointed out that these more serious side-effects are rare, and long-term acetazolamide is still sometimes used when no other means of controlling the intraocular pressure is available.

If the intraocular pressure remains uncontrolled by safe medical treatment and there is evidence of continued loss of visual field, then surgical treatment is indicated. A large number of operations have been devised for the management of chronic open angle glaucoma and most of these entail allowing the aqueous to drain subconjunctivally through an artificial opening made in the sclera. The operation which is usually preferred at the present time is the trabeculectomy. In this operation a superficial 'trapdoor' of sclera is raised and the deeper layer, including the trabecular meshwork, is removed. The trapdoor is then sewn back into position. Aqueous drains around the edge of this scleral flap (Fig. 11.4). Although most of these operations may reduce the intraocular pressure very effectively and often for many years, they all tend to increase the rate of formation of cataract. This is the main reason why surgery is never considered the first line of treatment in chronic open angle glaucoma.

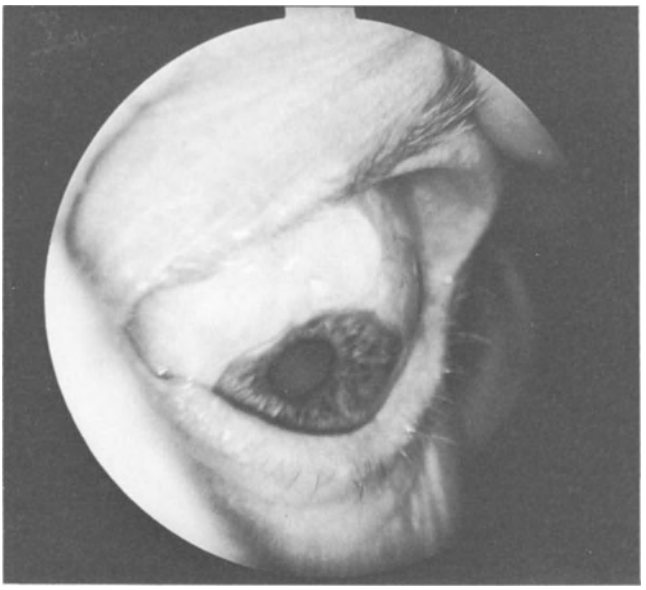

Fig. 11.4. A drainage bleb. Aqueous drains subconjunctivally following this type of glaucoma surgery.

Management

Most eye units now run special clinics for dealing with glaucoma patients. From what has been said, it should be clear that patients with chronic glaucoma require much time and attention. Initially the nature of the disease must be explained and they must realise that the treatment is to arrest the progress of the condition and not to cure it. Furthermore, any visual loss that occurs is irretrievable, so that regular follow-up visits are essential for checking the intraocular pressure and carefully assessing the visual fields.

Acute Narrow Angle Glaucoma

This condition is less common than chronic open angle glaucoma, comprising about 5% of all cases of primary glaucoma. It is a much more dramatic condition than the chronic disease and fits in more closely with the popular lay idea of 'glaucoma'. It tends to affect a slightly younger age group than chronic glaucoma and only occurs in predisposed individuals. There is a particular type of eye which is liable to develop acute glaucoma; this is a small hypermetropic eye with a shallow anterior chamber. One never meets a myope with acute glaucoma. This fact bears a special importance when attempting to diagnose a subacute attack.

Pathogenesis and Natural History

The eye which is going to develop narrow angle glaucoma has a shallow

anterior chamber and is hypermetropic. The forward bowing of the iris, known as iris bombé, which has already been mentioned, may be more evident in these individuals. Another factor is the gradual but slight increase in size of the lens which takes place with ageing. The rise in intraocular pressure which occurs is due to occlusion of the angle by the iris root and it may be precipitated by dilating the pupil. An uncontrolled acute attack of glaucoma can lead to rapid and permanent loss of the sight of the affected eye. Although it is known that occasionally patients recover spontaneously from such an attack, they may be left with chronic angle closure and a picture similar to that of chronic open angle glaucoma. About half the patients with narrow angle glaucoma will develop a similar problem in the other eye if steps are not taken to prevent this, and it will be seen that prophylactic surgery for the other eye is now the rule.

Symptoms

The Subacute Attack. Here it might be helpful to consider a typical patient, who might be a male or female, aged about 50. Such a patient would have a moderate degree of hypermetropia and rather a narrow gap between iris and cornea as shown by the shallow anterior chamber. During the autumn months, this patient's pupil might be noted to be slightly wider, as one might expect with the dimmer illumination, and one evening the pupil dilates sufficiently to allow the iris root to nudge across the angle and obstruct the flow of aqueous. Immediately the intraocular pressure rises acutely, perhaps to 30 or 40 mmHg, and pain is felt over the eye. At the same time the acute rise of pressure causes the cornea to become oedematous. Since it is evening, the patient observes that street lights when viewed through the oedematous cornea appear to have coloured rings around them, as if they were being viewed through frosted glass. At this point the patient retires to bed and on sleeping the pupil becomes small and the intraocular pressure rise is relieved. After several of these attacks the patient may seek attention from the family doctor. Patients present as healthy people with evening headaches associated with blurring of the vision and they are wearing convex lenses in their spectacles. Subacute glaucoma is easily missed, partly because it is rare amongst the large number of sufferers from headache. If attention is not sought at this stage or if the diagnosis is missed, then one evening the acute attack develops.

The Acute Attack. After a number of subacute attacks an irreversible turn of events may occur. The iris root becomes congested, raising the intraocular pressure further and producing further congestion. The headache becomes much worse and the vision becomes seriously impaired. The doctor, who may be called in the following morning, is confronted with a patient who is nauseated and vomiting and at first sight an acute abdominal problem may be suspected, until the painful red eye should make the diagnosis obvious. Sometimes acute glaucoma does not cause much pain or nausea and in these cases the physical signs in the eye become especially important (Fig. 11.5).

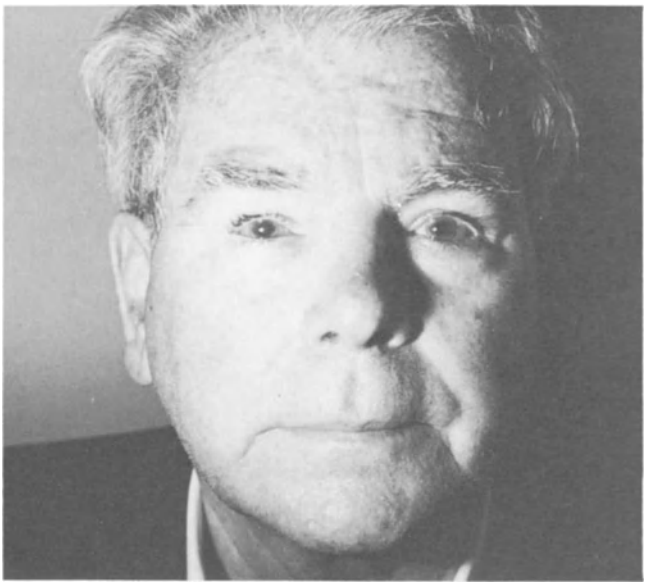

Fig. 11.5. Patient with narrow angle glaucoma affecting his right eye. The pupil was semidilated and the patient had been vomiting. Note the narrower palpebral fissure on the right side.

Signs

The most obvious physical sign is the semidilated fixed pupil. The iris and the constricting sphincter muscle of the pupil are damaged by the raised intraocular pressure. The pupil is not able to constrict and after a day or two the iris becomes depigmented, taking on the grey atrophic colour which gave glaucoma its name. Prompt and effective treatment should prevent any damage to the iris. The eye is red and a pink frill of engorged deeper capillaries is seen around the corneal margin; this important sign, as opposed to conjunctival inflammation, is known as ciliary injection. Corneal oedema can usually be detected without optical aids by observing the lack of lustre in the eye and any attempts to assess the hardness of the eye by palpating it through the eyelids will elicit another sign, that of tenderness of the globe. The visual acuity may be reduced to 'hand movements' in a severe attack. There are two rather subtle signs that often persist permanently after the acute attack has been resolved. The first is the presence of a white irregular microscopic deposit on the anterior surface of the lens, and the second is the presence of whorl atrophy in the iris. The pattern of the iris becomes twisted as if the sphincter has been rotated slightly. Both these signs may provide useful evidence of a previous attack which has resolved spontaneously.

Measurement of the intraocular pressure at this point may reveal a reading of 70 mmHg or more, but it is probably advisable to avoid placing any instruments on the oedematous cornea. Very gentle palpation of the globe is

Table 11.1 Signs of acute glaucoma

1. Ciliary injection
2. Semidilated oval pupil
3. Corneal oedema
4. Tenderness of globe
5. Poor vision
6. Shallow anterior chambers
7. Hard eye

usually enough to confirm that the eye has the consistency of a brick, especially when the pressures of the two eyes are compared. It should be realised that digital palpation of the globe can be very misleading and the method cannot be used to detect smaller rises in intraocular pressure with any degree of reliability (Table 11.1).

Examination of the other eye will reveal a shallow anterior chamber. The depth of the anterior chamber can be assessed by shining a focused beam of light obliquely through the cornea and noting the width of the gap between where the light strikes the cornea and where it strikes the iris. After inspecting a few normal eyes in this way, the observer can soon learn when an anterior chamber is abnormally shallow. This facility is important to anyone who intends to instil mydriatic drops into an eye. A shallow anterior chamber does not contraindicate mydriatic drops but it does indicate the need for extreme caution and care that the pupil is afterwards restored to its normal size. The angle of the anterior chamber itself is not exposed to direct inspection and it can only be seen through a gonioscope (Fig. 11.6). This instrument is a contact

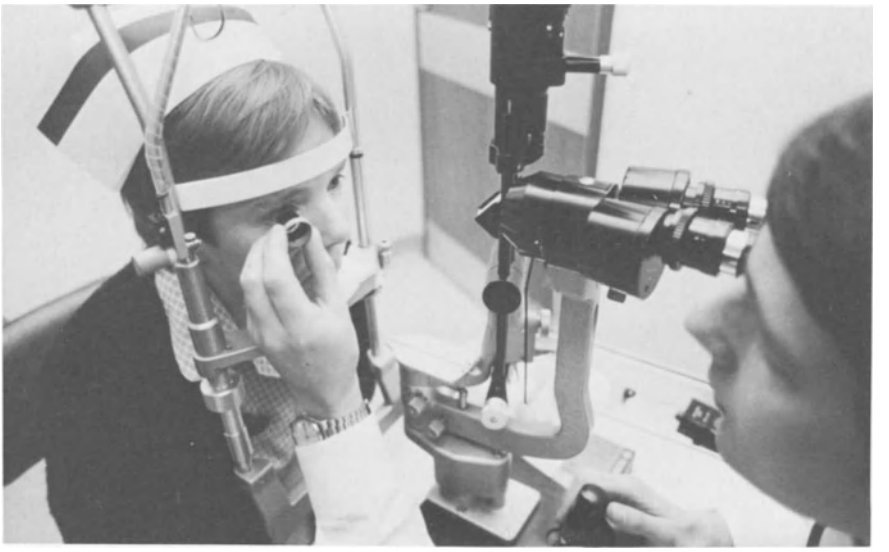

Fig. 11.6. Preparing for gonioscopy.

lens with a mirror mounted on it and through it the width of the angle can be estimated; if the angle is open, the various structures adjacent to the iris root and inner surface of the peripheral cornea can be identified. Gonioscopy forms a routine part of the examination of any patient with glaucoma, although in acute narrow angle glaucoma the presence of a closed angle can often be presumed by the presence of the other physical signs. Where there is any doubt, it may be necessary to apply a drop of hypertonic glycerol to the cornea to clear the oedema before applying the gonioscope.

The sooner narrow angle glaucoma is diagnosed and treated, then the better are the results of treatment. Unfortunately it is in the early subacute stage of the disease that the diagnosis may be difficult. A number of provocative tests have been devised for the patient who presents with suspicious symptoms but a normal intraocular pressure. The simplest test is the 'dark room test': the patient's intraocular pressure is measured before he or she is seated in a darkened room for half an hour. The intraocular pressure is again measured immediately after this, and a rise in pressure of more than 5 mmHg may be taken to be significant. Certain drugs can have a similar effect by having a mildly mydriatic action when taken by mouth. The phenothiazines have been incriminated in this respect. Of course, such drugs will have no adverse effect on patients who have already been treated and identified as cases of glaucoma. Only in the unsuspected cases of subacute narrow angle glaucoma is there a real risk of precipitating an acute attack.

Treatment

Acute narrow angle glaucoma is a surgical problem and any patient suffering from the condition requires urgent admission to hospital. To do less than this is to undertreat the condition and run the risk of producing chronic narrow angle glaucoma. On admission the affected eye is treated with intensive miotic drops. A typical regime would be the application of pilocarpine 4% every minute for 5 minutes then every 5 minutes for an hour followed by instillation every hour. This treatment is supported by an injection of acetazolamide. If the renal function is unimpaired, acetazolamide may be given intravenously (250 or 500 mg) followed by an oral dose of 250 mg q.i.d. In many cases these measures relieve the acute attack within hours. During this period the patient is kept in bed and analgesics are given if required. It is important that the other eye is also treated with pilocarpine 2% q.i.d. in order to prevent a second disaster.

Once the intraocular pressure has been controlled, the cure is maintained by performing a peripheral iridectomy. By this simple operation a small triangular hole is made in the peripheral iris. This allows the bulging iris bombé to sink backwards like a punctured ship's sail and is a sure means of preventing further acute attacks. The following week it is usual to carry out a similar prophylactic operation on the fellow eye. In some patients, the angle of the anterior chamber remains partially occluded by peripheral adhesions from the iris. In these cases a simple peripheral iridectomy may not be adequate and it may be necessary to carry out a drainage operation such as a

trabeculectomy. Most patients with acute narrow angle glaucoma are cured by surgery, although a small proportion develop cataracts in later years. The prognosis in adequately treated narrow angle glaucoma is therefore very good, but in the absence of treatment the result is disastrous.

The treatment of narrow angle glaucoma seems likely to undergo a small revolution over the next few years. This is because a new generation of lasers is appearing, which make it possible to perforate the iris quite simply. The results in narrow angle glaucoma are yet to become apparent. Some surgeons feel that intensive miotic treatment is unnecessary and this may certainly be true in less severe cases.

Secondary Glaucoma

The intraocular pressure may become raised as the result of a number of different disease processes in the eye quite apart from the causes of primary glaucoma which have just been described.

Secondary to Vascular Disease in the Eye

Central Retinal Vein Thrombosis. This is a common cause of sudden blurring of the vision of one eye in the elderly. The retinal veins can be seen to be dilated and surrounded by haemorrhages. In some cases recovery is marred by a rise in intraocular pressure which is delayed for about 3 months. The prompt appearance of this very painful complication has given it the name of 'hundred day glaucoma'. This type of glaucoma is usually difficult to control and even surgical measures may prove ineffective. A typical feature is the appearance of a vascular membrane over the anterior surface of the iris and sometimes across the angle of the anterior chamber. This vascularised tissue lends a pinkish hue to the iris and is termed *rubeosis iridis.* Patients with a central retinal vein thrombosis followed by secondary glaucoma have another problem because there is a recognised association between chronic open angle glaucoma and central retinal vein occlusion. This means that some patients who present with an occluded vein are found to have chronic glaucoma in the other eye.

Diabetes. Patients with severe diabetic retinopathy may also develop rubeosis iridis and secondary glaucoma. The vascular occlusive features of diabetic eye disease give it many resemblances to central retinal vein thrombosis and the secondary glaucoma which develops is also very resistant to treatment. The ultimate outcome is sometimes a blind and painful eye which has to be removed.

Secondary to Uveitis

During an attack of acute iridocyclitis the intraocular pressure is often below normal because the production of aqueous by the ciliary body is reduced.

When the normal production of aqueous is resumed it may induce a rise in pressure because the outflow channels have been obstructed by inflammatory exudate. This type of secondary glaucoma responds to vigorous treatment of the iridocyclitis, and here it is essential to dilate and not constrict the pupil and to apply steroid treatment. Acetazolamide and timolol may also be required. The type of secondary glaucoma which develops after the iridocyclitis of herpes zoster infections can be particularly insidious. The intraocular pressure may remain high without obvious pain and with relatively slight inflammatory changes in the eye. Secondary glaucoma usually responds well to treatment and once the underlying inflammation has subsided the eye returns to normal.

Secondary to Tumours

Malignant melanoma of the choroid and retinoblastoma may cause glaucoma. The raised intraocular pressure can be an important diagnostic feature when a suspected lesion is seen in the fundus. When a patient presents with a blind glaucomatous eye the possibility of malignancy must always be in the back of one's mind.

Secondary to Trauma

Trauma may precipitate a rise in intraocular pressure in a number of different ways. Sometimes, especially in children, bleeding may occur into the anterior chamber after a contusion injury. This can seriously obstruct the flow of aqueous both through the pupil and into the angle. Such an episode of bleeding may occur on the 2nd or 3rd day after the injury, turning a slight event into a very serious problem. On other occasions a contusion injury may cause splitting or recession of the angle which is associated with glaucoma. The iridocyclitis which follows perforating injuries tends to be complicated by glaucoma and the ophthalmologist must be constantly aware of such a complication.

Drug-Induced Glaucoma

Local and also systemic steroids can cause a rise in intraocular pressure and this is more likely to occur in patients with a family history of glaucoma. Steroid glaucoma is now a well-recognised phenomenon and 'steroid reactors' can be identified by measuring the intraocular pressure before and after instilling a drop of dexamethasone. The less potent steroids, hydrocortisone and prednisolone, are less likely to cause this problem and new steroids are now being produced with the claim that they have the anti-inflammatory effect of dexamethasone but no effect on intraocular pressure.

The possibility of inducing primary closed angle glaucoma by drugs has already been mentioned.

Secondary to Abnormalities in the Lens

A cataractous lens may become hypermature and swell up, pushing the iris diaphragm forwards and obstructing the angle of the anterior chamber. The situation is relieved by removing the lens. Phacolytic glaucoma occurs when a mature cataract causes a type of uveitis. This is thought to result from leakage of lens proteins through the lens capsule. A dislocated or subluxated lens, either the result of trauma or as a congenital abnormality, can be associated with a rise in intraocular pressure.

Congenital Glaucoma

This type of glaucoma is extremely rare and it is often, though not always, inherited. This means that the affected child may be brought to the ophthalmologist by the parents because they are aware of the condition in the family. Children may be born with raised intraocular pressure and for these cases the prognosis is not so good as in those where the pressure rise does not occur until after the first few months of life.

The glaucoma is due to defective development of the angle of the anterior chamber, and gonioscopy shows that the normal features of the angle are obscured by a pinkish membrane. Raised intraocular pressure in infancy has a dramatic effect because it causes enlargement of the globe. This can best be observed by noting an increase in the corneal diameter. The enlarged eye has given the condition the name of buphthalmos or 'bull's eye' (Fig. 11.7). Other

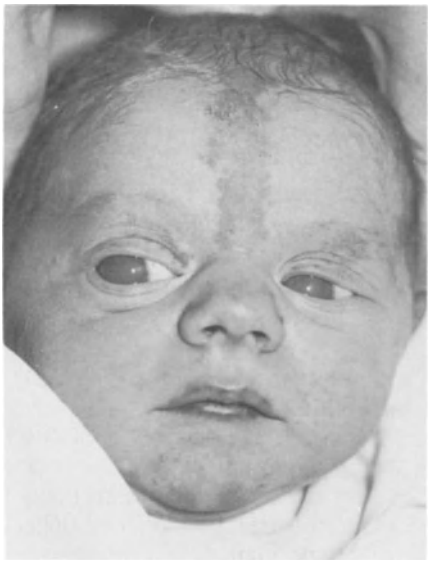

Fig. 11.7. Congenital glaucoma in a newborn infant.

important signs are photophobia and corneal oedema. The diagnosis is confirmed by an examination under anaesthesia which includes measuring the corneal diameters and the intraocular pressure. Surgical treatment is nearly always required and this involves passing a fine knife through the peripheral cornea so that the point reaches the opposite angle of the anterior chamber. Once in the angle, it is moved gently to and fro to open up the embryonic tissue which covers the trabecular meshwork (goniotomy).

12 Retinal Detachment

Detachment of the retina signifies an inward separation of the sensory part of the retina from the pigment epithelium. The retina bulges inwards like the collapsed bladder of a football. Once detached, the retina can no longer function and in humans it tends to remain detached, unless treatment is available.

Although the condition is relatively rare in the general population, it is important for several reasons. First, it is a blinding condition which can be treated very effectively and often dramatically by surgery. Second, retinal detachment may on occasions be the first sign of malignant disease in the eye and, finally, nowadays the condition may often be prevented by prophylaxis.

Incidence

Retinal detachment is rare in the general population but an eye unit serving a population of 500 000 might expect to be looking after three or four cases at any one time and each case remains in hospital for about 1 week. It can be seen, therefore, that a doctor in general practice might see a case once in every 2 or 3 years, especially if we consider that some retinal detachment patients go directly to eye casualty departments without seeking non-specialist advice. Although children are sometimes affected, the incidence increases with age and reaches a maximum in the 50–60 age group. There is a smaller peak in the mid-twenties due to traumatic detachments in young males.

Certain groups of people are especially liable to develop detachment of the retina: severely short-sighted patients have been shown to have an incidence as high as 3.5% and about 1% of aphakic patients have detachments.

In just under a quarter of cases, the other eye becomes affected at a later date. This means that the sound eye must be examined with great care in every instance.

Pathogenesis

There is an embryological explanation for retinal detachment in that the separating layers open up a potential space during the early development of the eye. The inner lining of the eye develops as two layers. In its earliest stages of development, the eye is seen as an outgrowth of the forebrain, the optic vesicle, the cavity of which is continuous with that of the forebrain. The vesicle becomes invaginated to form the optic cup, and the two-layered cup becomes the two-layered lining of the adult eye. Anteriorly in the eye the two layers line the inner surface of the iris and ciliary body. Posterior to the ciliary body the outer of the two layers remains as a single layer of pigmented cells, known as the pigment epithelium. The inner of the two layers becomes many cells thick and develops into the sensory retina. In the adult the sensory retina is closely linked, both physically and metabolically, with the pigment epithelium and, in particular, the production of visual pigment relies on this juxtaposition. When the retina becomes detached and the sensory retina is separated from the pigment epithelium, the retina can no longer function and the sight is lost in the detached area. Both pigment epithelium and sensory retina are included in the term 'retina' and in this sense 'retinal detachment' is a misnomer.

The retina receives its nourishment from two sources: the inner half deriving its blood supply from the central retinal artery, and the outer half from the choroid. The important macula region is supplied mainly by the choroid. When the retina is detached, the central retinal artery remains intact and continues to supply it since it is also detached with it. The outer half of the retina is deprived of nourishment, being separated from pigment epithelium and choroid. Eventually degenerative changes appear, the macula being affected at an early stage. It is interesting that after surgical replacement the retina regains much of its function during the first few days but further recovery may occur over as long a period as 1 or even 2 years.

Classification

Detachment of the retina may occur as the result of:

1. *Holes in the sensory retina* due either to degeneration or to trauma.
2. *Traction from within:* fibrous strands may form in the vitreous and subsequently contract causing the retina to be tented up. This is seen after injury or in diabetic retinopathy.
3. *Pressure from outside:* an expanding choroidal tumour is usually associated with retinal detachment; very rarely, inflammatory exudate may be the cause.

The Formation of Holes in the Retina

It was noticed as long ago as 1853, only a short time after the invention of the ophthalmoscope, that many detached retinae have minute holes in them, but it was not until the 1920s that the full significance of these holes as the basic cause of the detachment became realised. The holes may be single or multiple and are more commonly situated in the anterior or more peripheral part of the retina. They are usually horseshoe shaped, though sometimes rounded. In order to understand how these tears or holes occur, it is necessary to understand something of retinal degeneration and the vitreous.

Retinal Degeneration

When examining the peripheral retina of otherwise normal subjects, it is surprising to find that from time to time there are quite striking degenerative changes. Perhaps this is not so surprising when one considers that the retinal arteries are end arteries and these changes occur in the distal part of the circulation. Different types of degeneration have been described and named and certain types are recognised as being the precursors to hole formation. Peripheral retinal degeneration is more commonly seen in myopic patients and it is always seen after cataract surgery.

The Vitreous

The normal vitreous is a clear gel which occupies most of the inside of the eye. Its consistency is similar to that of raw white of egg and, being a gel, it takes up water and salts. It is made up of a meshwork of collagen fibres whose interspaces are filled with molecules of hyaluronic acid. The vitreous is adherent to the retina at the ora serrata (junction of ciliary body and retina) and around the optic disc and macula. If we move our eyes, the vitreous moves, and, being restrained by its attachments, swings back to its original position again. The vitreous is not usually perfectly transparent and most people become aware of small particles of cellular debris which can be observed against a clear background such as a blue sky or an x-ray screen. These particles can be seen to move slowly with eye movement and appear to have momentum, just as one would expect if one considers the way the vitreous moves.

These vitreous floaters are commonplace and tend to increase in number as the years pass. They often become more evident to the individual when under stress; the anxious student may observe a floater following the gaze across the page of a book and this may set in train a series of worries about possible eye disease. Patients quite commonly present with this symptom when their real problem is anxiety or stress. But the vitreous undergoes a more dramatic change with age; often in the late fifties it collapses from above, coming away from its normal position against the retina and eventually lying as a contracted mobile gel in the inferior and anterior part of the cavity of the globe. The rest

of the globe is occupied by clear fluid. When this happens the patient may complain of something floating in front of the vision and also the appearance of flashing lights. This is because the mobile shrunken vitreous sometimes causes slight traction on the retina. As a rule, the same symptoms are then experienced subsequently in the other eye. It is very common to find a detached vitreous in an elderly person's eye in the absence of any symptoms. This then is the condition known as vitreous detachment. It is common and usually of no pathological significance. One must be careful about the use of the term 'detachment' in front of the patient because for many people this means only one thing, a detached retina.

Unfortunately it is true that when the vitreous detaches it may, on rare occasions, cause the formation of a retinal tear or hole, possibly at a point of abnormal attachment of vitreous to retina. In myopic subjects the vitreous tends to be more fluid and this may be an important factor to account for the higher incidence of retinal detachment in such patients.

Signs and Symptoms

Let us now consider a typical patient, possibly a myope in the middle fifties, either male or female, who suddenly experiences the symptoms of 'flashes and floaters', sometimes spontaneously or sometimes after making a sudden head movement. The symptoms are similar to those produced by a vitreous detachment but tend to be more pronounced and obvious to the patient. Proper interpretation of such symptoms can save sight and they will therefore be considered in more detail.

Flashes ('photopsiae'). When questioned, the patient usually says that these are probably present all the time but are only noticeable in the dark. They seem to be especially apparent before going to sleep at night. The flashes are usually seen in the peripheral part of the visual field. They must be distinguished from the flashes seen in migraine which are quite different and are usually followed by headache. The migrainous subject tends to see zig-zag lines which spread out from the centre of the field and last for about 10 minutes. Elderly patients with a defective vertebrobasilar circulation may describe another type of photopsia in which the flashing lights tend to occur only with neck movements or after bending.

Floaters. It has already been explained that black spots floating in front of the vision are commonplace but often called to our attention by anxious patients. When the spots are large and appear suddenly, they may be of pathological significance. For some reason patients often refer to them as tadpoles or frogspawn or even a spider's web. It is the combination of this symptom with flashing lights that makes it important.

Flashes and floaters appear because the vitreous has tugged on the retina producing the sensation of light and often when the tear appears there is slight bleeding into the vitreous, causing the black spots. When clear-cut symptoms of this kind appear they must not be overlooked; the eyes must be examined

fully until the tear in the retina is found. Sometimes a small tear in the retina is accompanied by a large vitreous haemorrhage and thus sudden loss of vision. In such a case the proper treatment is bedrest for 2 or 3 days to hasten the clearing of the haemorrhage, after which the retinal tear can often be seen and treated. Other causes of vitreous haemorrhage, such as diabetes, hypertension, retinal vasculitis or trauma, must also be considered.

Traumatic Holes

A perforating injury of the eye can produce a tear at any point in the retina, but contusion injuries commonly produce tears in the extreme retinal periphery and in the lower temporal quadrant. This is because the lower temporal quadrant of the globe is most exposed to injury from a flying missile such as a squash ball. The threatened eye makes an upward movement as the lids attempt to close. Tears of this kind often take the form of a dialysis, the retina being torn away in an arc from the ora serrata. Warning symptoms in these patients are usually masked by the symptoms of the original injury, and they tend to present some months, or occasionally years, after the original injury with the symptoms of a retinal detachment. This is unfortunate because the tear can be treated if it is located before the detachment occurs.

When the presence of a retinal tear is suspected, the pupils of both eyes must be widely dilated and the fundi examined by direct and indirect ophthalmoscopy. The triple mirror gonioscope is also used to obtain a microscopic view of the peripheral fundus. At this stage a drawing is usually made of the location of any tears or weak areas. Many tears can be found with the direct ophthalmoscope alone, but using this instrument for the peripheral fundus demands some extra practice.

Studies on post-mortem eyes show that some patients have flat retinal tears and do not develop retinal detachments. Probably only a small percentage of tears lead to a detachment, but certain types of tear are very likely to cause trouble. These are: tears with symptoms, tears that are large, and tears that are situated in the upper part of the retina. However, all retinal tears must be suspect.

Signs and Symptoms

Once a retinal tear has appeared, the patient may seek medical attention, and effective treatment of the tear may ensue. Unfortunately some patients do not seek attention, or, if they do, the symptoms may be disregarded. Indeed, in time the symptoms may become less, but after a period of a month or two (this period may vary between minutes and years), a black shadow is seen encroaching from the peripheral field. This may appear to wobble. If the detachment is above, the shadow encroaches from below and it may seem to improve spontaneously with bedrest, being at first better in the morning.

Inspection of the fundus at this stage shows that fluid seeps through the retinal hole, raising up the surrounding retina like a blister in the paintwork of a car. Such a shallow detachment of the retina may be difficult to detect but the affected area tends to look slightly grey and, most important, the choroidal pattern can no longer be seen. The analogy is with a piece of wet tissue stuck against grained wood. If the tissue paper is raised slightly away from the wood, the grain is no longer visible. As the detachment increases, the affected area looks dark grey and the retinal vessels look black. The retina can be seen ballooning into the vitreous cavity and it often wobbles with eye movement. The tear in the retina shines out red as one views the pigment epithelium and choroid through it.

Once a black shadow of this kind appears in front of the vision, the patient usually becomes alarmed and seeks immediate medical attention. Urgent admission to hospital and retina surgery are needed.

Traction Retinal Detachment

The retina may be pulled away by the contraction of fibrous bands in the vitreous, especially after perforating injuries of the eye. The symptoms and signs may be similar but are complicated by those of the original injury. Advanced proliferative diabetic retinopathy may be complicated by detachment of the retina when a contracting band either pulls away a piece of retina to produce a hole or simply tents up the retina by direct traction. When such a diabetic patient experiences sudden loss of vision in one eye, the most likely cause is vitreous haemorrhage. It is important to distinguish this from the much less common but treatable retinal detachment. Vitreous haemorrhages can, of course, be treated if they persist for more than a few months but immediate action is not usually indicated.

Retinal Detachment Due to Tumours or Exudate

A malignant melanoma of the choroid may present as a retinal detachment. Often the melanoma is evident as a black lump with an adjacent area of detached retina. If the retina is extensively detached over the tumour, the diagnosis may become difficult. It is important to avoid performing retina surgery on such a case because of the risk of disseminating the tumour. Suspicion should be raised by a balloon detachment without any visible tears, and the diagnosis may be confirmed by transilluminating the eye to reveal the tumour.

Exudative retinal detachments are too rare to warrant more than a mention. They are seen in patients with severe hypertensive retinopathy and toxaemia of pregnancy. Harada's disease is a symptom complex which includes exudative uveitis with retinal detachment, patchy depigmentation of the skin, meningitis and deafness. Its cause is unknown. Exudative detachments tend to resolve spontaneously, sometimes after 2 or 3 months.

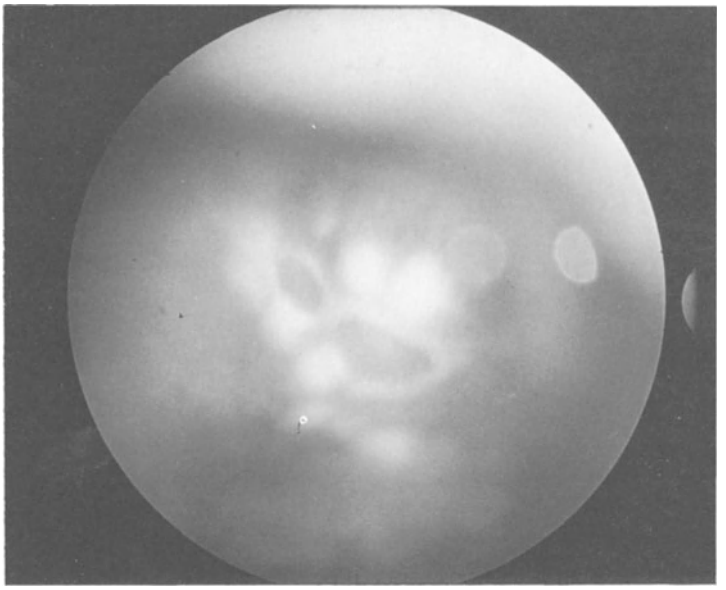

Fig. 12.1. A flat retinal tear undergoing light coagulation treatment.

Management

Prophylaxis

Flat retinal tears can be sealed by means of light coagulation. A powerful light beam from a laser or xenon arc is directed at the surrounds of the tear (Fig. 12.1). This produces blanching of the retina around the edges of the hole and, after some days, migration of pigment cells occurs from pigment epithelium into retina and the blanched area becomes pigmented. At the same time a bond is formed across the potential space and a retinal detachment is prevented. This procedure can be carried out, without any anaesthetic, in a few minutes. A wider and more diffuse area of chorioretinal bonding can be achieved by cryopexy, which entails freezing from the outside. A cold probe is placed on the sclera over the site of the tear and an ice ball is allowed to form over the tear. A similar type of reaction develops following this treatment, but it tends to be uncomfortable for the patient and general anaesthesia is to be advised.

Retina surgery

In the early part of this century it was generally accepted that there was no known effective treatment for retinal detachment. It was realised that a period

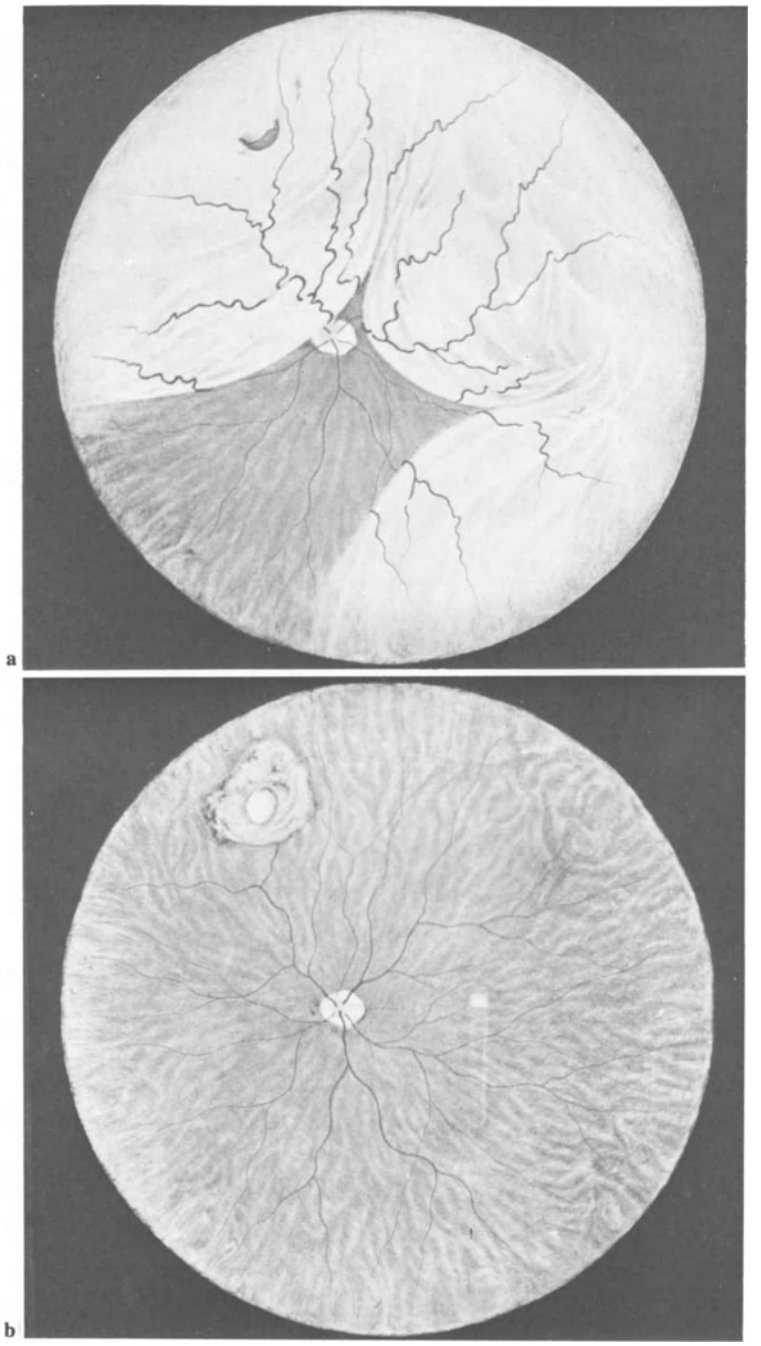

Fig. 12.2a, b. Retinal detachment. **a** Before treatment, **b** after treatment. (After J. Gonin, 1934.)

of bedrest resulted in flattening of the retina in many instances. This entailed a prolonged period of complete immobilisation with the patient lying flat with both eyes padded. This treatment can restore the sight but only temporarily because the retina redetaches when the patient is mobilised. It was also very dangerous for the patient in view of the risk of venous thrombosis and pulmonary embolism. In the 1920s it began to be realised that effective treatment of retinal detachment depends on sealing the small holes in the retina (Fig. 12.2). It was already known by then that the fluid under the retina could be drained off externally simply by puncturing the globe, but up till then no serious attempt had been made to associate this with some form of cautery to the site of the tear. Once it became apparent that cautery to the site of the tear combined with the release of subretinal fluid was effective, it also became evident that not all cases responded to this kind of treatment. It was almost as if the retina was too small for the eye in some cases, an idea which led to the design of volume-reducing operations which effectively made the volume of the globe smaller. This in turn led to the concept of mounting the tear on an inward protrusion of the sclera to prevent subsequent redetachment. Modern retina surgery involves the sewing of small inert pieces of material, usually silicone rubber, onto the outside of the sclera in such a way as to make a suitable indent at the site of the tear. Once a tear has been closed by such means, it is often necessary to drain off the subretinal fluid. In more difficult cases the eye may be encircled with a silicone strap to provide all-round support to a retina with extensive degenerative changes. In spite of the development of surgical techniques, some cases still provide difficulties. The retina can sometimes become so extensively torn that it folds up on itself and the damaged retina may become fixed by the growth of a fine fibrous membrane across its inner surface. A number of ingenious techniques have been developed in order to deal with such problems.

Prognosis

The retina can now be replaced by one operation in about 85% of cases. Of the successful cases, a proportion do not achieve a full restoration of their central vision, although usually the peripheral field recovers. The degree of recovery of central vision—that is, the ability to read the Snellen chart—depends largely on the duration of the detachment prior to surgery and whether the detachment was extensive enough to involve the macula region. Even when the retina has been detached for 2 or 3 years, it is still possible to restore useful navigational vision. When retina surgery has failed, further surgery may be required and for a few patients a series of operations is necessary. If it is thought that more than one operation is going to be needed, then it is helpful to the patient if he is warned about this before the treatment is started.

13 Squint

The word 'squint' refers to a failure of the visual axes of the eyes to meet at the point of regard. For normal vision each eye must be focused on and lined up with the object of regard. The fact that we have two eyes positioned some 60 mm apart means that we can accumulate considerably more data about our environment than would be possible with one eye alone. This can best be exemplified by considering what happens when one eye is suddenly lost as the result of injury or disease. Apart from the obvious loss of visual field which necessitates turning the head to the blind side, the patient experiences impaired distance judgement. The skilled craftsman notices a deterioration in the standard of his work and the elderly notice that they pour tea into the saucer rather than the cup. In time, depth perception improves and the patient adapts to the defect to some extent; children may adapt to one-eyed vision in a remarkable way. But it seems that modern civilised living does not have such great demands for binocular vision now that many fine tasks are carried out by machines. It is no coincidence that those animals whose survival depends on catching their food by means of accurate distance judgement have their eyes placed in front of their head, enabling the two eyes to be focused together on their prey.

Investigation of a normal human population reveals that although the eyes may be situated on the front of the face, they do not always work together, and it will be seen that there are a number of reasons why the mechanism may fail. The ability to use the eyes together is called binocular vision. It can be measured and graded by presenting each eye separately with a series of images by means of an instrument known as a synoptophore (Fig. 13.1).

1. *Simultaneous macular perception* is said to be present if the subject can see two dissimilar images which are presented simultaneously to each eye, e.g. a triangle to one eye, a circle to the other.

2. *Fusion* is present if the subject can see two parts of a whole image as one whole when each half is presented to a separate eye, e.g. a picture of a house to one eye, a picture of chimneys to the other eye, and the whole picture is maintained as one as the eyes converge. The range of fusion can be measured.

3. *Stereopsis*, the third grade of binocular vision, is present if, when

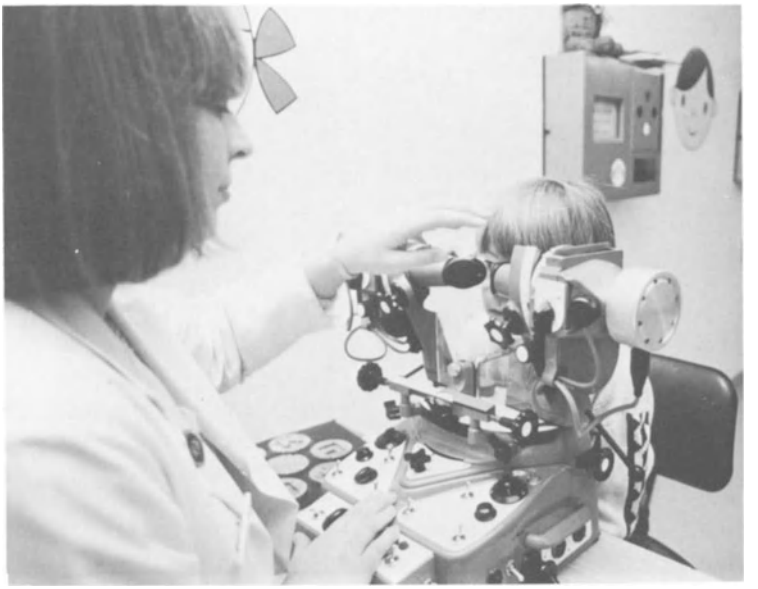

Fig. 13.1. The synoptophore.

slightly dissimilar views of an object are presented to each eye separately, a single three-dimensional view of the whole is seen. Stereopsis itself can also be graded if very fine degrees of impairment of binocular function need to be measured.

This ability of ours to fuse the images from each eye and to make stereoscopic pictures in our minds from them seems to develop during the early years of life and, furthermore, its development seems to be dependent on visual input. Below the age of 8 years, any misalignment of the eyes which disturbs binocular vision may permanently damage this function.

Another problem arises if the alignment of the eyes is disturbed during childhood. The child may at first, as one might expect, notice double vision but very quickly learns to suppress the image from one eye, thereby eliminating the annoyance of diplopia at the expense of binocular vision. In fact most, but not all, children learn to suppress when using monocular instruments, switching the other eye on again when the instrument is not being used. Prolonged suppression seems to lead to a more permanent state of visual loss called amblyopia of disuse. The word 'amblyopia' simply means blindness. Thus suppression is a temporary switching off of one eye when the other eye is in use, whereas amblyopia of disuse is a permanent impairment which seems to result from prolonged suppression. Amblyopia of disuse can also occur if the sight of one eye is defective as a result of opacities in the media, even though the alignment of the eyes has not been disturbed. Again this only occurs in children under the age of 8 years. Covering one eye of a baby can lead to permanent impairment of the vision of that eye as well as

impairment of the ability to use the eyes together. An adult may have one eye covered for many months or probably years without suffering visual loss.

Before considering the causes and effects of squint in children and adults, it is necessary to know something of the different kinds of squint.

Types of Squint

Sometimes the deviation from the normal line of sight by one eye only occurs when the patient is tired, although it can be induced during clinical testing. Such a squint is said to be latent. The deviation may be such that one eye is turned out or inwards. When one eye is permanently turned outwards the condition is termed an exotropia or divergent squint. A latent divergent squint is termed an exophoria. Convergent squints are classed esotropias and esophorias. Squints can also be usefully classified according to whether the deviation is the same in every position of gaze. When the angle of the squint is constant in all positions of gaze, the squint is said to be concomitant. When the angle of the squint increases when looking in one particular direction, the squint is said to be incomitant. In the case of the incomitant squint an extraocular muscle or group of extraocular muscles can usually be identified whose malfunction is the cause of the squint. Children who present with squints usually show concomitance, whereas adults more often present with incomitant squints. The sudden appearance of an incomitant squint is nearly always an indicator of serious underlying disease.

Squint in Childhood

During the first few weeks of life the eyes may seem to wander about aimlessly with limited ability to fix. Between the ages of 2 and 6 months, fixation becomes steadier even though the fovea is not fully developed, and by the age of 6 months convergence on a near object may be maintained for several seconds. Even at birth some degree of following movement of the eyes can be seen in response to a flashing light, but only the most gross squints can be diagnosed during the early months of life. If the eyes appear to be squinting at the age of 6 months, referral to an ophthalmologist is indicated. Squint appearing in childhood is important for various reasons:

1. The squint may be due to serious underlying disease either intracranial or intraocular.
2. The persistence of the deviation results in visual loss in one eye due to amblyopia of disuse.
3. The deviation may result in the failure of the development of normal binocular vision.
4. Cosmetic effect.

Amblyopia of Disuse

This is a very common cause of unilateral visual impairment. The casualty officer in an eye department becomes familiar with the patient who presents with a defect or injury in one eye when the other eye has been weak since childhood. The eye suffering from amblyopia of disuse shows certain characteristic clinical features:

1. An indefinite central scotoma which is difficult to assess by routine visual field testing.
2. Impaired visual acuity but usually able to decipher vertical lines of letters better than horizontal ones.
3. Normal fundus.
4. Small residual squint or, if not, the affected eye relatively hypermetropic.
5. Normal colour vision, in the absence of congenital colour blindness.
6. History of poor vision since childhood.

An eye suffering from amblyopia of disuse is sometimes referred to in lay parlance as a 'lazy eye'. This should never be accepted as a diagnosis until confirmed by careful examination.

In recent years there has been a considerable research interest in this type of amblyopia and some information is now available concerning its neurological background, even though the primary site of the defect is still uncertain.

Causes of Squint in Childhood

1. Refractive error — hypermetropia, myopia
2. Opaque media — corneal opacities, cataract, uveitis
3. Disease of retina or optic nerve — retinoblastoma, optic atrophy
4. Congenital or acquired weakness of extraocular muscles
5. Abnormalities of facial skeleton leading to displacement of extraocular muscles

Refractive Error

In order to understand how refractive error can cause squint, one must first understand how the act of accommodation is linked to the act of convergence. That is to say, we must realise that when we focus upon an object not only is each individual eye separately focused on it, but the eyes swivel together by the requisite amount to allow them both to view at once. Accommodation is thus closely linked to convergence because a given amount of accommodation must be associated with an equivalent amount of convergence. In hypermetropic subjects this relationship between accommodation and convergence may be upset. In order to overcome hypermetropia, the eyes must accommodate excessively and sometimes this excessive focusing needed to view things clearly

induces excess convergence and hence causes a squint. This type of accommodative squint may be fully corrected simply by wearing spectacles. If there is a difference in refractive error between the two eyes, and especially where the image on one retina remains blurred, amblyopia may develop with consequent weakening of binocular function and even squint. The commonest type of childhood squint is the convergent squint associated with hypermetropia.

Opaque Media

Congenital cataract can occasionally present as a squint. In a similar manner, if one cornea becomes opaque as the result of infection, for example herpes simplex keratitis, or as the result of injury, then a squint may develop within a few weeks. A completely blind eye from whatever cause tends to converge if the blindness occurs in early childhood. Blindness of one eye in an adult tends to result in a divergent squint. This is sometimes a useful indicator of the age of onset of blindness.

Disease of the Retina or Optic Nerve

Such a possibility provides an important reason for the careful examination of the fundus in every case.

Congenital or Acquired Muscle Weakness

Accommodation spasm is an interesting but unusual cause of squint in children. The patients are usually young girls aged 8–11 years and there is often a history of domestic upset or social problems at school. Such patients are referred because they have difficulty in reading the Snellen test type on routine testing at school; when asked to perform this test they exercise their accommodation, making everything appear blurred. The excessive accommodation is on occasions accompanied by overconvergence which may persist. Myasthenia gravis is extremely rare in children but it may present as a squint. Sixth, third or fourth cranial nerve palsies are sometimes seen after road traffic accidents and the surgeon must always bear in mind the possibility of a sixth or other cranial nerve palsy being associated with raised intracranial pressure.

Musculofascial Abnormalities

In some cases of squint there is a degree of facial asymmetry. These patients may also have 'asymmetrical eyes', one being myopic or hypermetropic relative to the other. Sometimes there is no refractive error but there is assumed to be an asymmetry of the insertions of the extraocular muscles which

may account for the failure to retain binocular vision. There is a group of conditions known as the musculofascial anomalies in which there is marked limitation of the eye movements in certain directions accompanied by abnormal eye movements such as retraction of the globe.

Diagnosis

History

When faced with a case of suspected squint there are certain aspects of the history which may be very helpful in assisting with the diagnosis. It is often useful to ask who first noticed the squint. Sometimes a mother has been made anxious by a well-wishing neighbour or relative, and in these cases there may be no true squint but merely the appearance of one. The mother is usually the best witness. Unfortunately some children have a facial configuration which makes the eyes look as though they are deviating when they are not and it is essential that the student or general practitioner should be able to make this distinction if he is to avoid sending unnecessary referrals to the hospital (Fig. 13.2). Concomitant squint in childhood tends to be inherited in a dominant pattern and the family history of squint provides a useful diagnostic pointer. From the point of view of prognosis, it is useful to find out whether the squint

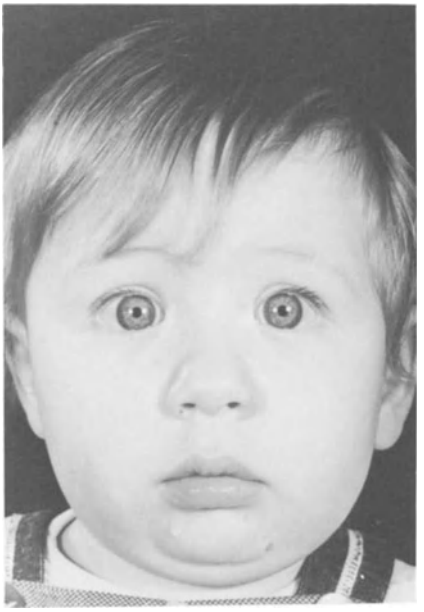

Fig. 13.2. Epicanthus.

is constant or intermittent and also the age of onset. A full ophthalmic history must be taken which should include the birth history and any illness which might have caused or precipitated the problem.

Examination

While the history is being taken from the parents, one should be making an assessment of the child. If the child is obviously shy or nervous, a useful method is to introduce something of interest to the child in the conversation with the parents. At this point it is important not to approach the child directly but to allow him or her to make an assessment of the doctor. It is quite impossible to examine an infant's eye in a noisy room, thus the number of people present should be minimal and they should not be moving about. The room lighting should be dim enough to enable the light of a torch to be easily seen. The first important part of the examination is to shine a torch at the patient so that the reflection of the light can be seen on each cornea. The position of these corneal reflexes is then noted carefully. The more mobile the child, the less time is there to observe this. If there is a squint, the reflexes will be positioned asymmetrically in the pupil. If the patient has a left convergent squint, the reflection from the left cornea is displaced outwards towards the pupil margin. A rough assessment of the angle of the squint can be made at this stage by noting the position of the abnormal reflection. One of the difficulties experienced at this point is due to the continuous movement of the child's eyes, which may make it difficult at first to know whether the light is being accurately fixated. By gently moving the torch slightly from side to side, it is usually possible to confirm that the child is looking, albeit momentarily, at the light.

Once the light reflexes have been examined, the cover test can be performed. Once again, the reflection of the light from each eye is noted but this time one of the eyes is smartly covered, either with the back of the hand or with a card. The latter method is preferable, at least for student examinations, if not in actual practice. If the fixating eye is covered, a movement of the non-fixing eye to take up fixation may then be observed (Fig. 13.3). After some practice it is possible to detect even very slight movements of this kind. The result of the test may be misleading if the non-fixing eye is too weak to take up fixation and, indeed, an assessment of the vision of the non-fixing eye can be made during the cover test. This basic cover test is then elaborated if necessary. If no deviation is detected, the cover can be quickly swapped from one eye to the other and any movement of the eye which had been covered can be noted. That is to say, the latent deviation produced by covering one eye is spotted by noting the small recovery movement made by the previously covered eye. Finally the cover test is performed when the patient is fixing a distant object. One type of squint in particular can be missed unless this is done. This is the divergent squint seen in young children which is often only present when viewing distant objects. The parents may have noticed an obvious squint and yet testing by the doctor in the confines of a small room reveals nothing abnormal, with ensuing consternation all round.

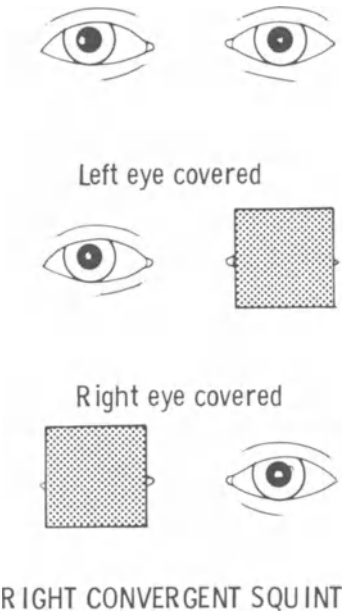

Fig. 13.3. The cover test.

After the cover test has been performed it is necessary to test the ocular movements to find out whether the squint is concomitant or incomitant, and at this stage in the preliminary examination it is usual to instil a mydriatic and cycloplegic drop (cyclopentolate 1%) in order to obtain a measure of the refractive error when the eyes are completely at rest. At the same time the optic fundus can be examined.

In most instances the nature of the squint becomes apparent by this stage and further testing of the binocular function and more accurate measurement of the angle of the squint are carried out using the synoptophore. In some cases further tests, which may include an examination under anaesthesia or even a CAT scan, are needed.

Management of Squint in Childhood

Glasses

Any significant degree of refractive error is corrected by the prescription of glasses. Sometimes the squint is completely straightened when glasses are worn but more often the control is partial, the glasses simply acting to reduce the angle of the squint. Glasses may be prescribed in a child as young as 6–9 months if really necessary. It is important that the parents have a full understanding of the need to wear glasses if adequate supervision is to be expected. When the spectacles are removed at bed time, a previous squint may

appear to become even worse and the parents should be warned about this rebound effect.

Orthoptic Follow-Up

The orthoptic department forms an integral part of the modern eye unit. It is run and manned by orthoptists who can be regarded as physiotherapists of the eyes. Once the patient has started wearing the glasses a further assessment in the orthoptic department is then arranged. This assessment includes the following.

1. *Measurement of visual acuity* with and without glasses, reading and distance. For infants, modifications of the Snellen test are used, such as the Stycar test. For very small children, the ability to fix minute test objects, such as polystyrene spheres, is measured.

2. *Prism bar cover test.* This is a modification of the cover test already described in which a series of graded prisms is placed in front of the deviating eye and the size of prism needed to displace the corneal reflection of the light to its proper position is noted.

3. *Grading of binocular function,* using the synoptophore, the presence of simultaneous macular perception, fusion, and stereopsis can be determined. When a squint has been present for some time, abnormal retinal correspondence (ARC) may have occurred. The eye which has turned in 'adapts' to its new relationship with the outside world instead of suppressing the image. This situation makes the treatment of the squint more difficult. Abnormal retinal correspondence is only observed when the eyes are being used together. When the squinting eye is used alone, macula fixation is restored. A more entrenched situation arises with eccentric fixation. Here the retina's idea of positions in space becomes permanently maladjusted and the eye remains misaligned even when the non-squinting eye is closed. By this stage it will have become apparent whether the squint is unilateral or alternating. In the latter case either one or the other eye is used more or less at random with convergence of the non-fixing eye. In the case of a unilateral squint the one eye remains permanently turned in and, in the absence of treatment, becomes amblyopic. There is a type of alternating convergent squint which is seen at birth; the infants may develop the habit of preserving the right eye to look at the left side of the visual field and vice versa. Early surgery is often needed in such cases of cross-fixation.

4. *Head posture.* The existence of head posture in a case of squint may indicate that good binocular vision is present but requires a turn of the head to be maintained. Surgery may be needed to eliminate the head posture. It is therefore important to distinguish ocular torticollis from true torticollis which is due to an abnormality of the sternomastoid muscle. A head posture always suggests that a group of extraocular muscles is not functioning. The simplest example of this is the head turn which occurs in the presence of a lateral rectus palsy; the position of the head gives a clue to the exact nature of the palsy.

Once the orthoptic assessment has been made, the question of treatment by occlusion of the good eye has to be considered. By covering the good eye for a limited period, the sight of the amblyopic eye can be improved. The younger the child, the better are the chances of success. In older children beyond the age of 7 or 8, not only is amblyopia more resistant to treatment, but the treatment itself can interfere seriously with school work. The type and amount of occlusive treatment have to be planned and discussed with the parents. Orthoptic exercises may also be used in an attempt to strengthen binocular function.

Surgery

If the squint is not controlled by glasses, surgery should be considered. Some parents are under the misapprehension that surgery is an alternative to glasses, but of course this is never the case, at least in younger children. From the cosmetic point of view, surgery is highly effective. The appropriate extraocular muscles are shortened or lengthened, measurements being made in millimetres to correspond with the angle of the squint in degrees. Sometimes two operations or more are needed due to occasionally unpredictable results, but from the cosmetic point of view, nobody need suffer the indignity of a squint. Once the eyes have been put straight or nearly straight by surgery, the functional result depends on the previous presence of good binocular vision and good vision in each eye.

Squint occurs in about 2% of the population and so it is a very common problem, but it is only a small proportion of these cases that eventually require surgery. The commonest type of squint in childhood is the accommodative convergent squint associated with hypermetropia and here surgery is indicated only when spectacles prove inadequate. Divergent squints are less common but more often require surgery. Intermittent exotropia is a well-defined entity occurring in children where the squint is only seen when viewing distant objects, and these children tend to close one eye when out walking in order to avoid diplopia. Binocular vision is usually quite normal in these patients and early surgery is effective.

Squint in Adults

Adults who present with a squint have usually suffered defective action of one or more of the extraocular muscles. It is important to have a basic understanding of the action of these muscles.

Anatomy of the Extraocular Muscles

These can be conveniently divided into three groups.

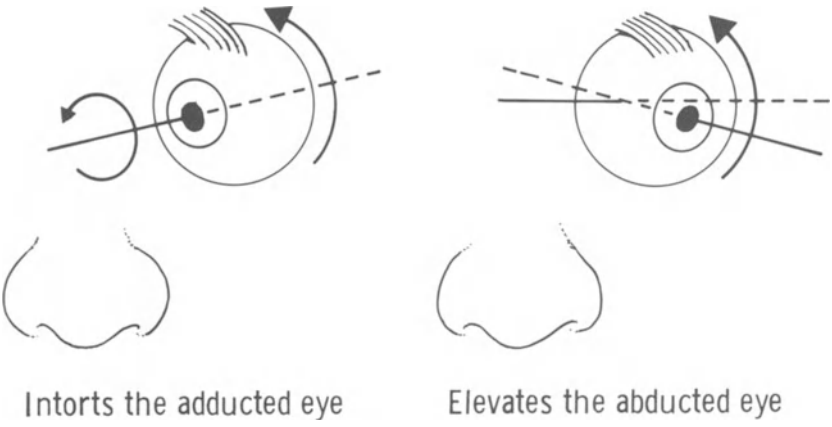

Fig. 13.4. Primary and secondary action of the superior rectus.

The Horizontal Recti. The medial and lateral recti act as yoke muscles, like the reins of a horse. They rotate the eye about a vertical axis. The lateral rectus abducts the eye (turns it out) and the medial rectus adducts the eye (turns it in).

The Vertical Recti. These act as vertical yoke muscles but they run diagonally from their origin at the apex of the orbit to be inserted 7 or 8 mm behind the limbus. The action of these muscles depends on whether the eye is abducted or adducted in the first place. The primary action of the superior rectus is to elevate the abducted eye and the inferior rectus depresses the abducted eye. The secondary action of the superior rectus is to adduct and intort the adducted eye; the inferior rectus adducts and extorts the adducted eye. Intorsion and extorsion refer to rotation about an anterior–posterior axis through the globe. The important thing to realise is that the action of these vertical muscles depends on the position of the eye (Fig. 13.4).

The Obliques. These are also vertical yoke muscles but they are positioned in the opposite diagonal to the recti. The inferior oblique elevates and the superior oblique depresses the adducted eye. When the eye is abducted, the superior oblique intorts and abducts it and the inferior oblique extorts and abducts it.

When a patient has a fourth cranial nerve palsy on the right side, the right eye can no longer look down when it is turned in (or adducted) due to the defective superior oblique muscle. Double vision is experienced which is maximal (i.e. widest displacement of images) when looking down to the left.

When a patient has a sixth cranial nerve palsy on the right side, the right eye can no longer abduct. A right convergent squint is seen and the patient experiences double vision except when looking to the left. There may be a head turn to the right.

When a patient has a third nerve palsy on the right, the right eye is turned down and to the right and, if the palsy is complete, the upper lid droops and

the pupil is dilated. Movement of the eye is very limited but some torsion may be seen if the superior oblique is working.

Causes

Adults who *present* with a squint usually have a well-defined ocular muscle palsy. This may be due to a pathological process at any point from the brain, through the nerve to the muscle; these will be discussed elsewhere. Two important causes are disseminated sclerosis in the younger age groups and hypertensive vascular disease in older patients. Diabetes is another important cause that must be excluded in all age groups.

Some adult squints prove to be neglected childhood concomitant squints and sometimes a latent squint which has been well controlled throughout childhood breaks down in adult life.

In adult life a blind eye tends to turn outwards and a divergent squint may be due solely to impaired vision.

Diagnosis

In contrast to the situation with children, who very rarely complain of their squints, the sudden onset of a squint in adult life is extremely disabling because of intractable double vision. The double vision is less apparent when the lesion is more central, involving the level of the cranial nerve nucleus or above. Here the patient tends to complain more of blurred vision and confusion.

A carefully taken history may reveal the diagnosis. First it is necessary to ensure that the double vision is only present with both eyes open and then the patient can be questioned about the position of the second image and whether the separation of the images is maximal in any particular direction of gaze. The duration and constant or intermittent nature of the squint must be determined as must the history of any relevant associated disease, past or present.

Once the history has been obtained, the nature of the squint can be investigated by the cover test and measured by the Maddox wing and Maddox rod. An accurate record of the impaired muscle action can be recorded on the Hess screen.

Maddox Wing

This ingenious but simple device is held in the patient's hand. By looking through the eyepieces, one eye is made to look at an arrow and the other at a row of numbers. If the eyes are straight, the arrow points at zero, and if not, the arrow indicates the angle of the squint.

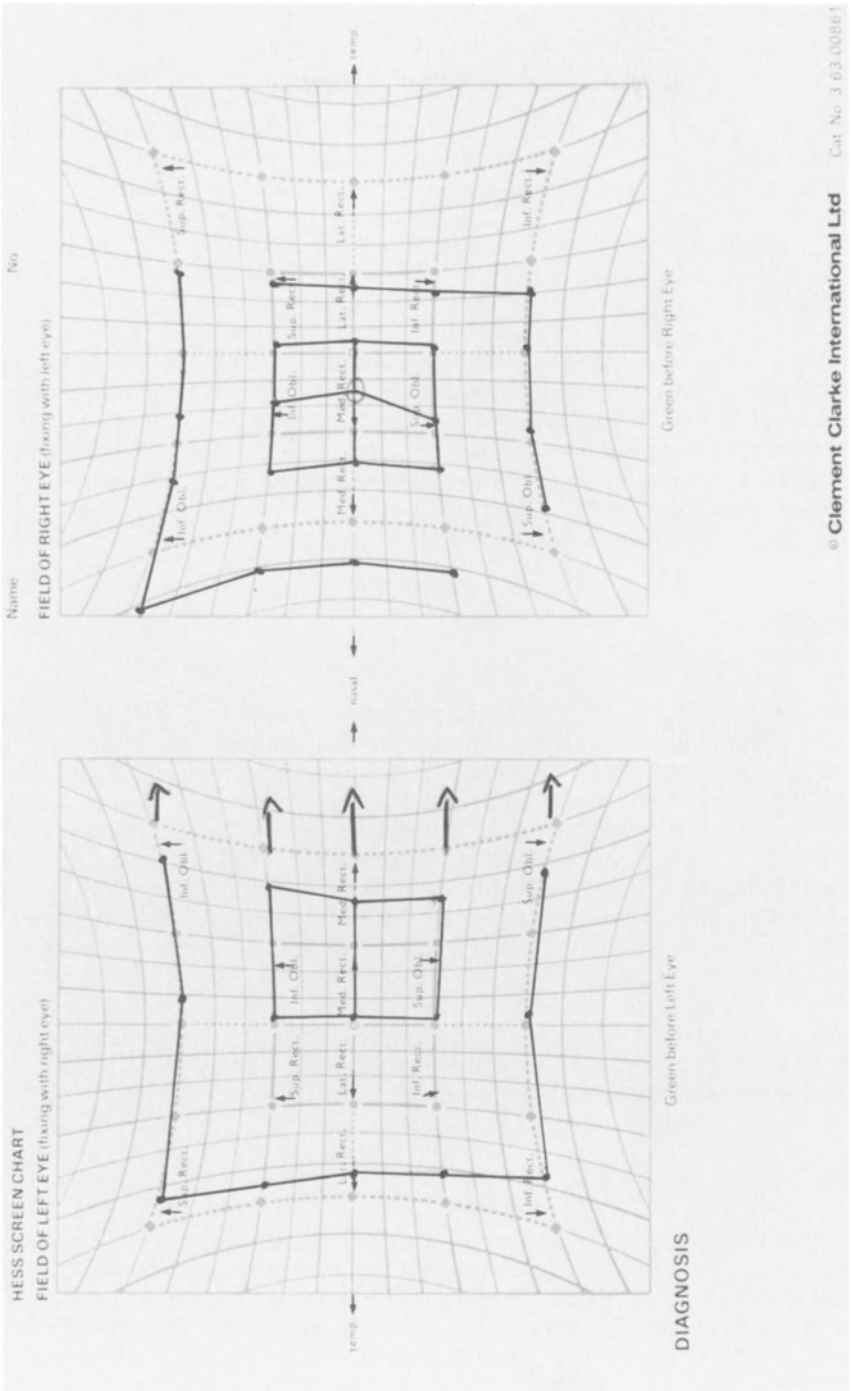

Fig. 13.5. Right lateral rectus palsy as recorded on Hess screen.

Maddox Rod

The Maddox wing measures the deviation at reading distance and the rod is a similar device to measure the deviation when viewing a distant object. A special optic glass is placed in front of one eye which turns the image of a light source into a line image. One eye, therefore, views the point source of light and the other a line, and the separation of these two images can be measured on a scale.

Hess Screen

Here the eyes are dissociated by placing a green filter in front of one and a red filter in front of the other. Two small torches are then used which project spots of coloured light complementary to those seen through the goggles. The patient holds one of the torches and the observer the other. The observer projects his light onto a special screen and the patient is asked to project his light onto what appears to him to be the position of the observer's light. Any discrepancy is recorded on a chart which is kept in the patient's notes (Fig. 13.5).

Treatment

Many cases of adult squint recover spontaneously within a period of 3–6 months. The immediate treatment entails eliminating the diplopia by occluding one or other eye. This may be conveniently achieved by applying adhesive tape to the spectacle lens. If the angle of the squint is sufficiently small, it may be possible to regain binocular vision by means of a prism. Fresnel prisms are thin and flexible and can be simply stuck onto the spectacle lens as a temporary measure during the recovery period. When the squint shows no sign of recovery over a period of 9 months or more, then surgery is usually required to restore binocular vision. Before applying these principles of management it is essential to treat the underlying cause of the squint. It would be a serious error to treat the diplopia due to raised intracranial pressure by means of prisms without instituting a full neurological investigation, just as it would not help the patient with myasthenia gravis to undergo squint surgery before medical treatment has been started.

Ocular Muscle Imbalance

Mild latent squints may sometimes go undetected until a period of stress or excessive reading precipitates symptoms of eye strain and headache. Under normal circumstances the eyes are perfectly straight, but it is the effort to keep them straight that produces the symptoms. Heterophoria is the overall term applied to such a problem and the latent deviation may be inwards (esophoria), outwards (exophoria), upwards or downwards (hyperphoria or

hypophoria). Because most people's eyes assume a slightly divergent position when completely at rest, a degree of exophoria is almost the rule and is usually of no significance. Vertical muscle imbalance is less well tolerated and even a slight deviation may cause symptoms. Small but significant degrees of vertical muscle imbalance are seen in otherwise normal individuals who show a marked difference in refractive error between the two eyes, or in those with facial asymmetry. The provision of a small prism incorporated into the spectacle lenses of such patients may produce dramatic relief, but we must always remember that the appearance of an ocular muscle imbalance may be the first indication of more serious underlying disease. A small vertical deviation may be the first sign of a tumour of the lacrimal gland or thryotoxic eye disease and a wide range of investigations may be needed before one can be satisfied with the excellent results of symptomatic treatment.

14 Tumours of the Eye

In this chapter the more important ocular tumours will be considered. There are a considerable number of other rare tumours and the interested student should refer to one of the more specialised and comprehensive textbooks of ophthalmology for further reading.

The Globe

Expanding tumours in the eye present diagnostic problems because it is not usually possible to biopsy them.

Malignant Melanoma. The commonest intraocular tumour is the malignant melanoma of the choroid, which differs from melanoma of the skin in that it grows more slowly and metastasises late. At first it is seen as a raised pigmented area which may be anywhere in the fundus (Fig. 14.1). As the

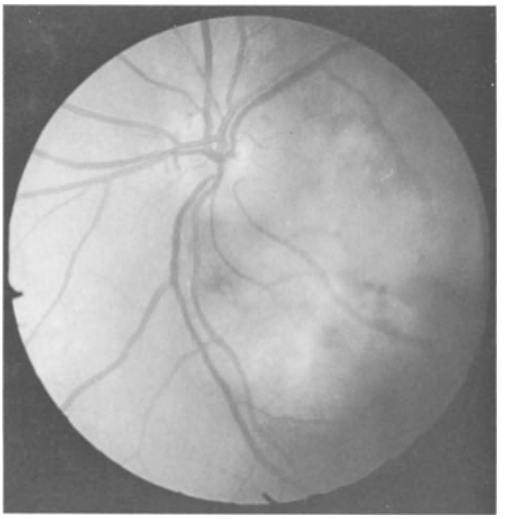

Fig. 14.1. Choroidal melanoma.

tumour enlarges there may be an associated retinal detachment or, less often, secondary glaucoma. Initially the appearance may resemble a choroidal haemorrhage and serial photography may be needed to confirm the diagnosis. The appearance of metastases may be delayed for several years and may occur even if the eye has been removed. Most cases are still treated by removal of the eye but radiotherapy and local removal have been advocated and justified by the ultimate poor prognosis following enucleation (surgical excision of the eye). Untreated, the tumour may extend into the orbit and provide an unpleasant problem for the patient.

Retinoblastoma. This is a rare tumour of childhood which arises not from the choroid but, as its name suggests, from the retina. It shows certain rather strange and unusual features. It is usually present from birth, is usually inherited as an autosomal dominant, and in a quarter of cases appears in both eyes. Initially it may be seen in an individual, suspected on account of the family history, as a small, white, raised mass. Examination under anaesthesia is essential in such cases because the tumour may be in the extreme periphery of the fundus. A larger tumour may present as a white mass in the pupil ('leucocoria') and such an appearance in infancy demands immediate referral to an ophthalmologist. Extension tends to occur locally up the optic nerve and enucleation is often life saving. Smaller tumours can be treated effectively by radiotherapy and many patients now survive into adult life. Genetic counselling is essential for these patients in order to prevent the increasing incidence of the tumours which will result from effective medical treatment.

Melanoma of the Iris. This rare pigmented iris tumour causes distortion of the pupil, which may be a warning sign. It is extremely slow growing and probably much less malignant than was originally supposed.

The Eyelids

Benign vascular tumours of the eyelids fall into three types.

Capillary Haemangioma of the Newborn ('Strawberry Naevus'). This is seen quite frequently and nearly all examples regress spontaneously. The tumours appear as red, slightly raised marks on the skin. Even very extensive tumours of this kind can show a dramatic improvement over several years and conservative management is usually indicated unless the tumour is associated with a fold of skin which occludes the eye. The possibility of amblyopia of disuse must always be borne in mind.

Cavernous Haemangioma. This tumour lies more deeply in the skin and appears as a bluish swelling in the lid which expands when the child cries. These lesions may also disappear spontaneously or, if persistent, they may be treated by freezing.

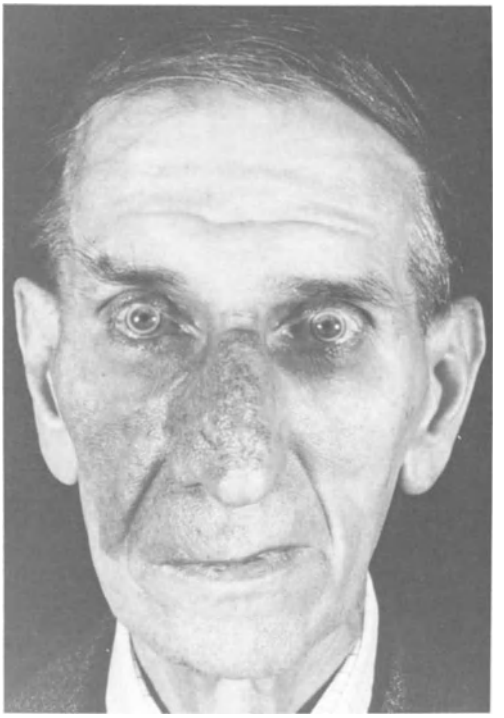

Fig. 14.2. Port wine stain.

Telangiectatic Haemangioma. Also known as the port wine stain, this tumour tends to be distributed over the area supplied by one or more of the branches of the fifth cranial nerve and usually remains throughout life as a dark red discoloration in the skin (Fig. 14.2). The importance of this particular appearance is its association with secondary glaucoma and haemangioma of the meninges. The latter produces calcification and a characteristic x-ray appearance. The combination of lesions is known as the Sturge–Weber syndrome.

Basal Cell Carcinoma. This is the commonest malignant tumour of the eyelids in adults. The tumour begins as a small insignificant nodule which turns into a small crater-like lesion with a slightly raised edge. At this stage it is a simple matter to remove the lesion and confirm the diagnosis by biopsy, but if left the tumour tends to spread and the prognosis becomes worse once the underlying bone is invaded (Fig. 14.3). Extensive neglected basal cell carcinomata are best treated palliatively by radiotherapy; they very rarely metastasise.

Squamous Cell Carcinoma. This tumour may initially resemble the basal cell carcinoma although it is distinguishable histologically. Spread tends to occur

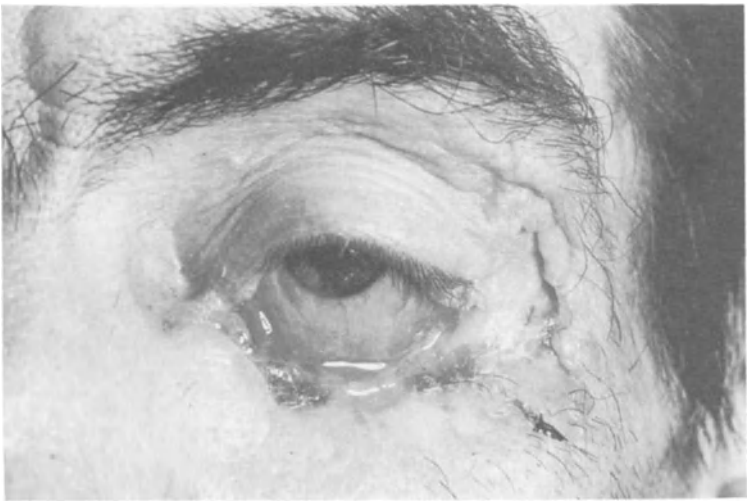

Fig. 14.3. Extensive basal cell carcinoma involving bone.

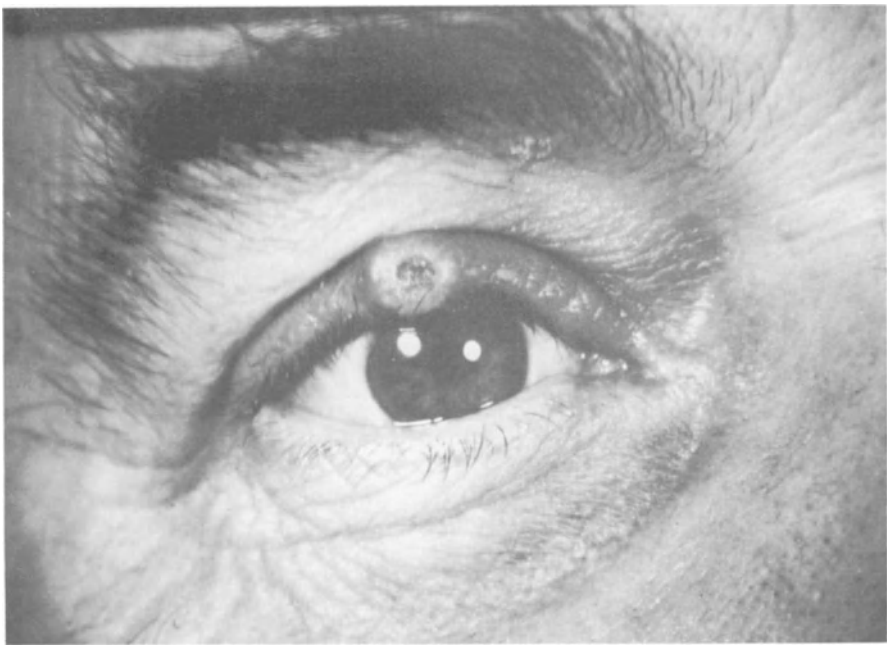

Fig. 14.4. Keratoacanthoma of upper lid.

to the local lymph nodes, preauricular for the upper lid and submandibular for the lower lid, and the tumour is liable to metastasise.

There are a number of benign neoplasms which occur on the eyelids and which may resemble basal cell or squamous cell carcinomata in their early stages.

Keratoacanthoma. This is an example of such a lesion which grows rapidly, too rapidly for a tumour, over a period of a few weeks and then resolves spontaneously (Fig. 14.4). Small lumps on the eyelids should be removed and biopsied. Larger lumps may be biopsied by taking a small segment from them prior to total excision if this proves necessary. Special care should be taken with the excision of any lesion on the eyelid in view of the risk of causing distortion of the lid margin or exposure keratitis.

Papilloma. This is the name used to describe a rather common virus-induced lump or nodule often seen on the lid margin.

Malignant Melanoma and Conjunctival Naevus. Malignant melanoma of the eyelids is similar to malignant melanoma elsewhere, appearing as a raised, often shiny black lump. It metastasises at a very early stage and the prognosis does not seem to be altered by excision. Malignant melanomata may also occur on the conjunctiva (Fig. 14.5), but they should not be confused with the relatively common benign conjunctival naevus. The latter is a slightly raised pigment-stippled lesion often seen at the limbus on the temporal side. Closer examination with the hand lens or microscope reveals one or two minute cysts. It is generally accepted that these lesions should be excised if they become irritable or sometimes simply on cosmetic grounds, but they very rarely become malignant.

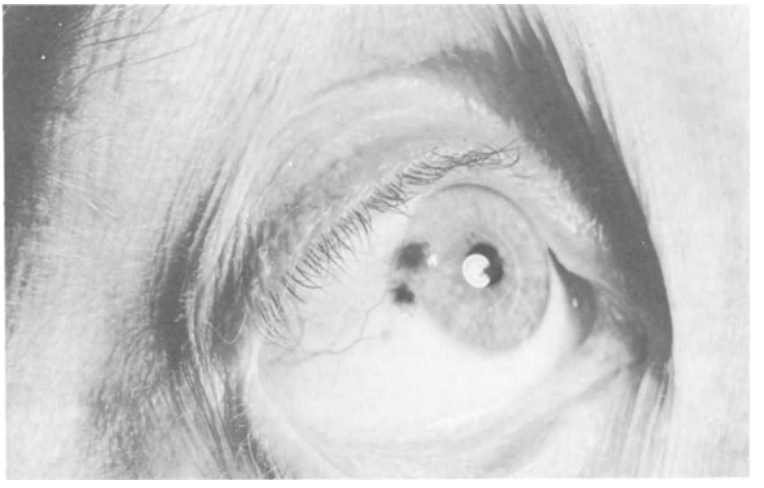

Fig. 14.5. Malignant melanoma of the conjunctiva.

The Orbit

Dermoid Cyst. This cystic swelling is usually seen at the level of the eyebrow in the upper outer part of the orbit. It is smooth and fluctuant and often fixed to bone. Sometimes a deeper part of the cyst may occupy a cavity in the bone and an x-ray with tomograms is advisable when this is suspected. Excision on cosmetic grounds and for diagnosis is usually indicated.

Pseudotumour. This is an inflammatory swelling in the orbit of unknown cause which may present with proptosis and diplopia. A mass may be palpable in the orbit and biopsy reveals non-specific inflammatory tissue. Diagnosis may eventually be by exclusion of other causes of proptosis. In severe cases a course of systemic steroids is usually very effective.

Haemangioma. This is the commonest primary neoplasm of the orbit. It is benign and more often seen in children. It is unusual for surgery to be necessary in such cases.

Glioma of the Optic Nerve. This rare tumour causes progressive proptosis and optic atrophy but it may be very slow growing. There is an association with Von Recklinhausen's disease, and the presence of pigmented patches in the skin should make one suspect this.

Rhabdomyosarcoma. This rare but highly malignant orbital tumour is seen in children. Its growth is so rapid that it may be misdiagnosed as orbital cellulitis. If a correct diagnosis is made at an early stage, there is some hope of reaching a cure by radiotherapy. The tumour arises from striated muscle and the histological diagnosis is confirmed by finding striation in the tumour cells.

Metastatic Tumours and Tumours from Neighbouring Sites. A wide variety of tumours may invade the orbit and produce proptosis and often diplopia. Lymphoma is one example; it may present as an isolated lesion or in association with Hodgkin's disease or leukaemia. Examples of local spread from adjacent structures include carcinoma of the nasopharynx, carcinoma of the lacrimal gland and meningioma.

Exophthalmos and Proptosis

Both these terms mean forward protrusion of the eyes but traditionally exophthalmos refers to the bilateral protrusion in thyroid disease. Proptosis refers to unilateral forward displacement of the globe from whatever cause. In practice the terms tend to be used rather loosely and are now almost synonymous.

Causes of Proptosis

When one eye seems to bulge forward the doctor may have a serious problem on his hands and he should be able to consider the likely causes.

Pseudoproptosis. An apparent bulging forward of the eye occurs if the eye is too big, as in unilateral high myopia, or if the other eye is sunken following a blow-out fracture of the maxilla.

Muscle Palsy. A slight degree of proptosis, about 1 or 2 mm, accompanies palsies of the extraocular muscles.

Thyrotoxicosis. This is the commonest cause of unilateral or bilateral proptosis, diagnosis being achieved from the history and tests of thyroid function. (See later for the 13 signs of thyroid disease in the eyes.)

Infection. Orbital cellulitis, usually from neighbouring sinuses, requires urgent otorhinological opinion.

Trauma. Proptosis can occur as a result of retro-orbital haemorrhage. Diagnosis should be possible from the patient's history.

Haemangioma. This may expand after bending down. The diagnosis may be confirmed by angiography.

Pseudotumour. Biopsy should be carried out if possible, and other causes excluded, after which a trial of steroids should be considered.

Mucocele of Sinuses. Diagnose by x-ray.

Lymphomatous Tumour. A biopsy, full blood count and sternal marrow test should be carried out.

Others. There is a large number of possible rare causes for proptosis.

Assessment of Proptosis

In the clinic, proptosis is best assessed by standing behind the seated patient and asking him to look down. The position of each globe in relation to the lids and face can be best seen by this means. Proptosis can be measured by means of an exophthalmometer. A number of such instruments are on the market and they depend on measuring the distance from the rim of the outer margin of the orbit to the level of the anterior part of the cornea. These measurements are not always very accurate but best results are achieved by ensuring that they are made by the same person on each occasion for a given patient.

Once thyroid disease and trauma have been excluded, the patient may have to be admitted to the ward for further investigations including CAT scan, possibly angiography and sometimes orbital biopsy.

15 Ocular Trauma

That injuries to the eye and its surrounding region demand special attention and create great concern for patient and doctor is self-evident when the eye alone is involved, but when other life-threatening injuries are present the eye injury, seeming slight at the time, may be overlooked. Sometimes the eyelids may be so swollen that it is difficult to examine the eyes and a serious perforating wound may be obscured. When other injuries are present and an anaesthetic is needed, it is essential that the eyes are examined carefully, if possible under the same anaesthetic. As in the case of injuries elsewhere, those to the eye demand urgent and immediate treatment, and neglect can result in tragedy from what initially seems a very slight problem.

Injuries to the Globe

Contusion

The eye casualty officer comes to recognise a familiar pattern of contusion, the effect of squash-ball injuries and golf balls in sport and the result of blows from large flying objects in industry or after criminal assault. Injuries in industry have now been better controlled by providing protective clothing and making machinery safer. Sporting injuries are now also being controlled more effectively, although at the time of writing there are still frequent injuries from squash balls. The prospect of the wider use of protective goggles in sport may well change this. Rather ironically, a recent type of injury was the result of wearing a particular kind of swimming goggle but the fault in the goggle appears to have been corrected. Because of the protection provided by the surrounding orbital margin, footballs, and even tennis and cricket balls, do not usually cause serious contusion. The rare golf-ball contusion injury is especially dangerous, often resulting in loss of the eye.

The extent of damage to the eye from contusion depends on whether it has been possible to close the eyelids in time before the moment of impact. If the lids have been closed, bruising and swelling of the eyelids is marked and the injury to the eye may be minimal. It must be remembered though that this is not an infallible rule and the eyes themselves must always be carefully

examined, even when there is extreme swelling of the lids. It is always possible to examine an eye by retracting the eyelids under anaesthesia if necessary.

The important clinical features of contusion injury are best considered by looking at the anatomical parts of the eye from anterior to posterior:

Cornea

The epithelium may be abraded causing extreme pain and photophobia. Examination here is facilitated by instilling drops of local anaesthetic. However,the drops should not be continued as treatment, in spite of the relief that they bring, because they seriously delay healing.

Anterior Chamber

A small bleed into the anterior chamber is seen as a fluid level of blood inferiorly ('hyphaema'). Many ophthalmologists take this as a sign to admit the patient to hospital, mainly because of the risk of secondary bleeding after 2 or 3 days. This risk is especially serious in children where a secondary bleed is liable to cause secondary glaucoma with serious consequences. When the anterior chamber is filled with blood and the intraocular pressure rises, the cornea tends to become blood-stained. This can result in a permanent corneal opacity.

Iris

When confronted by a flying missile, the normal reaction is to attempt to close the eyelids and to rotate the eyes upwards. This is the reason why the commonest point of impact is the lower temporal part of the eye and it is in

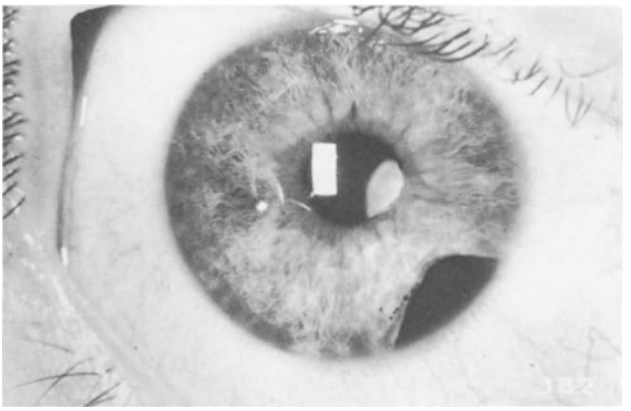

Fig. 15.1. Iridodialysis.

this region of the iris that one is most likely to see peripheral iris tears ('iridodialysis'). When the eye is compressed the iris periphery is torn at its root, leaving a crescentic gap which looks black, but through which the fundus and red reflex can be observed. Such an injury also provides an excellent view of the peripheral part of the lens and the zonular ligament (Fig. 15.1).

Contusion may result not in a tear of the iris root, but in a splitting of the iris and ciliary body from the sclera producing recession of the angle of the anterior chamber; this appearance is often associated with secondary glaucoma.

A sudden impact on the eye may also produce microscopic radial tears in the pupillary sphincter of the iris. This may be a subtle microscopic sign of previous injury when no other signs are present, or the damage may be more severe resulting in permanent dilatation of the pupil (traumatic mydriasis).

Lens

Any severe contusion of the eye is liable to cause cataract, but the lens may not become opaque for many years after the injury. The lens may also become subluxated (slightly displaced due to partial rupture of the zonular ligament) or even dislocated into the anterior or posterior chamber. Displacement of the lens also tends to be associated with secondary glaucoma.

Vitreous

The vitreous may become detached after a contusion injury if it has not already undergone this change as part of the normal ageing process. Vitreous haemorrhage is another sequel of contusion. Such haemorrhages usually clear completely in time but they tend to accompany more serious damage to the retina which may only be fully revealed when the vitreous has cleared.

Retina

Bruising and oedema of the retina are seen as grey areas with scattered haemorrhages. The macula region is susceptible to oedema after contusion injuries and sometimes the elevated oedematous retina around the macula gives the appearance of a 'macula hole'. Just as tears can occur in the peripheral iris, so a similar problem is seen in the peripheral retina. These crescentic retinal dialyses are also most common in the lower temporal quadrant and their importance lies in the fact that they may lead to retinal detachment unless the tear is sealed by cryopexy or some form of light coagulation. Any significant contusion injury of the eye must command careful examination of the peripheral fundus.

The total picture of retinal oedema and haemorrhage after injury is termed commotio retinae, or 'Berlin's oedema'.

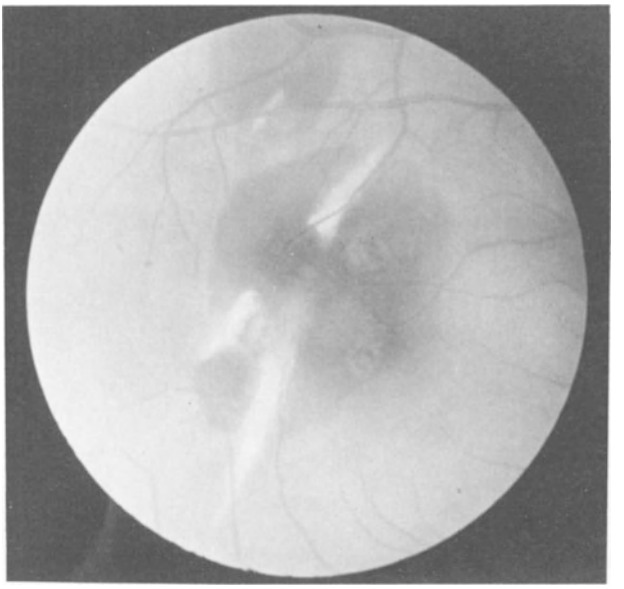

Fig. 15.2. Choroidal tear with associated retinal haemorrhage following trauma.

Choroid

Tears in the choroid following contusion injury have a characteristic appearance. They are concentric with the disc and are seen as white crescents where the sclera is exposed. When near the macula, there is usually permanent damage to the central vision (Fig. 15.2).

Optic Nerve

A variable degree of optic atrophy may become apparent a few weeks after a contusion injury. Blunt injuries to the eye may cause bleeding into the optic nerve sheath, and this can result in complete loss of vision on the affected side, the blindness being permanent.

Management

In most instances the management of these injuries involves observation and bedrest if there has been bleeding into the eye. The most important part of the management is to recognise and treat any complications that arise, in particular secondary glaucoma and retinal tears. Acetazolamide given by mouth and timolol eyedrops are useful in controlling the intraocular pressure.

Perforation

A perforating wound of the eye is a surgical emergency. As soon as the globe is penetrated there is a serious risk of infection. The vitreous is an excellent culture medium and in the pre-antibiotic era eyes were totally lost within 2 or 3 days as a result of this. Perforating injuries are seen in children from scissor blades, screwdrivers or darts, and in adults from road traffic accidents. It is likely that the introduction of the compulsory wearing of safety belts will eliminate the vast majority of perforating injuries from broken car windscreens.

The outcome of a perforating injury is dependent on the depth of penetration and the care with which the wound is cleaned and sutured. If the cornea alone is damaged, excellent results may be obtained by careful suturing under the operating microscope with the patient under general anaesthesia. Once the lens has been damaged the patient will sooner or later have to face the problems of aphakia. The use of intraocular acrylic implants gives some promise in this respect. Deeper penetration introduces the further risk of retinal detachment.

On admission or in the casualty department, the patient is given tetanus prophylaxis and both a systematic and local antibiotic. If the iris has prolapsed into the wound, then this must be carefully cut away so that iris tissue is not included in the healing wound, as this would increase the risk of sympathetic ophthalmia, a type of inflammation which can affect the undamaged eye. The recent developments in instrumentation for vitreous surgery have also altered the prognosis after deeper injuries because it is now possible to remove tissue in a much more meticulous and controlled manner.

Sympathetic Ophthalmia

This rare complication of perforation is most common in children. The injured eye remains markedly inflamed and the wound may have been cleaned inadequately or too late. Over a period of 2 weeks to several months, a particular type of inflammatory response appears in the uvea. The choroid is infiltrated with eosinophils, lymphocytes and giant cells. Subsequently a similar type of reaction is seen in the other eye so that ultimately the patient risks becoming completely blind. This strange granulomatous reaction which may follow perforating injury and also surgery is fortunately rare and responds quite successfully to treatment in most cases. It is thought to be the result of hypersensitivity to uveal pigment. The warning signs are the presence of photophobia or blurring of vision in the sound eye, and granulomatous keratic precipitates are seen on the posterior surface of the cornea with the slit-lamp microscope. Preventive treatment entails the removal of the injured eye and, if this is done within 2 weeks of the injury, the risk to the other eye is negligible. This means that any severely injured eye may need to be removed if it remains unduly inflamed. Modern surgery probably allows the preservation of many eyes that would have been removed in previous years. If sympathetic

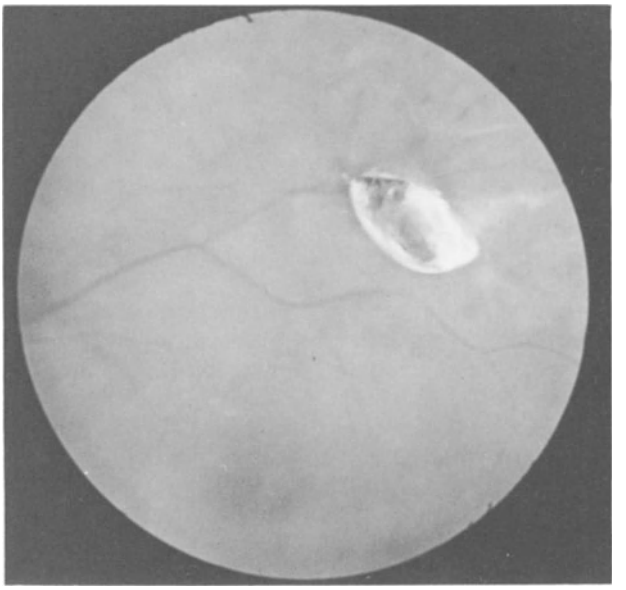

Fig. 15.3. Metallic intraocular foreign body lying against retina.

ophthalmia does occur some time after the injury, then the condition is treated with local and systemic steroids; although a good response is often obtained, the condition tends to recur.

Intraocular Foreign Body

Metallic foreign bodies tend to enter the eyes of those who operate high-speed grinders without goggles or those using a hammer and chisel on metal without protection. These injuries may seem slight at the time and the patients may not attach much importance to them. **Any such eye injury with this occupational history warrants an x-ray of the eye.** When ferrous metals remain in the eye they may cause immediate infection, or, if not, deposition of iron salts occurs in a process known as siderosis. This may lead eventually to blindness of the eye. Other metals also tend to give similar reactions, and for these reasons the metallic fragment should be removed (Fig. 15.3). Fortunately this can often be achieved using a magnet. The site of removal depends on the position of the foreign body in the eye. Localisation of the foreign body can be accurately made by suitable x-ray techniques. Airgun pellets cause particularly severe eye injuries and if the pellet remains in the eye, the eye is usually lost because of the extensive disruption produced at the time of injury. Some intraocular foreign bodies, such as some of the newer alloys or particles of glass, may well be tolerated by the eye and the decision must be made as to whether the act of removing the foreign body will cause more disruption than leaving it alone.

When a foreign body is found lying deeply in the cornea, its removal may result in loss of aqueous and collapse of the anterior chamber. It is prudent to arrange that removal should be done under full sterile conditions in the operating theatre where the corneal wound can be sutured if necessary. Careful slit-lamp examination is particularly important when a perforating injury is suspected. Under the microscope it may become quite obvious that a small corneal wound is full thickness.

Injuries to the Eyelids

Loss or destruction of eyelid tissue should always be treated as a threat to vision. The upper lid especially is important in this respect. The immediate concern is to ensure that the cornea is properly covered when the lids are closed. If more than one-third of the margin of the upper lid is lost, this must be replaced by grafting from the lower lid. When less than one-third is missing, the gaping wound can usually be closed directly. Up to one-third of the lower lid can also be closed by direct suturing, but when more than this is lost or when it has to be transferred to the upper lid, a slide of tissue from the lateral canthus can be effected, combined if necessary with a rotating cheek flap.

One of the most important features of the repair of lid injuries is the method of suturing. If the lid margin is involved, the repair should be made under the operating microscope. An untidy repair can result in a permanently watering eye due to kinking of the lid. The lid is best sewn in at least two layers, using catgut for the conjunctiva, 7-0 silk for the lid margin, and 4-0 silk for the skin. A horizontal mattress suture in the lid margin is a good way to prevent subsequent kinking. A tight wound should be avoided and on the relatively rare occasions when primary closure cannot be achieved, the wound is cleaned and debrided and then left until the swelling has subsided. Special attention must be paid when the medial part of the lid has been torn and this involves the canaliculus. Tears of the upper canaliculus do not seem to be so important but inadequate treatment of the lower canaliculus can leave a permanently watering eye. The proper treatment of this kind of injury entails suturing under general anaesthesia after threading a nylon thread through the upper canaliculus, back round the common canaliculus and into the medial end of the lower canaliculus and out into the wound. The end of the thread is then inserted through the remainder of the lower canaliculus and tied to the other end which is projecting from the upper canaliculus. This procedure sounds complex but in practice, and with the right instrument, is surprisingly simple.

Contusion of the eyelids, otherwise known as a black eye, is of course a common Saturday night problem for the casualty officer. Usually the presence of a black eye is an indication that the afflicted was smart enough to close his eye in time to avoid injury to the globe and it is unusual to find damage to the eyes after Saturday night fist fights, for example unless a weapon was involved. Broken beer glasses produce devasting injuries to the eyes as well as causing lid lacerations.

Injuries to the Orbit

Blows on the side of the cheek and across one or other eye occur as mining injuries or in road traffic accidents. The most common injury is the 'blow-out' fracture. Here the globe and contents of the orbit are forced backwards causing fracture of the orbital floor and displacement of bone downwards into the antrum. The inferior rectus muscle becomes tethered so that there is limitation of upward movement. The infraorbital nerve may also be injured producing anaesthesia of the skin of the cheek. Once the surrounding swelling has subsided, the posterior displacement of the globe becomes obvious and the globe may also show evidence of contusion. A considerable improvement from the functional and cosmetic point of view can often be obtained by positioning a plastic implant in the floor of the orbit after freeing the prolapsed tissue.

Fractures of the skull which extend into the orbit may be accompanied by retro-orbital haemorrhage and proptosis. Cranial nerve palsies affecting the ocular movements are also commonly seen in this type of injury and the vision may be affected by optic nerve damage. A blow on the eye may result in blindness with at first no other evidence of injury, but subsequently the optic disc becomes atrophic, usually after 2 or 3 weeks. The impaired pupil reaction to direct light is the only sign in these cases when they first present.

Foreign bodies may enter the orbit and, unless they are within the globe, it is usual to leave them alone rather than attempt a difficult exploration.

Radiational Injuries

The eyes may be exposed to a wide range of electromagnetic radiation from the shorter wavelength ultraviolet rays through the wavelengths of visible light to the longer infra-red waves, x-rays and microwaves. X-rays pass straight through the eye without being focused by the optical media and, in large enough doses, may cause generalised damage. It is especially important to realise that therapeutic but not diagnostic doses of x-rays tend to cause cataracts and the eye must be suitably screened during treatment. Excessive doses of x-rays also cause stenosis of the lacrimal canaliculi and retinal neovascularisation. As one might expect, visible light does not normally damage the eyes, although an intense light source may be absorbed by the pigment epithelium and converted to heat producing a macula burn. After eclipses of the sun there are usually a number of patients who arrive in the casualty departments of eye hospitals with macula oedema and sometimes serious permanent impairment of visual acuity. Sun gazing with consequent retinal damage has been reported after taking LSD. The laser beam provides a source of intense visible light which is used in ophthalmology as a deliberate means of producing burns in the retina. Some industrial and military lasers are extremely powerful and may produce accidental retinal burns even when reflected off shiny objects. Ultraviolet rays, which are shorter than visible

light, do not penetrate the eye but in large enough doses produce burns of the eyelids and cornea. On the skin this is seen as erythema and later pigmentation, and on the cornea a punctate keratitis is seen with the slit lamp. Ultraviolet damage of this kind is seen after gazing with unprotected eyes at welders' arcs, after exposure of the eyes to sunray lamps, and after exposure to the sun under certain conditions such as in snow on mountain tops. All these types of injury show a delayed effect, the symptoms appearing 2 or 3 hours after exposure and lasting for about 48 hours. There is usually severe pain and photophobia so that it may not be possible to open the eyes, hence the term 'snow blindness'. The use of locally applied steroid and antibiotic drops hastens recovery.

Unlike ultraviolet light, infra-red rays penetrate the eye and can cause cataract. A specific kind of thermal cataract has been well described in glass blowers and furnace workers but this is now rarely seen due to the use of protective goggles. Microwaves, in the form of diathermy, can cause cataract but the eye must be in the path of the beam if damage is to occur, and microwave ovens should not be expected to be dangerous in this respect unless the head were placed in the oven when it was switched on. Recently concern has been expressed about the possibility of radiation damage to the eyes from visual display units. Such damage has never been demonstrated, any more than it has from the face of a television set.

Further Reading

Bellows JG (1979) Management of glaucoma. Masson, USA
Duke-Elder Sir Stewart (1972) System of Ophthalmology, Vol. XIV: Injuries (2 vols.). Kimpton, London
Henderson JW (1980) Orbital tumours. Thieme Stratton, New York
Kolker AE, Hetherington J (1983) Diagnosis and therapy of the glaucomas, 5th edn (Becker and Shaffer). CV Mosby, St Louis
Reese AB (1976) Tumours of the eye, 3rd edn. Harper & Row, Philadelphia
Rosen ES, Haining WM, Arnott EJ, eds (1984) Intraocular lens implantation. CV Mosby, St Louis Toronto
Shields MB, ed. (1982) A study guide for glaucoma. Williams & Wilkins, London

Section IV

Problems of the Medical Ophthalmologist

16 Testing Visual Acuity

Measurement of visual acuity is the most important test of vision performed by the doctor and yet it is surprising how often it is omitted in examination by the non-specialist. It has already been shown that the differential diagnosis of the red eye can be simplified by noting the vision in the affected eye. After injuries of the eye it is just as important to note the vision in the uninjured eye as it is to note that in the injured eye. Simple measurement of the visual acuity is, of course, of limited value without a knowledge of the spectacle correction or whether the patient is wearing the appropriate spectacles.

Measuring the visual acuity means measuring the function of the macula, which is, of course, one small part of the whole retina. A patient may have grossly impaired visual acuity and yet have a normal visual field enabling him to walk about and lead a normal life apart from being able to read. This state of affairs is seen in patients with senile macula degeneration and can be compared with the situation in which a patient has grossly constricted visual fields but normal macula function, as is sometimes seen in retinitis pigmentosa or advanced chronic simple glaucoma. Here the patient appears to be blind, being unable to find his way about, but he may surprise the ophthalmologist by reading the test type from top to bottom once he has found it.

The simplest way to measure visual acuity might be to determine the ability to distinguish two points when placed very close together. Such a method was supposed to have been used by the Arabs when choosing their horsemen; they chose only those who were able to resolve the two stars which form the second 'star' in the tail of the Great Bear constellation. A point source of light such as a star, although it is infinitely small, forms an image with a diameter of about 11μ on the retina. This is because the optical media are not perfect and allow some scattering of the light. In practice it is possible for a person with normal vision to distinguish two points apart if they are separated by 1 mm when placed 10 m away. Two such points would be separated by 2μ on the retina. This might be surprising considering that a spot of light casts a minimum size of image of 11μ due to scatter, but such an image is not uniform, being brighter in the centre than at the periphery. In fact, the resolving power of the eye is limited by the size of the cones, which have a diameter of 1.5μ.

In the clinic the visual acuity is measured by asking the patient to read a standard set of letters, the Snellen chart. This is placed at a distance of 6 m

Fig. 16.1. "The newsprint these days isn't what it used to be...."

from the eye. The single large letter at the top of this chart is designed to be just discernible to a normally sighted person at a range of 60 m. If the patient's vision is so poor that only this and no smaller letter can be seen at 6 m, then the vision is recorded as the fraction '6/60'. The normal-sighted person who can read the chart down to the smaller letters designed to be discerned at 6 m is recorded as having a visual acuity of 6/6. The normal range of vision extends between 6/4 and 6/9.

The near visual acuity is also measured using a standard range of reading types and here care must be taken to ensure that the correct spectacles for near work are used if the patient is over the age of 45, (p. 102 Fig. 16.1). Normally the results of testing the near visual acuity are in agreement with those for measuring distance vision providing the correct spectacles are worn if needed. The visual acuity of each eye must always be measured by placing a card carefully over one eye and then transferring this to the other eye when the first eye has been tested. The visual acuity of both eyes together is usually the same or fractionally better than the vision of the better of the two eyes tested individually. In certain special circumstances the binocular vision may be worse than the vision of each eye tested separately.

A number of other tests have been developed to measure visual acuity. Infants below the reading age can be measured with surprising accuracy using the Stycar test. Here, letters of differing size are shown to the child, who is asked to point to the same letter on the card which is given to him. Up to the age of $1\frac{1}{2}$ or 2, the optokinetic drum may be used. This makes use of the phenomenon of optokinetic nystagmus produced by moving a set of vertically arranged stripes across the line of sight. When the stripes are sufficiently narrow, they are no longer visible and fail to produce any nystagmus. The eyes

are examined using a graded series of stripes. This kind of test can be used to measure visual acuity in animals other than man. The 'E' test is a way of measuring the visual acuity of illiterate patients. This is based on the Snellen type but the patient is presented with a series of letter 'E's of different sizes and orientations and is given a wooden letter 'E' to hold in the hands. He is than instructed to turn the wooden letter to correspond with the letter indicated on the chart.

The Snellen type has the great advantage of being widely used and well standardised, but it must be realised that it is a measure of something more complex than simply the function of the macula area of the retina. It involves a degree of literacy and also speech, and testing shy children or elderly patients may sometimes be misleading.

One final way of measuring the visual acuity is by assessing the ability to resolve a grating. Here, the word 'grating' refers to a row of black and white stripes where the black merges gradually into the white. Such a grating can be varied by altering either the contrast of black and white or the width of the stripes (the 'frequency'). Thus for a given individual, the threshold for contrast and frequency can be measured. This type of test has certain theoretical advantages over standard methods but it is only used for research purposes at the present time.

Measuring for Spectacles

If a patient has not been recently tested for spectacles, then not only may the measurement of visual acuity be inaccurate, but the symptoms may be due to the need for a correct pair of glasses. The measurement which determines the type of spectacles needed requires skill developed by practice and the use of the right equipment. The most obvious way to measure someone for a pair of glasses is to try the effect of different lenses and ask the patient whether the letters are seen better with one lens or another. This is known as subjective testing and by itself it is not a very accurate method because some patients' observations as to the clarity of letters may be unreliable. Furthermore, a healthy young person may see quite clearly with a wide range of lenses simply by exercising the ciliary muscle. Fortunately the refractive error of the eye can be measured by an objective method and an answer can be reached without consulting the patient. The method entails observing the rate of movement of the shadow of the iris against the red reflex from the fundus of the eye after interposing different strengths of lens. In order to make an accurate measurement of the spectacle requirement, both objective and subjective refractions are performed and the results compared.

Objective Refraction

The patient is fitted with a spectacle trial frame into which different lenses can

be slotted. In the case of young children it is usually advisable to instil a mydriatic and cycloplegic drop beforehand to eliminate focusing. The ophthalmologist then views the eye to be examined through an instrument known as a retinoscope, from a distance of about one arm's length. The red reflex can be seen and the instrument is then moved slightly so that the light projected from the retinoscope moves to and fro across the pupil. The shadow of the iris on the red reflex is then seen to move, and the direction and speed of movement depend on the refractive error of the patient. By interposing different lenses in the trial frame, the movement of the iris shadow can be 'neutralised' and the exact refractive error determined. The trial frame can accommodate both spherical and cylindrical lenses so that the amount of astigmatism can also be measured.

Subjective Refraction

Here, considerable skill is also needed because many patients become quite tense when being tested in this way and may not initially give accurate answers. Lenses both stronger and weaker than the expected requirement are placed in the trial frame and the patient is asked to read the letters of the Snellen chart and to say whether they are more or less clear. A number of supplementary tests are available which enable one to check the patients' answers.

It can be seen that the word 'refraction' refers to the total test for glasses, although the same word refers to the bending of the rays of light as they pass from one medium to another. Accurate refraction takes 10 or 15 minutes to perform, or longer in difficult cases, and it is an essential preliminary to an examination of the eye itself.

Automated Refraction

In recent years attempts have been made to develop an automated system of refraction, and instruments are now commercially available. They are, however, still expensive and not always accurate when there are opacities in the optical media. Some of these instruments are beginning to appear in ophthalmic departments.

One further way of measuring the refractive error without asking the patient any questions is by making use of the visually evoked response. This is the name given to the minute electrical changes detectable over the back of the scalp when the eyes are exposed to a repeated stimulus, usually a flashing chequerboard. When fine checks are viewed, the response can be modified by interposing different lenses. This method is of great interest but it is still not very reliable and takes time to perform.

Considering the importance of the measurement of visual acuity, it is not surprising that a number of tests have been developed for this, but the simple Snellen chart remains an essential part of any doctor's surgery. It must be remembered that this is a measure of the function of the centre of the visual field only and it is possible to have advanced loss of vision with normal visual

acuity, as is seen sometimes in patients with chronic glaucoma or retinitis pigmentosa. The assessment of the rest of the visual field has also been standardised and a number of instruments have been developed to measure it. These have already been described in Chapter 2 together with various other measurements of different aspects of vision.

17 The Inflamed Eye

In an earlier chapter we have already seen that 'the red eye' is an important sign in ophthalmology, and there are a number of reasons why the eye may become inflamed. When the exposed parts of the eye such as the conjunctiva and the cornea are the primary sites of inflammation, the cause is usually infection or trauma. Common examples are chronic conjunctivitis or a corneal foreign body. However, here we are going to consider a type of inflammation which arises deeper in the eye and primarily from the uvea. The uvea has a tendency to become inflamed for no apparent external reason and in this respect can be compared to a joint; indeed, there is a recognised association between uveitis and arthritis. In spite of the fact that the eye is open to microscopic examination, the exact cause of uveitis is usually obscure, although there is evidence to indicate a relationship with other kinds of autoimmune disease. Uveitis can be divided into anterior or posterior uveitis; anterior uveitis is the same entity as iridocyclitis, and posterior uveitis is the same as choroiditis. Apart from the uvea, the sclera and the episclera (that is, the connective tissue deep to the conjunctiva and overlying the sclera) may also be affected by similar inflammatory changes.

Anterior Uveitis

Symptoms

The patient suffering from acute anterior uveitis is usually aware that there is something seriously amiss with the eye. The vision is blurred and the eye aches and may often be extremely painful. Photophobia is usual and often pain on focusing on near objects is a feature. The age incidence is wide but a particular type of anterior uveitis is commonly seen in the early twenties, and every eye casualty officer becomes very familiar with this particular form. When the disease presents for the first time in the elderly, the underlying cause is likely to be different and age provides an important diagnostic feature. Acute anterior uveitis usually appears quite suddenly over a period of about 24 hours and

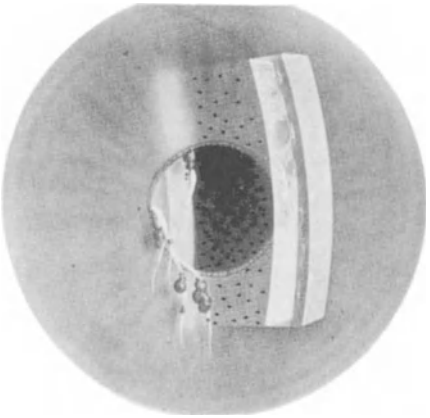

Fig. 17.1. Aqueous flare and cells in acute anterior uveitis.

then resolves on treatment in 2 or 3 weeks. A further exacerbation may occur during this period and there is a strong tendency towards recurrence after a few months or a year in the same or the other eye.

Signs

The eye is red, but of especial importance is the presence of a pink flush around the cornea (the ciliary flush) which indicates an inflammatory process either in the cornea or within the eye itself. The pupil is small because the sphincter goes into spasm. Thus the pupil of iritis is small and treatment is aimed at making it larger, whereas the pupil of acute glaucoma is large and treatment is aimed at making it smaller. Unless there is secondary glaucoma, the cornea remains bright and clear, but with a pen torch it may be possible to see that the aqueous looks turbid. That is to say, a beam of light shone through the aqueous resembles a beam of sunlight shining through a dusty room (Fig. 17.1). Normally, of course, the aqueous is crystal clear even when examined with the microscope. The presence of an occasional cell in the aqueous may be normal, especially if the pupil has been dilated for some other reason, but suspicion should be raised if more than three or four cells are seen. In fact the early diagnosis of anterior uveitis can entail very careful slit-lamp examination. It is usual to discriminate between the presence of cells in the aqueous and the presence of flare. The latter reflects a high protein content and is a feature of more long-standing disease. Because there are convection currents in the aqueous, inflammatory cells are swept down the centre of the posterior surface of the cornea and become adherent to it, often forming a triangular-shaped spread of deposits known as keratic precipitates, or 'KP' (Fig. 17.2). The microscopic appearance of the KP is determined by the type of cells. If a

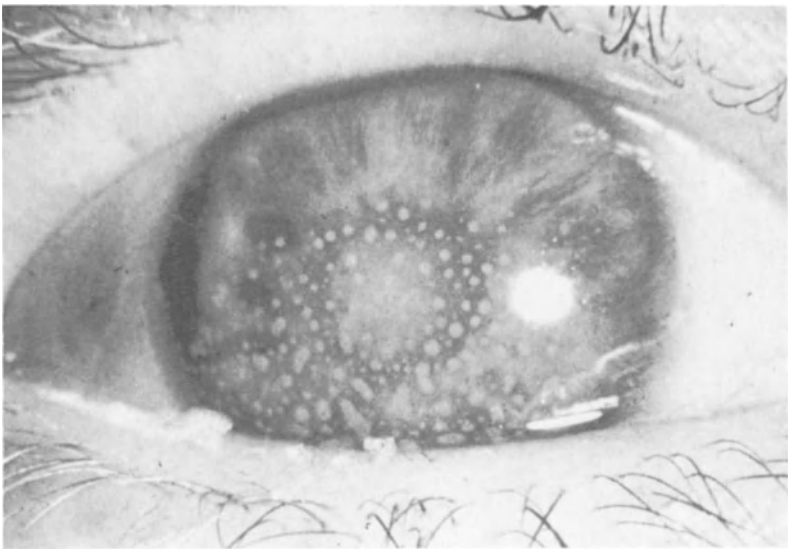

Fig. 17.2. Keratic precipitates.

granulomatous type of inflammatory reaction is taking place involving giant cells, then the KP may be large, resembling oil droplets ('mutton fat KP'). This form of KP is seen in uveitis associated with sarcoidosis and also tuberculosis and leprosy. When the inflammation is non-granulomatous, a fine dusting of the posterior surface of the cornea may be evident. KP tend to become absorbed but they may remain more permanently as pigmented spots on the endothelium. Anterior uveitis is often associated with the formation of adhesions between the posterior surface of the iris and the lens. These are called posterior synechiae and become evident when attempts are made to dilate the pupil since parts of the iris remain stuck to the pupil giving it an irregular appearance. In severe cases of anterior uveitis, pus may collect in the anterior chamber to the extent that a fluid level may be seen where the layer of pus has formed inferiorly. This is known as hypopion — literally, 'pus below' (Fig. 17.3). A hypopion is an indication of severe disease in the eye and the patient should be treated in hospital. Hypopion tends to occur in certain specific types of anterior uveitis. It is occasionally seen in elderly diabetics with inadequately treated corneal ulcers, particularly those with vascular occlusive disease. It is also seen in Behçet's disease which is a rare disorder characterised by hypopion uveitis, and ulceration of the mouth and genitalia. A hypopion is occasionally seen following cataract surgery and it appears to be a fortunately rare complication of the use of intraocular acrylic lenses.

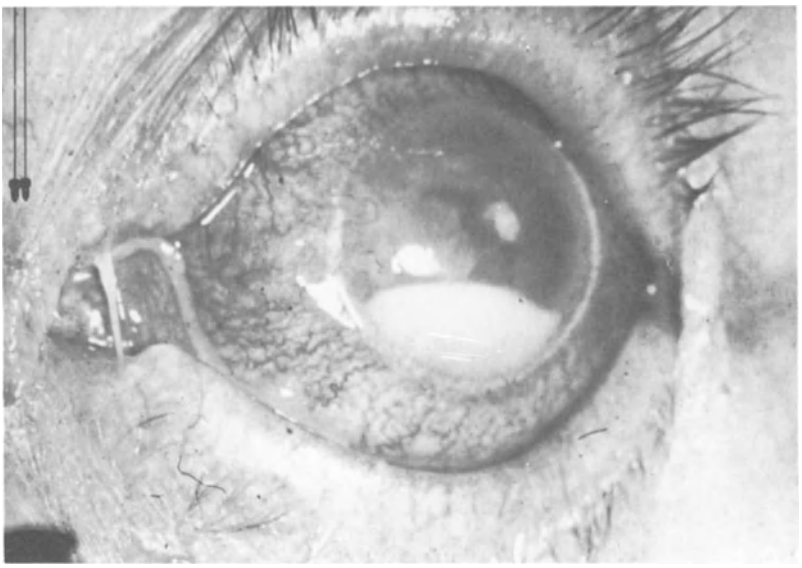

Fig. 17.3. Hypopion uveitis.

Complications

The visual prognosis of acute anterior uveitis as commonly seen in young people is usually good unless recurrences are frequent. Chronic uveitis is more prone to complications. Secondary glaucoma may cause serious problems and a careful check on the intraocular pressure must be maintained. The rise in intraocular pressure may be due to direct obstruction of the outflow of aqueous by inflammatory cells or by the presence of adhesions between the peripheral part of the iris and the posterior surface of the cornea (peripheral anterior synechiae). Sometimes, especially when treatment has been inadequate, the posterior synechiae sticking the pupil margin to the anterior surface of the lens become extensive enough to obstruct the passage of aqueous through the pupil. The iris bulges forwards giving the appearance known as iris bombé. Cataract is a further serious complication which may appear after repeated attacks of anterior uveitis. It nearly always first affects the posterior part of the lens and unfortunately interferes with the vision at an early stage.

Causes

For the majority of patients who present to eye out-patient departments with this condition, no specific cause is found. However, there are many known causative agents. The ophthalmologist is obliged to exclude at least some of these even though he knows that more often than not a negative result will be

obtained. It is necessary to explain this to patients otherwise considerable anxiety may be created by the fact that 'no cause can be found' for their complaint. When we say no cause can be found we really mean that there is no evidence of any associated systemic disease and this should be of some reassurance to the patient.

It has already been mentioned that it can be helpful to consider the age of the patient when trying to eliminate the possibility of underlying systemic disease. Uveitis is very rare in young children, but when seen, the possibility of juvenile rheumatoid arthritis must be borne in mind. In young adults sarcoidosis, gonorrhoea, Reiter's disease and ankylosing spondylitis are all recognised associations. In former years tuberculosis was very high on the list of suspected causes but this would appear to be a rare cause nowadays. Septic foci in adjacent structures, such as dental sepsis or sinusitis, have also been under suspicion. In the case of the elderly, the onset of anterior uveitis may prove to be a recurrence of previous attacks and the same underlying causes must be suspected, but here there is also the possibility of lens-induced uveitis associated with cataract and also diabetes mellitus.

Three other special types of anterior uveitis must be mentioned at this stage.

Sympathetic Ophthalmia

This is a rare but dramatic response of the uvea to trauma. The significance of the condition rests in the fact that even though the trauma has only affected one eye, the inflammatory reaction occurs in both. It may follow perforating injuries, especially when uveal tissue has become adherent to the wound edges. The injured eye remains severely inflamed and, after an interval of between 2 weeks and several years, the uninjured eye becomes affected. Careful wound toilet can probably prevent many cases, as can also removal of blind, painful and inflamed eyes within the critical 2 week period following injury.

Heterochromic Iridocyclitis

This type of anterior uveitis presents in 20 to 40 year olds and is usually unilateral. The vision becomes blurred and the iris becomes depigmented. The eye usually remains white; the inflammatory reaction is low grade and chronic and does not usually respond at all to treatment. Cataracts occur very commonly. The condition has been mimicked by denervating the sympathetic supply of the eyes in experimental animals and it seems possible that there may be a neurological cause, unrelated and distinct from other types of uveitis.

Pars Planitis

This refers to a low-grade inflammatory response which is seen in young adults. There is minimal evidence of anterior uveitis and the patient complains of floating spots in front of the vision. Inspection of the fundus reveals

vitreous opacities and careful inspection of the peripheral retina shows whitish exudates in the overlying vitreous. The condition runs a chronic course and occasionally may be complicated by secondary glaucoma. The cause is unknown.

Treatment and Management

Once the diagnosis has been made, it is usual to embark on a number of investigations, guided in part by the history and taking into account especially any previous chest or joint disease. An x-ray of chest, skull and sinuses and a blood count including measurement of the ESR are routine in most clinics, but the expense of further investigations is now often spared if the patient appears completely fit and well in other respects. The history and background of the patient may lead one to suspect the possibility of venereal disease. In the case of some infective types of anterior uveitis, the diagnosis is made before the uveitis appears because the condition occurs as a secondary event. This is the case following herpes simplex keratitis and also in patients with herpes zoster affecting the upper division of the fifth cranial nerve.

The treatment involves the administration of local steroids and mydriatic drops. When the condition is severe, a subconjunctival injection of steroid should be given and relief of symptoms may be further achieved by local heat in the form of a warm compress. Atropine is the mydriatic of first choice except in the mildest cases when homatropine or hyoscine drops may be used. Steroid drops should be administered every hour during the acute stage and then gradually tailed off over a period of a few weeks. Systemic steroids are not usually indicated and should be reserved for those cases in which the sight becomes seriously jeopardised. If any underlying systemic disease is identified, then of course this should also be treated if effective treatment is available. The proper management of anterior uveitis demands the expertise of an ophthalmic surgeon and, when the condition is affecting both eyes, the patient is best treated in hospital. A special word of warning is needed for those patients who have undergone previous intraocular surgery. For these patients what is normally a mild infective conjunctivitis may lead to intraocular infection. The development of anterior uveitis, weeks, even years, after the operation, can indicate disastrous consequences if urgent and intensive antibiotic treatment is not applied.

Posterior Uveitis

Symptoms

When the choroid, as opposed to the ciliary body and iris, becomes inflamed, the eye is not usually painful or red and the patient complains of severe blurring or loss of vision. If the focus of choroiditis remains peripheral, then

the disease may remain unnoticed, as is witnessed by the relatively frequent observation of isolated healed foci in the fundi of asymptomatic patients. Often the inflammation spreads from choroid to retina and thence to the vitreous. When this happens the vision becomes markedly blurred, even when the original focus is remote from the macula region. Choroiditis at the macula itself usually leads to permanent loss of central vision.

Signs

In its early stages, choroiditis may be seen as a grey or yellowish raised area which may be discrete or multiple and anywhere in the fundus. A cellular reaction may appear in the overlying vitreous, seen as localised misting with the ophthalmoscope, and eventually the whole vitreous may become clouded, obscuring any view of the fundus and the original site of inflammation. The patient usually presents at this stage so that the origin of the problem only becomes apparent after the inflammation has subsided. When a patch of choroiditis heals, the margins become pigmented and a white patch of bare sclera remains, the result of atrophy of the pigment epithelium and choroid. Sometimes larger choroidal vessels survive as a clearly seen network overlying the white sclera surrounded by a pigment halo. During the active stage, inspection of the vitreous with the slit lamp reveals the presence of cells and very often the anterior chamber also contains cells. Posterior uveitis comes into the differential diagnosis of a white eye with failing vision. When the vitreous becomes cloudy the condition must be distinguished from vitreous haemorrhage. The latter nearly always occurs acutely over a period of hours, whereas the cloudiness following uveitis takes a few days to develop. Examination of the vitreous with the slit lamp can reveal whether the vitreous is filled with inflammatory cells or red cells.

Causes

As in the case of anterior uveitis, it is often impossible to pinpoint any systemic cause and the condition seems to be confined to the eye. However, a number of systemic associations have been recognised and often are related to specific types of posterior uveitis.

Toxoplasmosis

Toxoplasma gondii is a parasite, a protozoan carried by cats. Man and other intermediate hosts may be infected. In the adult the infection is usually mild but in the case of infected pregnant mothers the child in utero may be infected by the more severe congenital form of the disease. The organism enters the brain as well as the eyes and may cause mental deficiency and epilepsy. A characteristic type of calcification is seen on the skull x-ray. In the eye a focal type of choroiditis often affects both eyes and this is usually at the posterior

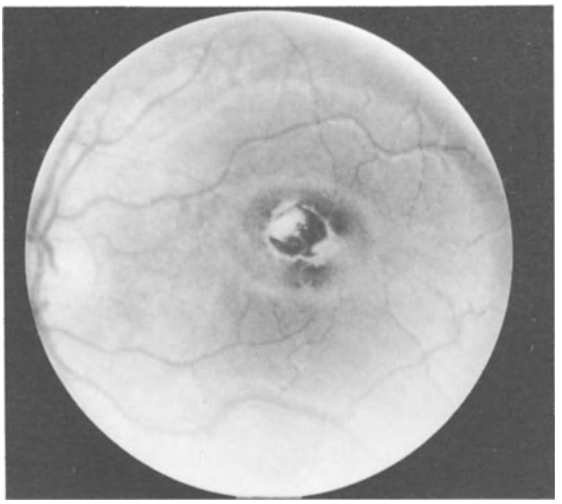

Fig. 17.4. Healed choroiditis.

pole in the macula region (Fig. 17.4). Histologically the *Toxoplasma* is found in the eye lesions. The diagnosis may be confirmed by sending some blood for the toxoplasmosis dye test. This test must be interpreted carefully because a high proportion of the population becomes infected at some point and the positive results increase with age, even in those with no clinical evidence of infection. For this reason the diagnosis may be less easy in acquired toxoplasmosis where evidence of systemic involvement may be slight or absent. It has been shown that there is a higher incidence of positive dye tests in patients with posterior uveitis than in the normal population, but in an individual case it is often necessary to demonstrate a changing titre in order to confirm the diagnosis.

Unfortunately the treatment of toxoplasmosis is disappointing. A combination of pyrimethamine and sulphadiazine has been recommended but its value in the management of the uveitis is doubtful and such treatment may cause a serious fall in the white cell count. It is generally accepted that systemic steroids have some beneficial effect and may help to clear the vitreous more rapidly, but this treatment may also be disappointing. In fact the majority of cases resolve spontaneously, leaving more or less chorioretinal scarring at the macula region. Recurrences are fairly common and the fresh choroidal inflammation tends to arise on the edge of a previous scar.

Toxocara canis

This nematode has been found in the enucleated eyes of young patients with a

severe type of chorioretinitis. The vitreous tends to be filled with a white mass of inflammatory cells so that the presence of a tumour may be suspected (e.g. retinoblastoma). Endophthalmitis tends to develop in these cases and the sight of one eye may be completely lost. During the acute stage the peripheral blood may show an eosinophilia. The disease is thought to be transferred from dogs by faecal contamination. Treatment is unsatisfactory.

Tuberculosis

In former years this was considered to be a common cause of posterior uveitis, clinicians having been impressed by the number of patients with a previous history of TB. The relationship seems less likely now that tuberculosis has been almost eliminated from the population. On the other hand, choroidal tubercles are a well-described entity: these raised yellowish granulomatous foci were used as a diagnostic feature of miliary tuberculosis. They are only seen in extremely ill patients and the yellowish tubercles become pigmented as they heal.

Sarcoidosis

The eye is very frequently involved in sarcoidosis, but involvement usually takes the form of an anterior uveitis. The choroiditis is more often peripheral and accompanied by inflammatory changes in the retinal veins. Sheathing of the veins may be seen and the vision may be impaired by macula oedema. When the diagnosis is suspected, the conjunctiva and skin should be searched for possible nodules which may be biopsied, and an x-ray of the chest may reveal enlargement of the hilar lymph nodes. The management of the ophthalmological problem may involve treatment with local and systemic steroids but the opinion of a physician specialising in sarcoidosis is essential and should be sought before embarking on treatment.

Histoplasmosis

A certain type of cystic haemorrhagic macula lesion combined with discrete foci of peripheral uveitis has been associated with infection by the fungus *Histoplasma capsulatum.* Infection with this organism occurs throughout the world but is more common in the Mississippi Valley. A severe pneumonitis may occur but most cases are asymptomatic. The evidence for infection in a particular case tends to be necessarily circumstantial — hence the expression 'presumed ocular histoplasmosis'.

Syphilis

Diffuse chorioretinal scarring is a feature of congenital syphilis. The active

stage of inflammation is not usually seen and probably occurs at an early age. One should take note of other possible features such as deafness and corneal scarring from previous interstitial keratitis. The scattered pigmentation in the fundus may suggest an inherited retinal degeneration but a careful family history together with electrodiagnostic testing of the eyes usually enables one to distinguish the two conditions. It is also important to carry out serological testing. The *Treponema pallidum* immobilisation test (TPI) and the fluorescent treponemal antibody test (FTABS) are the most sensitive and specific. Acquired syphilis may also cause chorioretinitis but this, too, is extremely rare.

Other Causes

A very wide variety of infective agents have been shown to cause posterior uveitis on rare occasions. Leprosy and the coxsackie group of viruses are two examples chosen from many. Sympathetic ophthalmia has already been mentioned as a specific form of uveitis following injury. An especially rare but intriguing form of uveitis is known as the Vogt–Koyanagi–Harada syndrome in which is seen the combination of vitiligo, poliosis, meningoencephalitis, uveitis and exudative retinal detachments. The syndrome of oral and genital ulceration combined with recurrent uveitis was originally described by Behçet but it is now recognised to be a more widespread disease involving inflammation of the blood vessels, joints and gastrointestinal tract.

The Role of Autoimmunity in Uveitis

Although it has been long recognised that bacterial and viral infection may account for some cases of uveitis, it has also been recognised that the majority of cases fail to show any evidence of this. Furthermore, in many instances the eye disease may be associated with known autoimmune disease elsewhere in the body. There are several different ways in which the uvea might be expected to become the focus of an antigen–antibody reaction; for example, a foreign agent such as a virus might reside in the uvea and cause an antibody response which coincidentally involves uveal tissue or, on the other hand, a foreign agent might react with specific marker on the cell membrane to produce a new active antigen. It is now recognised that patients who inherit certain of the human leucocyte antigens (HLA) are more susceptible to particular types of uveitis, for example the uveitis seen in ankylosing spondylitis and Reiter's disease. It has been suggested that HLA may act as the specific marker in these cases. A further way in which the uvea might become the centre of an immune response concerns the question of self-recognition. It now appears that there is a mechanism in the body which normally prevents antibodies in the serum from acting against our own tissues. This active suppression is maintained by a population of thymus-derived lymphocytes (T lymphocytes) known as

T-suppressor cells. There is evidence to suggest that sympathetic ophthalmitis might arise from inhibition of the T-suppressor cells after uveal antigens have been introduced into the bloodstream. Patients with juvenile rheumatoid arthritis occasionally develop uveitis, whereas rheumatoid disease in adults is more commonly associated with the dry eye syndrome and episcleritis.

Management

The great increase in interest in immunological diseases in recent years, which has accompanied advances in tissue grafting and cancer research, has led to attempts to treat uveitis with means other than steroids. Salicylates and immunosuppressive agents are now sometimes used to supplement or replace steroids in difficult cases, but the value of such treatment is still not clear.

If posterior uveitis is not due to any recognisable infective cause, it is usual to start treatment with systemic steroids if the visual acuity becomes significantly impaired or if the lesion is close to the macula. Systemic steroids are best administered on an in-patient basis, especially as the sight is jeopardised. This has the added advantage of allowing a more detailed pretreatment examination, and often the opinion of a general physician may be valuable at this stage. Secondary glaucoma may also need to be treated and salicylates and immunosuppressive agents may be administered to resistant cases. When posterior uveitis keeps recurring at the edge of previous healed foci, laser coagulation has been used in patients suspected of being infected with toxoplasmosis. The rationale of this treatment is to destroy any remaining encysted organisms.

Endophthalmitis and Panophthalmitis

When inflammatory changes in the posterior uvea extend into the vitreous and there is an extensive involvement of the centre of the globe, the patient is said to have endophthalmitis. Further extension of the inflammation into the anterior segment of the eye and into the sclera leads to panophthalmitis. Endophthalmitis is one of the feared results of infection after injury or surgery but it may prove reversible with intensive antibiotic treatment. Once the stage of panophthalmitis has been reached, the sight is usually lost permanently and the eye begins to shrink.

Episcleritis and Scleritis

Both these conditions form part of the differential diagnosis of the red eye. The episclera is the connective tissue underlying the conjunctiva and it may

become selectively inflamed, either diffusely or in localised nodules. Close inspection of the eyes shows that the inflammation is deeper than the conjunctiva and there is a notable absence of any discharge. The eye is red and painful. Episcleritis is seen from time to time in the casualty department and the patient may be otherwise perfectly fit and well. Such cases tend to recur and some develop signs of rheumatoid arthritis. The condition responds to local steroids, but aspirin may also prove effective. Scleritis is less common and more closely linked with rheumatoid arthritis. In severe cases the sclera may become eroded with prolapse of uveal tissue. Both episcleritis and scleritis are seen in other forms of collagen disease and notably in cases of gout.

18 The Ageing Eye

Although the eye undergoes a number of well-defined changes with age, the distinction between these involutionary changes and disease is not always clear cut. For the elderly patient it is often reassuring to know that the problem is part of a 'normal' process rather than the result of a specific illness and perhaps sometimes an artificial demarcation is drawn for the benefit of the patient.

The increase in number of elderly people presents problems in ophthalmology. A high proportion of elderly people instil drops into their eyes, either prescribed for them or as self-medication, and it is important that adequate advice is received. Advising the elderly is often time consuming and may entail speaking to a younger relative or neighbour, but an adequate explanation of the disease or problems will avoid anxiety and probably the need for further subsequent unnecessary consultation.

The three commonest diseases of the elderly eye are cataract, glaucoma and senile macula degeneration. The first can be cured, the second arrested or prevented, while the third tends to run a progressive course. Attempts to measure the incidence of these problems have produced a wide range of figures, but out of a population of elderly persons complaining of impaired vision about 30% turn out to have a cataract and a similar number to have senile macula degeneration, whereas 5% or less have chronic open angle glaucoma. Although there is an unexpectedly high incidence of cataract in patients with chronic simple glaucoma, the association of macula degeneration with cataract or glaucoma is more random.

Changes in the Eyes with Age

The External Eye

The eyelids tend to lose their elasticity and become less firmly opposed to the globe. The upper and lower lid margins become progressively lower so that whereas in the infant the upper lid may ride level with or slightly above the

corneal margin, in an elderly subject the upper lid may cross the upper margin of the pupil and an area of white may be seen between the lower margin of the cornea and the lower lid. Some limitation of the ocular movements is accepted as normal in the elderly, especially limitation of upward gaze. The conjunctiva tends to become more lax and a thin fold of conjunctiva may be trapped between the lids when blinking if this becomes excessive. In some elderly patients there is loss of connective tissue around the lacrimal puncta so that the opening is seen elevated slightly from the rest of the lid. Degenerative plaques are seen on the bulbar conjunctiva in the exposed region and the conjunctiva is especially prone to chronic inflammation.

The Globe

Arcus senilis is the name given to the circular white infiltrate seen around the margin of the cornea. The lens gradually loses its plasticity throughout life and this results in a progressive reduction in the focusing power of the eye. A child may be able to observe details of an object held 5 cm from the eye, but as a result of the hardening of the lens the nearest point at which an object can be kept in focus gradually recedes. This progressive degeneration tends to pass unnoticed until the eye is no longer able to focus to within the normal reading distance. This usually occurs at the age of 45 if the eyes are otherwise normal, and the phenomenon is called *presbyopia.* Some degree of opacity of the lens fibres is very common in old age and only when this becomes more extensive is the term 'cataract' used. The pupil becomes smaller with age and does not show the wide range of adjustment to illumination seen in younger people. The vitreous shows an increase in small opacities visible to the subject as 'vitreous floaters'. A more dramatic degenerative change which occurs in a high proportion of normal individuals in the 60–70 age group is detachment of the vitreous. The formed part of the vitreous separates from the retina, usually above at first, leaving a fluid-filled gap between the retina and posterior vitreous face. Movement of the vitreous face may cause troublesome symptoms but often a vitreous detachment goes unnoticed and is an incidental finding on examination of the eye. The important association between sudden vitreous detachment and subsequent retinal detachment has already been discussed in Chapter 12. The appearance of the fundus also shows gradual changes; the retinal arterioles become straighter and more narrow, as also do the venules. Colloid bodies are more commonly seen due to degenerative changes in Bruch's membrane and the pigment epithelium, and peripheral chorioretinal degeneration is more evident. The young retina is more shiny than the old retina and in the elderly the normal light reflex is less marked. The optic disc tends to become paler and a degree of optic atrophy is accepted by many clinicians as a senile change unrelated to disease.

Eye Disease in the Elderly

A recent survey in the United States has shown that the incidence of cataract in the 45–64-year-old population is 5.6% for males and 2.1% for females. The incidence is slightly higher in the negro population, and rises to 21.6% for males and 26.8% for females in the 65–75-year-old population. In the same age group the incidence of senile macula degeneration is 9.6% for males and 6.9% for females. Both these conditions are therefore very common and they demand time and medical expertise, both at the primary care level and in hospital.

Senile Macula Degeneration

Older patients with senile macula degeneration complain of blurring of their vision and inability to read. Younger or more observant patients notice that straight edges may look kinked. Usually one eye is considerably more affected than the other and, because the degenerative process is limited to the macula, the peripheral field remains unaffected and the patient can walk around quite normally. Difficulty in recognising faces or in seeing bus numbers is also a common complaint. About a third of the patients give a family history of similar problems.

In the early stages of the condition, inspection of the fundus shows spots of pigment in the macula and perimacula region. Drusen are also often seen (Fig. 18.1). These are small round yellowish spots often scattered over the posterior pole. Unfortunately the word 'drusen' is used rather loosely in ophthalmology to refer to two or three types of swelling seen in the fundus. It is used to

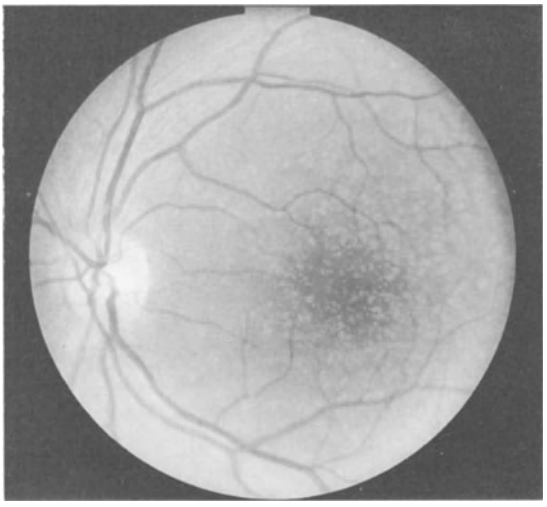

Fig. 18.1. Colloid bodies at the posterior pole of the fundus.

describe the very rare mulberry-like tumours seen around the optic nerve head in tuberose sclerosis and it is also used when referring to the multiple shiny excrescences seen on the optic disc as a congenital abnormality. Drusen seen at the posterior pole of the eye as a senile change are also known as 'colloid bodies' and perhaps this term is preferable. Under the microscope, colloid bodies are seen as a degenerative change in Bruch's membrane. In the early stages of senile macula degeneration, breaks in Bruch's membrane are also seen and in recent years it has been observed that a fan of new vessels may appear in the choroid in the perimacula region. The growth of these new vessels seems to be important because they invade the breaks in Bruch's membrane. Serous or haemorrhagic exudate tends to occur and this may be either under the pigment epithelium or subretinal. A sudden loss of central vision may be experienced as the result of such an episode.

Management

The most important aspect of senile macula degeneration is the possibility of treatment. Controlled trials of the effect of laser photocoagulation of the choroidal new vessels have shown that the progress of the condition can be significantly delayed. It is important that those cases that are likely to benefit from treatment are first identified. At the present time this entails photography of the optic fundus and fluorescein angiography. Very often patients present at the stage when new vessels have already advanced across the macula region or where the macula has already been permanently damaged by haemorrhage or exudate, making effective treatment impossible. Apart from photocoagulation, there are other practical measures which can be taken in the management of these patients. Telescopic lenses may be needed for reading or watching television and consideration should be given to the question of blind registration. It is important to explain the nature of the condition and prognosis to the patient. This can alleviate considerable anxiety and fear of total blindness and help the patient come to terms with the problem. In most cases one eye is involved first, the other following suit after 2 or 3 years. The vision, as measured on the Snellen chart, progressively deteriorates to less than 6/60, but the peripheral field remains unaffected so the patient is able to find his or her way about, albeit with some difficulty.

Cataract

This common condition in the elderly eye has already been considered, but it is important that every physician can identify and assess the density of a cataract in relation to the patient's vision. The physician must realise the potential of cataract surgery in the restoration of vision. The contraindications for cataract surgery are few and even in extreme old age the patient may benefit. Surgery may be delayed if the patient has only one eye or if there is some other pathology in the eye which is likely to affect the prognosis. The need for someone to assist the patient in the instillation of eyedrops and the domestic

chores during the postoperative period is also an important factor. About one-third of the population aged over 70 years suffer from cataract, but the quoted figures vary according to the diagnostic criteria. If an elderly person has an opaque lens which obscures any view of the fundus with the ophthalmoscope and the pupil reacts briskly, then he or she is likely to do well after surgery. It is useful to remember that the reading vision is usually fairly well preserved even when the cataract is quite dense, and if the patient is unable to read, there may be coincidental senile macula degeneration.

Glaucoma

The various types of glaucoma have also been considered already, and it should now be realised that glaucoma is simply the manifestation of a group of diseases, each of which has a different prognosis and treatment. Chronic simple or open angle glaucoma is the important kind in the elderly because it often remains undiagnosed. The physician can play a vital part in the screening of this disease by becoming familiar with the nature of glaucomatous cupping of the optic disc. About 1% of the population over the age of 55 are thought to suffer from chronic simple glaucoma and the figure may rise to as high as 30% in those over 75. In most instances the treatment is very simple but requires the cooperation and understanding of the patient. The treatment is preventative rather than curative. Chronic simple glaucoma is best managed in the glaucoma clinic of an eye unit on a long-term basis. By this means the visual fields and intraocular pressure can be accurately monitored and the treatment adjusted as required.

Deformities of the Eyelids

Both entropion and ectropion are common in the elderly, and a complaint of soreness and irritation in the eyes should always prompt a careful inspection of the configuration of the eyelids. Entropion is revealed by pressing the finger down on the lower lid so that the inverted lid becomes everted again to reveal the lash line. Sometimes entropion may be intermittent and not present at the time of examination, but usually, even under these circumstances, there is a telltale slight inversion of the lid which is made apparent by comparing the two sides. Ectropion is nearly always an obvious deformity due to the easy visibility of the reddened and everted conjunctiva, but slight degrees of ectropion may be less obvious. The lower punctum alone may be slightly everted, causing a watering eye, and the symptoms may be relieved by applying retropunctal cautery to the conjunctiva. Both ectropion and entropion respond very well to lid surgery and there is no reason why a geriatric patient should put up with the continued discomfort and irritation when a complete cure is readily available. These lid deformities may recur sometimes and require further lid surgery, but careful surgery in the first instance should largely prevent this.

Temporal Arteritis

This condition, seen only in the elderly, may rapidly cause total blindness unless it is treated in time. The disease is more common than was originally supposed but it is very rare under the age of 50. Medium-sized vessels, including the temporal arteries, become inflamed and the thickening of the vessel wall leads to occlusion of the lumen. Histologically the inflammatory changes are characterised by the presence of foreign body giant cells and the thickening of the vessel wall is at the expense of the inner layers so that the total breadth of the vessel may not be altered. The inflammatory changes tend to be segmental so that a single biopsy of a small segment of the temporal artery does not always reveal the diagnosis.

Patients with temporal arteritis usually present in the eye department with blurring of vision or unilateral loss of vision. Typically these symptoms are accompanied by headache and tenderness of the scalp so that brushing the hair may be painful. Often there is low-grade fever and there may be aches and pains in the muscles and joints as well as other evidence of ischaemia in the brain and heart. The blurring of vision is due to ischaemia of the optic nerve head. The diagnosis rests largely on finding a raised ESR and a positive temporal artery biopsy in an elderly patient with these symptoms. Palpation of the temporal arteries reveals tenderness and sometimes thickening and the absence of pulsation is a useful sign. Curiously, the pain of temporal arteritis is sometimes relieved by biopsy, perhaps because inflamed nerves around the artery are divided at the same time.

Inspection of the fundus in a patient with visual symptoms shows pallor and often swelling of the optic nerve head and narrowing of the retinal arterioles. Once the disease is suspected, a biopsy is essential and this should be done without delay before treatment is started. The symptoms disappear rapidly after administering systemic steroids, initially in a high dose, for example prednisolone 120 mg per day, and the dosage is then reduced rapidly according to the level of the ESR. Once the ESR is down to normal levels, a maintenance dose of systemic steroids is continued, if necessary for several months.

Temporal arteritis is recognised as a self-limiting condition which lasts for 2 or 3 months in most cases. About a quarter of all patients are liable to become blind and in some instances extraocular muscle palsies causing diplopia and ptosis may confuse the diagnosis. One might summarise the disease by saying it causes headache in patients aged over 70 with an ESR over 70, who require treatment with over 70 mg of prednisolone.

Stroke

Patients who complain of visual symptoms after a stroke quite often have an associated homonymous hemianopia, and the association between hemiplegia and homonymous hemianopia should always be borne in mind. A simple confrontation field test may be all that is required to confirm this in a patient with poor vision and normal fundi following a hemiplegic episode. The

vertical line of demarcation between blind and seeing areas is very well defined and may cut through the point of fixation. Fortunately the central 2° or 3° of the visual field are often spared. When there is so-called macula sparing, the visual acuity as measured by the Snellen chart may be normal. Patients tend to complain of difficulty in reading if the right homonymous field is affected rather than the left, and although they may be able to read individual words, they have great difficulty in following the line of print. Thus a patient with a right hemiplegia and a right homonymous hemianopia may have normal fundi and a visual acuity of 6/6 and yet be unable to read the newspaper. The picture may be further complicated by true dyslexia and the patient may admit to being able to see the paper and yet be unable to make any sense of it. Dyslexia may be suspected if other higher functions such as speech have been affected by the stroke. One of the features of a homonymous hemianopic defect in the visual field is the patient's complete lack of insight into the problem, so that even a doctor may fail to notice it in himself. It is unusual for a homonymous hemianopia to show any signs of recovery, but once patients understand the nature of the handicap they may learn to adapt to it to a surprising degree.

19 The Child's Eye

Examination

How Normal Features Differ From Those in the Adult

At birth the eye is large, reaching adult size at about the age of 2. One might expect that before the eye reaches its adult size, it would be long sighted, being to small to allow parallel rays of light to be brought to a focus on the retina. In actual fact the immature lens is more globular and thus compensates for this by its greater converging power. More than three-quarters of children aged under 4 years are slightly hypermetropic. The slight change of refractive error which occurs as they grow compares with the more dramatic change in axial length from 18 mm at birth to 24 mm in the adult. The slight degree of hypermetropia seen in childhood tends to disappear in adolescence. Myopia is uncommon in infancy and tends to appear between the ages of 6 and 9 years and then gradually increases over subsequent years. The rate of increase of myopia is maximal during the growing years and this may often be a cause of parental concern.

The iris of the newborn infant has a slate-grey colour due to the absence of stromal pigmentation. The normal adult coloration does not develop fully until after the 1st year. The pupil reacts to light at birth but it may not dilate very effectively in response to mydriatic drops. The fundus tends to look grey and the optic disc somewhat pale, deceiving the uninitiated into thinking that it is atrophic. The foveal light reflex is absent or ill-defined until the infant is 4–6 months old. By the age of 2 the fundus has taken on its adult appearance.

The ability to fix a light is seen at birth, although much of the time the eyes may seem to wander aimlessly. The eye movements may also be poorly coordinated during the first 2 or 3 months, giving rise to concern about the possibility of squint. By 6 months the movements of the eyes should be well coordinated, and referral to an ophthalmologist is essential if a squint is suspected.

Once children learn to identify letters, at the age of 4 or 5, the Snellen chart can be employed to measure visual acuity, which by this age is normally 6/6 or

6/9 or not far from the adult level. The Stycar test can be used for 3–4 year olds or sometimes younger children and a similar level of visual acuity is seen as soon as the child is able to cooperate with the test conditions. Stycar results tend to be slightly better than Snellen results when measured in the same child, perhaps because the Stycar test involves seeing a single letter rather than a line.

Method of Examination

The general examination of the eye has already been considered, but in the case of the child certain aspects require special consideration. Before the age of 3 or 4 years, it may not be possible to obtain an accurate measure of the visual acuity, but certain other tests which attempt to measure fixation are available. The rolling ball test measures the ability of the child to follow the movement of a series of white balls graded into different sizes. Another test makes use of optokinetic nystagmus which can be induced by making the child face moving vertical stripes on a rotating drum. The size of the stripes is then reduced until no movement of the eyes is observed. In practice a careful examination of the child's ability to fix a light, and especially the speed of fixation, is helpful. The behaviour of the child may also be a helpful guide, such as, for example, the response to a smile or the recognition of a parent. Sometimes grossly impaired vision in infancy is overlooked or interpreted as a psychiatric problem, but such an error can usually be avoided by careful ophthalmological examination. The reaction of the pupils is also an essential part of any visual assessment.

One of the difficulties in examining children is that they are rarely still for more than a few seconds at a time and any attempts at restraint usually make matters worse. Before starting the examination it is useful to gain the child's confidence by talking about things that might interest him or her, not directly but in conversation with the parent. In fact it is sometimes better to ignore the rather anxious child deliberately during the first few minutes of the interview. Once the young patient has summed you up, hopefully in a favourable light, then a gentle approach in a quiet room is essential for best cooperation. The cover test can only be performed well under such conditions and once this has been done the pupils and anterior segments can be examined, first with a hand lens but if possible with the slit-lamp microscope. Fundus examination and measurement of any refractive error demand dilatation of the pupils and paralysis of accommodation. Cyclopentolate 1% or tropicamide 1% are both used in drop form for this purpose. The indirect ophthalmoscope is a useful tool when examining the neonatal fundus, the wide field of view being an advantage in these circumstances. If the infant is asleep in his mother's arms, this may be an advantage since it is often a simple matter to raise one eyelid and peer in without waking up the patient. In the case of children between the ages of 3 and 6 years, fundus examination can be more easily achieved by sitting down and asking the patient to stand and look at some spot or crack on the wall while the optic disc is located.

On some occasions the child has become too excited or anxious to allow a proper examination and here one may have to decide whether it is reasonable

to postpone the examination for a week or whether the matter seems urgent enough to warrant proceeding with an examination under anaesthesia. A common casualty situation occurs when a child is brought in in distress with a suspected corneal foreign body or perhaps a perforating injury. Here it is simplest to wrap the patient in a blanket so as to restrain both arms and legs and then examine the cornea by retracting the lids with retractors. Particular care must be taken when examining an eye with a suspected perforating injury in view of the risk of causing prolapse of the contents of the globe. Any ophthalmological examination demands placing one's head close to that of the patient and this can alarm a child unless it is done sufficiently slowly and with tact. It is sometimes helpful to make the child listen to a small noise made with the tongue or ophthalmoscope to ensure at least temporary stillness.

Screening of Children's Eyes

It is likely that in future more and more work of this kind will be done on younger and younger children. In an ideal world all children's eyes would be examined at birth and again at 6 months to exclude congenital abnormalities and refractive errors. This is rarely achieved, although most children in Britain are examined by a non-specialist at these points. Most children are also screened routinely in school at the age of 6, and any with suspected poor vision are referred for more detailed examination. A further examination is often conducted at the age of 9 or 10 and again in the early teens. The commonest defect to be found is, of course, simple refractive error, that is, the need for spectacles. This ophthalmological screening is usually performed by a nurse and involves measurement of visual acuity by the Snellen chart or other method suited to the age of the child. If the screener could also check for family history of eye problems (e.g. amblyopia of disuse or retinoblastoma), this would be a great advantage, but the family history may not be available in the absence of the parent. When there is a difference in the visual acuity of each eye, the screener should suspect the possibility of a treatable medical problem rather than just a refractive error. A test of colour vision should also be included in the screening programme for older children and this is best done using the Ishihara plates. It is worth remembering that colour blindness affects 8% of men and 0.4% of women and it may have important implications on the choice of a job. It is also equally important to realise that colour blindness may vary considerably in degree and may often be so mild as to cause only minimal inconvenience to the sufferer. For this reason, true occupational tests of colour vision may be of more value than the Ishihara plates in individual cases.

Congenital Eye Defects

Lacrimal Obstruction

The watering of one or both eyes soon after birth is a common problem. The

obstruction is normally at the lower end of the nasolacrimal duct where a congenital plug of tissue remains. Infection should be prevented by the use of local antibiotic drops and the mother should be taught how to massage the tear sac. This manoeuvre causes mucopurulent material to be expressed from the lower punctum and can be used as a diagnostic test. If carried out regularly, this helps to relieve the obstruction. If spontaneous relief of the obstruction does not occur by the age of 6 months, probing and syringing of the lacrimal passageway under general anaesthesia are usually curative. It is important to remember that a watering eye may be due to excessive production of tears as well as inadequate drainage, and in a child, a corneal foreign body or even congenital glaucoma may be mistaken for lacrimal obstruction by the unwary.

Epicanthus

This relatively minor defect at the medial canthus is formed by a bridge of skin running vertically. The defect is seen normally in some oriental races. In Europeans it usually disappears as the bridge of the nose develops, but its importance lies in the fact that it may give the misleading impression that a squint is present.

Ptosis

Congenital ptosis may be unilateral or bilateral and sometimes shows a dominant inheritance pattern. When the ptosis is associated with other lid deformities, surgery should probably be avoided if the drooping lids are not causing any significant head tilt.

Structural Abnormalities of the Globe

There are a large number of rare and curious abnormalities of the globe to be seen. In anophthalmos the globe of the eyes may be completely absent or rudimentary on one or both sides. Sometimes the eye is excessively small (microphthalmos). Aniridia (congenital absence of the iris) is inherited as a dominant trait and may be associated with congenital glaucoma. The lens on one or both sides may be subluxated or dislocated from birth. This may be suspected if the iris is seen to be tremulous (iridodonesis). This strange wobbling movement of the iris may be seen after cataract surgery. If iridodonesis is evident after an injury to the eye, it indicates traumatic subluxation or dislocation of the lens. Congenital subluxation of the lenses may occur as a single congenital abnormality but it is also seen in Marfan's syndrome (congenital heart disease, arachnodactyly, high arched palate, subluxated lenses, etc.) and homocysteinuria. Congenital glaucoma has already been discussed; this may also be inherited in a dominant manner and is the result of persistent embryonic tissue in the angle of the anterior chamber. When the intraocular pressure is raised in early infancy, the eye becomes enlarged producing buphthalmos ('bull's eye').

Congenital Cataract

The lens may be partially or completely opaque at birth. Congenital cataract is often inherited and may be seen appearing in a dominant manner in families together with a number of other congenital abnormalities elsewhere in the body. The condition may also be acquired in utero, the most well known example of this being the cataract due to rubella infection during the first trimester of pregnancy: remember the triad of congenital heart disease, cataract and deafness after rubella. Minor degrees of congenital cataract are sometimes seen as an incidental finding in an otherwise normal and symptomless eye. The nature of the cataract usually helps with the diagnosis. It may be recalled that the lens fibres in the eye are being layed down from the outside of the lens throughout life. If the cataract appeared in utero, then quite often only the central part of the lens is opaque. There are a number of varieties and shapes and sizes of congenital cataract but most can easily be missed unless the lens is viewed against the background of the red reflex; one does not necessarily expect to see a white area in the pupil. The behaviour of a young infant does not always give such an obvious clue to congenitally defective vision as one might imagine, especially when the child suffers from other defects such as mental deficiency. Many children seem to cope very well with what may seem to be quite dense cataracts, even when they reach school age, and surgery should be avoided unless it is clear that the school performance is going to be significantly jeopardised. If dense cataracts are present at birth, surgery should be carried out as soon as the neonatal period is passed. If possible, contact lenses should also be fitted at this stage. Unfortunately, one of the eyes in patients with congenital cataracts tends to become severely amblyopic, even after early surgery. The surgery of congenital cataracts is highly effective in restoring the vision to a blind or potentially blind child. A high proportion of operated patients develop retinal detachments in middle age, although it is possible that modern techniques have reduced the risk. Congenital cataracts are usually removed by aspirating the contents of the lens, leaving the posterior capsule intact. The posterior capsule may subsequently become opaque, necessitating one or more 'needling operations' later on in childhood. When a congenital cataract appears on one side only, surgery may not be indicated in view of the high risk of amblyopia of disuse which tends to prevent a useful surgical result.

Coloboma

This term refers to the results of the failure of fusion of the foetal cleft of the optic cup. Coloboma of the iris is seen as a notch in the pupil going down and out or down and in which makes the pupil into a keyhole shape. The defect may be limited to the iris and as such slight and insignificant, or it may be associated with a similar fusional defect of the choroid. Inspection of the fundus reveals an oval white area extending inferiorly from the optic disc. Of course, if the macula area is involved, the vision may be seriously affected. Often these patients have one good eye with minimal involvement and one

weak eye with an extensive choroidal coloboma. Clefts are also seen in the eyelids as a rare congenital abnormality and these are also known as 'colobomata' although they are probably not true fusional defects.

Congenital Nystagmus

Children with congenital nystagmus are usually brought to the department because their parents have noticed that their eyes seem to be continuously wobbling about. Such abnormal and persistent eye movements may simply occur because the child cannot see (sensory nystagmus) or they may be due to an abnormality of the normal control of eye movements (motor nystagmus). It is important to distinguish congenital nystagmus from acquired nystagmus due to a space-occupying intracranial lesion.

Sensory Congenital Nystagmus

The roving eye movements are described as pendular, the eyes tending to swing from side to side. Examination of the eyes reveals one of the various underlying causes: congenital cataract, albinism, aniridia, optic atrophy, or any other cause of impaired vision in both eyes. A special kind of inherited retinal degeneration known as Leber's amaurosis may present as congenital nystagmus. The condition resembles retinitis pigmentosa but occurs at a particularly young age, leading to near blindness at school age. Patients with congenital nystagmus usually need to be examined under general anaesthesia, and electroretinography should be performed at the same time.

Motor Congenital Nystagmus

The exact cause of this type of nystagmus is usually never ascertained but a proportion of cases show recessive inheritance. Other abnormalities may be present, such as mental deficiency, but many children are otherwise entirely normal. The nystagmus tends to be jerky, with the fast phase in the direction of gaze to right or left. The distance vision is usually impaired to the extent that the patient may never be able to see a car number plate at 23 m. The near vision, on the other hand, is usually good, enabling many patients with this problem to graduate through university.

Spasmus Nutans

This term refers to a type of pendular nystagmus which is present shortly after birth and resolves spontaneously after 1 or 2 years. Like other forms of congenital nystagmus, it may be associated with head nodding.

Albinism

Albinos have pale-pink skin and white hair, eyebrows and eyelashes. They often have congenital nystagmus. The optic fundus appears pale and the choroidal vasculature is easily seen. The iris has a grey-blue colour but the red reflex can be seen through it giving the iris a red glow. Albinism is inherited in a recessive manner and may be partial or complete, or it may be limited to the eye in the condition known as ocular albinism. Albinos need strong glasses for their refractive error, which is usually myopic astigmatism. Dark glasses are also usually required because of photophobia. Tinted contact lenses may sometimes be helpful.

Other Rare Congenital Disorders

A large number of rare and curious congenital disorders can affect the eye and orbits. Gargoylism (Hurler's disease) is due to infiltration of the tissues by mucopolysaccharides and a characteristic speckling of the cornea may be seen with the slit lamp (Fig. 19.1). In Marfan's syndrome dislocation of the lens is associated with cardiac and skeletal abnormalities. Osteogenesis imperfecta is a dominantly inherited weakness of the bones in which multiple fractures are associated with a blueish colour of the sclera. The sclera is thin in these patients allowing the underlying pigment to show through. Crouzon's disease is one of several examples of failure of the normal development of the bones of the skull. In this particular example the eyes are set further apart than normal

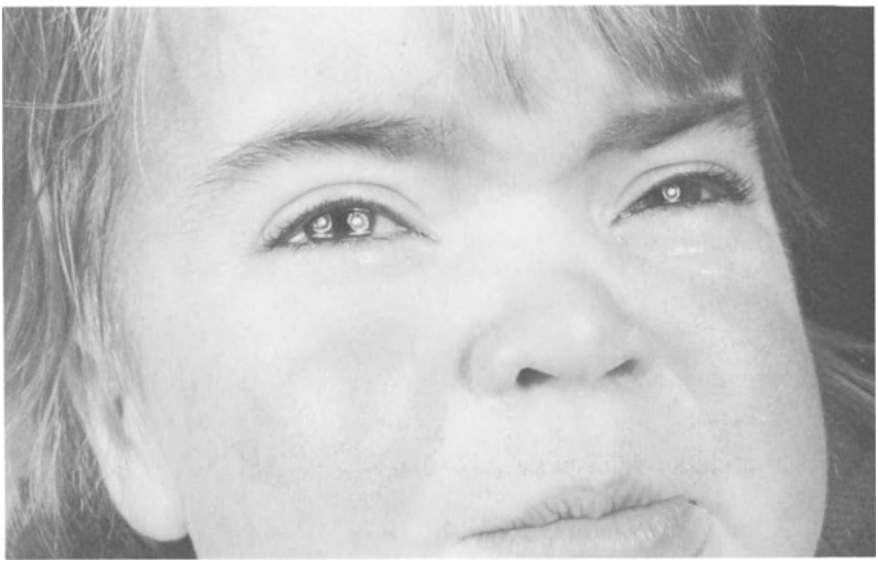

Fig. 19.1. Gargoylism.

and there is exophthalmos and often a divergent squint. The small optic foramina may lead to optic atrophy.

The Phakomatoses

The three conditions Von Recklinghausen's neurofibromatosis, tuberose sclerosis (Bournville's disease) and von Hippel–Lindau disease are classed together under this name. They all involve the eye but may not become evident until later in life. Often examination of the eye reveals the diagnosis. In Von Recklinghausen's neurofibromatosis, multiple neurofibromata are seen on the skin and the eyelids may be enlarged and distorted. Gliomata may develop in the optic nerves and scattered pigment 'cafe au lait' patches are seen in the skin. In tuberose sclerosis, mental deficiency and epilepsy are associated with a raised nodular rash on the cheeks and mulberry-like tumours in the optic fundus. Von Hippel–Lindau disease presents to the ophthalmologist as angiomatosis retinae. Vascular tumours appear in the peripheral retina, which may leak and expand and lead to detachment of the retina. Similar tumours may be present in the cerebellum.

Other Diseases in Childhood

Retrolental Fibroplasia

In the early 1940s, premature infants with breathing difficulties began to be treated with oxygen, and 12 years elapsed before it was realised that the 'retinopathy of prematurity' was due to this very treatment. During the course of excessive oxygen therapy in a premature infant, the retinal vessels become narrowed and the optic disc becomes pale. When the oxygen treatment is stopped, the retinal veins become engorged and new vessels grow from the peripheral arcades in the extreme periphery of the fundus. This growth of abnormal vessels leads to vitreous haemorrhage, retinal detachment and fibrosis of the retina. The infant may rapidly become blind, although some are minimally affected. The management of these cases now entails monitoring of the blood-oxygen level and regular inspection of the fundi of children at risk. The paediatrician who is managing the administration of oxygen therapy must balance the risk of endangering the life of the child against that of causing blindness. Recently it has been claimed that these children may be protected by treatment with vitamin E or by laser treatment to the peripheral retina.

Ophthalmia Neonatorum

It is important to realise that in the early part of this century, a large proportion of the inmates of blind institutions had suffered from ophthalmia

neonatorum. The disease affects primarily the conjunctiva and cornea and is the result of infection by organisms resident in the maternal birth passage. The gonococcus was the most serious cause of blindness but a number of other bacteria have been incriminated including staphylococci, streptococci and pneumococci. Subsequently it has been realised that chlamydial ('TRIC virus') infection of the genital tract may lead to the same problem, as may also infection by herpes simplex. The blindness which resulted from this condition was so serious that any excessive discharge from the eyes has been a notifiable disease in this country since 1914. Ophthalmia neonatorum is caused by unhygienic conditions at birth and its relative rarity nowadays is due to the fact that midwives are trained to instil a drop of 1% silver nitrate as soon as the eyes are open, and to a general improvement in hygiene at birth.

Bacterial conjunctivitis usually occurs between the 2nd and 5th day after birth, whereas chlamydial infection, known as inclusion blenorrhoea, tends to occur a little later, between the 6th and 10th day. Purulent or mucopurulent discharge is evident and the eyelids may become tense and swollen so that it is difficult to open them and carry out the all-important examination of the cornea. When the disease is suspected, the infant should be admitted to hospital and treated with intensive local antibiotics, for example tetracycline or penicillin and streptomycin drops every hour. If the cornea is threatened, systemic antibiotics are also needed. Diagnosis is achieved by taking a conjunctival culture before treatment is started and by looking for the inclusion bodies of the chlamydial virus in a smear. The mother should also have a vaginal smear and culture taken.

Leucocoria

This term means 'white pupil' and there are a number of conditions which may produce this effect in early childhood. The important thing to realise is that if a mother notices 'something white' in the pupil, the matter must never be overlooked and requires immediate investigation. The differential diagnosis includes congenital cataract, retrolental fibroplasia, endophthalmitis, some rare congenital abnormalities of the retina and vitreous and, not common but most important, retinoblastoma.

Abnormalities of Refraction

Nowadays children whose vision is impaired because they need a pair of spectacles are usually discovered by routine school testing of visual acuity. They may also present to the doctor because the parents have noticed them screwing up their eyes or blinking excessively when doing homework. Some children can tolerate quite high degrees of hypermetropia without losing visual acuity simply by exercising their accommodation, and unless it is felt that there is a risk of amblyopia or squint, glasses are better avoided in such cases. By contrast, even slight degrees of myopia, if both eyes are affected, can interfere with school work. Myopia does not usually appear until between the ages of 5 and 14, and most commonly at about the age of 11.

Squint

This exceedingly common inherited problem of childhood has already been considered, but it is worth summarising some of its main features. All cases of squint require full ophthalmological examination because they may be caused by treatable eye disease and may lead to impaired vision in one eye if neglected. There is no reason why any patient, child or adult, should suffer the indignity of looking 'squint eyed' because the eyes can be straightened by surgery. In spite of this, it is not always possible to restore the full simultaneous use of the two eyes (binocular vision). In general, the earlier in life that the treatment is started, the better the prognosis.

Amblyopia of Disuse

This condition has also been considered in a previous chapter. It has been defined as a unilateral impairment of visual acuity in the absence of any other demonstrable pathology in the eye or visual pathway. This rather negative definition fails to explain that there is a defect in nerve conduction due to inadequate usage of the eye in early childhood. The word 'amblyopia' means blindness and tends to be used rather loosely by ophthalmologists. It is most commonly used to refer to amblyopia of disuse (or amblyopia ex anopsia or 'lazy eye') but it is also used to refer to loss of sight due to drugs. Amblyopia of disuse is extremely common and some patients even seem unaware that they have any problem until they suffer damage to their sound eye. This weakness of one eye results when the image on the retina is out of focus or out of position for more than a few days or months during early childhood or, more specifically, below the age of 8 years. Amblyopia of disuse therefore arises as the result of squint or one-sided anomaly of refraction (anisometropia), or it may occur as the result of opacities in the optical media of the eye. A corneal ulcer in the centre of the cornea of a young child may rapidly lead to amblyopia. Once a clear image has been produced on the retina, either by the wearing of spectacles or the treatment of any optical defect, the vision in the weak eye may be greatly improved by occluding the sound eye. The younger the patient, the better are the chances of improving the vision by occlusion. Beyond the age of 8 years it is unlikely that any significant improvement can be achieved by this treatment and, by the same measure, it is unlikely that amblyopia will appear after the age of 8. An adult could suffer total occlusion of one eye for several months without experiencing any visual loss in the occluded eye.

Juvenile Macula Degeneration

Many families have now been described whose members show various types of macula degeneration and these conditions have been classified according to the age of onset. Best's disease is the name given to the form of heredomacula degeneration which appears at any time from birth to about the age of 6 (Fig.

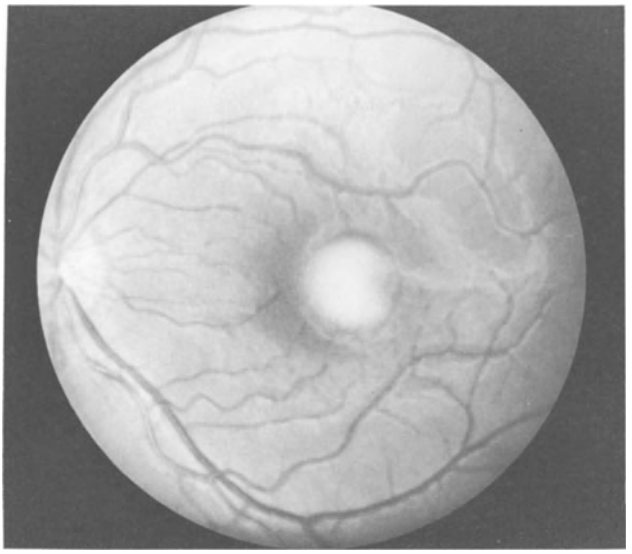

Fig. 19.2. Juvenile macula degeneration.

19.2). Often such a child will be brought for examination by parents who are concerned about a fall in school performance. It is often difficult to make an accurate assessment of the vision of a child at such an age and most patients who present in this way turn out to have normal eyes but have been unduly shy or nervous when tested. Juvenile macula degeneration is rare, but a missed diagnosis may result in untold distress to patient and parents.

Uveitis

Uveitis is rare in childhood; it may take the form of choroiditis, sometimes shown to be due to toxoplasmosis or toxocara, or of anterior uveitis sometimes associated with Still's disease. The management of these cases is similar to that in the adult, but recurrences may result in severe visual loss in spite of treatment.

Optic Atrophy

One must be rather wary about the diagnosis of optic atrophy in very young children because the optic discs tend to look rather pale normally. Occasionally unilateral visual loss with or without a squint is found to be associated with pallor of the disc on one side. Confirmed optic atrophy, either unilateral or bilateral, requires a full neurological investigation. The causes of optic atrophy in childhood are numerous but the important ones may be listed as follows.

Optic Atrophy Without Systemic Disease

Hereditary optic atrophy
Drug toxicity: streptomycin, isoniazid, ethambutal, chloramphenicol, sulphonamides, nalidixic acid

Optic Atrophy With Systemic Disease

Glioma of chiasm and craniopharyngioma
Post-meningitic
Post-traumatic after head injury
Hydrocephalus
Cerebral palsy
Disorders of lipid metabolism
Other rare causes

20 Systemic Disease and the Eye

Diabetes

Diabetes mellitus is an important cause of blindness: about 1000 people are registered blind from diabetes per year in the United Kingdom. Most of these patients are elderly, but diabetes is the commonest cause of blindness in young people in this country. The management of diabetic eye disease has improved greatly over the past 10 years so that much of this blindness can now be prevented. In spite of this, most general practitioners are aware of tragic cases of rapidly progressive blindness in young diabetics. The more serious manifestations of diabetes in the eye tend to affect patients in the prime of life. The tragedy is even greater when one considers that this blindness is largely avoidable.

Diabetes is therefore the most important systemic disease which gives rise to blindness. Many diabetics remain free of eye problems, but a diabetic is ten times more likely to become blind than other members of the population.

When taking an eye history from diabetic patients, it is especially important to note the duration of the diabetes and the age of onset.

Diabetic retinopathy is extremely rare under the age of 10 years; it does not usually appear until the disease has been present for some years. Juvenile onset diabetics usually take longer to show eye changes than those with a late onset.

Although diabetic retinopathy is the most serious ocular complication, the eye may be affected in a number of other ways and it is convenient to consider the various ocular manifestations of diabetes in an anatomical manner, beginning anteriorly.

Eyelids

It is usual to check the urine of patients presenting with recurrent styes but in practice it is unusual for diabetes to be diagnosed in this way. Xanthelasma of the eyelids is said to be slightly more common in diabetics.

Ocular Movements

Elderly diabetic patients are more prone to develop transient sixth cranial nerve palsies than non-diabetics of the same age group. Third cranial nerve palsies are less commonly seen.

Cornea and Conjunctiva

Some diabetics have recurrent subconjunctival haemorrhages. Although these haemorrhages are very common in otherwise healthy individuals, frequent recurrence may suggest diabetes. Corneal ulcers in diabetics may prove particularly troublesome. Minor trauma to the cornea may lead to the formation of an indolent chronically infected ulcer which responds very slowly to intensive treatment with local and systemic antibiotics. Inadequate treatment may lead to endophthalmitis and loss of the eye. This problem occurs especially in diabetics with severe vascular disease and typically in a patient who has had to have a gangrenous leg removed.

Anterior Chamber

A particular kind of iritis is occasionally seen in middle-aged diabetics. Sometimes fresh untreated cases may present to an eye casualty department complaining of blurred vision. Iritis may also occur secondary to corneal infection and ulceration.

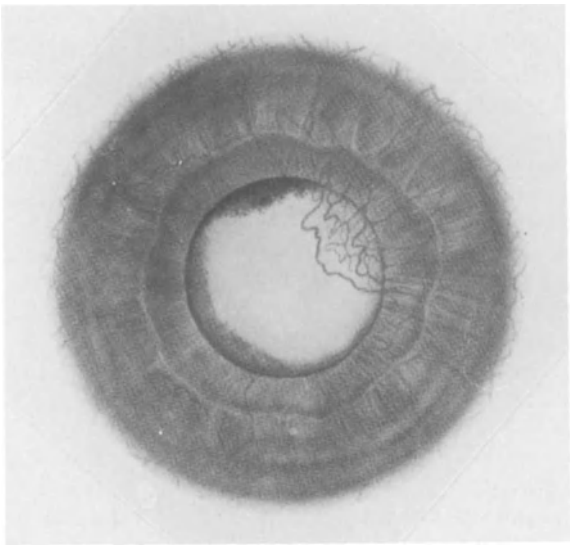

Fig. 20.1. Rubeosis iridis. (With acknowledgement to M.L. Berliner, 1949.)

Iris

The iris itself often shows degenerative changes in long-standing diabetics. The pupil may react sluggishly and fail to dilate very widely after the instillation of mydriatic drops. The surgeon can appreciate that pigment is easily lost from the iris when it is handled, and it is interesting that a characteristic vacuolation of the pigment epithelium lining the posterior surface of the iris is seen in histological sections. When diabetes seriously interferes with the circulation of the eye, the iris may become covered on its anterior surface by a fibrovascular membrane. To the naked eye, the iris takes on a pinkish colour, but examination with the slit-lamp microscope or a magnifying lens soon reveals the minute blood vessels on its surface. The appearance is known as 'rubeosis iridis'; very few eyes retain useful sight once this neovascularisation occurs (Fig. 20.1).

Lens

It was mentioned in an earlier chapter that diabetics tend to develop senile cataracts at an earlier stage than normal. In addition, a rapidly advancing type of cataract is seen in young poorly controlled patients. This cataract matures rapidly and is similar to the rare cataract seen in starvation from whatever cause. It is not possible to look at a cataract and deduce from its appearance that it is has been caused by diabetes — or, at least, it is not yet possible. On the other hand, the routine testing of urine of patients with cataracts produces a good return of positive results, making this an essential screening test.

It was also mentioned in a previous chapter that the refractive power of the lens may change in response to a rise in the blood-sugar level. Undiscovered diabetics quite often become short sighted due to this so-called index myopia. They may then obtain some distance glasses and subsequently consult their doctor, who treats their diabetes. By this time the glasses are made and, of course, turn out to be unsatisfactory, because the index myopia may improve with treatment. In some instances index myopia proves irreversible, being the first sign of cataract formation.

Retina and Vitreous

Diabetic retinopathy is the serious complication of diabetes in the eye and often reflects severe vascular disease elsewhere in the body. There are two kinds of diabetic retinopathy: exudative (or background) and proliferative. Background retinopathy is very common when diabetes has been present for some years and is less of a threat to the sight than the proliferative variety. It is important that the doctor should be able to recognise diabetic retinopathy and especially important that he or she should be familiar with the warning signs that indicate proliferative changes.

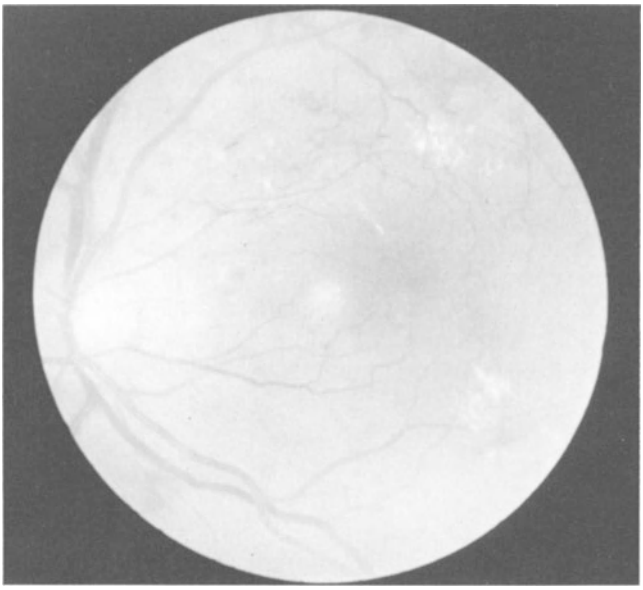

Fig. 20.2. Exudative diabetic retinopathy.

Exudative Retinopathy

There are usually no ophthalmic symptoms initially, but inspection of the fundi of most diabetics who have had the disease for a few years reveals, at first, a few *microhaemorrhages,* often on the temporal side of the macula but often scattered over the posterior pole of the fundus (Fig. 20.2). These may come and go over months and the overall picture may be unchanged after 10 years. The vision is not affected unless the dot haemorrhages are clustered round the macula region. *Hard exudates* are also seen and these tend to form rings around areas of diseased vessels, although only one part of the ring may be present at any given point. These are yellowish-white deposits with well-defined edges, the result of leakage from diseased blood vessels. *Capillary dilatation* is a more subtle sign of diabetic retinopathy. Histological examination of diseased retina has shown areas of capillary closure and capillary microaneurysms. The vessel walls have thickened basement membranes and loss of mural cells.

Proliferative Retinopathy

This poses a much more serious problem. Until recently, 50% of such cases became blind 5 years after diagnosis. In fact, a high proportion of patients with exudative retinopathy develop macula degeneration in later years but retain their ability to find their way about. The blindness which follows proliferative retinopathy may be complete and devastating. Proliferative

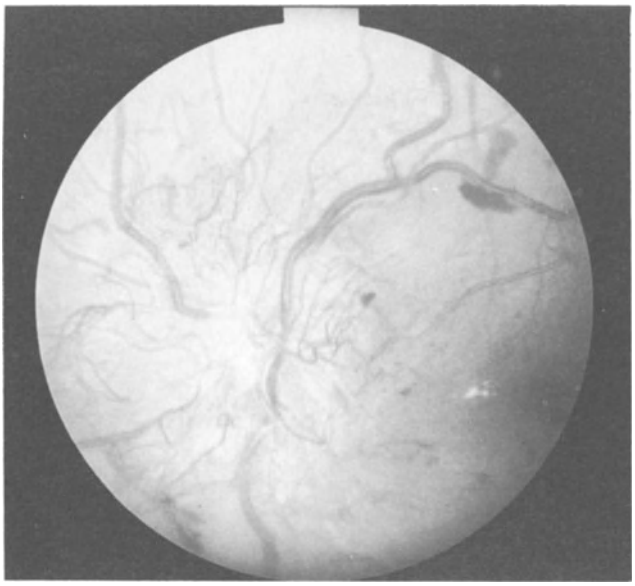

Fig. 20.3. Proliferative diabetic retinopathy.

retinopathy may be quite severe before the patient notices anything and the situation may have to be explained very carefully, in the absence of symptoms. Inspection of the fundus reveals sprouts of new vessels from the larger retinal veins, often from the region of the optic disc. These new vessels are thought to be a response to retinal ischaemia (Fig. 20.3). They grow between retina and vitreous and tend to leak, causing pre-retinal and vitreous haemorrhages. The vitreous often shrinks, drawing the vessels and associated fibrous tissue into the centre of the globe. Recurrent bleeding and fibrosis may lead to retinal detachment and secondary glaucoma.

The Preproliferative Stage. Proliferative retinopathy is typically seen in poorly controlled diabetics in their twenties and thirties. The situation may become very bad very quickly and it is important to be able to recognise the warning signs. There are three of them: a large number of haemorrhages, irregular calibre and dilatation of the retinal veins and, finally, the presence of soft exudates. Soft exudates are greyish white with ill-defined edges like patches of cotton wool. They represent an accumulation of white axoplasmic material around a retinal infarct. These warning signs may herald the appearance of the minute vessels, which should not be confused with normal disc capillaries or with widened collateral vessels.

Treatment

Control of Diabetes. This aspect of treatment may seem self-evident, but in the

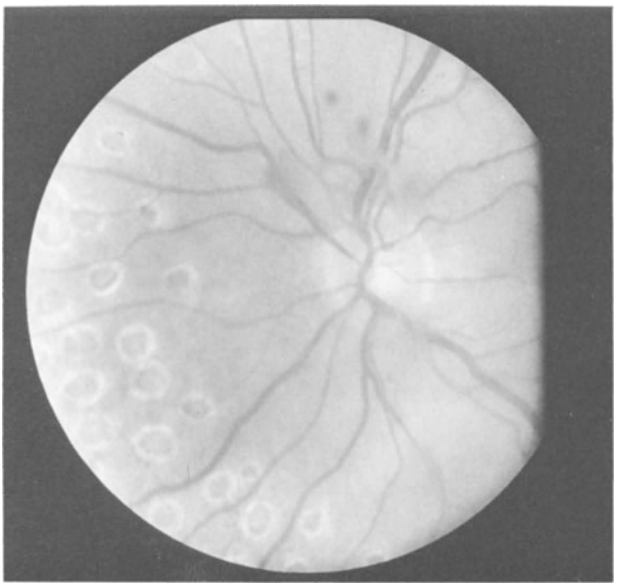

Fig. 20.4. Diabetic fundus after laser treatment.

past the value of careful control has not always been fully recognised. Some patients have the impression that 'eye problems develop anyway if the diabetes has been present long enough'. Nothing could be further from the truth.

Laser Coagulation. The use of a focused light beam to cauterise the retina has been practised for several years and the value of this treatment has been confirmed by extensive clinical trials. The exact mode of action is not known but it has been suggested that the cautery of ischaemic areas prevents the release of some, as yet unidentified, vasoformative factor. The treatment must be applied promptly in the early proliferative stage or sometimes before. The laser treatment of exudative retinopathy is less effective but good results have been claimed (Fig. 20.4).

Glaucoma Surgery. Drainage surgery may be needed if glaucoma is not controlled by medical means.

Retina Surgery. From time to time, an improvement in vision can be achieved by retinal detachment surgery and it should be remembered that detachment of the retina may also be a cause of sudden visual deterioration in these patients.

Vitreous Surgery. There have been dramatic advances in the technical side of vitreous surgery in recent years so that it is now possible to remove a persistent vitreous haemorrhage and to divide or remove fibrous tissue, even from the surface of the retina.

Prognosis

A better understanding of diabetic retinopathy has resulted from the use of more accurate methods of investigation, especially fluorescein angiography but also the routine use of indirect ophthalmoscopy and slit-lamp microscopy. Serial fundus photography and the use of ultrasound have also been important. This better understanding has led to more effective treatment so that the more severe ocular complications are now largely avoidable. Blindness tends to be limited to those cases where social or other circumstances make management difficult.

Thyrotoxicosis

Although the typical eye signs of hyperthyroidism (Grave's disease) are usually accompanied by the other systemic manifestations of the disease, they may also appear without any other clinical evidence of thyrotoxicosis. Furthermore, the eye signs may become worse even though the other clinical signs and symptoms are well controlled by medical treatment. The disease is thought to be due to an autoimmune process but the relationship between the eye and systemic changes is still obscure. The ophthalmologist sees two kinds of thyrotoxic eye disease: one in young people, usually females in their late teens or early twenties, and the other in late middle age in either sex.

Youthful Thyrotoxic Eye Disease (Fig. 20.5)

These patients may have other signs and symptoms of thyrotoxicosis such as excitability, weight loss, high pulse rate, dislike of warm weather, and fine

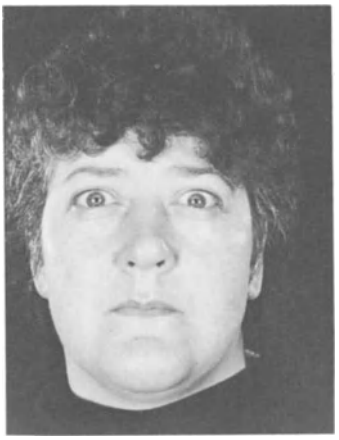

Fig. 20.5. Thyrotoxicosis.

tremor. The eye symptoms are usually limited to irritation and soreness and the signs may be as follows.

Lid Retraction. Eyelid drawn up slightly, more on one side than the other. Reveals white sclera above corneoscleral junction.

Lid Lag. When instructed to follow a pencil as it moves downwards, the upper lid appears to lag behind the rotation of the eye, revealing more of the white above. The upper lid shows jerky movements as the eye rotates smoothly down.

Lid Swelling. Puffiness of the eyelids may be present.

Chemosis. This means conjunctival oedema. To the naked eye it appears as though the eyes are brimming with tears, and the expression 'the tear that never drops' is sometimes used.

Proptosis. Lid retraction may give the false impression of proptosis but measurement of the position of the globe in relation to the bony orbit can be achieved by means of an exophthalmometer. Any relative protrusion can thus be measured for future reference.

Exposure Keratitis. Punctate staining with fluorescein across the lower part of the cornea is characteristic and due to inadequate closure of the retracted upper lid.

Management

As the symptoms are relatively mild, treatment is usually limited to that of the exposure keratitis; an antibiotic ointment instilled at night is often sufficient. Sometimes a small lateral tarsorrhaphy on each side can greatly improve the appearance of a young girl with lid retraction. Lid retraction may also be improved by the use of guanethidine eyedrops.

Middle-aged Dysthyroid Eye Disease

This can affect either sex and the patient may not be clinically thyrotoxic. It is seen, for example, in patients who have previously been treated with radioactive iodine and who are euthyroid. It must be borne in mind as a possible cause of diplopia, even in the absence of abnormal thyroid function. This type of thyrotoxic eye disease may be very severe. It usually lasts for 3 or 4 years before undergoing spontaneous resolution. During this time it is possible for the sight to be lost if treatment is inadequate. The symptoms and signs are at first the same as those experienced by the younger patients but more serious signs may become added as follows.

Extraocular Muscle Palsy. The muscles become infiltrated and thickened producing a characteristic appearance on CT scan which helps to distinguish this form from other causes of diplopia. The main restriction of movement is due to tethering of the inferior recti with limitation of upward gaze. The resulting pressure on the globe may cause the intraocular pressure to rise on looking up and this has been used as a diagnostic test.

Marked Proptosis. Forward protrusion of the globe may lead to severe exposure keratitis demanding urgent attention.

Optic Nerve Compression. This is due to the increased pressure within the orbit. The first sign may be swelling of the optic disc, followed by optic atrophy. It is therefore vitally important to *monitor the visual acuity* in these cases.

Management

A tarsorrhaphy may be needed as well as local antibiotics, but if there is visual deterioration, large doses of systemic steroids are probably the best line of treatment (e.g. prednisolone 120 mg/day). Initial recovery is usually dramatic and rapid but then the side-effects of systemic steroids ensue. The dose should be reduced as soon as feasible but it may be necessary to continue with a maintenance dose for many months. Some surgeons claim that surgical decompression of the orbits is a more satisfactory first line of treatment in these severe cases. If double vision persists beyond the acute stage, extraocular muscle surgery may be helpful and operations have also been designed to deal with lid retraction (Table 20.1).

Table 20.1. The 13 possible eye signs of thyroid disease

1. Proptosis
2. Raised intraocular pressure when looking up
3. Lid lag
4. Lid retraction
5. Lid swelling
6. Chemosis
7. Conjunctival congestion
8. Extraocular muscle palsy
9. Exposure keratitis
10. Corneal ulceration
11. Papilloedema
12. Impaired visual acuity
13. Constriction of visual field

Routine Tests for Thyrotoxicosis

Serum thyroxine (T_4)	Thyroid autoantibodies
T_3 resin uptake	T_3 assay

Hypertension

Even though the effects of raised blood pressure on the appearance of the fundus of the eye were recognised in the 19th century, the nature of the detailed changes is still disputed. Certain characteristic features, such as the nipping of the veins at arteriovenous crossings, narrowing of the arterioles, haemorrhages, papilloedema and exudates, are beyond doubt. Some confusion can be avoided if it is realised that the effects of raised blood pressure are modified by other changes in the eye due to natural ageing. It is now accepted that the exact cause of the raised blood pressure does not of itself influence the fundus appearance, in spite of the persistence of old-fashioned terms such as 'renal retinitis' referring to hypertensive retinopathy with renal disease.

The Effect of Age on the Retinal Blood Vessels

This has already been discussed in Chapter 18. In older patients the retinal arteries are seen to be narrower and straighter and the veins are also narrower than in young patients. The term 'retinal arteriosclerosis' is used to describe these changes.

The Effect of Raised Blood Pressure on the Retinal Vessels

In younger patients, irregular narrowing of the retinal arterioles is seen, and is thought by many to be due to spasm of the vessel walls. This hypertonicity

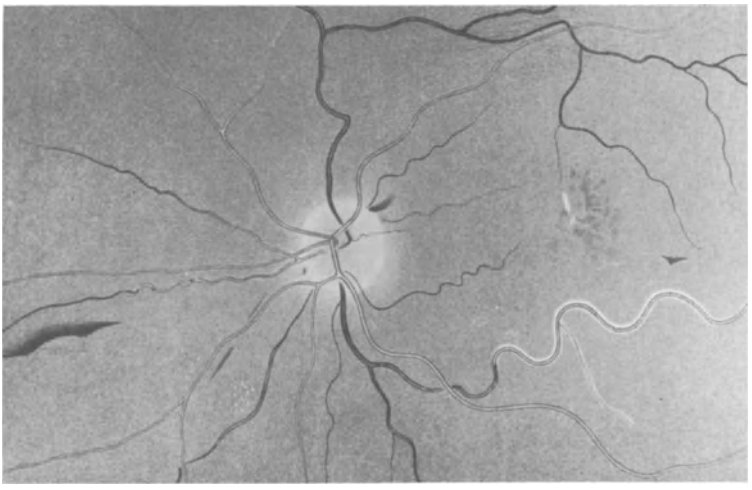

Fig. 20.6. The effect of raised blood pressure on the retinal vessels. (After R.M. Gunn, 1898.)

leads in time to more permanent changes in the vessel walls so that the vessels resemble those of an older patient. Nipping of the veins at arteriovenous crossings is seen and on the distal side of the crossing the vein may be distended. Occasional flame haemorrhages and some slight swelling of the optic disc may indicate more severe vascular damage but do not necessarily lead to 'malignant' hypertension (Fig. 20.6).

In older patients, the already narrowed vessels tend to show less dramatic changes. Hypertonicity of the vessel walls is not seen but arteriovenous nipping remains an important sign and haemorrhages may be present in more severe cases. The exudates of hypertension are 'soft' or 'cotton wool' exudates and reflect ischaemic damage to the nerve fibre layer. Swelling of the disc may also occur.

'Malignant' Hypertension

Occasionally patients with a severe hypertensive problem present directly to the ophthalmologist because their main symptom is blurring of the vision, the other more usual symptoms being less evident. On examination, the visual acuity may be only slightly reduced and there may be some constriction of the visual fields. Inspection of the fundus reveals marked swelling of the optic disc, the oedema often extending well away from the disc with scattered flame-shaped haemorrhages. If the diastolic blood pressure is above 130 mmHg, there is little doubt about the diagnosis, but below this level it is essential to bear in mind the possibility of raised intracranial pressure. When hypertension is as severe as this the patient should be treated as an acute medical emergency and referred without delay to the appropriate physician.

Grading of the Degree of Hypertensive Retinopathy

Keith and Wagener graded the fundoscopic signs in the following manner more than 40 years ago. The system is still used by clinicians.

Grade I. Attenuation of arteries and increase of light reflex (an appearance sometimes called 'silver wiring').

Grade II. More severe degree of Grade I with the addition of 'angling' at the arteriovenous cross-overs and compression of the veins.

Grade III. The above two grades combined with the addition of retinal oedema and often a star figure at the macula due to oedema. Cotton-wool spots and flame-shaped haemorrhages are also seen (Fig. 20.7).

Grade IV. All the changes of Grade III combined with the presence of papilloedema.

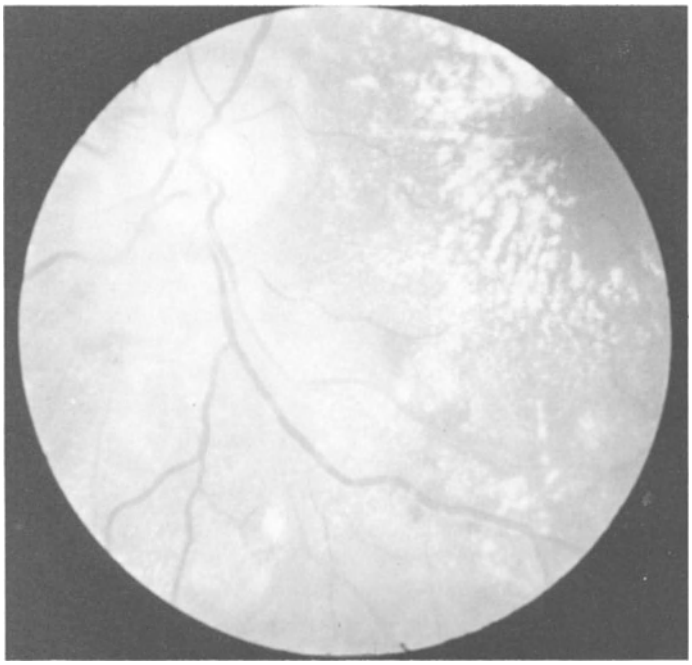

Fig. 20.7. Severe hypertensive retinopathy.

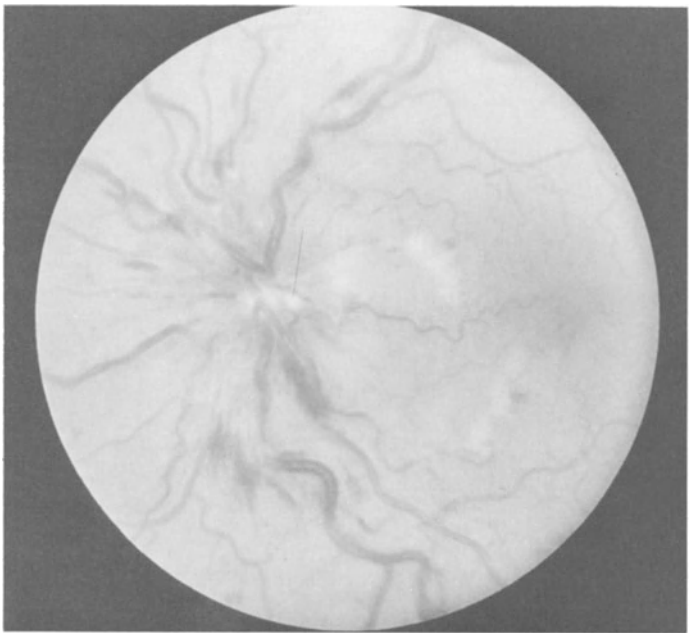

Fig. 20.8. Central retinal vein occlusion.

Other Associated Vascular Changes

Retinal Vascular Occlusion

This is more common in hypertensive patients. The most frequent occurrence is the central retinal vein occlusion in which the visual prognosis is fair and some recovery can often be expected. The fundus appearance is dramatic with numerous scattered haemorrhages and swelling of the optic disc and the patient experiences sudden blurring of vision in one eye (Fig. 20.8). This can be compared with occlusion of the central retinal artery which is less common and in which the prognosis is uniformly bad. Here the fundus appears pale and the arteries are narrowed. There is a cherry-red spot at the macula.

Emboli

Cholesterol emboli may be seen in the retinal arteries, sometimes in association with arterial occlusion. These usually arise from atheromatous plaques in the carotid artery. Calcified emboli may be seen in association with diseased heart valves and platelet or fibrin emboli may also be observed.

Ischaemic Optic Neuropathy

Some elderly patients complaining of visual loss in one eye are found to have a pale swollen optic disc and sometimes evidence of branch artery occlusion, giving an altitudinal defect of the visual field. This appearance should suggest the possibility of temporal arteritis and an ESR and a temporal artery biopsy should be considered as urgent investigations (Fig. 20.9).

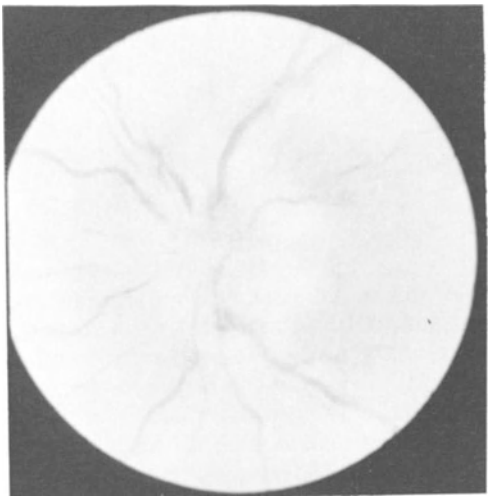

Fig. 20.9. Ischaemic papilloedema.

Anaemia

When the haemoglobin concentration in the blood is abnormally low, this becomes apparent in the fundus. The retinal vessels become pale and the difference between arteries and veins becomes less apparent. The fundus background also appears pale but this sign is dependent upon the natural pigmentation of the fundus and may be misleading. In severe cases small haemorrhages are usually seen, mainly around the optic disc, and a special feature of anaemic retinopathy is the presence of white areas in the centre of some of the haemorrhages. The haemorrhages tend to be flame shaped. Examination of the conjunctiva is perhaps of more value — or at least is certainly an easier way of assessing the haemoglobin level — and this part of the examination of the eye should, of course, precede ophthalmoscopy.

The Leukaemias

Fundus changes are common in both lymphatic and myeloid leukaemia, and it is not possible to distinguish the two from examination of the eyes. The main features are dilatation and tortuosity of the veins; leukaemic deposits may be seen as greyish areas amongst scattered haemorrhages, some of which show grey centres. Sometimes the veins are sheathed by grey leukaemic infiltrate and the optic discs may be swollen. Apart from the fundus changes, the vision may be impaired by leukaemic infiltrates elsewhere in the visual pathway with corresponding defects in the visual field.

Sickle Cell Anaemia

This condition is mentioned separately because of the severe and devastating effect it may have on the vision. The sickle cell haemoglobinopathies are inherited, the affected person having abnormal haemoglobins recognised by the electrophoretic pattern and labelled alphabetically. Haemoglobins S and C are the most important ophthalmologically. Thalassaemia (persistence of foetal haemoglobin) can also cause retinopathy. Abnormal blood flow in the smaller retinal vessels due to the abnormal haemoglobins leads to the formation of microaneurysms and haemorrhages. In severe cases new vessels appear producing a retinopathy not unlike that of diabetes, although it tends to start in the peripheral part of the fundus.

Other Blood Disorders

Polycythaemia is characterised by a marked increase in the number of red blood cells in the circulation. It may be primary (polycythaemia vera), or secondary to pulmonary insufficiency or congenital heart disease. In both instances the fundus takes on a florid appearance, the vessels being engorged and tortuous and often the optic disc is swollen. Central retinal vein occlusion is a recognised complication. The conjunctiva of these patients also appears congested and, very rarely, they may present with unexplained red eyes.

The various haemorrhagic disorders may sometimes show retinal haemorrhages but specific changes have not been described.

21 Neuro-ophthalmology

It is found in most ophthalmic departments that it is necessary to retain a close liaison with neurological and neurosurgical departments, and neuro-ophthalmology is now in itself becoming a subspecialty. Retrobulbar neuritis, for example, is a condition which presents quite commonly to eye casualty departments and usually requires further investigation by a neurologist. Less common but equally important are the pituitary tumours which, it will be seen, can present in a subtle way to the ophthalmologist and which may require urgent neurosurgical attention. There are many other, sometimes rare, conditions which find common ground between the disciplines.

The Optic Disc

Normal Disc

One must be familiar with some of the variations found in otherwise normal individuals before being able to diagnose pathological changes. The optic discs mark the entrance of the optic nerves to the eye and this small circular part of the fundus is non-seeing and corresponds with blind spots in the visual field. When examining an optic disc, five important features are to be noted; the colour, the margins, the vessel entry, the central cup, and the presence or absence of haemorrhages.

Colour. The disc is pink but often slightly paler on the temporal side. That of the neonate may be deceptively pale and some elderly discs appear atrophic without evidence of disease. Pallor of the disc is due to loss of nerve tissue which causes exposure of the underlying sclera. The myopic disc is relatively pale, whereas the hypermetropic disc is pinker than normal (Fig. 21.1).

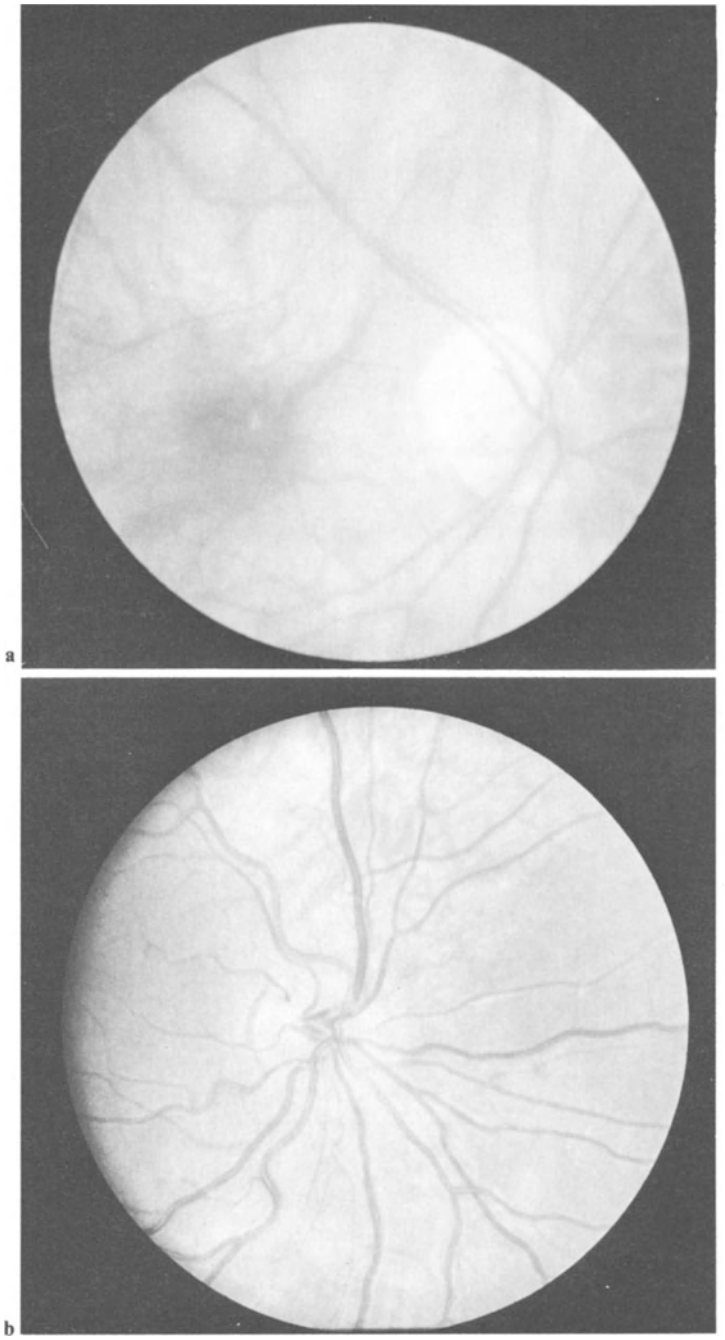

Fig. 21.1a, b. The normal optic disc: **a** of a myope, **b** of a hypermetropic patient.

Margins. These are better defined in myopic than hypermetropic subjects. In hypermetropes the edges of the disc may appear raised, sometimes resembling papilloedema. It is common to see a crescent of pigment on the temporal side of the disc.

Vessel Entry. In general, a central retinal artery and vein divide into upper and lower branches which in turn divide into nasal and temporal branches close to the disc margin. Many variations in the pattern are seen normally. The veins are darker and wider than the arteries and, unlike the arteries, can be seen to pulsate if examined carefully.

Central Cup. The centre of the disc is deeper, i.e. further away from the observer, than the peripheral part. This central cup occupies about a third of the total disc diameter in normal subjects.

Haemorrhages. Haemorrhages are never seen on normal discs.

A number of minor congenital abnormalities are seen on the disc. In an astigmatic eye the disc is often oval. The central cup may be filled in by 'drusen' — small hyaline deposits which may be found on the surface or buried in the substance of the disc. Alternatively, the central cup may be hollowed out by a congenital pit in the disc. Myelinated nerve fibres are recognised by their strikingly white appearance, which obscures any underlying vessels, and their fluffy margin. The central cup may be filled in by persistent remnants of the hyaloid artery which runs in the embryo from disc to lens. Some of these and other congenital abnormalities of the disc may be associated with visual field defects which are not progressive but which can cause diagnostic confusion.

Pale Disc

Optic Atrophy

Optic atrophy means loss of nerve tissue on the disc, and the resulting abnormal pallor of the disc must be accompanied by a defect in the visual field, but not necessarily by a reduction in the visual acuity. It must be remembered that the disc tends to be somewhat pale in short-sighted eyes and care must be taken in diagnosing optic atrophy in such cases. The number of small vessels which can be counted on the disc is sometimes used as an index of atrophy in different cases.

Classification of the causes of optic atrophy usually includes the term 'consecutive optic atrophy', referring to atrophy following retinal degeneration. The terms primary and secondary atrophy are also used but because these terms are confusing a simple aetiological classification will be used here. It should be borne in mind that it is not usually possible to determine the cause of optic atrophy by the appearance of the optic disc. Even the cupped pale disc of chronic glaucoma may be mimicked by optic atrophy due to chiasmal compression. When optic atrophy follows swelling of the optic

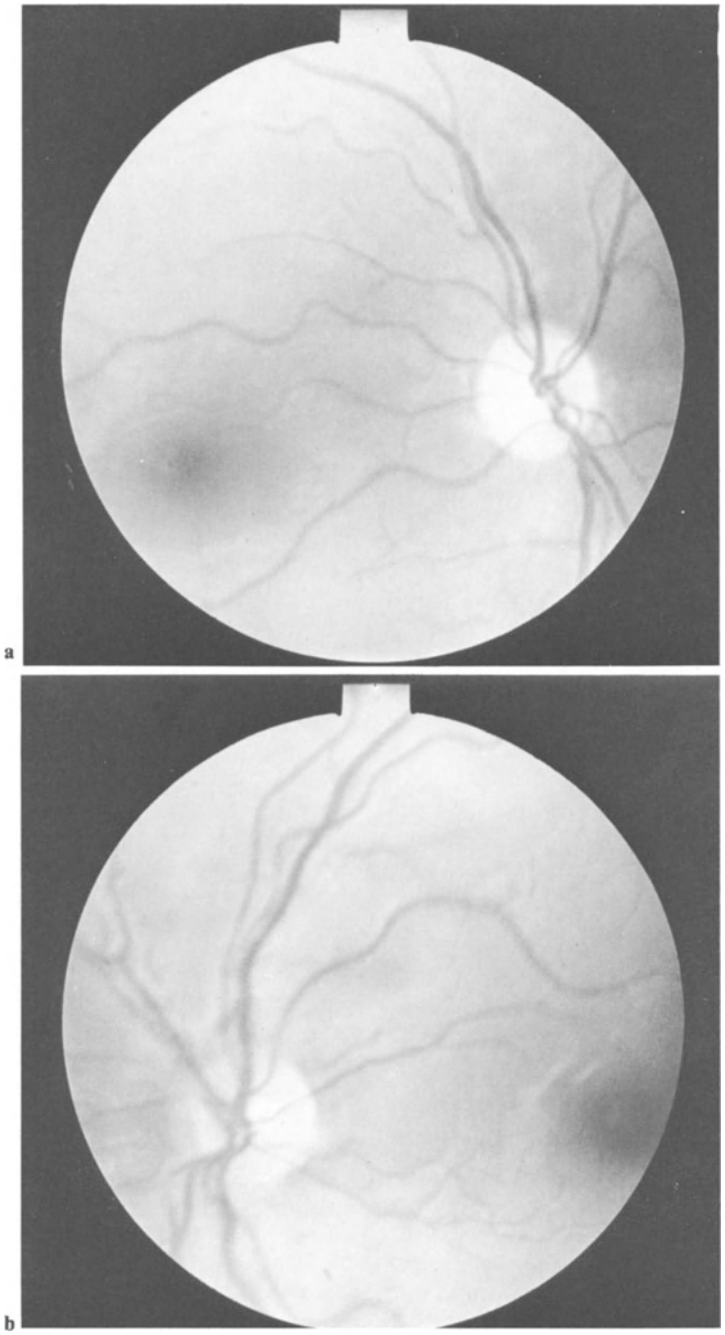

Fig. 21.2a, b. Optic atrophy in a patient with a pituitary adenoma. **a** The atrophic right optic disc. **b** The normal left optic disc.

disc, there is more gliosis than when it is 'primary', that is, due to disease in the nerve itself. Gliosis makes the appearance of the disc more grey or yellowish-grey than white and the cribriform marking often seen in optic atrophy may not be evident.

The following are the important causes of optic atrophy.

1. *Glaucoma.*

2. *Vascular.* Following obstruction of the central retinal artery or vein.

3. *Following disease in the optic nerve,* for example optic neuritis or compression of the nerve by an aneurysm or tumour (Fig. 21.2).

4. *Following papilloedema.* The disc may become atrophic as a direct result of the swelling, irrespective of its cause.

5. *Inherited.* Retinitis pigmentosa is an inherited retinal degeneration in which there is a progressive night blindness, constriction of the visual field and scattered pigmentation in the fundus. As the condition advances towards blindness, the discs become atrophic. Optic atrophy may also appear in certain families without any other apparent pathology, for example Leber's optic atrophy and congenital or infantile optic atrophy. It is also seen in the rare but distressing cerebro-macula degeneration which presents with progressive blindness, epilepsy and dementia.

6. *Toxic.* A number of poisons can specifically damage the optic nerve; methyl alcohol is a classical example. Tobacco amblyopia is a type of progressive atrophy due to excessive smoking of coarse tobaccos, usually in a pipe and often in association with a high ethyl alcohol intake. Reversal is achieved by abstention.

7. *Trauma.* The optic nerve may be damaged by indirect injury if bleeding occurs into the dural sheath. This may result from a fracture in the region of the optic foramen or, rarely, from contusion of the eye itself. After the nerve has been damaged, a period of a few weeks elapses before the nerve head becomes atrophic, so that initially the eye may be blind but the fundus normal. The pupil reaction to direct light is impaired from the time of the injury. Such an injury may result in complete and permanent blindness in the affected eye but a degree of recovery is achieved in a small proportion of cases.

Swelling of the Optic Disc

This is a serious sign because it may be due to raised intracranial pressure and an intracranial space-occupying lesion. There are, however, a number of other more common causes.

Apparent Swelling. The margins of the optic disc may be ill-defined and even appear swollen in hypermetropic eyes. Other congenital abnormalities of the disc such as drusen or myelination of the nerve fibres may also be mistaken for true swelling (Fig. 21.3).

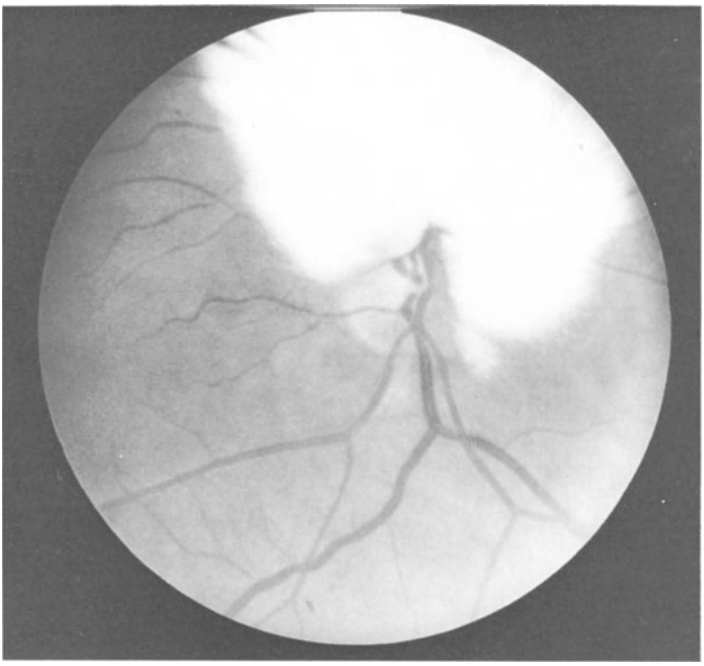

Fig. 21.3. Myelinated nerve fibres.

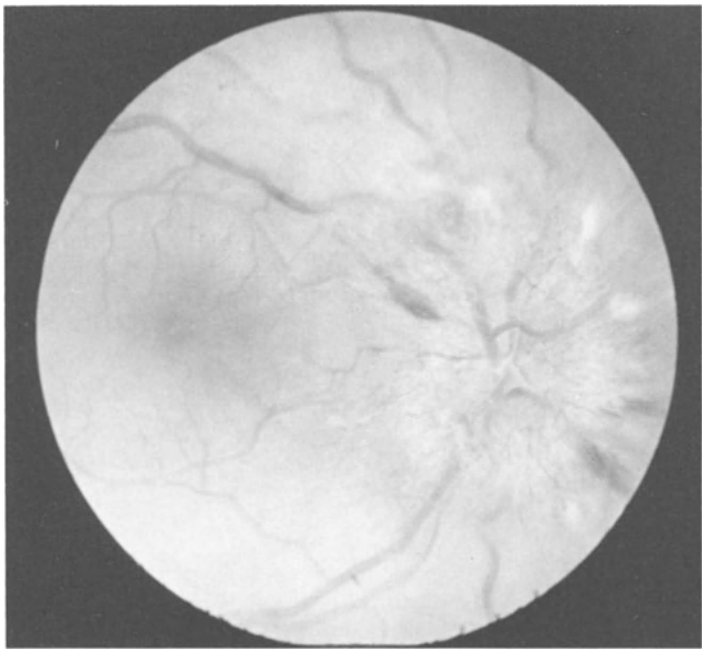

Fig. 21.4. Papilloedema.

Vascular. The disc may be swollen in congestive cardiac failure or in patients with severe chronic emphysema. Marked swelling of the disc with numerous haemorrhages is seen in occlusion of the central retinal vein and this compares with the pale and less haemorrhagic swelling that is seen in ischaemic optic neuropathy. In the latter instance, swelling of the disc occurs in association with arterial disease and one must take pains to exclude temporal arteritis in the elderly.

Postoperative. Swelling of the disc is quite common in the immediate postoperative period after intraocular surgery. It may persist for longer periods if the intraocular pressure remains low but it is not usually regarded to be of serious significance.

True Papilloedema. Every doctor must be aware of the triad of headache, papilloedema and vomiting as an important feature of raised intracranial pressure. The optic disc may be markedly swollen and haemorrhages are present around it, but not usually in the peripheral fundus (Fig. 21.4). In chronic papilloedema, the disc is paler and haemorrhages may be few or absent. Although these patients may complain of transient blurring of the vision, the visual acuity is usually normal and testing the visual fields shows only some enlargement of the blind spots. It is important to realise that the word 'papilloedema' refers to the non-inflammatory swelling of the disc which results from raised intracranial pressure. The most common causes of raised intracranial pressure are cerebral tumours, malignant hypertension, cerebral abscess, subdural haematoma, hydrocephalus and benign intracranial hypertension.

Diagnosis of papilloedema entails careful examination of the optic disc which must be backed up with visual field examination and colour fundus photography. The latter is especially helpful when repeated to show any change in the disc appearance. Fluorescein angiography may also be of great diagnostic help in difficult cases.

Optic Neuritis. This most commonly occurs in association with a plaque of demyelination in the optic nerve in patients with multiple sclerosis. The central vision is usually severely affected, in contrast with papilloedema, but optic neuritis occurs in many instances without any visible swelling of the disc.

Other Causes. In severe diabetic retinopathy, the early stages are sometimes marked by disc swelling. In severe cases of thyrotoxic exophthalmos, the orbital congestion may cause disc swelling. In both instances the doctor should be warned that serious consequences may ensue unless prompt treatment is applied.

Multiple Sclerosis

This common and important neurological disease may often present initially as

an eye problem and its proper management requires careful coordination at the primary care level. It is important to realise that multiple sclerosis cannot be diagnosed after one single attack of retrobulbar neuritis since this could cause unnecessary alarm about something which may never happen. The diagnosis of multiple sclerosis should be made by a neurologist and is based on finding additional evidence of the disease elsewhere in the body.

The cause of multiple sclerosis is not known but the disease is characterised by the appearance of multiple inflammatory foci in relation to the myelin sheaths of nerves throughout the central nervous system. The optic nerve between globe and chiasm is commonly involved at an early stage and there may be a delay of several years before other features of the disease appear. Young or middle-aged people are mainly affected and the prognosis is worse when the disease is acquired at an early age.

Ocular Findings

Retrobulbar Neuritis

This is an important cause of unilateral sudden loss of vision in a white eye in a young person. The patient complains of pain behind the eye on attempting to move it and there is often a grey or coloured patch in the centre of the field of view. In bad cases the sight of the affected eye may be lost completely. On examination the pupil reaction is diminished on the affected side but his may be the only objective evidence of disease. It is essential to test the pupil before dilating it with eyedrops. The fundus is often normal initially, although there may be a slight swelling of the optic disc. After 2 or 3 weeks the optic disc starts to become pale. The visual prognosis is good. Most patients make a complete or nearly complete recovery after 3 months. There is a risk that the other eye may be affected at a later date and recurrent attacks may cause permanent damage to the vision. Fortunately it is extremely rare for a patient to be made blind by multiple sclerosis.

The diagnosis at the time of the acute attack relies on the history and noting the pupil reaction. It is often important to make the diagnosis in retrospect. The patient may give a history of visual loss in one eye which has recovered and at a later date presents with other non-ocular signs and symptoms of demyelinating disease. If it can be confirmed that the patient has had a previous attack of optic neuritis, this may help in the confirmation of the diagnosis of disseminated sclerosis. Under these circumstances the pallor of the disc may be helpful, but careful assessment of the colour, vision and measurement of the visually evoked potential may provide conclusive evidence. At the time of the acute attack, testing the visual field may reveal a centrocaecal defect. The size of this defect diminishes as healing occurs, often leaving a small residual defect between blind spot and central area.

Nystagmus

This usually appears at a later stage than optic neuritis and may only be evident on lateral gaze. It is often horizontal.

Internuclear Ophthalmoplegia

Whereas double vision is a common symptom in multiple sclerosis, it is unusual to see an obvious defect of the ocular movements. Sometimes it can be seen that one medial rectus fails to adduct when the patient is asked to look to one side, and yet when the patient is made to converge the eyes on a near object, the medial rectus moves normally. This failure of the muscle action with certain eye movements only is termed an 'internuclear ophthalmoplegia' and it is very characteristic of multiple sclerosis when seen in young people, but usually has a vascular cause in the elderly.

Other Features

Other types of ocular muscle palsy, for example a lateral rectus palsy or ptosis, are rare. Careful inspection of the fundi in some cases reveals inflammatory changes around the retinal vessels.

Defects in the Visual Fields

The pattern of a visual field defect gives useful localising information for lesions in the visual pathway. Because the right half of *each* retina is linked by nerves to the right occipital cortex and because the splitting of nerve fibres from each half occurs at the chiasm, lesions in the optic nerve anterior to the chiasm tend to cause unilateral defects whereas those posterior to the chiasm produce hemianopic or quadrantianopic defects (Fig. 21.5). Cortical lesions

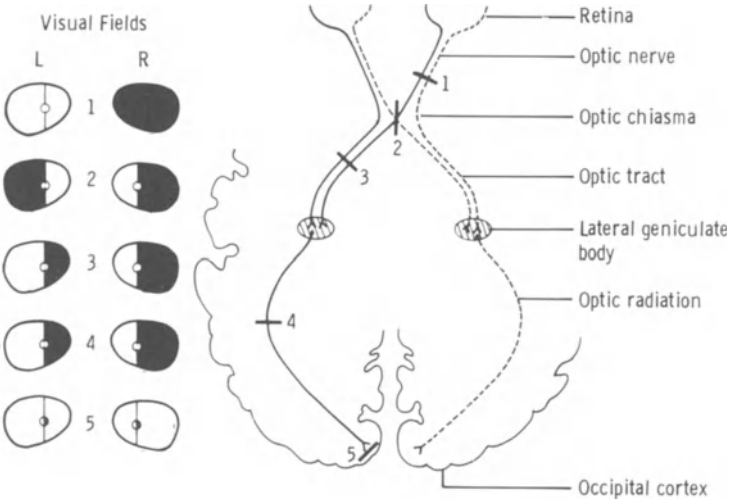

Fig. 21.5. Diagram of visual pathway.

Fig. 21.6. "My car keeps knocking my gate post. (Hemianopes should never drive)."

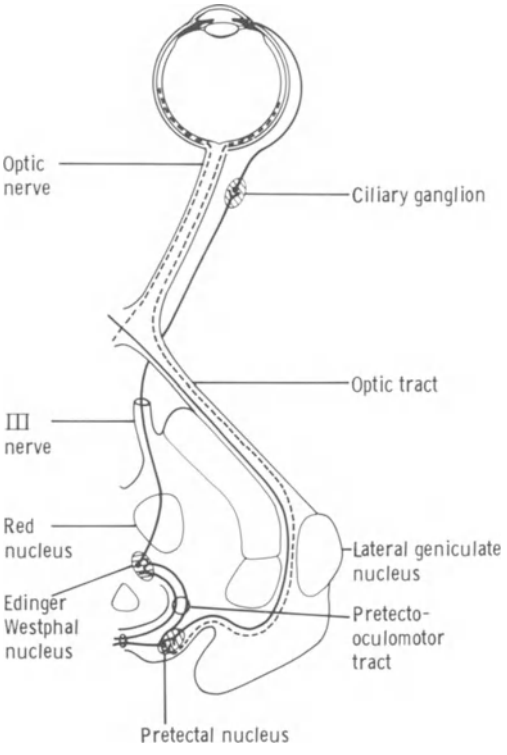

Fig. 21.7. Diagram of pupillary pathway.

tend to be more congruous, that is, the blind areas on each side tend to be similar in shape and size. Cortical lesions also show better preservation of central vision ('macula sparing'). A special type of field defect is seen with expanding pituitary tumours, the resulting pressure on the centre of the chiasm producing a bitemporal defect. Localised defects in the retina produce equivalent localised defects in the visual field on the affected side. Defects due to retinal disease are relatively common, for example glaucoma in the elderly, and care must be taken to interpret field defects with this possibility in mind. Notice from the diagram in Fig. 21.5 that the right half of the visual field is represented in the left half of each retina and thus the left half of the brain. This complies with the general rule that events occurring on the right side of the body are represented on the left side of the brain. It is surprising how patients may be unaware of a severe visual field defect, especially in hemianopia (Fig. 21.6).

Abnormalities of the Pupil

The pupil constricts and dilates largely under the action of the sphincter muscle which lines the pupil margin. It is supplied by parasympathetic fibres from the midbrain which relay in the ciliary ganglion having been conveyed along the third cranial nerve (Fig. 21.7). The dilator muscle is arranged radially within the iris and responds to the sympathetic nerves conveyed in the sympathetic plexus overlying the internal carotid artery. These fibres in turn arise from the superior cervical ganglion. The sympathetic supply to the dilator muscle therefore runs a long course from the hypothalamus to the midbrain and spinal cord and then up again from the root of the neck with the internal carotid artery.

Miosis refers to a small pupil, mydriasis to a large pupil (big word, big pupil). The pupil grows smaller with age. In young children pupils are relatively large and sometimes anxious parents bring up their children because they are concerned about this. During sleep the pupils become small. The pupils react to afferent stimuli conveyed along the optic nerves. The nerve fibres leave the optic nerves posterior to the chiasm and pass to the midbrain. When examining the eye with the ophthalmoscope it is evident that the pupil constricts more vigorously when the macula is examined than when the more peripheral fundus is stimulated with the ophthalmoscope light. When an eye is totally blind, usually there is no pupil reaction but, as a general rule, the sight of an eye must be very poor before the pupil reaction is affected. It should be apparent from Fig. 21.7 that the patient with cortical blindness may have a normal pupil reaction. We must also remember that a pupil may not react to light because it is mechanically bound down to the lens by adhesions. When both maculae are damaged by senile macula degeneration, the pupils may be slightly wider than normal and may show sluggish reactions.

The Abnormally Dilated Pupil

The commonest reason for unilateral mydriasis is *drugs* in the form of locally administered eyedrops, either prescribed by an ophthalmic department or obtained from a friend's medicine cabinet. The next commonest cause is probably the *Holmes–Adie syndrome,* a condition which is more common in young female patients. The affected pupil is usually dilated and contracts very slowly in response to direct and indirect stimulation. In bright light the pupil may be constricted on the affected side and take some time to dilate in the dark. This tonic pupil reaction may be combined curiously with absent tendon jerks in the limbs. When the vision is blurred and the pupil widely dilated, the symptoms may be partially relieved by the use of a weak miotic. After a delay of months or years the other eye may become affected. The overall disability is minimal and the condition has not so far been related to any other systemic disease. Acute narrow angle glaucoma can occasionally present in this manner and confusion may arise if the eye is not very red; however, closer examination of the eye should make the diagnosis obvious. Since the nerve fibres which cause constriction of the pupil are conveyed in the oculomotor nerve, oculomotor palsy if complete is associated with mydriasis. For this reason dilatation of the pupil may be a serious sign of raised intracranial pressure after head injury. One pupil may be wider than the other as a congenital abnormality (congenital anisocoria).

The Abnormally Constricted Pupil

Again, drugs are a common cause. Meiotic drops are still widely used for the treatment of chronic simple glaucoma and the constricted pupils of the morphine addict are well known if not so commonly seen. When a constricted pupil on one side is observed it is important to note the position of the eyelids. A slight degree of associated ptosis indicates the possibility of Horner's syndrome. The total syndrome comprises miosis, narrowing of the palpebral fissure due to paralysis of the smooth muscle in the eyelids (Müller's muscle), loss of sweating over the affected side of the forehead and a slight reduction of the intraocular pressure. Horner's syndrome may be caused by a wide diversity of lesions anywhere along the sympathetic pathway but quite often it is noted in the elderly as an isolated finding and investigation fails to reveal a cause. The Argyll Robertson pupil is a very rare but famous example of the meiosed pupil which responds to accommodation but not to direct light. This type of pupil reaction was originally described as being closely associated with syphilis of the central nervous system.

Double Vision

Double vision (diplopia) may be monocular or binocular. Monocular diplopia,

that is, diplopia that is still present when one eye is closed, is quite common and is usually due to cataract. Some patients say that they can see double when they mean that the vision is blurred. Double vision of recent onset should always be treated as a serious symptom. It is usually disabling, preventing the patient from working or even walking about. Some patients discover that the symptoms are relieved by placing a patch over one eye. Slight degrees of double vision may be compensated by a head tilt or turn and the nature of the adopted head posture can help to identify the cause of the double vision. In the same way, if the history is elucidated carefully, noting, for example, whether the diplopia is worse for near or distant vision or whether there is horizontal or vertical displacement of the second image, then a possible cause may be suspected even before examining the patient.

Normal Eye Movements

The complaint of double vision suggests that the separate eyes are not both fixed on the point of regard. The eye which is 'off line' sees the object of regard but it appears displaced. This failure of the eyes to work together is due to malfunction of one or a group of eye muscles or the neurological mechanisms which control them.

From the clinical point of view it is convenient to divide the eye muscles into horizontal and vertical groups. The horizontal muscles, the medial and lateral recti, are easy to understand because their actions are in one plane and they simply adduct (turn in) or abduct (turn out) the globe. The vertical recti are best considered as having primary and secondary actions. It is important to realise that the action of the vertical recti changes with the position of the globe. For example, when the eye is abducted the superior rectus elevates the

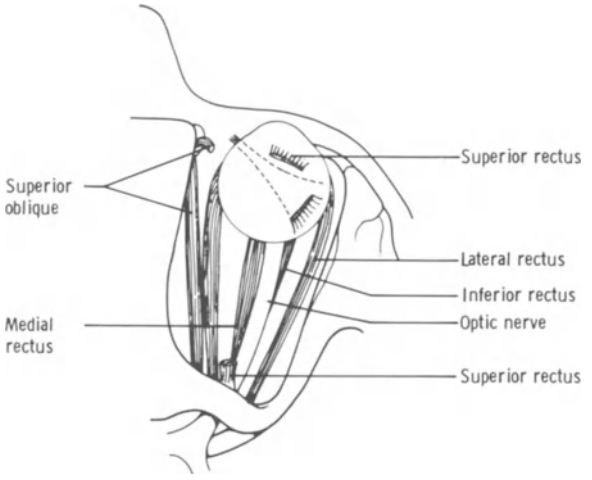

Fig. 21.8. The extraocular muscles.

globe, but when the eye is adducted the superior rectus rotates the eye inwards round an anterior–posterior axis (intorts). In a similar manner the inferior oblique elevates the adducted eye and extorts the abducted eye (Fig. 21.8). In order to test the action of the superior oblique muscle, one must first ask the patient to adduct the eye and test for depression in adduction. That is to say, a superior oblique palsy prevents the eye from looking down when it is turned in. The main line of action of the vertical recti is seen when the eye is abducted and that of the obliques is seen when the eye is adducted.

Examination of a patient with double vision entails first of all testing the gross eye movements in the cardinal positions of gaze and then noting the degree of separation of the images in these various positions. The Hess chart is one of several ingenious methods of recording the abnormal eye movements: the principle is to place a green filter before one eye and a red filter before the other and to ask the patient to look at a screen on which are placed a number of small illuminated white dots. The patient is then asked to localise the dots with a pointer. The amount of false localisation can then be measured in all positions of gaze. This technique is invaluable when assessing the recovery of an ocular muscle palsy.

Young children adapt to double vision very rapidly by suppressing the image from one eye, and under the age of 8 years the suppression may lead to more permanent amblyopia if the situation is not relieved. In adults the double vision may persist and be disabling for months or even years if not treated by incorporating prisms into the spectacles or by muscle surgery.

Causes

Ocular Muscle Imbalance. It will be recalled from the chapter on squint that some patients have a latent squint which is controlled much of the time but sometimes becomes overt. A typical example is the hypermetrope with esophoria who begins to complain of double vision when working for an examination. This problem may be solved simply by prescribing suitable spectacles. Sometimes anxious patients who have had a squint since childhood begin to notice their double vision again, having suppressed one image for many years. The symptoms are usually relieved with the cause of the anxiety.

VIth Cranial Nerve Palsy. The affected eye is converged due to weakness of the lateral rectus muscle. It occurs most commonly as an isolated episode in hypertensive elderly patients and heals spontaneously in 3–6 months. Elderly diabetics are also more prone to VIth cranial nerve palsies. In young patients the possibility of multiple sclerosis or even raised intracranial pressure must be borne in mind.

IVth Cranial Nerve Palsy. The eye fails to look down when it is turned in and may be turned slightly up when the other eye is looking straight ahead. Trauma (a blow over the eye) is an important cause in younger patients but a full investigation for an intracranial space-occupying lesion is usually needed.

IIIrd Cranial Nerve Palsy. The eye is turned out and slightly down, the pupil is dilated and ptosis is usually severe enough to close the eye. Trauma is an important cause in young people but carotid aneurysm and diabetes should also be considered.

Thyrotoxicosis. Patients with this condition develop double vision because the extraocular muscles become infiltrated with inflammatory cells. The action of the inferior recti in particular becomes impaired and diplopia on upward gaze is a common sign. Spontaneous recovery tends to occur over a period of years but muscle surgery may be required.

Myasthenia Gravis. This disease presents sometimes with diplopia with or preceded by ptosis which becomes worse as the day goes by. Any one muscle or group of muscles may be affected. The symptoms and signs show a transient improvement seconds after the intravenous injection of edrophonium chloride (Tensilon).

Blow-Out Fracture of the Orbit. A special cause of double vision following injury is the trapping of extraocular muscles, usually the inferior rectus, in the line of fracture. The patient experiences double vision on looking upwards and the limitation of movement is evident.

22 Genetics and the Eye

Many types of eye disease are inherited and it is always important to enquire about the family history when interviewing a patient with ophthalmological complaints. Some types of inherited eye disease lead to blindness and relatives of patients with such conditions often seek advice concerning their risk of contracting the disease.

Genetic Mechanisms

In order to be able to give advice about the appearance of inherited disease in future generations, it is essential to have a knowledge of the mechanism of genetic transmission.

The nucleus of each cell in the body contains 46 chromosomes arranged as 23 pairs. The 23rd pair comprises the sex chromosomes (the remainder being known as autosomes). These are responsible for the transmission of sex characters but also carry a number of other genes unrelated to sex. In a woman the sex chromosomes are the same length but in a man one is shorter than the other. The shorter one is known as the 'Y' chromosome and the longer one, which is the same as the female sex chromosome, is the 'X' chromosome. When the sperm or ova are formed in the body, the pairs of chromosomes separate and the nuclei of the gametes (i.e. sperm or ova) contain only 23 chromosomes. When fertilisation occurs the 23 chromosomes from each gamete reunite as pairs. Genetic material is thus equally provided from each parent. Genes are discoid elements arranged along the length of a chromosome and each one is known to bear special influence on the development of one or more individual characteristics. Genes are arranged in pairs on adjacent chromosomes. The two genes of the pair may be similar (homozygous) or different (heterozygous). If different, one may exert an overriding influence and is said to be dominant. The gene which is overridden is said to be recessive.

Pathological genes can carry abnormalities which are transmitted to the offspring in the same way as other normal characteristics. In a given individual, the abnormal gene may be recessive and masked by the other one

of the pair. The individual would thus not appear to have the disease but could transmit it. The three important patterns of inheritance are:

1. Autosomal recessive
2. Autosomal dominant
3. Sex-linked recessive

Autosomal Recessive Inheritance

If an abnormal recessive gene is paired with another abnormal one on the opposite chromosome, it will have an effect, but if the opposite gene is normal, the abnormality will not become manifest. Recessive disease in clinical practice usually results from the mating of heterozygous carriers. If the abnormal gene is represented by 'a', then the disease will appear in the individual with genetic configuration 'aa' (homozygote) and not with the configuration 'aA' (heterozygote). When two heterozygotes mate the likely offspring can be considered as in the diagram (Fig. 22.1). If a patient has recessively inherited disease, his parents are likely to be normal but there may be brothers or sisters with the disease. It is important to enquire whether the parents are blood relatives because this greatly increases the likelihood of transmission. If an individual with recessive disease marries someone with the same recessive disease, then all the offspring will be affected. If one spouse is a carrier and the other has the disease, then there is a risk that 50% of the offspring would be carriers and 50% would be affected. When a carrier

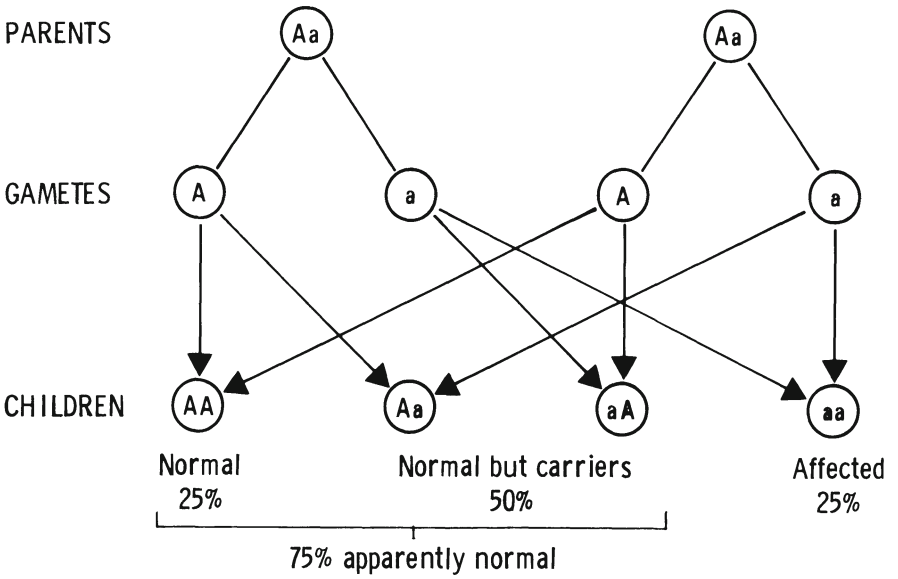

Fig. 22.1. Recessive inheritance.

marries a normal individual, 50% of the offspring are carriers. These expected findings can be calculated quite easily using the type of diagram shown in Fig. 22.1.

Autosomal Dominant Inheritance

When a gene bearing a defect or disease gives rise to the disease even though the other one of the pair is normal, it is said to be dominant. An affected heterozygote may therefore have 50% of affected children when married to a normal spouse. Of course, if both spouses carry the abnormal dominant gene, then all the offspring will be affected. Dominant inheritance can only be shown with certainty if three successive generations show the disease and if about 50% of individuals are affected. Also one sex should not be affected more than the other (Fig. 22.2).

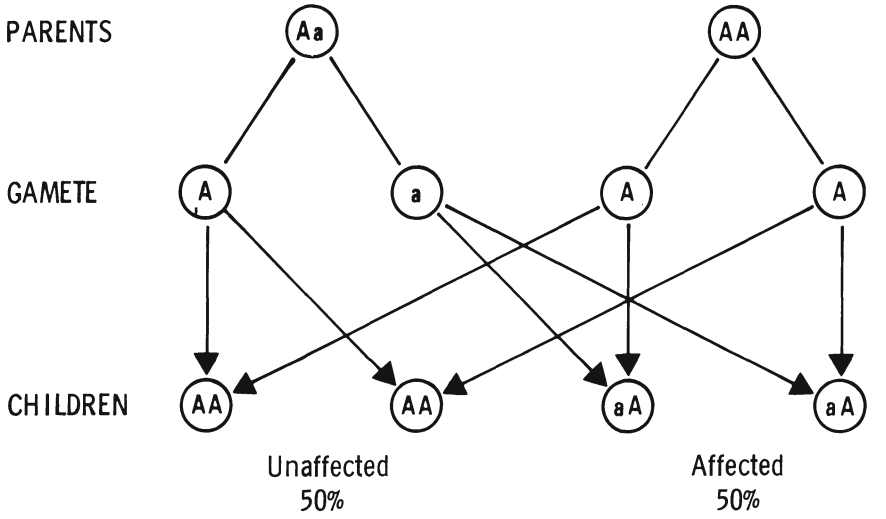

Fig. 22.2. Dominant inheritance.

Sex-Linked Recessive Inheritance

It has been mentioned already that males have the 'XY' configuration of sex chromosomes whereas females have 'XX'. Because of the unpaired nature of much of the male sex chromosomes, some recessive genes may have an effect in males when they do not do so in the female. Certain important eye conditions are carried in this way in pathological genes on the X chromosome and the pattern of inheritance is termed X-linked recessive. Examples of this type of inheritance are seen in ocular albinism and colour blindness. Retinitis pigmentosa may also show this pattern in some families. When inheritance is

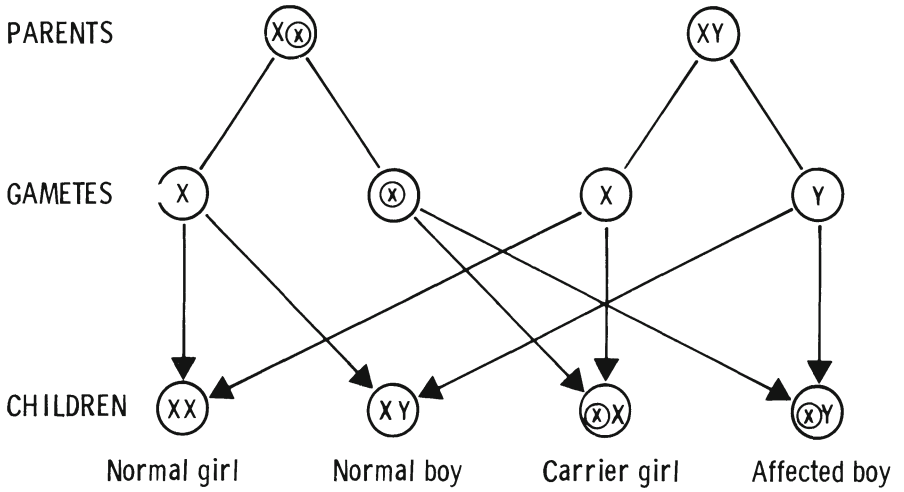

Fig. 22.3. X-linked inheritance.

X-linked, only males are affected and there is no father-to-son transmission of the disease. Instead it is conveyed through a carrier female to the next generation (Fig. 22.3).

This description of the three important modes of inheritance should make it apparent that it is possible to predict the likely disease incidence in offspring. It should also be realised that such predictions can only be based on careful and extensive investigation of the family. Although some eye diseases are known to follow a fixed pattern of inheritance, others, notably retinitis pigmentosa, may be inherited in different ways in different families. In most large centres genetic clinics are now in existence in which time is devoted specifically to the investigation of families and also to the detection of carriers (Table 22.1).

So far in this chapter we have considered how abnormal genes are inherited. Microscopic studies of the chromosomes themselves have revealed that abnormal numbers of chromosomes may be produced by a fault at the

Table 22.1 Examples of types of genetic disease

Autosomal dominant	*Autosomal recessive*	*X-linked recessive*	*Chromosomal dysgenesis*
Retinitis pigmentosa	Retinitis pigmentosa	Colour blindness	Down's syndrome
Retinoblastoma	Tay–Sach's disease	Ocular albinism	Turner's syndrome
Congenital glaucoma	Generalised albinism	Retinitis pigmentosa	Klinefelter's syndrome
Marfan's syndrome			Patan's syndrome
Neurofibromatosis			
Sturge–Weber syndrome			
Von Hippel–Lindau disease			

moment of fertilisation. A common example of this is Down's syndrome. Cytogenetic studies have shown that these patients have an extra chromosome which is indistinguishable from chromosome 21. Down's syndrome is more common in children born to older women and the eye changes include narrow palpebral fissures with a characteristic slant, cataract, high myopia and rather intriguing grey spots on the iris known as Brushfield's spots. Brushfield's spots are sometimes seen in otherwise normal individuals. Turner's syndrome (one missing X chromosome) and Klinefelter's syndrome (an extra X chromosome) are further examples of diseases in which there are known to be abnormalities of the chromosomes which are visible under the microscope. People with these last two diseases are of interest to the ophthalmologist on account of the abnormal but predictable manner in which they inherit colour blindness.

23 Drugs and the Eye

Fortunately, because they are absorbed through the cornea, it is possible to achieve a high concentration of most drugs in the eye by applying them in the form of eyedrops. In this way a high local concentration can be obtained with minimal risk of systemic side-effects. Certain drugs may produce systemic side-effects, for example the slowing of the pulse after the use of timolol drops or sweating and nausea after frequent use of pilocarpine. The action of local applications may be prolonged by incorporating them in an ointment or, more effectively, by the use of a plastic slow-release device which can be lodged under the upper lid (for example the Ocusert). For most purposes, eyedrops are prescribed in 10-ml amounts in a multiple-dose container. After the container has been opened it should not be kept for longer than a month. In order to avoid undue stinging, drops may be buffered to near the pH of tears and they contain a preservative such as phenyl mercuric nitrate or benzalkonium chloride. It must be borne in mind that patients who develop an allergic reaction to drops may be reacting to the preservative. Single-application containers are also available and these may not contain preservative.

Eye lotions are usually prescribed in 200-ml quantities and are used to irrigate the conjunctival sac. Sodium chloride eye lotion is used in first aid in order to flush out foreign bodies or irritant chemicals. Fresh mains tap water is an adequate substitute. Antibiotics and steroids are sometimes administered subconjunctivally. This is a good way of achieving high concentrations in the eye but, because only 1 ml can be given at a time, the drug must be sufficiently soluble to be contained in this dose. When an eye becomes infected it is usual to give systemic antibiotics as well as local applications.

Treatment of Infection

It is generally considered better to use local antibiotics which are not in general use for systemic infections elsewhere in the body. Chloramphenicol, for

example, is rarely used for systemic infections because of its side-effects; but it has been used for many years in the form of eyedrops. Other wide-spectrum antibiotics which are used in drop form are gentamicin, neomycin and framycetin. When an infection of the eye is suspected, a culture is taken from the conjunctival sac and treatment started with a wide-spectrum antibiotic. Systemic and subconjunctival administration is essential if the infection is intraocular. A number of antiviral drugs are now available. Trachoma responds well but slowly to tetracycline hydrochloride ointment. Herpes simplex infections of the cornea may be treated with idoxuridine 0.5% ointment applied every 4 hours or vidarabine 3% ointment applied five times daily. The newer antiviral agent acyclovir is now being used with effect for herpes simplex and herpes zoster. It is available as an ointment (Zovirax) and it can also be given orally as tablets.

Drops Which Widen the Pupil

Routine mydriasis to allow examination of the fundus is probably best achieved by the short-acting tropicamide drops. These last for 3 hours, reaching a maximal effect after 10–15 minutes. Cyclopentolate, homatropine and hyoscine last for 24 hours but have a more potent cycloplegic effect, that is, they relax the accommodation and allow more accurate retinoscopy in young subjects. Atropine is a very potent and useful mydriatic which is used when it is necessary to break down or prevent adhesions between the iris and lens in acute iritis. Its effect lasts for 7 days. In rare instances it is possible to induce an attack of acute glaucoma in susceptible subjects by the instillation of a mydriatic. Susceptible subjects are usually over the age of 50, have shallow anterior chambers and are hypermetropic. Pilocarpine drops should be used to reverse the mydriasis in these cases. Atropine and homatropine may cause a type of contact dermatitis on the skin of the eyelids and may also produce toxic systemic effects in some patients.

Drops Which Constrict the Pupil

Meiotics are most commonly used in the treatment of glaucoma; pilocarpine has proved the most useful drug in this respect. It lasts for about 4 hours and although it is very effective in reducing intraocular pressure, the constriction of the pupil and spasm of accommodation cause dimming of vision and myopia. Both these side-effects are maximal after 30 minutes. Physostigmine is more potent and is still occasionally used. Ecothiopate (Phospholine Iodide) is also a potent miotic whose effect lasts for 12 hours. The more powerful miotics can, in addition to their other side-effects, increase the rate of formation of cataracts.

Treatment of Glaucoma

Although in the past pilocarpine has been the routine first-line treatment of chronic open angle glaucoma, the pattern of treatment has changed with the introduction of timolol maleate. This beta-blocking agent can be administered as eyedrops twice daily and by itself can control many cases of chronic glaucoma. It is remarkably free of local side-effects, but systemic absorption may be significant and timolol should be avoided in patients with asthma or heart failure. Adrenaline when given as drops can reduce the intraocular pressure. Miotics act by 'opening up' the trabecular meshwork and allowing the better outflow of aqueous from the eye. Adrenaline and timolol reduce the production of aqueous.

Certain drugs can reduce the intraocular pressure when given by mouth. The most widely used of these is acetazolamide (Diamox). This is a sulphonamide and a carbonic anhydrase inhibitor. It was introduced originally as a diuretic but it became apparent that it is a potent ocular hypotensive agent. Its use is limited by side-effects: most patients taking acetazolamide experience paraesthesiae in the fingers and toes and some feel tired and listless. Elderly people may become disorientated and complain of indigestion. Renal colic is also a recognised complication, especially in younger men, and renal failure has been described. The long-term use of acetazolamide leads to potassium depletion. Acetazolamide is used for the short-term control of acutely raised intraocular pressure and often prior to surgery. Apart from its use in glaucoma, it is also used to soften the eye before cataract surgery.

Local Anaesthesia in Ophthalmology

Proxymetacaine (Ophthaine) is a useful short-acting anaesthetic drop which is comfortable to instil. Amethocaine is also widely used. Cocaine drops are still used when carrying out surgery to the eye under local anaesthesia and they have a much longer lasting effect. Local anaesthetic drops should not be prescribed to patients as pain relievers because the anaesthetic cornea is likely to become ulcerated. Lignocaine with or without adrenaline is injected into the eyelids for lid surgery. Local anaesthesia for intraocular procedures is effected by injecting lignocaine behind the globe. For this technique of retrobulbar injection, a special needle is required which is passed inferior to the globe and then through the cone of rectus muscles.

Drugs and Contact Lenses

As a rule, contact lenses should not be worn when the eye is being treated with

drops. The exception is when the contact lenses themselves are being used for some therapeutic purpose. Soft hydrophilic contact lenses may take up and store the preservative from some kinds of drop. The preservative benzalkonium chloride is especially liable to be absorbed onto a lens. A number of different proprietary preparations are used for the storage of contact lenses. Hydrophilic lenses can be stored in 0.9% sterile saline and the case, lens and solution should be heated at regular intervals to reduce contamination by micro-organisms (daily heating to 80°C for 40 min has been recommended). When it is essential that drops are administered to a patient wearing a soft lens, it is often possible to prescribe in the form of single-dose containers which do not contain preservative.

A number of systemically administered drugs have been suspected of causing contact lens intolerance; however, direct proof is lacking for all of these, although it is widely recognised amongst contact lens practitioners that tolerance is reduced by the contraceptive pill and also by pregnancy.

Artificial Tears

When it has been shown that the production of tears is inadequate, some relief from symptoms can be obtained by the frequent use of artificial tear drops. Hypromellose forms the basis of most such solutions. Liquid paraffin ointment and polyvinyl alcohol are also used. The efficacy of such treatment is often limited by the patient's inability to instil the drops sufficiently frequently. These drops have the advantage of being free of side-effects over long periods of use but they tend to leave a white deposit on the eyelids.

Steroids and the Eye

Local steroids are widely used in the treatment of eye disease; systemic steroids are not used unless the sight of the eye is threatened and they are required on a sight-preserving basis. It must be remembered that systemic steroids give the patient a sense of well-being which may give a false impression of the real benefit obtained. Furthermore, systemic steroids can have serious and life-threatening side-effects (Fig. 23.1).

Local steroids should also be applied with caution, and it is a good rule always to have a specific reason for giving them. That is to say, they should not be prescribed just to make red eyes turn white without a clear diagnosis. The reasons for this are twofold: first, local steroids enhance the multiplication of viruses, especially herpes simplex; second, they can cause glaucoma in certain predisposed individuals. In such individuals, the instillation of one drop of steroid may cause a temporary rise in intraocular pressure. The most potent steroid in this respect is dexamethasone followed by

Fig. 23.1. "Steroids give a patient a feeling of well-being."

betamethasone, prednisolone and hydrocortisone. It has been claimed that clobetasone and fluorometholone are relatively safe in this respect.

Damage to the Eyes by Drugs Administered Systemically

It has been mentioned that steroids may cause glaucoma; systemic steroids may also lead to cataract formation. Pilocarpine drops are also thought to hasten the onset of a cataract. It is important to realise that the long-term use of maximum doses of chloroquine can lead to irreversible blindness. Smaller doses of this drug produce reversible corneal opacities but if more than a total of 100 g is given in any one year, then the risk of retinal degeneration becomes significant. Many other drugs have been shown to cause specifically ocular side-effects: ethambutol may cause retinal oedema and optic atrophy, chlorpromazine in large doses can cause cataract. The list of drugs with ocular

side-effects is large and the reader should consult a specialised textbook for more information.

Further Reading

Caird FI, Pirie A, Ramsek TG (1968) Diabetes and the eye. Blackwell, Oxford
Duke-Elder Sir Stewart (1962) System of ophthalmology, Vol. III: The foundations of ophthalmology (hereditary, pathology, diagnosis and therapeutics). Kimpton, London
Duke-Elder Sir Stewart (1963) System of ophthalmology, Vol III: Normal and abnormal development, Part 1 — Embryology; Part 2 — Congenital deformities. Kimpton, London
Duke-Elder Sir Stewart (1966) System of ophthalmology, Vol. IX: Diseases of the uveal tract. Kimpton, London
Ellis PP (1981) Ocular therapeutics and pharmacology, 6th edn. CV Mosby, London
Garner A, Klintworth GK (1982) Pathology of ocular disease. Marcel Dekker, New York
Glaser JS (1978) Neuro-ophthalmology. Harper & Row, Hagerstown
Krill E, Archer DB (1977) Krill's hereditary retinal and choroidal diseases. Harper & Row, Hagerstown
L'Esperance FA, James WA (1981) Diabetic retinopathy. CV Mosby, St Louis
Lim AS, Constable J (1979) Colour atlas of ophthalmology. Kimpton, London
Michaelson IC (1980) Textbook of the fundus of the eye. Churchill Livingstone, New York
Miller N, ed. (1982) Walsh & Hoyt's neuro-ophthalmology, Vol. 1. Williams & Wilkins, Baltimore
Peyman GA, Sanders DR, Goldberg MF (1980) Principles and practice of ophthalmology, Vols II and III. Saunders, Philadelphia

Section V

Blindness

24 Blindness

Blindness marks the failure or inefficacy of ophthalmological treatment. Once a patient becomes permanently blind, he or she may be lost from the care of the ophthalmologist. This means that the ophthalmologist may not have personal experience of the size of the problem and may not be in a position to experience the relative incidences of different causes of blindness. The keeping of accurate statistics is of great importance, and in order to keep statistical records it is necessary to have a clear definition of blindness. Many people who dread blindness imagine having no perception of light in each eye. Fortunately this situation is uncommon, but many people are severely debilitated by visual loss.

Definition

In Britain the statutory definition of blindness refers to persons who 'are so blind as to be unable to perform any work for which eyesight is essential'. It is generally accepted that if the visual acuity is worse than 6/60, the patient will fit into this category, but allowance is made for those who are able to see to work in spite of such poor acuity and also for those who cannot work when their visual acuity may be rather better than 6/60. It is recognised by law that other factors such as the state of the visual field or the age of the patient must be taken into account. If the ophthalmic surgeon finds that a patient comes under this definition of blindness, then he is obliged to complete a form which is processed by the local community health and social services departments. Once registered blind, a patient is entitled to additional income tax relief and the blind welfare services become available to him. Patients who are not blind but suffer significant visual handicap may be registered as 'partially sighted'. They may also make use of the blind welfare services if this is felt to be necessary. Patients sometimes erroneously claim the benefits of the partially sighted because they have only one eye, even though the other eye is normal. When the vision with one or both eyes is 6/18 or better, the patient is not

usually considered to be partially sighted. When one eye is completely lost through injury or disease, then the amount of incapacity is set for medicolegal purposes at about 10%. In actual fact the amount of incapacity depends a great deal on the age of the patient. A child may adapt to a remarkable degree to being one eyed, even to the extent of being able to perform with skill at ball games. Adults who become one eyed find difficulty in judging distances or performing fine manual tasks. More importantly, a number of occupations are specifically barred to those whose vision is poor in one eye.

Standards of Vision for Various Occupations

The statutory definition of blindness refers to 'any work for which eyesight is essential'. Failure to comply with the requirements of a specific occupation does not, of course, indicate that the person is blind or partially sighted. The standards for various occupations can vary from year to year and are more or less exacting, depending on the occupation. In Britain, in order to drive a private motor vehicle, it is necessary to be able to read a number plate containing letters 3 1/2 inches high at a distance of 75 feet. In order to drive a heavy goods vehicle, the new applicant for a licence must have a visual acuity of at least 6/12 in the better eye and 6/36 in the worse eye.

Colour Blindness

This is not blindness in any sense of the word and indeed some colour blind individuals are unaware of any problem until their colour vision is tested. The screening of schoolchildren for colour blindness is now widely practised because of the occupational implications. Occupations which entail the reading of coloured warning lights or the matching of colours usually demand some form of colour vision test on entry. It is an advantage to the child to be aware of any defect during the early years of schooling.

Incidence and Causes

In the western world, the incidence of blindness varies from 50 to 200 blind persons per 100 000 of the population — an incidence of less than 1%. The figures vary considerably from country to country but this may be due partly to variations in the definition of blindness and the method of statistics. The causes of blindness also vary depending on climate and living conditions. In Britain the commonest cause of blindness in young people is diabetic retinopathy. For the elderly, the commonest cause is senile macula degeneration. The incidence of blindness from glaucoma is on the decline due

to earlier recognition of the disease. Cataract is an extremely rare cause of blindness because of the free availability and the high success rate of surgery. Although at one time ophthalmia neonatorum took a significant toll of eyesight, blindness from this cause is now prevented by the use of antibiotics, and blindness from other forms of infection is also very rare. In other countries, particularly India and Africa, infection still has a major role as a cause of blinding disease. Trachoma is still the commonest cause of blindness in the world, accounting for about 2 million blind people. Onchocerciasis (river blindness) is endemic in tropical Africa and South America. It is due to a roundworm which invades the eye as well as the rest of the body and is conveyed to the host by the blackfly. The third common cause of blindness in the world is xerophthalmia due to lack of vitamin A.

It is apparent that the problems of blindness in Europe and North America are very different from those in poorer parts of the world where much could be done by improving nutrition and living conditions.

Aids for the Blind

The most widely recognised aid and symbol of blindness is the white stick. It is also one of the most useful aids because it identifies the patient as blind and encourages others to give him assistance. Many blind people are concerned that they appear ill-mannered when failing to recognise someone and are grateful for some indication of their handicap. The long cane is simply a longer stick which can be used as a feeler. It has been used in Britain for 20 years and has been helpful in allowing some completely blind individuals to obtain better mobility. A range of electronic devices has been tried but these have not so far gained acceptance amongst blind people. Most of these devices rely on the patient's ears to hear an audible warning signal, but blind people usually prefer to use their undistracted sense of hearing as an important clue to their whereabouts. Another factor is that many blind people are elderly and find it difficult to cope with aids of this kind. Guide dogs are expensive to train and the patient must also take part in the training. Some young people find that a guide dog can expand their mobility to a great degree. Certain tactile aids are also useful, the best known of which is Braille. This system of reading for the blind was introduced from France more than 100 years ago. The letters of the alphabet are represented by numbers of raised dots on stiff paper. Blind children can learn Braille very rapidly and develop a high reading speed. Some adults find that their fingers are not sufficiently sensitive and this applies especially to diabetics. Books in Braille are now available in many different languages. Tape recordings of books are now becoming more popular than the older tactile methods of assimilation and the Talking Book Service provides a comprehensive library for the use of blind and partially sighted persons.

There are numerous other gadgets which can be helpful to the blind and partially sighted; a popular one is the device that can indicate whether a teacup is full or not. For those with some residual vision, a special telephone dial with

large numbers on it can be very helpful. Other ingenious devices range from relief maps which can be felt by the blind person to a telephone which speaks back through the earpiece the digit that has just been dialled. Research has also been carried out on aids which signal the position of objects by means of electrical stimuli to the skin and even by means of implanted electrodes in the visual cortex.

When the patient has a visual acuity of better than 6/60, then much can be achieved by the use of optical magnification. A suitable hand magnifying glass is the simplest and may often be the most effective form of assistance. If this is not adequate and the patient has been a keen reader, then a telescopic lens may be fitted to a spectacle frame with advantage. Apart from special telescopic lenses, closed circuit television aids are now available: a small television camera is held over the page and a magnified view of the written material is then presented on a television screen.

The well-being of a blind or partially sighted person may be greatly aided by relatively simple social measures. Advice in the home about the use of gas or electricity may be important and the patient can be made aware of the availability of local social clubs for the blind or keep-fit classes and bus outings. An elderly patient who plays the piano may be helped by the provision of an enlarged photocopy of a favourite piece of music. In spite of all these various possibilities, one must not forget that the simplest and most useful reading aid for a partially sighted person is a good light.

Artificial Eyes

These may be made of glass or plastic moulded to the shape of the eye socket and painted to match the other eye. Usually they are removed and washed at night by the patient and replaced the following morning. A slight degree of discharge from the socket is the rule but excessive discharge may indicate that the socket is infected, which may be due to roughening of the artificial eye with wear. Under these circumstances arrangements should be made for the eye to be polished and a course of local antibiotic treatment provided. It should always be borne in mind that a patient with an artificial eye may have had one eye removed because it contained a malignant tumour, in which case one must consider the possibility of local or systemic spread of the tumour. Alternatively the eye may have been removed through fear of sympathetic ophthalmia following injury; here the possibility of inflammation in the good eye must be considered. A well-made artificial eye is almost undetectable to the untrained eye but normal movement of the eye is restricted. When an eye has to be removed it is possible to stitch the eye muscles to an acrylic implant which remains buried under the conjunctiva. This provides movement to the posterior wall of the socket and consequently to the artificial eye placed in the socket. Such implants may occasionally extrude and they are not usually required in elderly patients when the demand for a fine cosmetic result is not so great.

Surgical removal of an eye (enucleation) is considered in the following circumstances:

1. When the eye is blind and painful
2. When the eye contains a malignant tumour
3. When the eye is nearly blind and sympathetic ophthalmia is a risk following a perforating injury

Removal of an eye requires a general anaesthetic and the patient remains in hospital for 4 days to 1 week. Two days or more after the operation the socket, which is lined with conjunctiva, is fitted with a plastic 'shell'. This is used to shape the socket until the artificial eye is fitted 3 or 4 weeks later.

Further Reading

Dobree JH, Boulter E (1982) Blindness and visual handicap: the facts. Oxford University Press, New York Oxford.

Subject Index