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Common Eye Diseases and their Management

Third Edition

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With 146 Figures

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Preface to Third Edition

It is a pleasure to welcome two new authors who have contributed to the third edition of "Common Eye Diseases": Peter Galloway and Andrew Browning. Six years have passed since the last edition but even in this relatively short time there have been significant advances in the diagnosis and management of eye disease and an update has become necessary. Each author has taken a block of chapters for revision and, where needed, illustrations have been added or removed. Apart from the four main authors, I am indebted to Mr Roland Ling for his invaluable work on the chapter on the retina and once again to Professor Rubinstein for his help with the chapter on contact lenses.

The original aims of the book have not been changed. It remains as a textbook for medical students and those starting a career in ophthalmology, but also for those in primary care who are likely to deal with eye problems, including nurses, optometrists and general practitioners.

It has been the intention to keep explanations as simple and nontechnical as possible without losing scientific accuracy; more detailed accounts should be sought in the larger textbooks. An updated reference list for further reading is given at the end of the book. An internet version of this edition is being planned and, in order to keep down the retail price, some financial help is needed. For this we are grateful for the interest of Pfizer Ltd, whose policy of educational support has allowed this edition to go forward at its present low price.

Acknowledgements

Although it is now many years since the first edition appeared, I still owe a great debt to my former secretary, Mrs A. Padgett, for her original help in preparing the basis for these further editions. No amount of word processing can replace this painstaking work. In this new edition, I have kept Geoffrey Lyth's original cartoons, which will perhaps lighten the heaviness of the text for those with an artistic bent. The two new authors have revised a number of chapters and their fresh input to an ageing textbook has been essential and much appreciated.

Finally, I would like to acknowledge the help and encouragement from Melissa Morton of Springer-Verlag, who has kept the ball bouncing back into my court with great efficiency and thereby played an important part in ensuring the birth of this new edition.

Preface to Second Edition

Like the first edition, this textbook is intended primarily for medical students, but it is also aimed at all those involved in the primary care of eye disease, including general practitioners, nurses and optometrists. The need for the primary care practitioner to be well informed about common eye conditions is even more important today than when the first edition was produced. A recent survey from North London has shown that 30% of a sample of the population aged 65 and over are visually impaired in both eyes and a large proportion of those with treatable eye conditions were not in touch with eye services. It is clear that better strategies for managing problems of eyesight need to be set up. One obvious strategy is the improved education of those conducting primary care and it is hoped that this book will contribute to this. For this second edition, I am grateful for the help of my coauthor Winfried Amoaku, whose personal experience in teaching medical students here in Nottingham has been invaluable. His expertise in the management of macular disease, now a major cause of sensory deprivation in the elderly, is also evident in these chapters.

The format of the book has not changed but some of the chapters have been expanded. For example, there is now a section dealing with the eye complications of acquired immune deficiency syndrome (AIDS). This problem barely existed at the time of the first edition. Cataract surgery has changed a great deal in this short time and is becoming one of the commonest major surgical procedures to be performed in a hospital. The management of glaucoma has also changed with the introduction of a range of new medications. Our aim has been to keep the original problem-oriented layout and to keep it as a book to read rather than a book to look at. There are a number of good atlases on eye disease and some of these are mentioned in the section at the end on further reading. Although the title of the book is "Common Eye Diseases", some less common conditions are mentioned and it is hoped that the reader will gain some overall impression of the incidence of different eye diseases.

Contents

1

The Scope of Ophthalmology

Although the eye and its surrounding structures would seem to provide an ideal anatomical and functional basis for specialisation, ophthalmology can no longer regard itself as a specialty on its own but more the heading for a group of subspecialties. There are those who know all about the pigment epithelium of the retina and yet bow to those who have a special knowledge of the bipolar cells in the retina. Over the past 100 years the science has advanced at an unbelievable rate and with the increase in our knowledge has come the development of treatments and cures, which have had a great impact on our everyday lives.

The importance of the eye and its function is sometimes underrated, but a consideration of the part played by vision in our consciousness makes us soon realise its value. If we think of dreams, memories, photographs and 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 to us in the clinic. Nevertheless, in a modern European community the effects of blindness are not so apparent as in former years, and blind people tapping their way about the street or begging for food are less in evidence to remind us of the deprivation that they suffer. This is due to the effective application of preventive medicine and the efficacy of modern surgical techniques. However, in the western world we have a new and increasing problem related to the increasing number of elderly people in the population. The problem is that of sensory deprivation owing to degenerative disease. Degenerative changes in the eye are now a major cause of blindness and although support services are being developed there is still no effective cure.

The broad and detailed scientific interest in the eye and vision is witnessed by the large number of journals, conferences and meetings that now exist, possibly more than in any other specialty. There are several hundred ophthalmological journals all contributing to the scientific literature on the subject and many are now accessible through the internet or on CD-ROM. As an organ of clinical specialisation, the eye does have a special advantage; it can be seen. Using the slit-lamp microscope it is possible to examine living nerves, including nervous system tissues and blood vessels, in a manner that is not possible in other parts of the body without endoscopy or biopsy. So much are the component parts of the eye on display to the clinician that when a patient presents to a casualty department with symptoms, the explanation of the symptoms should be made evident by careful examination. Compare this with the vague aches and pains that present to the gastroenterologist or the neurologist, symptoms that might 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 or she must look

again, because it is likely that something has been missed.

Most of the work of the ophthalmologist is necessarily centred on the globe of the eye itself, and there are a number of conditions that are limited to this region without there being any apparent involvement of the rest of the body. Ophthalmology is usually classified as a surgical specialty but it provides a bridge between surgery and medicine. Most of the surgery is performed under the microscope and here the application of engineering principles in the design of finer and finer instruments has played an important part. There is overlap with the fields of the plastic surgeons and the neurosurgeons. On the medical side, the ophthalmologist has links with the physicians and particularly the diabetic specialists and cardiologists, not to mention paediatricians and dermatologists.

Historical Background

In 1847, the English mathematician and inventor Charles Babbage showed a distinguished ophthalmologist his device for examining the inside of the eye, but unfortunately this was never exploited and it was not until 1851 that Hermann von Helmholtz published his classic description of his instrument, the ophthalmoscope. He developed the idea from his knowledge of optics and the fact that he had previously demonstrated the "red reflex" to medical students with a not dissimilar instrument. In principle, he had, for the purposes of his demonstration, looked through a hole in a small mirror, which reflected light from a lamp into the subject's eye. This produced the red reflex in the pupil well known to photographers and night drivers and no doubt this fascinated medical students at that time. Von Helmholtz worked out that a similar device could be used to inspect the inside of the eye. According to correspondence of the time, it took him about a week to learn the technique of examining in detail the structures within the eye and he wrote a letter to his father telling him that he had made a discovery that was "of the utmost importance to ophthalmology". Soon after this, a mass of descriptive information on the optic fundus appeared in the scientific literature and modern clinical ophthalmology was born. The changes in the eye associated with systemic diseases such as hypertension and anaemia became recognised. Several blinding conditions limited to the eye itself, such as glaucoma and macula degeneration, were also described at this time.

But we must not belittle the developments that had occurred before the invention of the ophthalmoscope. In the eighteenth 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 better correction of refractive errors in the eye. If we go back to the seventeenth century, the existing ophthalmological services were definitely limited, as 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.

Although records of eye surgical techniques go back as far as 3000 years, modern eye surgery was largely developed thanks to the introduction of cocaine and then of general anaesthesia at the end of the nineteenth century. The use of eserine eye drops to reduce the intraocular pressure in glaucoma was introduced at the same time, this being the forerunner of a number of different medical treatments that are now available. Cataract surgery saw great advances at the beginning of the twentieth 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-part" 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, as will be seen in Chapter 11. 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. This

in turn has led to small incision techniques and sutureless surgery, which has made the day-case cataract operation a routine. Forty years ago, the vitreous was a surgical no-man's land, but instruments have now been developed that can cut, aspirate and inject fluid simultaneously, all these procedures being carried out through fine-bore needles. Membranes, blood or foreign bodies can now be removed from the vitreous as a routine. Much important eye disease is inherited and it is not surprising that very important advances have occurred recently in the field of ophthalmic genetics. The gene controlling the development of the eye has now been identified and perhaps the answer to the tragic problem of inherited degenerative retinal disease is on the horizon.

In the early days of the development of the specialty, a number of specialised hospitals were built throughout the UK. 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 this country, often the result of pressures of local needs such as the treatment of industrial accidents. In recent years, there has been a tendency for eye departments to become incorporated within the larger district general hospitals, although individual eye hospitals remain and are still being built.

Making a Career in Ophthalmology

Ophthalmology is a popular specialty and so the aspiring eye surgeon can expect considerable competition. There are certain essential requirements. First, an initial interest in physics and optics is helpful and most important is a considerable degree of manual dexterity. Good binocular vision goes along with the manual dexterity demanded by microscopic surgery. That is to say, the future surgeon should see well out of each eye and should be able to use the eyes together to give proper stereoscopic vision.

In many cases, an interest in the subject is aroused in medical school by a mentor or a good teacher. By and large, those who see ophthalmology as a soft option are not happy in their career. Those who, as most doctors do, set out to improve the lot of the patient, find the specialty very rewarding because it is undoubtedly extremely effective in this respect.

In the UK, medically qualified graduates can start their eye training with a senior house officer (SHO) job and thence look for a specialist registrar post in one of the training centres. A question sometimes asked is what jobs as an SHO, other than ophthalmology, are best suited to an eventual career in ophthalmology. Obvious ones are in plastic surgery, neurology or neurosurgery but sometimes a seemingly unrelated one can prove to be good experience. The membership part of the FRCOphth qualification is needed at this point and once on the training ladder there is an exit examination before training is completed. The rules about training arrangements can vary from time to time and advice on this can be provided by the Royal College of Ophthalmologists. A handbook for trainees is supplied by the college on application. When the doctor is fully trained, he or she can decide whether to start applying for consultant posts or whether to gain a fellowship in a subspecialty and perhaps obtain a higher degree. At the present time consultant posts are often advertised as requiring some special expertise, such as paediatric ophthalmology or retina surgery.

2

Basic Anatomy and Physiology of the Eye

Introduction

The eye is the primary organ of vision. Each one of the two eyeballs is located in the orbit, where it takes up about one-fifth of the orbital volume (Figure 2.1). The remaining space is taken up by the extraocular muscles, fascia, fat, blood vessels, nerves and the lacrimal gland.

The eye is embryologically an extension of the central nervous system. It shares many common anatomical and physiological properties with the brain. Both are protected by bony walls, have firm fibrous coverings and a dual blood supply to the essential nervous layer in the retina. The eye and brain have internal cavities perfused by fluids of like composition and under equivalent pressures. As the retina and optic nerve are outgrowths from the brain, it is not surprising that similar disease processes affect the eye and central nervous system. The physician should constantly remind himself or herself of the many disease conditions that can simultaneously involve the eye and the central nervous system.

Basic Structure of the Eye and Supporting Structures

The Globe

The eye has three layers or coats, three compartments and contains three fluids (Figure 2.2).

1. The three coats of the eye are as follows:
   (a) Outer fibrous layer:
      - cornea
      - sclera
      - lamina cribrosa.
   (b) Middle vascular layer ("uveal tract"):
      - iris
      - ciliary body – consisting of the pars plicata and pars plana
      - choroids.
   (c) Inner nervous layer:
      - pigment epithelium of the retina
      - retinal photoreceptors
      - retinal neurons.
2. The three compartments of the eye are as follows:
   (a) Anterior chamber – the space between the cornea and the iris diaphragm.
   (b) Posterior chamber – the triangular space between the iris anteriorly, the lens and zonule posteriorly, and the ciliary body.
   (c) Vitreous chamber – the space behind the lens and zonule.
3. The three intraocular fluids are as follows:
   (a) Aqueous humour – a watery, optically clear solution of water and electrolytes similar to tissue fluids except that aqueous humour has a low protein content normally.
   (b) Vitreous humour – a transparent gel consisting of a three-dimensional

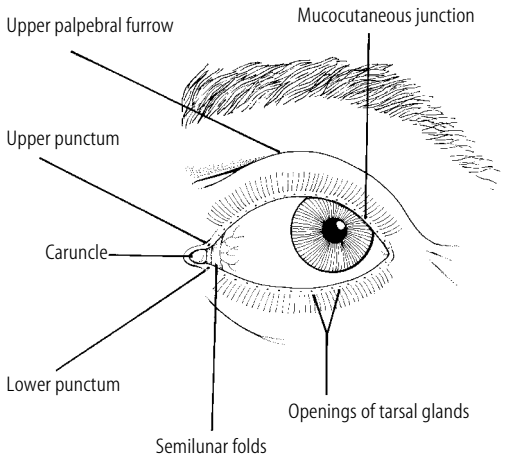


Figure 2.1. Surface anatomy.

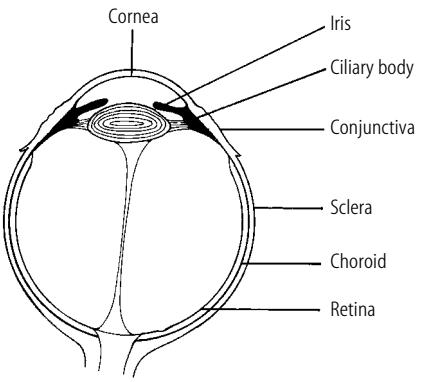


Figure 2.2. Layers of the globe.

network of collagen fibres with the interspaces filled with polymerised hyaluronic acid molecules and water. It fills the space between the posterior surface of the lens, ciliary body and retina.

(c) Blood – in addition to its usual functions, blood contributes to the maintenance of intraocular pressure. Most of the blood within the eye is in the choroid. The choroidal blood flow represents the largest blood flow per unit tissue in the body. The degree of desaturation of efferent choroidal blood is relatively small and indicates that the choroidal vasculature has functions beyond retinal nutrition. It might be that the choroid serves as a heat exchanger for the retina, which absorbs energy as light strikes the retinal pigment epithelium.

Clinically, the eye can be considered to be composed of two segments:

1. Anterior segment – all structures from (and including) the lens forward.
2. Posterior segment – all structures posterior to the lens.

The Outer Layer of the Eye

The anterior one-sixth of the fibrous layer of the eye is formed by the cornea. The posterior five-sixths are formed by the sclera and lamina cribrosa. The cornea is transparent, whereas the sclera, which is continuous within it, is white. The junction of cornea and sclera is known as the limbus. The cornea has five layers anteroposteriorly (Figure 2.3):

1. Epithelium and its basement membrane – stratified squamous type of epithelium with five to six cell layers of regular arrangement.
2. Bowman's layer – homogeneous sheet of modified stroma.
3. Stroma – consists of approximately 90% of total corneal thickness. Consists of lamellae of collagen, cells and ground substance.
4. Descemet's membrane – the basement membrane of the endothelium.
5. Endothelium – a single layer of cells lining the inner surface of Descemet's membrane.

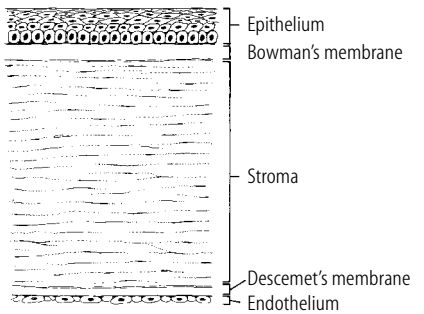


Figure 2.3. The cornea.

In the region of the limbus, the epithelium on the outer surface of the cornea becomes continuous with that of the conjunctiva, a thin, loose transparent nonkeratinising mucous membrane that 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. Although the conjunctiva is continuous, it can be divided descriptively into three parts: palpebral (tarsal), bulbar and fornix.

The sclera consists of irregular lamellae of collagen fibres. Posteriorly, the external two-thirds of the sclera become continuous with the dural sheath of the optic nerve, while the inner one-third becomes the lamina cribrosa – the fenestrated layer of dense collagen fibres through which the nerve fibres pass from the retina to the optic nerve. The sclera is thickest posteriorly and thinnest beneath the insertions of the recti muscles. There is a layer of loose connective tissue deep to the conjunctiva, overlying the sclera, called the episclera.

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, as this middle layer, which is called the uvea, is heavily pigmented as well as being vascular. The anterior part of the uvea forms the bulk of the iris body and hence inflammation of the iris is called either anterior uveitis or iritis. The posterior part of the uvea is called the choroid.

The iris is the most anterior part of the uvea. It is a thin circular disc perforated centrally by the pupil. Contraction of the iris sphincter muscle constricts the pupil, while contraction of the dilator pupillae muscle dilates the pupil.

The ciliary body is part of the uveal tissue and is attached anteriorly to the iris and the scleral spur; posteriorly it is continuous with the choroid and retina. The ciliary body is also referred to as the intermediate uvea.

The ciliary body is triangular in cross-section. The anterior side of the ciliary body is the shortest and borders the anterior chamber angle; it gives origin to the iris. The outer side of the triangle (mainly ciliary muscles) lies against the sclera. The inner side is divided into two zones: (1) the pars plicata forms the anterior 2 mm and is covered by ciliary processes and (2) the pars plana constitutes the posterior 4.5-mm flattened portion of the ciliary body. The pars plana is continuous with the choroid and retina.

The choroid consists of the following:

- Bruch's membrane – membrane on the external surface of the retinal pigment epithelium (RPE). It consists of the basement membrane of RPE cells and choriocapillaris. Between the two layers of basement membrane are the elastic and collagenous layers. Small localised thickenings of Bruch's membrane (which increase with age) are called drusen.
- The choriocapillaris – a network of capillaries supplying the RPE and outer retina.
- Layer of larger choroidal blood vessels external to the choriocapillaris.
- Pigmented cells scattered in the choroid external to the choriocapillaris.

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 pigment epithelium. These same layers can be traced into the retina, which is composed of an outer pigment epithelium and an inner sensory part, which contains the rods and cones, bipolar cells and ganglion cells (Figure 2.4). The junction of the retina and the pars plana forms a scalloped border known as the ora serrata.

It is important to note that the photoreceptor cells are on the external side of the sensory retina. The relationship of the retinal elements can be understood most readily by following the formation of the optic cup. As the single-cell layer optic vesicle "invaginates" to form the two-cell layered optic cup, the initially superficial cells become the inner layer of the cup. The RPE develops from the outer layer of the cup, facing the photoreceptors across the now obliterated cavity of the optic vesicle. The neurons of the sensory retina differentiate from the inner layer of the optic cup.

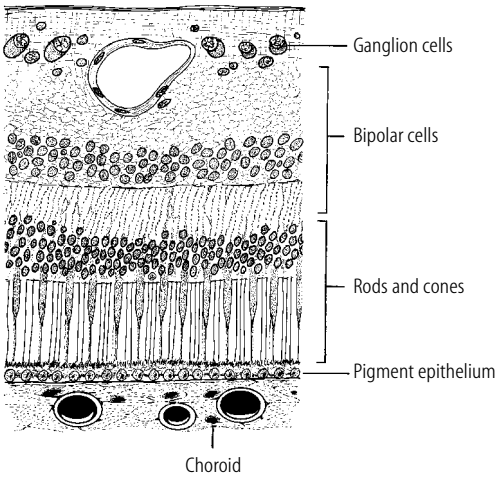


Figure 2.4. The retina.

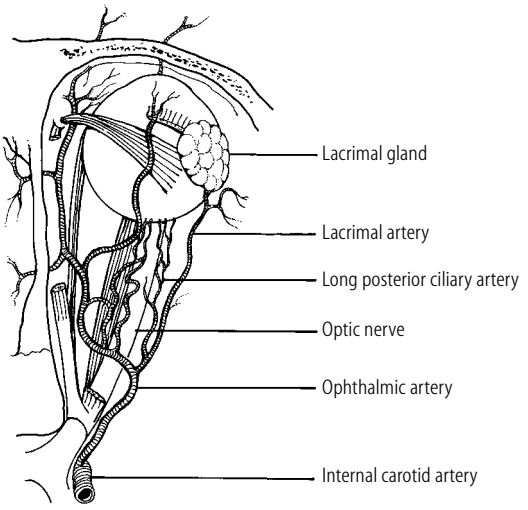


Figure 2.5. Blood supply of the eye.

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 (Figure 2.5).

Optic Nerve

The optic nerve meets the posterior part of the globe slightly nasal to the posterior pole and 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 that 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 (Figure 2.6). Nerve fibres sweep across the innermost part of the retina to reach

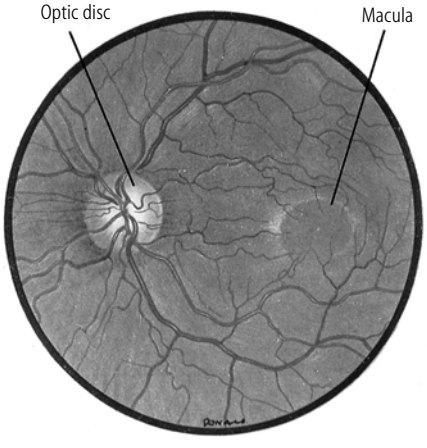


Figure 2.6. The optic fundus.

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 (Figure 2.7). 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 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 muscles). 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 chorioretina – this would seem to prove the existence of sensory fibres in the iris and choroid. The cornea is extremely sensitive, but again, the only sensory endings are those for pain.

The visual pathways include the following:

1. The retina:
   - rods and cones
   - bipolar cells
   - ganglion cells.
2. Axons of the ganglion cells visual and pupillary reflex pathways:
   - nerve fibre layer of retina
   - optic nerve
   - optic chiasm
   - optic tract.
3. Subcortical centres and relays:
   - superior colliculus – reflex control of eye movements
   - pretectal nuclei – pupillary reflexes
   - lateral geniculate body – cortical relay.
4. Cortical connections:
   - optic radiations
   - visual cortex (area 17) – vision and reflex eye movements
   - association areas (areas 18 and 19)
   - frontal eye field – voluntary eye movements.

If the rods and cones are considered analogous to the sensory organs for touch, pressure, temperature, etc. then the bipolar cells may be compared to the first-order sensory neurons of the dorsal root ganglia. By the same token, the retinal ganglion cells can be compared to the second-order sensory neurons, whose cell bodies lie within the spinal cord or medulla.

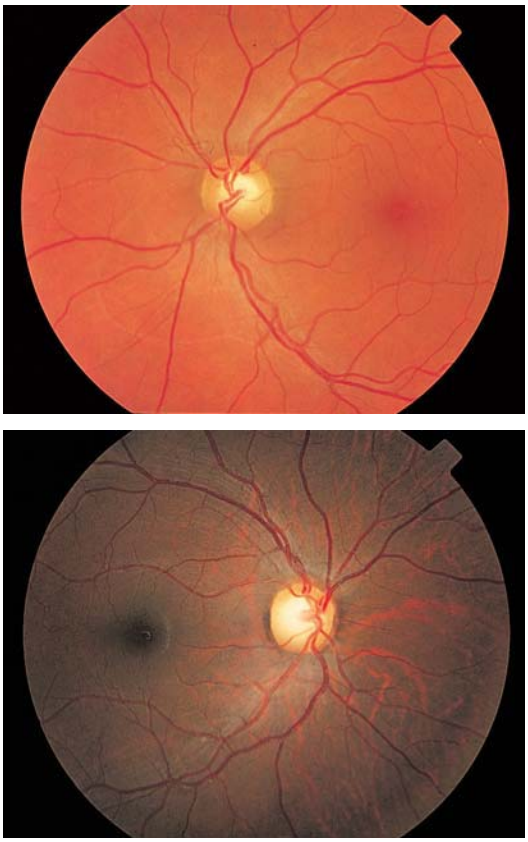


Figure 2.7. The normal fundus of **a** a Caucasian and **b** an African. The background is darker in the African owing to increased pigment in the retinal pigment epithelium (RPE). The nerve fibre layer is noticeable, especially along the superior and inferior temporal arcades. 🕮

The Eyelids

The eyelids may be divided into anterior and posterior parts by the mucocutaneous junction – the grey line (Figure 2.8). The eyelashes arise from hair follicles anterior to the grey line, while the ducts of the meibomian glands (modified sebaceous glands) open behind the grey line. The meibomian glands are long and slender, and run parallel to each other, perpendicular to the eyelid margin, and are located in the tarsal

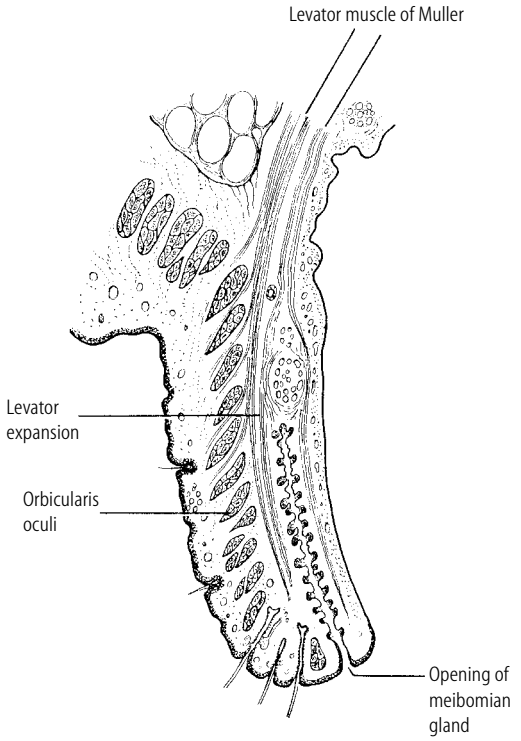


Figure 2.8. The eyelid.

plate of the eyelids. The tarsal plate gives stiffness to the eyelids and helps maintain its contour. The upper and lower tarsal plates are about 1 mm thick. The lower tarsus measures about 5 mm in height, while the upper tarsus measures about 10–12 mm.

The orbicularis oculi muscle lies between the skin and the tarsus and serves to close the eyelids. It is supplied by the facial nerve. The skin and subcutaneous tissue of the lids are thin. The inner surface of the eyelids is lined by the palpebral conjunctiva.

The Lacrimal Apparatus

The major lacrimal gland occupies the superior temporal anterior portion of the orbit. It has ducts that open into the palpebral conjunctiva above the upper border of the upper tarsus.

Tears collect at the medial part of the palpebral fissure and pass through the puncta and the canaliculi into the lacrimal sac, which terminates in the nasolacrimal duct inferiorly. The nasolacrimal duct opens into the inferior meatus of the nose.

The Extraocular Muscles

There are six extraocular muscles that help to move the eyeball in different directions: the superior, inferior, medial and lateral recti, and the superior and inferior obliques. All these muscles are supplied by the third cranial nerve except the lateral rectus (supplied by the sixth nerve) and superior oblique (fourth nerve).

All the extraocular muscles except the inferior oblique originate from a fibrous ring around the optic nerve (annulus of Zinn) at the orbital apex. The muscles fan out towards the eye to form a "muscle cone". All the recti muscles attach to the eyeball anterior to the equator while the oblique muscles attach behind the equator. The optic nerve, the ophthalmic blood vessels and the nerves to the extraocular muscles (except fourth nerve) are contained within the muscle cone (Figure 2.9).

The levator palpebrae superioris is associated with the superior rectus. It arises from just above the annulus of Zinn, runs along the roof of the orbit overlying the superior rectus and attaches to the upper lid skin and anterior surface of the tarsal plate of the upper lid. Tenon's capsule is a connective tissue covering that surrounds the eye and is continuous with the fascial covering of the muscles.

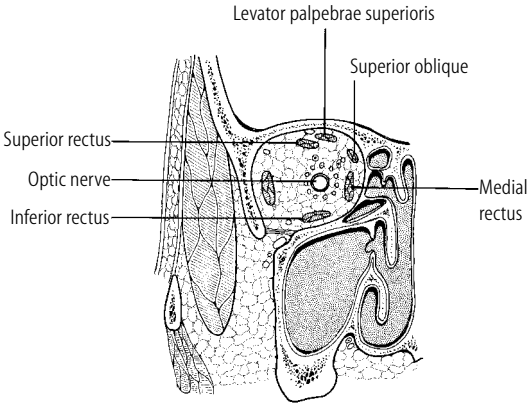


Figure 2.9. Anatomy of the orbit.

Physiology of the Eye

The primary function of the eye is to form a clear image of objects in our environment. These images are transmitted to the brain through the optic nerve and the posterior visual pathways. The various tissues of the eye and its adnexa are thus designed to facilitate this function.

The Eyelids

Functions include: (1) protection of the eye from mechanical trauma, extremes of temperature and bright light, and (2) maintenance of the normal precorneal tear film, which is important for maintenance of corneal health and clarity.

Normal eyelid closure requires an intact nerve supply to the orbicularis oculi muscles (facial nerve). Eyelid opening is affected by the levator palpebrae superioris supplied by the IIIrd cranial nerve.

The Tear Film

The tear film consists of three layers: the mucoid, aqueous and oily layers.

The mucoid layer lies adjacent to the corneal epithelium. It improves the wetting properties of the tears. It is produced by the goblet cells in the conjunctival epithelium.

The watery (aqueous) layer is produced by the main lacrimal gland in the superotemporal part of the orbit and accessory lacrimal glands found in the conjunctival stroma. This aqueous layer contains electrolytes, proteins, lysozyme, immunoglobulins, glucose and dissolved oxygen (from the atmosphere).

The oily layer (superficial layer of the tear film) is produced by the meibomian glands (modified sebaceous glands) of the eyelid margins. This oily layer helps maintain the vertical column of tears between the upper and lower lids and prevents excessive evaporation.

The tears normally flow away through a drainage system formed by the puncta (inferior and superior), canaliculi (inferior and superior), the common canaliculus (opening into the lacrimal sac) and the nasolacrimal duct (which drains into the nose).

The Cornea

The primary function of the cornea is refraction. In order to perform this function, the cornea requires the following:

- transparency
- smooth and regular surface
- spherical curvature of proper refractive power
- appropriate index of refraction.

Corneal transparency is contributed to by anatomical and physiological factors:

1. Anatomical:
   - absence of keratinisation of epithelium
   - tight packing of epithelial cells
   - mucous layer providing smooth lubricated surface
   - homogeneity of membranes – Bowman's and Descemet's
   - regular arrangement of corneal lamellae (parallel collagen fibres within each lamella, with adjacent lamellae being perpendicular). Regularity produces a diffraction grating
   - paucity of corneal stromal cells, which are flattened within lamellae
   - interspaces – absence of blood vessels.
2. Physiological
   - active dehydration of the cornea through Na^+/HCO_3^- metabolic pump located in the corneal endothelium. This dehydration is supplemented by the physical barrier provided by the corneal epithelium and endothelium.

The Aqueous Humour

The aqueous humour is an optically clear solution of electrolytes (in water) that fills the space between the cornea and the lens. Normal volume is 0.3 ml. Its function is to nourish the lens and cornea.

The aqueous is formed by active secretion and ultrafiltration from the ciliary processes in the posterior chamber. The fluid enters the anterior chamber through the pupil, circulates in the anterior chamber and drains through the trabecular meshwork into the canal of Schlemm, the aqueous veins and the conjunctival episceral veins.

The aqueous normally contains a low concentration of proteins, but a higher concentration of ascorbic acid compared with plasma. Inflammation of the anterior uvea leads to leakage of proteins from the iris circulation into the aqueous (= plasmoid aqueous).

The Vitreous Body

The vitreous consists of a three-dimensional network of collagen fibres with the interspaces filled with polymerised hyaluronic acid molecules, which are capable of holding large quantities of water. The vitreous does not normally flow but is percolated slowly by small amounts of aqueous. There is liquefaction of the jelly with age, with bits breaking off to form floaters. This degeneration occurs at an earlier age in myopes.

The Lens

The lens, like the cornea, is transparent. It is avascular and depends on the aqueous for nourishment. It has a thick elastic capsule, which prevents molecules (e.g., proteins) moving into or out of it.

The lens continues to grow throughout life, new lens fibres being produced from the outside and moving inwards towards the nucleus with age.

The lens is comprised of 65% water and 35% protein. The water content of the lens decreases with age and the lens becomes less pliable.

The lens is suspended from the ciliary body by the zonule, which arises from the ciliary body and inserts into the lens capsule near the equator.

The Ciliary Body

The ciliary muscle (within the ciliary body) is a mass of smooth muscle, which runs circumferentially inside the globe and is attached to the scleral spur anteriorly. It consists of two main parts:

1. Longitudinal (meridional) fibres – form the outer layers and arise from the scleral spur and insert into the choroid. Contraction of this part of the muscle exerts traction on the trabecular meshwork and also the choroid and retina.
2. Circular fibres – form the inner part and run circumferentially. Contraction moves the ciliary processing inwards towards the center of the pupil leading to relaxation of the zonules.

Accommodation

Accommodation is the process whereby relaxation of zonular fibres allows the lens to become more globular, thereby increasing its refractive power. When the ciliary muscles relax, the zonular fibres become taut and flatten the lens, reducing its refractive power. This is associated with constriction of the pupil and increased depth of focus.

Accommodation is a reflex initiated by visual blurring and/or awareness of proximity of the object of interest. The maximum amount of accommodation (amplitude of accommodation) is dependent on the rigidity of the lens and contractility of the ciliary muscle. As the lens becomes more rigid with age (and contractions of the ciliary body reduce), accommodation decreases. Reading and other close work become impossible without optical correction – presbyopia.

The Retina

This is the "photographic film" of the eye that converts light into electrical energy (transduction) for transmission to the brain. It consists of two main parts:

1. The neuroretina – all layers of the retina that are derived from the inner layer of the embryological optic cup.
2. The RPE – derived from the outer layer of the optic cup. It is comprised of a single layer of cells, which are fixed to Bruch's membrane. Bruch's membrane separates the outer retina from the choroid.

The retinal photoreceptors are located on the outer aspect of the neuroretina, an arrangement that arose from inversion of the optic cup and allows close proximity between the photosensitive portion of the receptor cells and the opaque RPE cells, which reduce light scattering. The RPE also plays an important role in regeneration/recycling of photopigments of the eye and during light–dark adaptation.

In order for the light to reach the photoreceptors to form sharp images, all layers of the retina inner to the photoreceptors must be transparent. This transparency is contributed to by the absence of myelin fibres from the retinal neurons. The axons of the retina ganglion cells normally become myelinated only as they pass through the optic disc to enter the optic nerve.

There are two main types of photoreceptors in the retina – the rods and the cones. In the fovea centralis the only photoreceptors are cones, which are responsible for acute vision (visual details) and colour vision. Outside the fovea, rods become more abundant towards the retinal periphery. The rods are responsible for vision in poor (dim) light and for the wide field of vision.

The retinal capillary network (derived from the central retinal artery) extends no deeper than the inner nuclear layer and nourishes the neuroretina from inside up to part of the outer plexiform layer. It is an end-arterial system. The choroid serves to nourish the RPE and the photoreceptors (by diffusion of nutrients). There are no blood vessels in the outer retina. The central fovea is completely avascular and depends on diffusion from the choroidal circulation for its nourishment. Thus, normal functioning of the retina requires normal retinal and choroidal circulation.

3

Examination of the Eye

As in all other medical examinations, examination of a patient with an eye problem should include history, physical examination and special investigation. The age as well as social history, including the occupation of the patient, should not be forgotten in such evaluation. A summary of such evaluation is provided in Table 3.1.

How to Find Out What a Patient Can See

One obvious way to measure sight is to ask the patient to identify letters that are graded in size. This is the basis of the standard Snellen test for visual acuity (Figure 3.1). 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, 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, whereas 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 can be considered as equivalent to the television 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, age-related macular degeneration, the patient might 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 might 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, although very useful, is not an adequate measure of vision on its own. For a proper clinical examination, we need to assess the visual fields and 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 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

Table 3.1. 🕮

History	
Age	
Ophthalmic:	
Subnormal vision	Duration. Difference between eyes
Disturbances of vision	Distortion, haloes, floaters, flashing lights, momentary losses of vision – field defects
Pain/discomfort	Increase/decrease
Discharge	Change in appearance – discolouration
Change in lacrimation	Swelling/mass
Diplopia	Displacement
General medical:	Diabetes/ hypertension/ COAD/dysthyroid/ connective tissue disease
Drugs	FH social/ occupational
Examination	
VA: distance/near (with and without glasses)	
Colour vision	
Visual fields	
Orbit	Proptosis/ enophthalmos
Ocular movements – conjugate and convergence	Eyelids and lacrimal apparatus
Pupils	Intraocular pressure
Position of eyes	
Conjunctiva, cornea	
AC	
Iris	
Media – lens/vitreous	
Fundus – retina/choroid, optic disc	
Special investigations	
Fluorescein angiography	
Radiological and ultrasound	
Haematological/biochemical	
Bacteriological/immunological	
Diagnosis	
Anatomical	E.g., cataract
Aetiological	E.g., diabetes

Figure 3.1. The Snellen chart. 🕮

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 ("PL").

Figure 3.2. The Stycar test. 🕮

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 (Figure 3.2), in which the child is asked to point at the letter on a card that is the same as the one held up at 6 m. Other ways of measuring visual acuity are discussed in Chapter 17.

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 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 was 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 was then introduced, and this instrument allows both central and peripheral fields to be plotted out on one chart. The Humphrey field analyser is a further development in field testing. It provides an automated visual field recording system (Figure 3.3). It also records the reliability of the patient by showing false-positive and false-negative errors. In practice this is very useful, as poor reliability is often an explanation for poor performance.

Figure 3.3. The Humphrey field analyser.

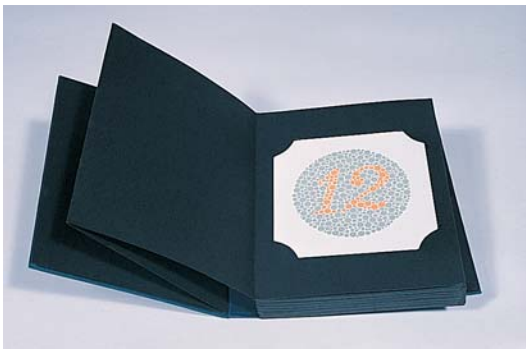


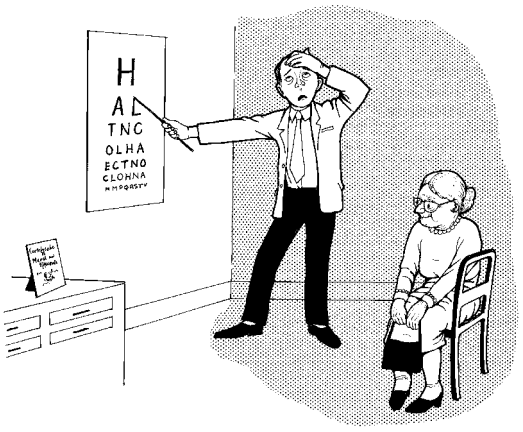
Figure 3.4. Ishihara plates for colour vision.

Colour Vision

The Ishihara plates provide a popular and effective method for screening for colour vision defects (Figure 3.4). The patient is presented with a series of plates on which are printed numerous coloured dots. The normal-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. There are other tests available that will measure blue–green defects, for example, the City University test. Other tests, such as the Farnsworth 100 Hue test, are available for the more detailed analysis of colour vision.

Spectacles

Measurement of the visual acuity might 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. As these glasses are designed for close work, the chart might be largely obscured and the uninitiated doctor might be surprised at the poor level of visual acuity (Figure 3.5). If the



I borrowed my husband's glasses. . . .

Figure 3.5. The uninitiated might be surprised at the poor level of visual acuity. 🕮

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. Similarly, an appropriate spectacle correction (near) must be worn when testing visual fields and colour vision. In an ophthalmic department, a check of the spectacle prescription is a routine part of the initial examination. Figure 3.6 shows how the converging power of the optical media and the length of the eye are mismatched to produce the need to wear spectacles (the dotted lines indicate the paths for rays of light without any corrective lens).

How to Start Examining an Eye

Evaluating the Pupil

Examination of the pupil is best performed in a dimly lit room.

Size and symmetry of pupils is assessed by asking the patient to fixate on a distant object, such as a letter on the Snellen chart. A dim light is then directed on to the face from below so that both pupils can be seen simultaneously in the diffuse illumination. Normally, the two pupils in any individual are of equal size, although slight differences in size might be observed in up to 20% of the population. Usually, physiological unequal pupils (anisocoria) remain unaltered by changing the background illumination.

In order to assess the pupil light reflex, a strong focal light is shone on the pupils, one after the other. The direct reaction and the consensual reaction (other pupil) are observed. If the afferent arc of the pupil pathway were normal, the direct and consensual reactions would be equal.

To assess the near response of the pupil, ask the patient to gaze at a distant object (e.g., Snellen chart), then at a near object (e.g., his own finger tip just in front of his nose). Observe the pupil as the patient changes gaze from distant to near fixation and vice versa. Generally, if the pupil light reflex is intact, the near reflex is normal.

External Eye and Lids

The eyelids should be inspected to make sure that the lid margins and puncta are correctly

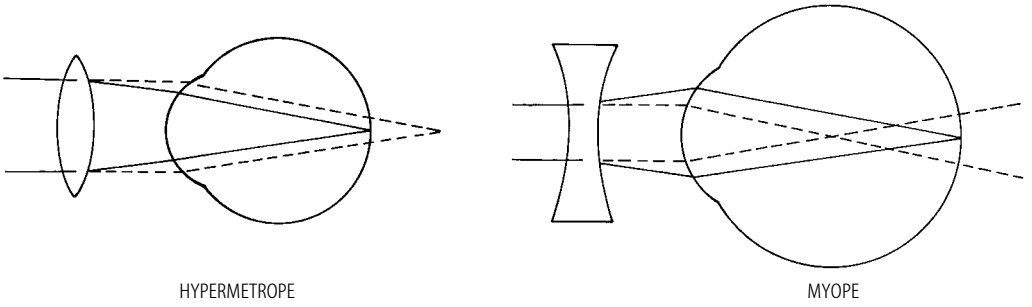


Figure 3.6. Optical defects of the eye. 🕮

aligned against the globe and that there are no ingrowing lashes. Early basal cell carcinomas (also known as rodent ulcers) on eyelid skin 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 can reveal an orbital tumour or 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 can result from inflammation of the conjunctiva and this might be accompanied by evidence of purulent discharge in the lashes. The presence of tear overflow and excoriation of the skin in 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 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 that are otherwise invisible. One might inspect the glass sides and water of a fish tank using a strong, focused torch in the same manner (Figure 3.7). Many ophthalmoscopes incorporate a focused beam of light

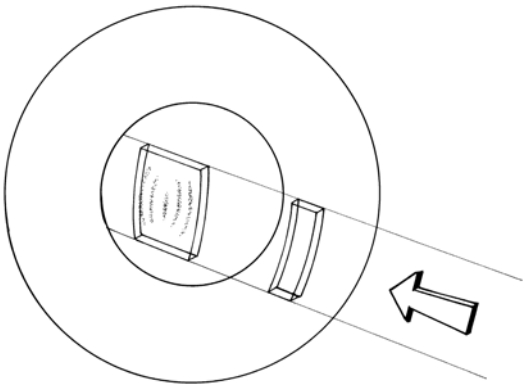


Figure 3.7. Focal illumination. 🕮

that can be used for this purpose. A magnified image of the anterior segment of the eye can be viewed with a direct ophthalmoscope held about 1/3 m away from the eye through a +10 or +12 lens. The principle has been developed to a high degree in the slit-lamp (Figure 3.8). 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

Figure 3.8. Slit-lamp examination. 🕮

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 might be of significance. Closer inspection of the iris might show that it is atrophic or fixed by adhesions. Turbidity or cells in the aqueous might 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 "gold-standard" method of measurement is to use the Goldmann tonometer (Figure 3.9), which relies on the principle of "applanation". In essence, the application of this principle provides a derived measurement of intraocular pressure by flattening a small known area of cornea with a variable force. The amount of force required to flatten a specific area is proportional to the intraocular pressure reading, and this is read from a dial. The readings provided by this measurement are highly reproducible and are given in millimetres of mercury (mmHg).

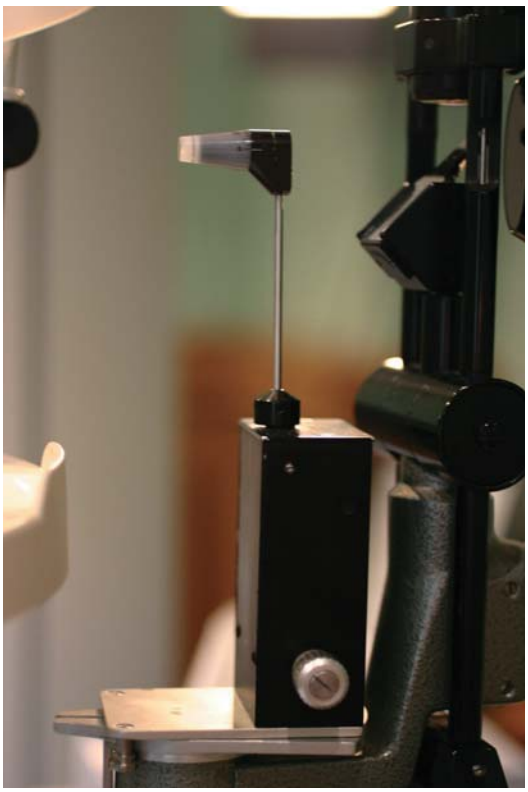


Figure 3.9. The Goldmann tonometer.

Figure 3.10. The Tonopen.

Some optometrists, however, employ "air-puff" tonometers, which are more portable and do not require attachment to a slit-lamp. These instruments are excellent for screening but are generally not as accurate as applanation tonometers. A convenient hand-held instrument (the Tonopen) is available (Figure 3.10) and is commonly used by ophthalmologists when a slit-lamp is not available.

At this stage, the pupil can be dilated for better examination of the fundi and optical media. A short-acting mydriatic is preferable, for example tropicamide 1% (Mydriacyl). These particular drops take effect after 10 min and take 2–4 h 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 nineteenth 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 von 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 that can be interposed between the eye of the patient and that of the observer, thereby overcoming any refractive problems that might defocus the view. These lenses are positioned by rotating a knurled wheel at the side of the ophthalmoscope. A number on the face of the instrument indicates the 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 might 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 with the left eye using the left hand (Figure 3.11). 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, slight and subtle opacities or defects in the optical media can 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 vessels and then, finally, the lower nasal vessels. Having examined the vessels, ask the patient to look directly at the ophthalmoscope light and the macular region should come into view. At first, this might look unremarkable, like a minute dot of light that follows our own light. More careful examination will reveal that it has a yellowish colour. To obtain a highly magnified view of the macular region, it is usually necessary to examine it with a special contact lens on the slit-lamp microscope, the Goldmann fundus lens. A fundus photograph is also helpful. After viewing the macula, the general fundus background should be observed. The appearance here depends on 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 that has an angled mirror attached to it. A view through this mirror is obtained using the slit-lamp microscope.

Figure 3.11. Direct ophthalmoscopy.

There are a number of other methods of examining the fundus. The ophthalmoscope described above is known as the direct ophthalmoscope. The indirect ophthalmoscope was introduced shortly after direct ophthalmoscopy. If one examines an eye with the pupil dilated through a mirror with a hole in it, the patient

Figure 3.12. Indirect ophthalmoscopy.

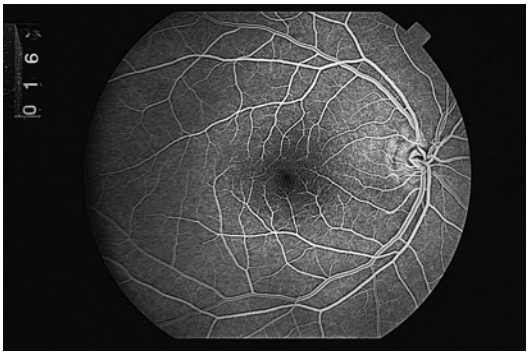


Figure 3.14. Fluorescein angiogram of normal fundus.

being at arm's length from the observer and the mirror being held close to the observer's eye, 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 retinal surgeon (Figure 3.12). If we want a highly magnified view of the fundus, the slit-lamp microscope can be used. However, a special lens must be placed in front of the patient's eye. This can be in the form of the triple-mirror contact lens (Figure 3.13). In recent years, it has become a routine practice to examine the fundus with the slit-lamp and strong convex lenses (e.g., VOLK +60, +78 or +90DS aspheric lenses). These high-power convex lenses provide inverted reversed images like the indirect ophthalmoscope. Another useful way of examining the fundus is by means of fundus photography. The photographs provide a permanent record of the fundus. 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 (Figure 3.14). Indocyanine green angiography (ICG) is more useful in assessing the choroidal circulation as ICG-A fluorescence is transmitted through the retinal pigment epithelium (RPE; compared with fluorescein [Figure 3.15]). Video filming is

Figure 3.13. The Goldmann triple mirror.

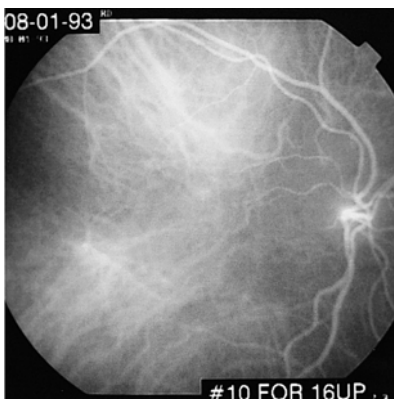


Figure 3.15. Indocyanine green angiography of normal fundus.

becoming an important method for observing changing events in the fundus and it is now possible to view a real-time image of the optic fundus on a television screen using the scanning laser ophthalmoscope. This type of equipment will undoubtedly become a routine tool for the ophthalmologist.

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 14). 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 (Figure 3.16). 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, an exophthalmometer is used (Figure 3.17). X-rays of the eye and orbit are still used. An X-ray is essential if an intraocular foreign body is suspected and it is useful for detecting bony abnormalities in the walls of the orbit caused by tumours. Computed tomography (CT) scanning has become an important diagnostic technique, especially for lesions in the orbit (Figure 3.18), particularly those involving bony tissues. This specialised X-ray has surpassed plain X-rays for most ophthalmic purposes. Magnetic resonance imaging (MRI) is more useful in assessing soft tissues of the orbit and cranium. Ultrasonography is a technique for measuring the length of the eye (which is a prerequisite for all cataract surgery); it can also be used to depict tissue planes within the eye, showing, for example, the size of intraocular tumours or the presence of vitreous membranes. It can be used to determine the presence or absence of retinal diseases, especially in eyes with opaque media (e.g., cataract or vitreous haemorrhage). Electroretinography provides a measure of the electri-

Figure 3.17. The exophthalmometer. 🕮

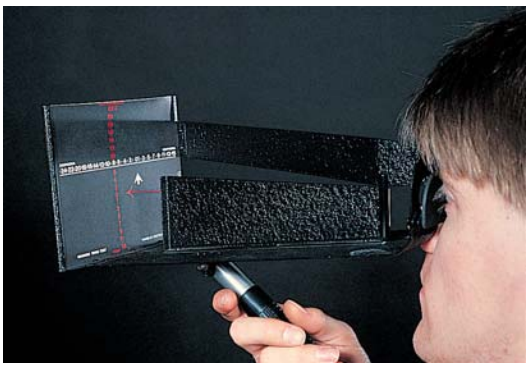


Figure 3.16. The Maddox wing. 🕮

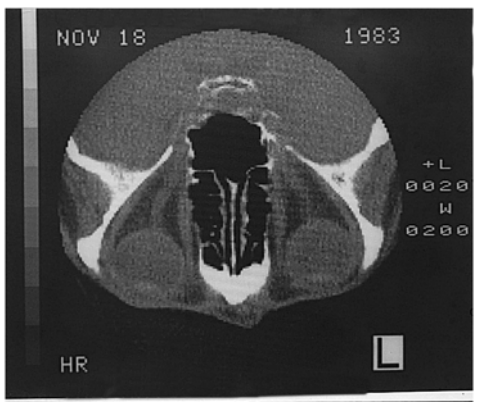


Figure 3.18. Computed tomography (CT) scan of eyes and orbit (normal). 🕮

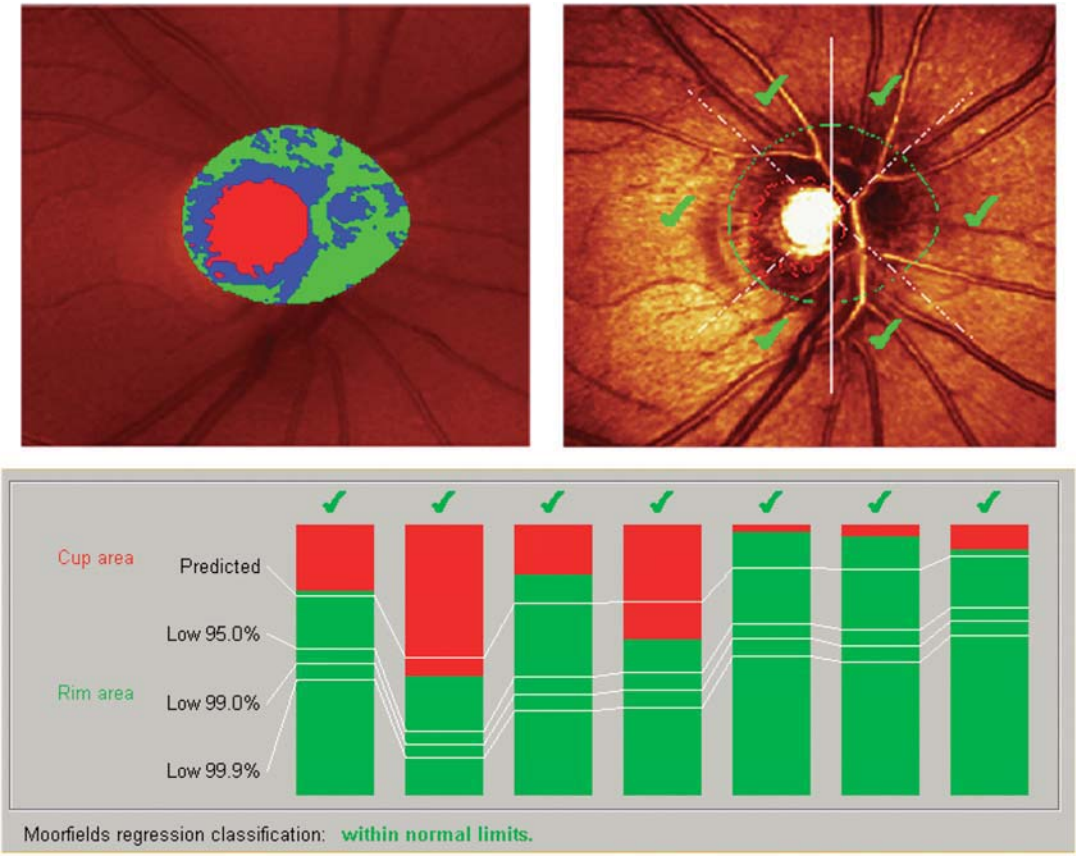


Figure 3.19. The Heidelberg retina tomograph.

cal changes that 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 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.

Technological advances have led to increasing dependence on imaging devices, such as digital fundus cameras for retinal screening in patients with diabetes. In addition, recent laser technologies, such as the Heidelberg retina tomograph, allow for a quick and easy way of scanning the optic nerve head in three dimensions (Figure 3.19) and the retinal nerve fibre layer. This is especially helpful in evaluating changes in patients with glaucoma.

4

Long Sight, Short Sight

It is useful to distinguish between long-sighted and short-sighted patients as you will see later in this chapter, but straight away we come across a problem with terminology. Think of the "short-sighted" old man who cannot see to read without glasses and, at the same time, the "short-sighted" young lady who cannot see clearly in the distance. The term "short sight" is used in these instances unwittingly by the layman to mean two different situations; either it can mean presbyopia (caused by diminished focusing power with ageing, as in the case of the old man) or it can mean myopia (caused by a larger eyeball, as in the case of the young lady).

Leaving aside presbyopia for the time being, we need to realise that the myopic person has physically larger than normal eyes, with an anteroposterior diameter of more than 24 mm, and, by contrast, the hypermetropic (or long-sighted person) person has smaller than usual eyes, with an anteroposterior diameter of less than 24 mm. To obtain a clear image, this abnormal length of the eye needs optical correction with a lens to bring light rays to a focus on the retina. The hypermetropic requires a convex lens to converge the rays, whereas the myopic person requires a concave lens to make light rays diverge before reaching the eye.

Glasses with convex lenses in them make the eyes look bigger and glasses with concave lenses in them make the eyes look smaller. Figure 4.1 shows a long-sighted (hypermetropic) patient whose glasses seem to enlarge the eyes and Figure 4.2 shows a short-sighted (myopic) patient. The clinical importance of this is that with a little practice the physician can tell the difference at a glance as the patient enters the room. This often helps with the diagnosis because certain eye diseases are associated with myopia and others with hypermetropia.

The nature of the spectacle correction can be verified by moving the lens from side to side 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 (Figure 4.3). The spectacles of the myopic patient contain concave, or diverging, lenses and, if these are moved to and fro in front of one's hand, the hand appears to move in the same direction as the movement. As a further clue, when we look at the hypermetrope from a slight angle, the line of the cheek goes out behind the magnifying lenses and *vice versa* for the myope (see Figures 4.1 and 4.2).

Here, again, let us remind ourselves that hypermetropia and myopia have nothing to do with presbyopia, which is the failure of the eyes to focus on near objects, appearing in middle age. This is nothing to do with the length of the eyeball but is related to a diminished ability to change the shape of the lens. It is corrected in otherwise normal eyes by using a convex lens. Obviously myopes, hypermetropes and those with no refractive error are all susceptible to presbyopia.

When we examine hypermetropic and myopic eyes with the ophthalmoscope, we find that there are physical differences between the

Figure 4.1. A long-sighted person.

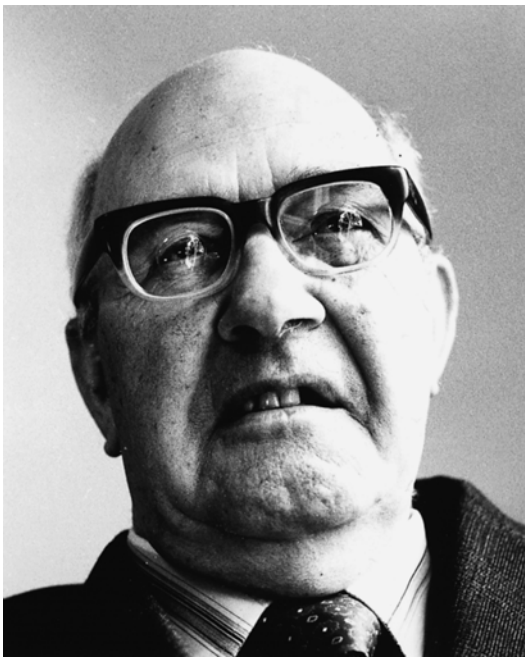


Figure 4.2. A short-sighted person.

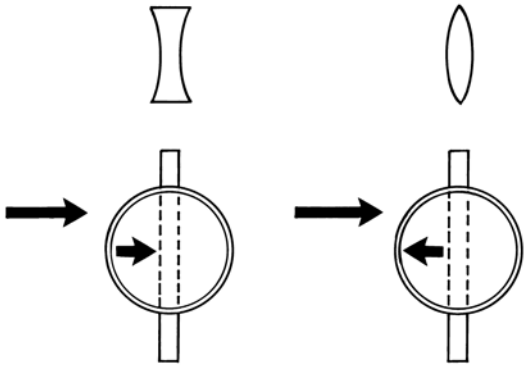


Figure 4.3. Concave lens "with"; convex lens "against". Try this for yourself in the clinic.

two. The optic disc of the hypermetrope tends to be smaller and pinker, and in extreme cases, especially in children, the disc can appear to be swollen when in fact it is quite normal. By contrast, the optic disc of the myope is larger and paler with well-defined margins and can be mistaken for an atrophic disc.

Hypermetropia is associated with certain eye conditions, notably narrow-angle glaucoma and childhood amblyopia of disuse. Myopia is associated with other conditions, particularly retinal detachment, cataract and myopic retinal degeneration. You must be aware, though, that whereas

Table 4.1. Eye disease and refractive error.

Myopia ("short sight")	Hypermetropia ("long sight")
Conditions associated with myopia	Conditions associated with hypermetropia
Retinal detachment Macula haemorrhages Cataract Myopic chorioretinal degeneration Down's syndrome Keratoconus (conical cornea)	Narrow-angle glaucoma Concomitant squint Amblyopia of disuse
Conditions causing myopia	Conditions causing hypermetropia
Large eye Cataract Diabetes mellitus Accommodation spasm, or "pseudomyopia" Congenital glaucoma	Small eye Retinal detachment Orbital tumours Macula oedema

refractive errors are extremely common, these particular conditions are relatively rare in the general population. Table 4.1 shows a more comprehensive list of these associations.

Having observed the nature of the spectacle lenses, we have now made a small step towards diagnosing the eye condition. If the patient is middle aged and complaining of evening headaches, seeing haloes around street lights and, at the same time, blurring of vision, narrow-angle glaucoma is the wrong diagnosis if the patient is myopic. It could well be the right diagnosis if the patient is hypermetropic. If the patient in Figure 4.2 were to complain of the sudden appearance of black spots combined with seeing flashes of light, he may be about to have a retinal detachment.

If we take note of whether a patient is long sighted or short sighted at an early stage, this information can influence the type of questions that are best asked when taking a history.

Finally, it is worth remembering that the myopic patient can see objects close at hand and read without glasses at any age, whereas the hypermetropic patient has to focus to see at all distances. If the hypermetrope has good focusing power (i.e., the younger patient), the distance vision may be clear without glasses but when hypermetropia is more severe, the unaided vision is poor at all ranges.

5

Common Diseases of the Eyelids

The Watering Eye

Quite often, patients present at the clinic or surgery complaining of watering eyes. It could be the golfer whose glasses keep misting up on the fairway, the housewife who is embarrassed by tears dropping on food when cooking, or the six-month-old baby whose eyes have watered and discharged since birth. Sometimes an elderly patient might complain of watering eyes when on examination there is no evidence of tear excess but the vision has been made blurred by cataracts. Some degree of tear overflow is, of course, quite normal in windy weather, and the anxious patient can overemphasise 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 can water because the tears cannot drain away adequately or because there is excessive secretion of tears.

Impaired Drainage of Tears

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

The Lacrimal Passageway

Most of the 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 (Figure 5.1). 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 of the lower punctum can appear to project upwards like a miniature volcano. Inadequate drainage of tears can result from displacement of the punctum; the lower lid in the elderly sometimes becomes turned inwards (entropion) because the whole tarsal plate rotates on a horizontal axis (Figure 5.2). This, in turn, is caused by slackening of the fascial attachments of the lower margin of the tarsal plate. At first, the eyelid turns in whenever the patient screws up the eyes but, eventually, the lid becomes permanently turned in so that the lashes are no longer visible externally and rub on the cornea. Such patients complain of watering, sore eyes and the matter can be corrected effectively by eyelid surgery. Entropion can also result from scarring and contracture of the conjunctiva on the inner surface of the eyelid.

Not only can the punctum become turned inwards, but it can also be turned outwards. Sometimes the eversion is slight, but enough to cause problems. The patient might have been using eyedrops, which, combined with the overflow of tears, sometimes causes excoriation and contracture of the skin of the lower eyelid. This leads to further eversion or ectropion of the lower eyelid (Figure 5.3). Often, the ectropion arises as the result of increasing

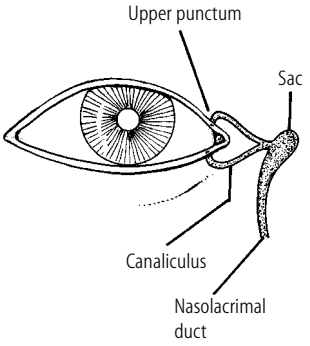


Figure 5.1. The lacrimal passageway.

laxity of the skin in the elderly but it might also result from scarring and contracture of the skin caused by trauma (cicatricial ectropion). Ectropion can be corrected effectively by suitable lid surgery.

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 complain of a watering eye even though they appear to have otherwise made a complete recovery.

Figure 5.2. Bilateral entropion. The inwardly turned lower eyelids are largely obscured by purulent discharge.

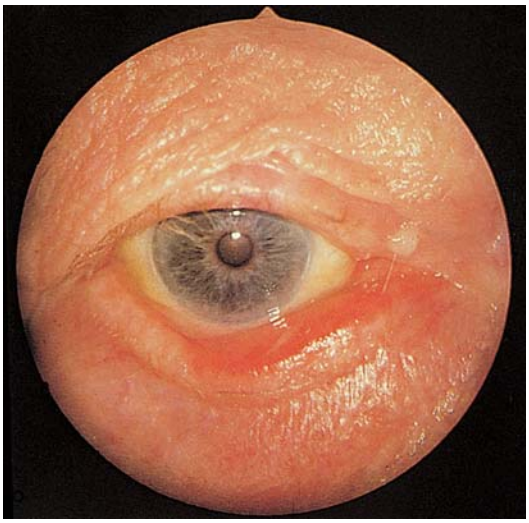


Figure 5.3. Ectropion.

Misplacement of the drainage channels, particularly of the punctum, can thus affect the outflow of tears, but perhaps more commonly the drainage channel itself 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 that has failed to become naturally perforated. In these cases, there is nearly always some purulent discharge, which can 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 once or twice daily and is instructed to instil antibiotic drops three or four times daily. This treatment alone can resolve the problem and many cases undoubtedly resolve spontaneously. Sometimes it is necessary to syringe and probe the tear duct under a short anaesthetic. Usually one waits until the child is at least nine months old before considering probing. In adults, the

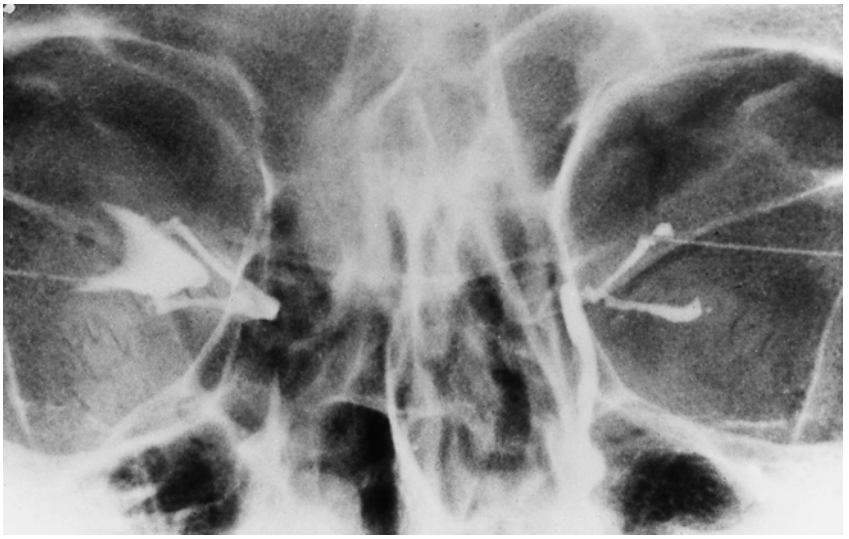


Figure 5.4. Dacryocystogram (with acknowledgement to Mr R. Welham).

obstruction is more often in the common canaliculus or nasolacrimal duct. In these cases the tear duct can be syringed after the instillation of local anaesthetic drops. This procedure is simple, although it must be done with care to avoid damaging the canaliculus, and even if the obstruction is not cleared, it can allow the surgeon to identify the site of the obstruction. Sometimes a permanent obstruction is identified at the lower end of the nasolacrimal duct, which can be relieved by surgery under general anaesthesia or the more recently introduced laser treatment applied through the nose. 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. This is injected into the lower canaliculus with a lacrimal syringe (Figure 5.4). The technique is known as dacryocystography.

Acute Dacryocystitis

Sometimes the lacrimal sac can become infected. This can occur in children or adults but is more common in adult females. The condition might present initially as a watering eye and, in its early stages, the diagnosis can 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 can point and burst. In its early stages, the condition can be aborted by the use of local and systemic antibiotics, but once an abscess has formed this can point and burst on the skin surface. Surgical incision and drainage of a lacrimal abscess can lead to the formation of a lacrimal fistula (Figure 5.5).

Rarely, the lacrimal canaliculi can become infected by the fungus Actinomycosis and a small telltale bead of pus can be expressed from the punctum. The condition is resistant to ordinary treatment with local antibiotics, 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 duct with a suitable antibiotic.

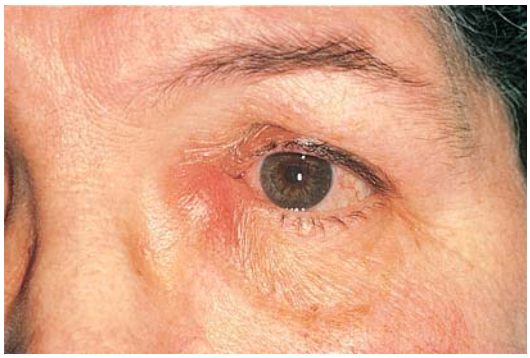


Figure 5.5. Acute dacryocystitis (with acknowlegement to Mr R. Welham).

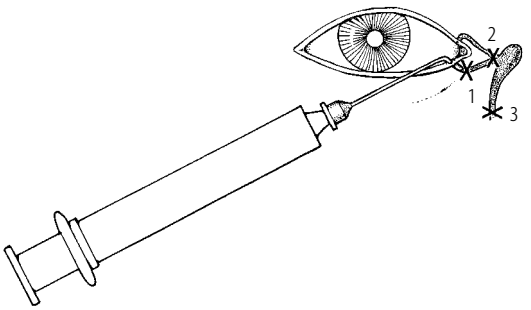


Figure 5.6. Diagnostic use of lacrimal syringing. (1) Obstruction in canaliculus shown by regurgitation of saline back through punctum. (2) Common canaliculus obstruction shown by return of saline through upper punctum. (3) Obstruction in nasolacrimal duct shown by filling of lacrimal sac. 🕮

The diagnosis of lacrimal obstruction therefore depends firstly on an examination of the eyelids, secondly on syringing the tear ducts, and then if necessary dacryocystography. Figure 5.6 illustrates the diagnostic use of lacrimal syringing.

Excessive Secretion of Tears

A wide range of conditions affecting the eye can cause an excessive production of tears, from acute glaucoma to a corneal abscess, but these do not usually present as a watering eye because the other symptoms, such as pain or visual loss, are more evident to the patient. Occasionally the unwary doctor can 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 can present in this way. Not uncommonly, a loose lash may float into the lower lacrimal canaliculus where it might become lodged, causing chronic irritation at the inner canthus. Its removal after weeks of discomfort produces instant relief and gratitude.

The Dry Eye

A patient might complain of dryness of the eyes simply because the conjunctiva is inflamed, but when the tear film really is defective, the patient might complain of soreness and irritation rather than 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 complains of dryness, or even if the symptoms appear to be improved by artificial tears.

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

Causes

- Systemic disease with lacrimal gland involvement:
  - sarcoidosis
  - rheumatoid arthritis (Sjögren's syndrome).
- Trachoma (chlamydial conjunctivitis and keratitis – see next chapter).
- Neuroparalytic keratitis.
- Exposure keratitis.
- Old age.
- Other rare causes.

Signs

Slit-lamp Examination

In a normal subject, the tear film is evident as a rim of fluid along the lid margin and a deficiency of this can be seen by direct examination. Prolonged deficiency of tears can 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, ocular pemphigoid and Stevens–Johnson syndrome, keratinisation of the cornea and conjunctiva with the formation of contracting adhesions between the opposed surfaces of the conjunctiva occurs. A similar change is apparent following chemical or thermal burns of the eyes.

Schirmer's Test

One end of a special filter paper strip is placed between the globe and the lower eyelid. The

Figure 5.7. Schirmer's test.

other end projects forward and the time taken for the tears to wet the projecting strip is measured. The test is not an accurate measure of tear secretion but it provides a useful guide (Figure 5.7).

Tear Film Break-up Time

Using the slit-lamp microscope, 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.

Management of the Dry Eye

This, of course, depends on the cause of the dry eye and the underlying systemic cause might require treatment in the first place. Artificial tear drops are a mainstay in treatment and various types are available, their use depending on which component of the tear film is defective. In severe cases, it might be necessary to consider temporary or permanent occlusion of the lacrimal puncta.

Deformities of the Eyelids

The Normal Eyelid

Figure 5.8 is a diagram of the normal eyelid in cross-section. The lids contain two antagonistic voluntary muscles: the more superficial orbicularis oculi, supplied by the seventh cranial nerve, which closes the eye, and the tendon of the levator palpebrae superioris, supplied by the third cranial nerve, which opens the eye. We must not forget that there is also some smooth muscle in the 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 seen in Horner's syndrome; increased tone is seen in thyrotoxic eye disease. These muscles (that in the upper lid is known as Muller's muscle) are attached to the skeleton of the lid, which is the tarsal plate, a plate of fibrous tissue (not cartilage) that contains the meibomian glands.

Epicanthus

Figure 5.9 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 might appear to the uninitiated to be

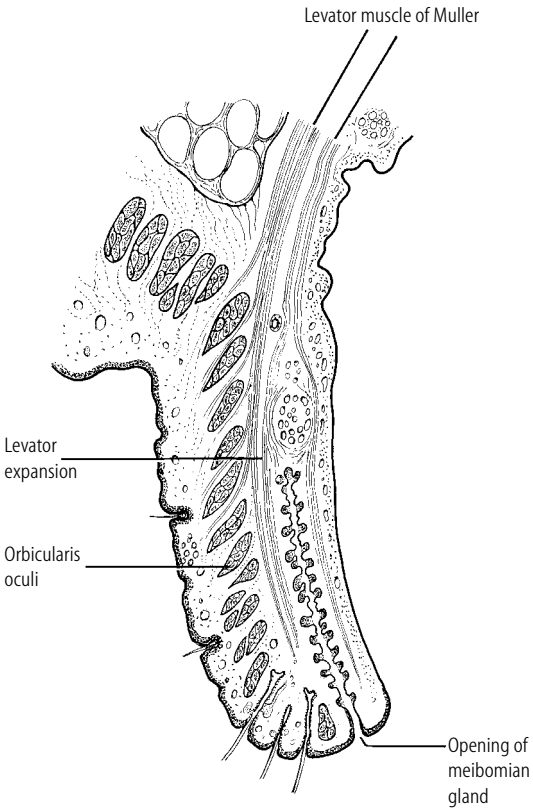


Figure 5.8. Cross-section of a normal eyelid.

Figure 5.9. Epicanthus. 🕮

squinting and this can cause considerable parental anxiety. It is important to explain that the squint is simply an optical illusion once the absence of any true deviation of the eyes has been confirmed. Epicanthus persists into adult life in Mongolian races, and occasionally it is seen in European adults. It can also be associated with other eyelid deformities.

Entropion

This is an inversion of the eyelid. The common form is the inversion of the lower eyelid seen in elderly patients. Often, the patient does not notice that the eyelid is turned in but complains of soreness and irritation. Closer inspection reveals the inverted eyelid, which can be restored to its normal position by slight downward pressure on the lower eyelid, only to turn in again when the patient forcibly closes the eyes. The inwardly turned eyelashes tend to rub on the cornea and, if neglected, the condition can lead to corneal scarring and consequent loss of vision. 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 eyelid 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 eyelid surgery. Entropion can also be seen following scarring of the conjunctival surface of the eyelids and one must mention, in particular, the entropion of the upper eyelid caused by trachoma. This is rare in the UK but still common in the Middle East and countries where trachoma is still rife.

Ectropion

This commonly seen outward turning of the lower eyelid in the elderly is eminently treatable and responds well to minor surgery. Senile ectropion can begin with slight separation of the lower eyelid from the globe, and the malposition of the punctum leads to overflow of tears and conjunctival infection. Irritation of the skin by the tears and rubbing of the eyes lead to skin contracture and further downward pulling of the eyelids. Like entropion, ectropion can be cicatricial and result from scarring of the skin of the eyelids. It can also follow a seventh cranial nerve palsy caused by complete inaction of the orbicularis muscle; this is called paralytic ectropion.

Lagophthalmos

This is the term used to denote failure of proper closure of the eyelids caused by inadequate blinking or lid deformity. In all these cases, the cornea is inadequately lubricated and exposure keratitis can develop. 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 might appear. This, in turn, can lead to the spread of infection into the eye and without prompt treatment with antibiotics, the eye might eventually be lost.

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

Blepharospasm

Slight involuntary twitching of the eyelids is 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 can

be unilateral or bilateral and cause great inconvenience and worry to the patient. It tends slowly to become more marked over many years. A small proportion of patients eventually develops Parkinsonism. Cases of recent onset need to be investigated because they might result from an intracranial space-taking lesion. In most cases, though, no underlying cause can be found. Patients with this type of blepharospasm (essential blepharospasm) can often be treated quite effectively by injecting small doses of botulinum toxin into the eyelids, but these need to be repeated every few months.

Redundant Lid Skin

Excessive skin on the eyelids is commonly seen in elderly people, often as a family characteristic. It might result from chronic oedema of the eyelids caused, for example, by thyrotoxic eye disease or 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 might 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 caused by 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 eyelid really is drooping or whether the lid on the other side is retracted. The upper lid might 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 and a dilated pupil is an oculomotor palsy, whereas slight ptosis, often not noticed by the patient or sometimes by the doctor, is more likely to mean Horner's syndrome. This syndrome is caused by 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 tendon is effective in some cases of congenital ptosis and sometimes in long-standing third cranial nerve palsies. Before embarking on surgery, it is important to exclude myasthenia gravis and corneal anaesthesia. Children with congenital ptosis need to be assessed carefully before considering surgery. In young children, ptosis surgery is indicated where the drooping lid threatens to cover the line of sight and where the ptosis causes an unacceptable backwards tilt of the head. In one rather strange type of congenital ptosis, the problem disappears when the mouth is opened and the patient might literally wink unavoidably when chewing. Careful consideration is needed before making the decision for surgery in these cases.

Causes of Ptosis

- Pseudoptosis: small eye, atrophic eye, lid retraction on other side.
- Mechanical ptosis: inflammation, tumour, and excess skin.
- Myogenic ptosis: myasthenia gravis.
- Neurogenic ptosis: sympathetic – Horner's syndrome, third cranial nerve palsy, any lesion in the pathway of these, carcinoma of the lung can cause Horner's syndrome.
- Drugs: guanethidine eye drops cause ptosis.
- Congenital: ask for childhood photograph, ask for family history.

Ingrowing Eyelashes (Trichiasis)

The lashes could grow in an aberrant manner even though the eyelids themselves are in good position. This might be the result of chronic infection of the lid margins or follow trauma. Sometimes one or two aberrant lashes appear for no apparent reason (Figure 5.10). The lashes tend to rub on the cornea producing irritation and secondary infection. The condition is referred to as "trichiasis". When one or two

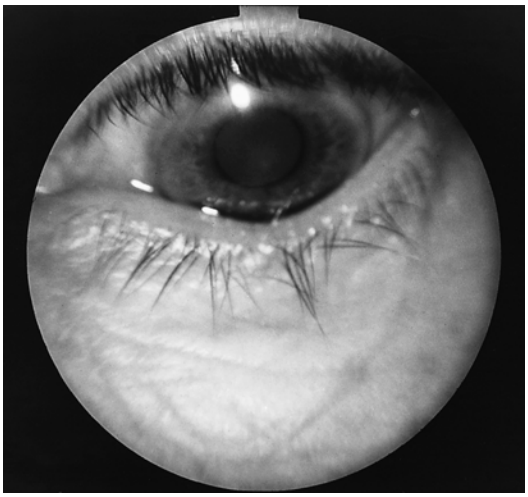


Figure 5.10. Trichiasis. This ingrowing eyelash on the lower eyelid has been causing a sore eye for three months.

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. At this stage, the best treatment is to destroy the lash roots by electrolysis before 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. 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 might be necessary to transpose or excise the lashes and their roots.

Infections of the Eyelids

Meibomian Gland Infection

The opening of the meibomian glands could become infected at any age, resulting in meibomitis, seen initially as redness along the line of a gland when the eyelid is everted. A small abscess might then form, with swelling and redness of the whole eyelid, and this can point and burst either through the conjunctiva or less often through the skin. The orifice of a gland could become occluded and the gland then becomes distended and cystic. The retained secretions of the gland set up a granulating reaction and the cyst itself might become infected. The patient might complain of soreness and swelling of the eyelid, which subsides, leaving a pea-sized swelling that 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 systemic or local antibiotics. Antibiotics might be required if the patient has several recurrences or if there are signs and symptoms of septicaemia. Once a pea-sized cyst remains in the tarsal plate, this can be promptly removed under a local anaesthetic unless the patient is a child, in which case a general anaesthetic might be required. The method of removal involves everting the eyelid and incising the cyst through the conjunctiva and then curetting the contents. Postoperatively, local antibiotic drops or ointment are prescribed (Figure 5.11).

Styes

These are distinct from meibomian infections, being the result of infection of the lash root. The eyelid might swell up and become painful and at this stage, the site of the infection can be uncertain. However, a small yellow pointing area is eventually seen around the base of an eyelash.

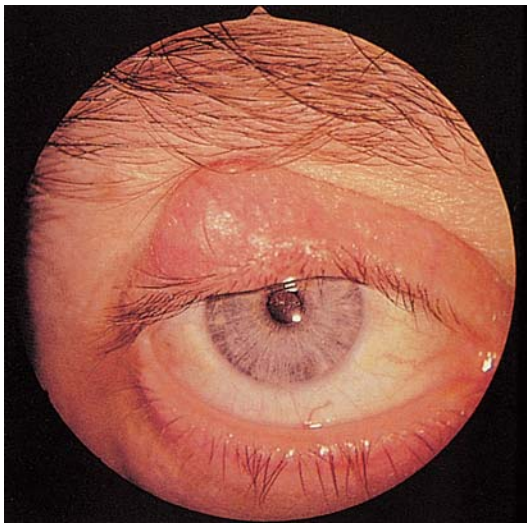


Figure 5.11. A meibomian cyst.

Hot steaming, again, is 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 from about six to ten years sometimes seem to go through periods of their lives when they can be dogged by recurrent styes and meibomian infections, much to the distress of the parents. Under these conditions, frequent baths and hairwashing are advised and sometimes a long-term systemic antibiotic might be considered. Recurrent lid infections can raise the suspicion of diabetes mellitus but in practice, this is rarely found to be an underlying cause.

Eyelid infections such as these rarely cause any serious problems other than a day or two off work and it is extremely unusual for the infection to spread and cause orbital cellulitis. Recurrent swelling of the eyelid in spite of treatment can indicate the need for a lid biopsy because some malignant tumours can, on rare occasions, present in a deceptive manner.

Blepharitis

This refers to a chronic inflammation of the lid margins caused by 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 could 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 can cause difficulties, but in more severe cases, the discomfort and irritation can interfere with work. Severe recurrent infection can 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 can be applied to the lid margins twice daily with good effect in many, but not all, cases. In severe cases with ulceration of the lid margin, it might be necessary to consider prescribing a systemic antibiotic, preferably after identifying the causative organism by taking a swab from the eyelids. Local steroids when combined with a local antibiotic are very effective treatment, 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.

Molluscum Contagiosum

This is a viral infection usually seen in children. The lesions on the eyelids are discrete, slightly raised and umbilicated and usually multiple. There are also likely to be lesions elsewhere on the body, especially the hands, and brothers or sisters might have the same problem. It is rare for the eye itself to be involved. In persistent cases, an effective form of treatment with children is careful curettage of each lesion under a general anaesthetic; in adults, cryotherapy is used for individual lesions, especially if they are adjacent to the lid margin with the propensity to cause conjunctivitis.

Orbital Cellulitis

Although this is not strictly a lid infection, it may be confused with severe meibomitis. The infection is deeper and the implications much more serious. In a child, where the condition is more common, there is eyelid swelling, pyrexia and malaise; urgent referral is needed. This applies especially if there is diplopia or visual loss, because a scan will be required to decide whether surgical intervention is going to be needed to drain an infected sinus.

Lid Tumours

Benign Tumours

Papilloma

Commonly seen on lids near or on the margin, these can be sessile or pedunculated, and are

Figure 5.12. Lid margin papilloma.

sometimes keratinised. These lesions are caused by the papilloma virus and are easily excised, but care must be taken if excision involves the lid margin (Figure 5.12).

Naevus

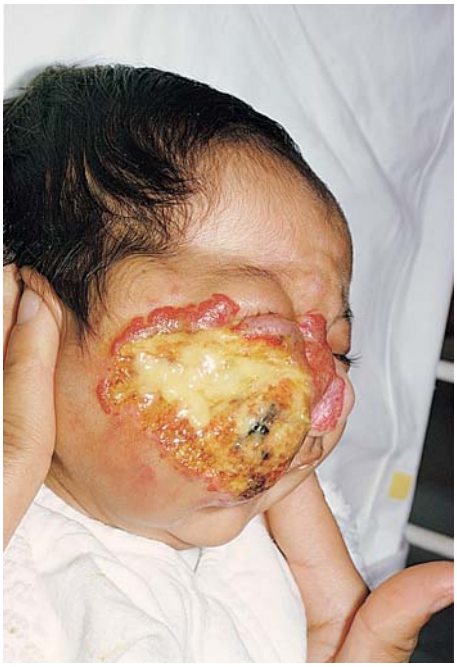
This is a flat brown spot on the skin; it might have hairs, and rarely becomes malignant.

Haemangioma

Seen as a red "strawberry mark" at or shortly after birth, this lesion can regress completely during the first few years of life. Figure 5.13 shows a gross example of the rare cavernous haemangioma, which might be disfiguring. This also can regress in a remarkable way. "Port wine stain" is the name applied to the capillary haemangioma. This is usually unilateral and when the eyelids are involved, there is a risk of association with congenital glaucoma, haemangioma of the choroid and haemangioma of the meninges on the ipsilateral side (Sturge–Weber syndrome). Children with port wine stains involving the eyelids need full ophthalmological and neurological examinations.

Dermoid Cyst

These quite common lumps are seen in or adjacent to the eyebrow. They feel cystic and are sometimes attached to bone. Typically, they present in children as a minor cosmetic problem. The cysts are lined by keratinised



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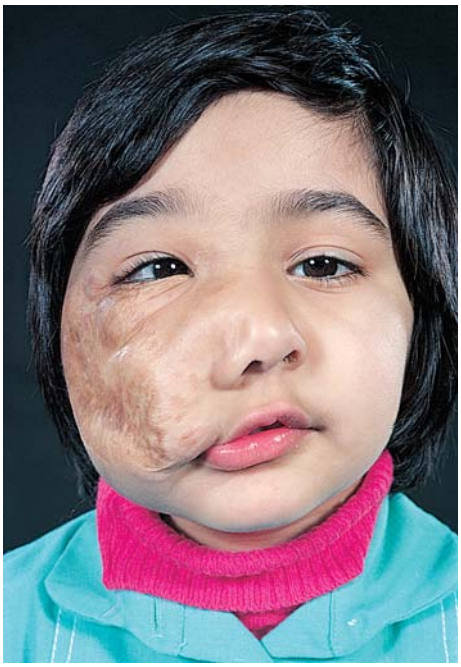


b

Figure 5.13. **a** Large disfiguring haemangioma in infancy. **b** The same lesion, which in this case had remained untreated, showing spontaneous regression.

epithelium and can contain dermal appendages and cholesterol. A scan might be needed before removal because some extend deeply into the skull.

Xanthelasma

These are seen as yellowish plaques in the skin; they usually begin at the medial end of the lids. They are rarely associated with diabetes, hypercholesterolaemia and histiocytosis. Usually, there is no associated systemic disease.

Malignant Tumours

Basal Cell Carcinoma

This is the most common malignant tumour of the lids, usually occurring on the lower lid. It appears as a small lump, which tends to bleed, forming a central crust with a slightly raised hard surround. The tumour is locally invasive only but should be excised to avoid spread into bone. Even large lesions can be approached surgically (Figure 5.14) and "Mohs" micrographic surgery is recognised as a tissue-sparing gold-standard approach in many centres. Radiotherapy is only occasionally used with a greater risk of recurrence than formal surgical excision.

Squamous Cell Carcinoma

This tends to resemble basal cell carcinoma and biopsy is needed to differentiate. It can also be mimicked by a benign self-healing lesion known as keratoacanthoma.

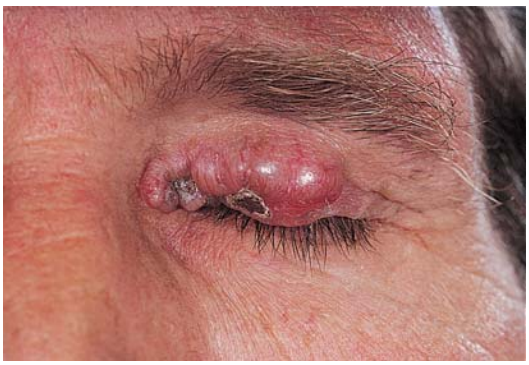


Figure 5.14. Cystic basal cell carcinoma that has extended to involve most of the upper eyelid.🕮

Malignant Melanoma

This raised black-pigmented lesion is highly malignant, but rare.

Allergic Disease of the Eyelids

This can present as one of two forms or a mixture of both. The more dramatic is acute allergic blepharitis in which the eyelids swell up rapidly, often in response to contact with a plant or eyedrops. The cause must be found and eliminated and treatment with local steroids might be needed. Chronic allergic blepharitis is seen in atopic individuals, for example hay fever sufferers or patients with a history of eczema. The diagnosis might require a histological examination of the conjunctival discharge. Drop treatment to alleviate symptoms includes mast cell stabilisers (such as lodoxamide) and histamine antagonists (such as emedastine), and these agents could take weeks to take effect. Patients with seasonal allergic conjunctivitis might require medication for a prolonged period over the spring and summer months each year.

Lid Injuries

One of the commonest injuries to the eyelids is caused by the presence of a foreign body under the eyelid – 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 medical 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 at the same time placing a glass rod horizontally across the lid. 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 to remove it using a cotton-wool bud (Figure 5.15).

Cuts on the eyelids can be caused by broken glass or sharp objects, such as the ends of screwdrivers. The important thing here is to realise that cuts on the lid margin can leave the patient with a permanently watering eye if not sewn up

Figure 5.15. Everting the upper eyelid.🕮

with proper microscopic control and using fine sutures. The lids can also be injured by chemical burns or flash burns. Exposure to ultraviolet light, as from a welder's arc or in snow blindness, can cause oedema and erythema of the eyelids. This might appear after an hour or two but resolves spontaneously after about two days.

6

Common Diseases of the Conjunctiva and Cornea

Subconjunctival Haemorrhage

This is common and tends to occur spontaneously or sometimes after straining, especially vomiting. It can also occur in acute haemorrhagic conjunctivitis caused by certain viruses and occasionally bacterial conjunctivitis. The eye becomes suddenly red and although the patient might experience a slight pricking, the condition is usually first noticed in the mirror or by a friend. The haemorrhage gradually absorbs in about 14 days and investigations usually fail to reveal any underlying cause. 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 (Figure 6.1).

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 diseases 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. In the chapter on the red eye (Chapter 7), you will find a plan of approach to the red eye that deals with the importance 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 could also be present, especially in cases of allergic conjunctivitis. The discharge around the eyelids tends to make vision only intermittently blurred (if at all) and the patient may volunteer that blinking clears the sight.

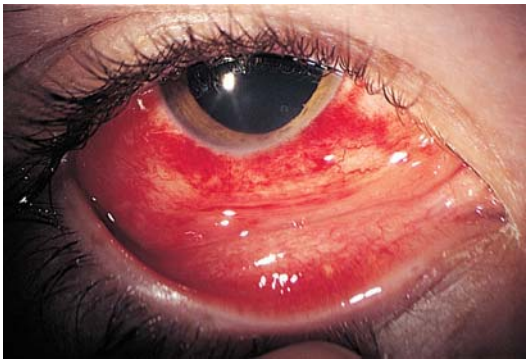


Figure 6.1. Subconjunctival haemorrhage.

Signs

Visual acuity is usually normal in conjunctivitis. The conjunctiva appears hyperaemic and there can 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 might lead one to suspect conjunctivitis results from inadequate tear secretion. Drooping of one or both upper lids is a feature of some types of viral conjunctivitis and this can be accompanied by enlargement of the preauricular lymph nodes. The ophthalmologist should train himself or herself to feel for the preauricular node as a routine part of the examination of such a case. Closer inspection of the conjunctiva might 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 might also reveal follicles or lymphoid hyperplasia. Being deep to the epithelium, they are small, pale, raised nodules and 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 at the site of maximal inflammation is gently scraped with the blade of a sharp knife or a Kimura spatula. The material obtained is placed on a slide and stained with Gram's stain and Giemsa stain. The infecting organism can thus be revealed or the cell type in the exudate might indicate the underlying cause.

Conjunctival Culture

In most cases of conjunctivitis, it might be good medical practice to take a culture from the conjunctival sac and the eyelid margin, but such a measure might not always be possible if a microbiological service is not near at hand. The cultures can be taken with sterile cotton-tipped applicators and sent to the laboratory, in an appropriate medium, as soon as possible.

Causes

- Bacterial.
- Chlamydial.
- Viral.
- Other infective agents.
- Allergic.
- Secondary to lacrimal obstruction, corneal disease, lid deformities, degenerations, systemic disease.
- Unknown cause.

Bacterial Conjunctivitis

In the UK, the commonest organisms to cause conjunctivitis are the pneumococcus, *Haemophilus* spp. 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 can also be caused by *Moraxella lacunata* but this organism is rarely isolated from cases nowadays. An important but rare form of purulent conjunctivitis is that caused by *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 perma-

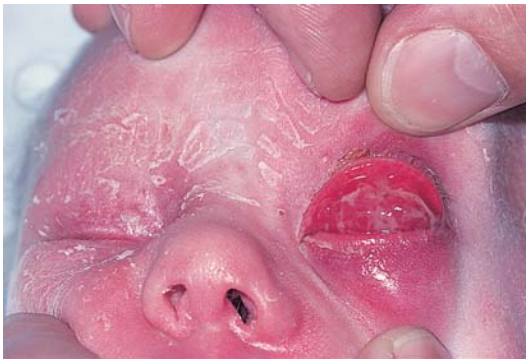


Figure 6.2. Ophthalmia neonatorum. 🕮

nent loss of vision. Purulent discharge, redness and severe oedema of the eyelids are features of the condition, which is generally known as ophthalmia neonatorum (Figure 6.2). Ophthalmia neonatorum can also be caused by staphylococci and the chlamydia (see inclusion conjunctivitis of the newborn). The disease is notifiable and any infant with purulent discharge from the eyes, particularly between the second and twelfth day postpartum, should be suspect. At one time, special blind schools were filled with children who had suffered ophthalmia neonatorum. 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 the UK but there is still a low incidence of ophthalmia neonatorum. Those affected require treatment with both topical medication (e.g., chloramphenicol 0.5% eye drops) and intramuscular benzylpenicillin (a cephalosporin, such as cefotaxime, is an alternative). Both parents of the child should also be assessed.

Pink eye is the name given to the type of acute purulent conjunctivitis that 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 three to five days. More severe cases might warrant the prescription of antibiotic drops instilled hourly during the day for three days followed by four times daily for five days. A conjunctival culture should be taken before starting treatment. Commonsense 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 to the inner and outer canthi is known as angular conjunctivitis with follicles on the superior tarsal conjunctiva. 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 *M. lacunata*. Often, zinc sulphate drops and the application of zinc cream to the skin at the outer canthus are sufficient treatment in such cases. Tetracycline ointment might be more effective.

Chlamydial Conjunctivitis

The chlamydia comprise a group of "large viruses" that are sensitive to tetracycline and erythromycin and that cause relatively minor disability to the eyes in northern Europe and the USA when compared with 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 and is caused by serotype D to K of *Chlamydia trachomatis*. The condition is usually, but not always, sexually transmitted. The conjunctivitis typically occurs one week after exposure. It can cause a more severe type of conjunctivitis in the newborn child, which can also involve the cornea. The infection is usually self-limiting but often has a prolonged course, lasting several months. The diagnosis depends on the results of conjunctival culture and examination of scrapings and the association of a follicular conjunctivitis with cervicitis or urethritis.

Chlamydial conjunctivitis responds to treatment with tetracycline. In children and adults, tetracycline ointment should be used at least four times daily. In adults, the treatment can be supplemented with systemic tetracycline, but this drug should not be used systemically in pregnant mothers or children under seven years of age. Azithromycin and other macrolide antibiotics are known to be particularly effective

in treating systemic chlamydial infection; azithromycin can be given conveniently as a one-off dose. A referral to genitourinary medicine is advisable on presentation, as a screening measure, because reinfection from partners can trigger a recurrent infection.

Trachoma

Although a doctor practicing in the UK might rarely see a case of trachoma, and even then only in immigrants, it is the commonest cause of 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. Trachoma is caused by *C. trachomatis* serotypes A, B and C and affects underprivileged populations living in conditions of poor hygiene. The disease begins with conjunctivitis, which, instead of resolving, becomes persistent, especially under the upper lid where scarring and distortion of the lid can result. The inflammatory reaction spreads to infiltrate the cornea from above and ultimately the cornea itself can become scarred and opaque (Figure 6.3). At one time, trachoma was common in the UK, especially after the Napoleonic wars at the end of the eighteenth century. It had been eliminated by improved hygienic conditions long before the introduction of antibiotics.

Adenoviral Conjunctivitis

Acute viral conjunctivitis is common. Several of the adenoviruses can cause it. Usually, the eye symptoms follow an upper respiratory tract infection and, although nearly always bilateral, one eye might be infected before the other. The affected eye becomes red and discharges; characteristically, the eyelids become thickened and the upper lid can droop. The ophthalmologist's finger should feel for the tell-tale tender enlarged preauricular lymph node. In some cases, the cornea becomes involved and subepithelial corneal opacities can appear and persist for several months (Figure 6.4). If such opacities are situated in the line of sight, the vision can 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 can result from the use of improperly sterilised ophthalmic instruments or even contaminated solutions of eye drops, and poor hand-washing techniques.

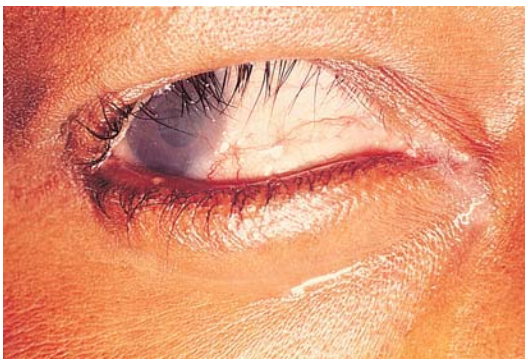


Figure 6.3. Trachoma trichiasis of upper lid and corneal vascularisation (with acknowledgement to Professor D. Archer). 🕮

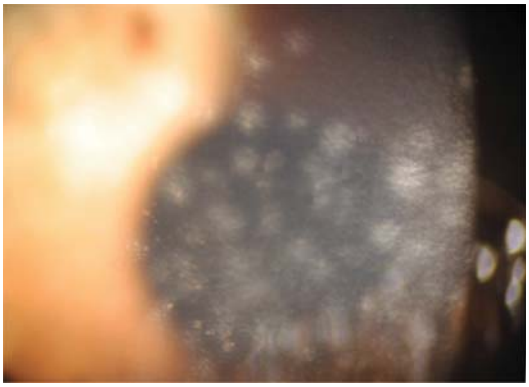


Figure 6.4. Adenoviral keratoconjunctivitis.

Herpes Simplex Conjunctivitis

This is usually a unilateral follicular conjunctivitis with preauricular lymph node enlargement. In children, it might be the only evidence of primary herpes simplex infection.

Acute Haemorrhagic Conjunctivitis

Acute haemorrhagic conjunctivitis is caused by enterovirus 70 (picornavirus) and usually occurs in epidemics. The disease is hugely contagious but self-limiting.

Other Infective Agents

The conjunctiva can 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 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 can 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 can 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 might be conjunctival oedema. If treatment is needed, vasoconstrictors, such as dilute adrenaline or naphazoline drops, can be helpful; sodium cromoglycate eye drops can be used on a more long-term basis. Systemic antihistamines are of limited benefit in controlling the eye changes.

Atopic Conjunctivitis

Unfortunately, patients with asthma and eczema can experience recurrent itching and irritation of the conjunctiva. Although atopic conjunctivitis tends to improve over a period of many years, it might result in repeated discomfort and anxiety for the patient, especially as the cornea can become involved, showing a superficial punctate keratitis or, in the worst cases, ulcer 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; the condition is also associated with the corneal dystrophy known as keratoconus or conical cornea. They are likely to develop skin infections and chronic eyelid infection by staphylococcus. The recurrent itch and irritation (in the absence of infection) 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 and aggravate herpes simplex keratitis.)

Vernal Conjunctivitis (Spring Catarrh)

Some children with an atopic history can 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 can interfere with schooling. Eversion of the upper lid reveals the raised papillae, which have been likened to cobblestones. In severe cases, the cobblestones can coalesce to give rise to giant papillae (Figure 6.5). Occasionally, the cornea is also involved, initially by punctate keratitis but sometimes it can become vascularised. It is often necessary to treat these cases with local steroids, for example, prednisolone drops applied if needed every two hours for a few days, thus enabling the child to return to school. The dose can then be reduced as much as possible down to a maintenance dose over the worst part of the season. More severe cases can derive some benefit from

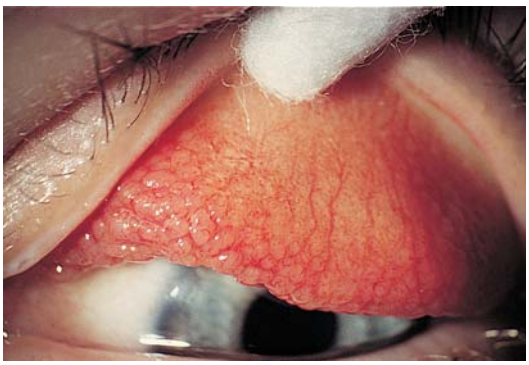


Figure 6.5. Vernal conjunctivitis (spring catarrh) papillary reaction.🕮

topical cyclosporin drops, or eyelid injections of triamcinolone to control the inflammatory response. Less severe cases can respond well to sodium cromoglycate drops; these can be useful as a long-term measure and in preventing but not controlling acute exacerbations. Other medications with a similar modest benefit in symptoms include lodoxamide (a mast cell stabiliser) and emedastine (a topical antihistamine).

Secondary Conjunctivitis

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

- Lacrimal obstruction
- Corneal disease
- Lid deformities
- Degenerations
- Systemic disease.

Lacrimal obstruction can cause recurrent unilateral purulent conjunctivitis and it is important to consider this possibility in recalcitrant cases because early resolution can 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, can 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 can 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 can 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 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 into the cornea, drawing a triangular band of conjunctiva with it. The eye becomes irritable because of 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 (Figure 6.6). Pterygium is more common in Africa, India, Australia, China and the Middle East than in Europe. It is rarely seen in white races living in temperate climates. Treatment is by surgical excision if the cornea is significantly affected with progression towards the visual axis; antibiotic drops might be required if the conjunctiva is infected. Non-infective inflammation of pterygium is treated with topical steroids.

Finally, when considering secondary causes of conjunctivitis, one must be aware that redness and congestion of the conjunctiva with secondary infection can be an indicator of systemic disease. Examples of this are the red eye of renal failure and gout, and also polycythemia rubra. The association of conjunctivitis, arthritis and nonspecific 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 rheumatoid arthritis. However, there are other rarer diseases that upset the quality or production of tears, such as sarcoidosis, pemphigus and Stevens–Johnson syndrome. Thyrotoxicosis is a more common

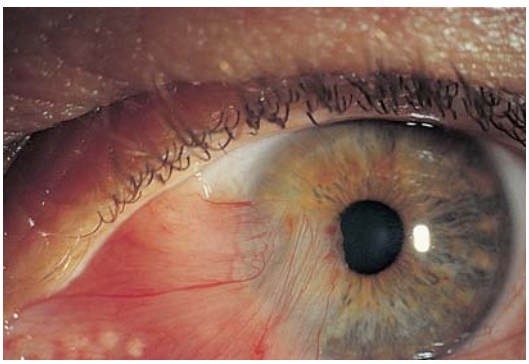


Figure 6.6. Pterygium.

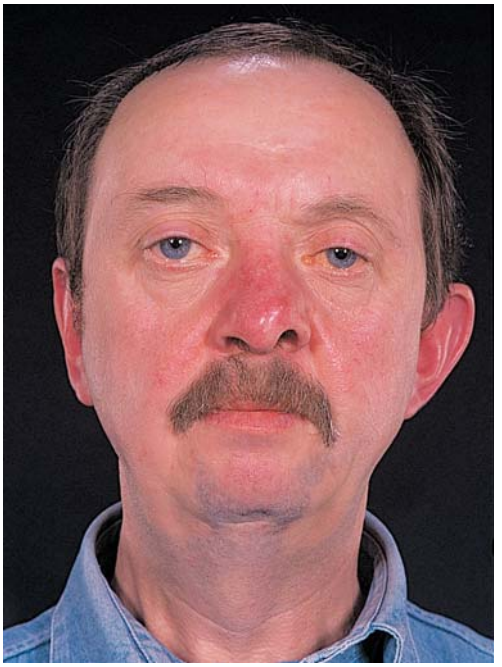


Figure 6.7. Acne rosacea.

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 (Figure 6.7). The cornea becomes invaded from the periphery by wedge-shaped tongues of blood vessels associated with recurrent corneal ulceration. Severe rosacea keratoconjunctivitis is seen less commonly now, perhaps because it responds well to treatment with the combination of systemic doxycycline, lubricants for associated dry eye and the judicious use of weak topical steroids. Usually, it is also necessary to instruct the patient to clean the lids and perform "lid hygiene", as such patients are often also affected by blepharitis.

Corneal Foreign Body

Small particles of grit or dust 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 can 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 h. 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 unnecessarily by overzealous use of surgical instruments, a permanent scar might 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.

The stroma of the cornea is surprisingly tough, permitting some degree of boldness when removing deeply embedded foreign bodies. It should be remembered that if the cornea has been perforated, 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 might 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 6.8 demonstrates the great advantage of the focused beam, and, in fact, this 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

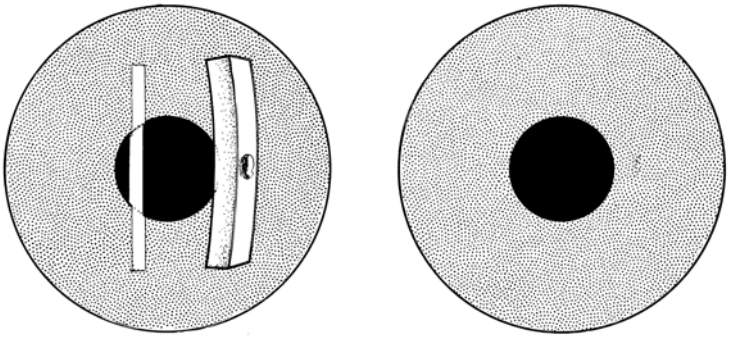


Figure 6.8. Focal illumination of corneal foreign body. 🕮

caused by 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.

Treatment

The aim of treatment is, of course, to remove the foreign body completely. Sometimes this is not as easy as it might 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 can be prudent to leave the rust ring for 24 h, after which it becomes easier 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 (Figure 6.9). The doctor will also usually require some optical aid in the form of special magnifying spectacles, for example "Bishop Harman's glasses" or the slit-lamp. Many foreign bodies can be easily removed with a cotton-wool bud (particularly those lodged under the upper lid), but otherwise at the slit-lamp a 25-gauge orange needle angled nearly perpendicular to the plane of the iris can be used to lift off the foreign body. When the foreign body is more deeply embedded, a battery-powered handheld blunt-tipped drill can be used to clean any rust deposits that remain, again under the careful control of the slit-lamp microscope.

Once the foreign body has been removed, an antibiotic drop is placed in the eye and the lids are then splinted together by means of a firm pad. 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. The visual acuity of the patient should always be checked before final discharge.

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, rarely, the vision can be permanently impaired by scarring. Also, on rare occasions, the site of corneal damage becomes infected and if

Figure 6.9. Removing corneal foreign body. 🕮

neglected, the infection can 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 frequent 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.

It is important to 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 can be quite unaware of such an injury and this might 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 might not set up an inflammatory reaction or irreversible degenerative changes until several weeks or even months have elapsed (Figure 6.10).

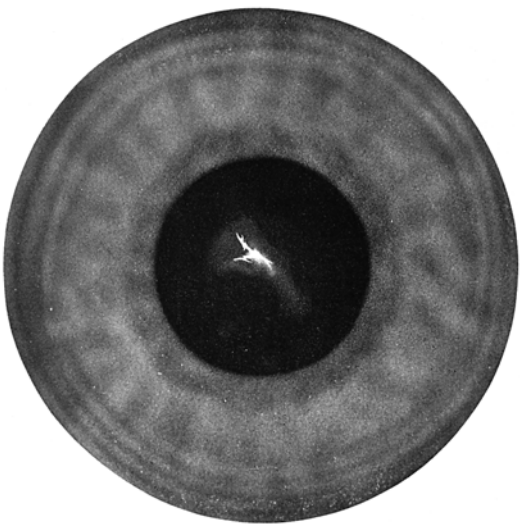


Figure 6.10. Beware of the full-thickeness corneal scar, when in doubt do an X-ray. 🕮

Corneal Ulceration

Corneal ulcers can arise spontaneously (primary) or they might 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. An 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 might be ciliary injection.

Types of Corneal Ulcer

Owing 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, or 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 can be difficult to persuade the patient that there is no foreign body. A denuded area of cornea is seen, which stains with fluorescein. It might 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 a "sore eye". This is because healing is impaired and serious damage to the eye could 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%) and an antibiotic ointment (such as chloramphenicol 0.5%), after which special 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. A pad is then

placed over the closed eyelids. 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 instill 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. The pain wears off after an hour or two and when the patient presents to the doctor there might be no obvious cause for the symptoms. In fact, careful examination with the slit-lamp reveals 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.

Owing to Bacteria

The commonest ulcer of this type is known as a "marginal ulcer" (Figure 6.11). 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 often seen as a white crescent-shaped patch near the corneal margin but there is usually, but not always, a small gap of clear cornea between it and the limbus (the corneoscleral junction). Such marginal ulcers are thought to be caused by exotoxins from *S. aureus*, mainly because they are often associated with *S. 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 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.

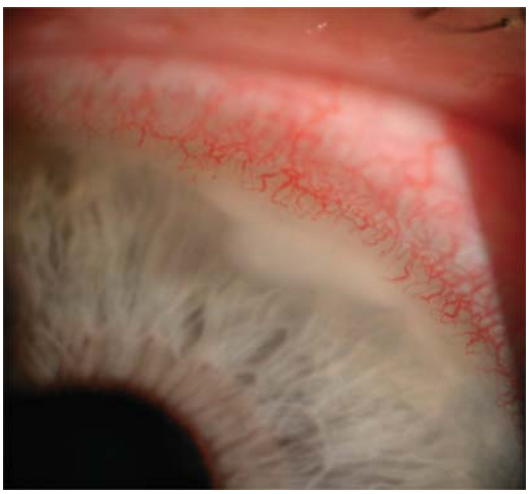


Figure 6.11. Marginal ulcer caused by bacterial infection.

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 defenses of the cornea are impaired (e.g., by underlying corneal disease, trauma, bullous keratopathy, dry eyes or contact lens wear).

There are three bacteria that can produce corneal infection despite healthy epithelium: *N. gonorrhoea*, *Neisseria meningitidis* and diphtheria. Pathogens most often associated with corneal infections, however, are *S. aureus*, *Streptococcus pneumoniae*, *Pseudomonas aeruginosa* and the enterobacteria (*Escherichia coli*, *Proteus* spp. and *Klebsiella* spp.). *Pseudomonas* spp. is an especially virulent bacterium as it can cause rapid corneal perforation if inadequately treated.

Usually there is pain, photophobia, watering and discharge in addition to redness. Examination reveals ciliary injection and a corneal defect, which might have a greyish base (infiltration). There is most often an associated (secondary) iritis, which can be severe, giving rise to a hypopyon (layer of pus in the anterior chamber).

Bacterial corneal ulcers are sight threatening and require urgent treatment. The causative organism needs to be identified by corneal scrapes. Appropriate antibiotics, usually a

combination of gentamicin and cefuroxime, which are applied frequently in hospital, provide a broad spectrum until the organisms are identified.

Owing to *Acanthamoeba* spp.

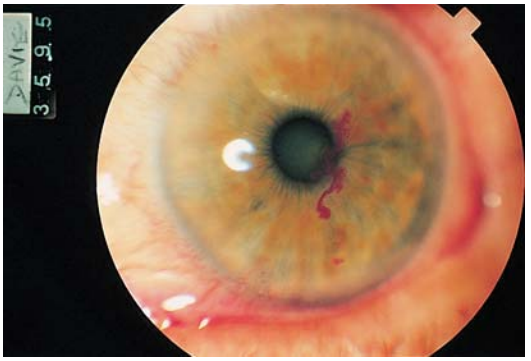
Acanthamoeba spp. are a free-living genus of amoeba that has been increasingly associated with keratitis. The keratitis is usually chronic and can follow minor trauma. Contact lens wearers are particularly at risk of this infection.

Owing 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 a few patients with this debilitating condition, which can put a patient off work for many months. Fortunately, it is only a few cases that cause such a problem, and most instances of this common condition give rise to a week or ten days of incapacity. Herpes simplex is thought to produce a primary infection in infants and younger children, which is transferred from the lips of the mother and might 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 tends to appear in association with other illnesses. Most people are familiar with the cold sores that appear on the lips because of herpes simplex. Sometimes, after a cold, one eye becomes sore and irritable and inspection of the cornea shows the characteristic corneal changes of herpes simplex infection. A slightly raised granular, star-shaped or dendriform lesion is seen, which takes up fluorescein (Figure 6.12a). 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 that 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 can occur or an inflammatory reaction can appear in the stroma deep to the infected epithelium. The eye remains red and irritable to an incapacitating degree and further dendritic ulcers might subsequently appear. In worse cases, the cornea can become anaesthetic so that, although the eye might 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

Antiviral agents are usually the first line of treatment. Examples of currently used antiviral agents are idoxuridine, trifluorothymidine, cytarabine and acyclovir. The most effective is acyclovir. 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. The removal of virus-containing epithelial cells (debridement) is now indicated only in cases that are resistant to antiviral agents, where there is toxicity to the drugs, or there is difficulty in acquiring or applying the antiviral agents. An antibiotic drop and cycloplegic are instilled and a firm pad and bandage applied. Touching the debrided area with iodine is now obsolete. Following this procedure, the eye can become very sore and the patient is given an analgesic. Often the corneal epithelium will heal after 48 h and the condition will be cured. Larger ulcers might not respond satisfactorily to this treatment. Steroids should not be used in the treatment of dendritic ulcers of the cornea (Figure 6.12b). It is well recognised that steroid drops enhance the replication of the herpes simplex virus (Figure 6.12c). They reduce the local inflammatory reaction and could give the false impression that the eye is improving. However, persistent use of local steroids in such cases could 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 can complicate the picture and require special treatment. The decision



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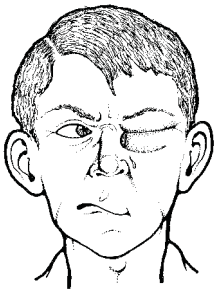

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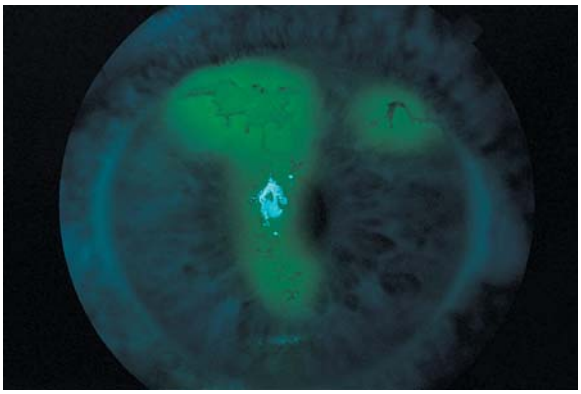

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Figure 6.12. **a** Dendritic ulcer of cornea. **b** Use of steroid drops in herpes simplex keratitis. **c** Progression of herpes simplex keratitis following use of steroid eye drops (with acknowledgement to Professor H. Dua). 🕮

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 might 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. An alternative is to induce drooping of the eyelid by an injection of botulinum toxin into the levator muscle. Surprisingly, the keratitis seems to heal usually in one to two 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. When herpetic keratitis has taken its toll, leaving a scarred cornea, the sight can eventually be restored again by a corneal graft. Unfortunately, recurrences still often occur and dendritic ulcers might appear on the graft.

Owing to Damage to the Corneal Nerve Supply

When the ophthalmic division of the trigeminal nerve is damaged by disease or injury, the cornea can become numb and there is a high risk of corneal ulceration. Such neurotropic ulcers are characteristically painless and easily become infected, with possible disastrous results. A tarsorrhaphy might be needed to save the eye but sometimes a soft contact lens can suffice, provided the ulcer is not infected at the time. Before embarking on the treatment of an anaesthetic cornea, the cause should be established and this may involve a full neurological investigation.

Owing 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, the surface of the cornea can 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 eye pad and local antibiotic ointment at night, might be needed. Botulinum toxin injection into the lid may obviate the need for surgery; this has the effect of dropping the upper lid for approximately three months, and is a useful temporising measure in some cases. It is important to bear in mind that the same risk of corneal exposure is evident in patients with severe thyrotoxic exophthalmos.

Corneal Dystrophies

There are a number of specific corneal dystrophies, most 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. A list for reference is shown in Table 6.1.

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 can be inherited as an autosomal recessive trait, although most patients do not have a positive family history. 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 can 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, 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 (Figure 6.13). 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 or corneal topography, that is, using an instrument to measure the curvature of the cornea in different meridians. Keratoconus tends to progress slowly and contact lenses can be helpful. Sometimes a corneal graft is required. Less common corneal dystrophies include Fuch's endothelial, stromal and anterior dystrophies.

Table 6.1. Corneal dystrophies. 🕮

Anterior dystrophies (corneal epithelium and Bowman's membrane):
Microcystic
Reis Buckler's
Stromal dystrophies:
Lattice
Macular
Granular
Posterior dystrophies (corneal endothelium and Descemet's membrane):
Fuch's
Posterior polymorphous
Ectatic dystrophies:
Keratoconus
Keratoglobus

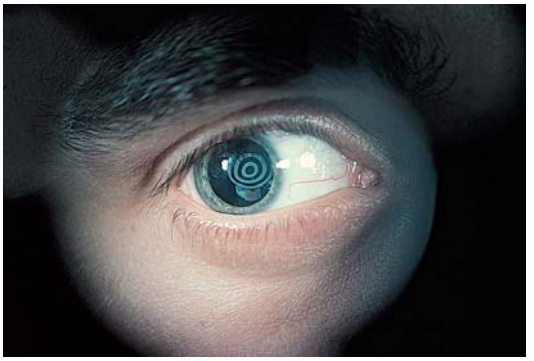


Figure 6.13. Keratoconus; Placido's disc image. 🕮

Corneal Degenerations

Apart from the inherited corneal dystrophies, 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 can gradually extend

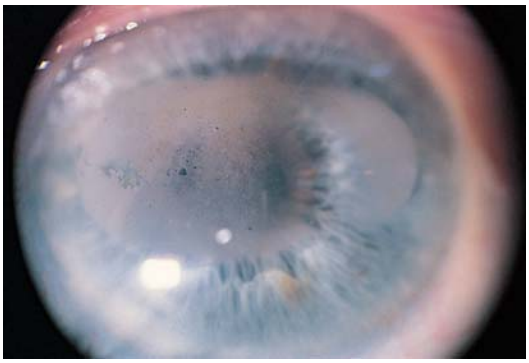


Figure 6.14. Band keratopathy. 🕮

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 that has become degenerate or in cases of long-standing corneal disease (Figure 6.14). Although band degeneration 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. Other corneal degenerations include Salzmann's nodular dystrophy and lipid keratopathies.

Corneal Oedema

To the naked eye, corneal oedema might not be obvious but careful inspection will reveal a lack of luster 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 becomes less well defined. Microscopically, a bedewed appearance is seen, minute droplets being evident in the epithelium. When the stroma is also involved, this can seem misty and might also be 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:

- Acute narrow-angle glaucoma
- Virus keratitis
- Trauma
- Contact lenses
- Postoperative
- Fuch's endothelial dystrophy.

When the intraocular pressure is suddenly raised from any cause, the cornea becomes oedematous. The normal cornea needs 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. The mechanism is impaired not only by raising the intraocular pressure, but also by infection or trauma. Senile degenerative changes might also be the sole underlying cause because of failure of the endothelial pumping mechanisms. Contact lenses, if ill fitting and worn for too long a period, can 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 can respond, in its early stages, to local steroids and sometimes a clear cornea can 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 can be necessary to consider a tarsorrhaphy, or in some instances, a corneal graft can 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. Patients with cataracts also see haloes, so that defects in other parts of the optical media of the eye might 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. Asking the patient to gaze straight ahead and then lightly touching the cornea with a fine wisp of cotton-wool can

assess corneal anaesthesia. 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 might 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 can 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 can ultimately result in anaesthesia. Herpes zoster is especially liable to lead to this problem and, because this condition can often be treated at home rather than in the ward, it will be considered in more detail here.

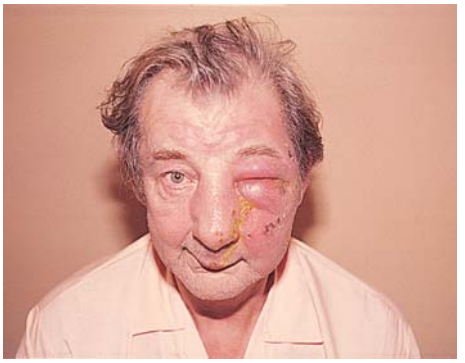


Figure 6.15. Herpes zoster ophthalmicus. 🕮

Herpes Zoster Ophthalmicus

This is caused by 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 three or four days the vesicles multiply and appear on the distribution of one or all of the branches of the fifth cranial nerve. The patient can develop a raised 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 might be vesicles on the lids and conjunctiva and, when the cornea is affected, punctate-staining areas are seen, which become minute subepithelial opacities. After four days to a week, the infection reaches its peak; the eyelids on the affected side might be closed by swelling, and oedema of the lids might spread across to the other eye (Figure 6.15). The vesicles become pustular and form crusts, which are then shed over a period of two or three weeks. In most cases, complete resolution occurs with remarkably little scarring of the skin considering the appearance in the acute stage. However, the cornea can be rendered permanently anaesthetic and the affected area of skin produces annoying paresthesiae, amounting quite often to persistent rather severe neuralgia, which can dog the patient for many years. Other complications include extraocular muscle palsies or rarely, encephalitis. Iridocyclitis is fairly common and glaucoma can develop and lead to blindness if untreated. At present, there is no known effective treatment other than the use of local steroids and acyclovir for the uveitis, and acetazolamide or topical beta-blockers for the glaucoma. Administration of systemic acyclovir or famciclovir early in the disease is known to reduce the severity of the neuralgia, but these medications need to be administered as soon as possible after the onset of symptoms for best effect. The disease has to run its course and the patient, who is usually elderly, could 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 anaesthetized patient because corneal ulceration and infection will soon result if this is neglected.

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

7

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 others are mild and of little consequence. Occasionally, the red eye can be the first sign of important systemic disease. It is important that every practicing 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 nonspecialist general practitioner.

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 might 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 patient, it can be a misleading symptom. Disease of the conjunctiva alone is not usually painful, whereas disease of the cornea or iris is generally painful.

The red eye will, therefore, be considered under three headings: the red eye that sees well and is not painful, the red painful eye that can see normally, and the red eye that does not see well and is acutely painful.

Red Eye That 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 might be noticed by someone other than the patient. The condition is common and resolves in about 10–14 days. It is extremely unusual for a blood dyscrasia to present with subconjunctival haemorrhages. Although vomiting or a bleeding tendency can also be rare causes, the normal practice is to reassure the patient rather than embark on extensive investigations, because the majority of cases are caused by spontaneous bleeding from a conjunctival capillary. This might be spontaneous and can result from a sudden increase in venous pressure, for example after coughing.

Conjunctivitis

Examination of the eye reveals 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 Conjunctivitis

This is usually infective and caused by a bacterium; it is more common in young people. It can 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 if the condition is unilateral. Sometimes an ingrowing lash might 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 eye drops could simply encourage growth of resistant organisms.

Chronic Conjunctivitis

This is a common cause of the red eye and almost a daily problem in nonspecialised ophthalmic practice. If we consider that the conjunctiva is a mucous membrane that is exposed daily 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 might 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 checklist of causes mentioned below.

The key symptoms of chronic conjunctivitis are as follows:

- Environmental factors, especially eye drops, make-up or foreign bodies.
- Lids stick in mornings?
- Do the eyes itch?
- Emotional stress or psychiatric illness?

The following is a checklist of causes of chronic conjunctivitis:

- 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 unusual to be able to incriminate a specific allergen for chronic conjunctivitis, unlike allergic blepharitis. On the other hand, hay fever and asthma could be the background cause.
- Infection: chronic conjunctivitis can begin as an acute infection, usually viral and usually following an upper respiratory tract infection.
- Drugs: the long-term use of adrenaline drops can cause dilatation of the conjunctival vessels and irritation in the eye. In 1974, it was shown that the beta-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 beta-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, can all cause vascular congestion and irritation of the conjunctiva. Migraine can also be associated with redness of the eye on one side and chronic alcoholism is a cause of bilateral conjunctival congestion.

Episcleritis

Sometimes the eye becomes red because of inflammation of the connective tissue underlying the conjunctiva, that is, the episclera. The condition can 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 or nonsteroidal anti-inflammatory agents. The underlying cause is often never discovered, although there is a well-recognised link with the collagen and dermatological diseases, especially acne rosacea. Episcleritis tends to recur and might persist for several weeks, producing a worrying cosmetic blemish in a young person (Figure 7.1).

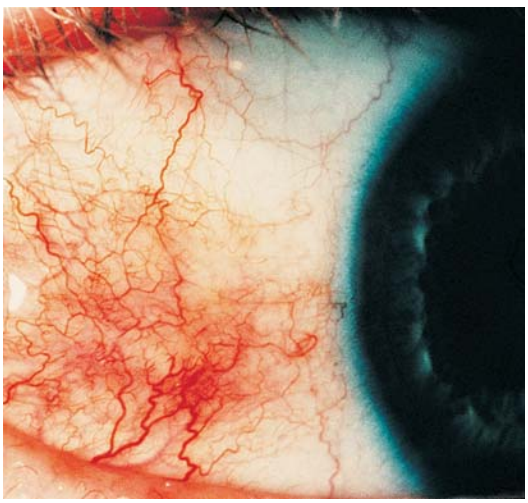


Figure 7.1. Episcleritis (with acknowledgement to Professor H. Dua).

Figure 7.2. Scleritis.

Red Painful Eye That Can See Normally

Scleritis

Inflammation of the sclera is a less common cause of red eye. There is no discharge but the eye is painful. Vision is usually normal, unless the inflammation involves the posterior sclera. It is most often seen in association with rheumatoid arthritis and other collagen diseases and sometimes can become severe and progressive to the extent of causing perforation of the globe (Figure 7.2). For this reason, steroids must be administered with extreme care. Treatment normally is with systemically administered nonsteroidal anti-inflammatory agents, for example flurbiprofen (Froben) tablets.

Red Painful Eye That 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 outpatient 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 around lights in the evenings. The raised intraocular pressure damages the iris sphincter and for this reason, the pupil is semi-dilated. Oedema of the cornea causes the eye to lose its luster and gives the iris a hazy appearance (Figure 7.3). The eye is extremely tender and painful and the patient could be nauseated and vomiting. Immediate admission to hospital is essential, where the intraocular pressure is first controlled medically and then bilateral laser iridotomies or surgical peripheral iridectomies are performed to relieve pupil block. Mydriatics should not be given to patients with suspected narrow-angle glaucoma without consultation with an ophthalmologist.

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 (Figure 7.4). Acute iritis is seen from time to time mainly in the 20–40-year age group, whereas acute glaucoma is extremely rare at these ages. Unless severe and bilateral, acute iritis is treated on an outpatient basis with local steroids and mydriatic drops. 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. The condition lasts for about two 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 might form slowly.

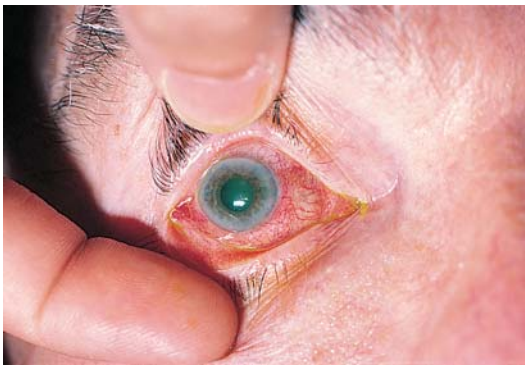


Figure 7.3. Acute angle-closure glaucoma.

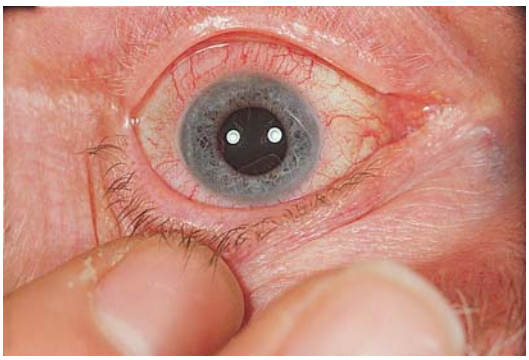


Figure 7.4. Acute iritis. The pupil has been dilated with drops.

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 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 Chapter 6 and the management of corneal injuries will be considered in Chapter 16.

Neovascular Glaucoma

The elderly patient who presents with a blind and painful eye and who might also be diabetic should be suspected of having neovascular glaucoma. Often, a fairly well-defined sequence of events enables the diagnosis to be inferred from the history, as in many cases secondary neovascular glaucoma arises following a central retinal vein occlusion. Following retinal vein occlusion, patients typically notice that the vision of one eye becomes blurred over several

hours or days. Some elderly patients do not seek attention at this stage and some degree of spontaneous recovery can seem to occur before the onset of secondary glaucoma. Fortunately, only a modest proportion of cases develops this severe complication, which usually occurs, surprisingly enough, after 100 days, hence the term "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. This form of secondary glaucoma remains as one of the few indications for surgical removal of the eye, if measures to control intraocular pressure are unsuccessful.

8

Failing Vision

Failing vision means that the sight, as measured by the standard test type, is worsening. The patient might say "I can't see so well doctor" or they might feel that their spectacles need changing. Some patients might not notice visual loss, especially if it is in one eye. Sometimes, more specific symptoms are given; the vision might be blurred, for example in a patient with cataract, or objects might appear distorted or straight lines bent if there is disease of the macular region of the retina. Disease of the macular can also make objects look larger or smaller. Double vision is an important symptom because it can be the result of a cranial nerve palsy, but if monocular, it could be caused by cataract. Patients quite often complain of floating black spots. If these move slowly with eye movement, they might be caused by some disturbance of the vitreous gel in the centre of the eye. If they are accompanied by seeing flashing lights, the possibility of damage to the retina needs to be kept in mind. "Vitreous floaters" are common and in most instances are of little pathological significance. Patients quite often notice haloes around lights and, although this is typical of an attack of acute glaucoma, haloes are also seen by patients with cataracts. Like many such symptoms, they are best not asked for specifically. The question "do you ever see haloes?" is likely to be followed by the answer "yes". Night blindness is another such symptom. No one can see too well in the dark, but if a patient has noticed a definite worsening of his or her ability to see in dim light, an inherited retinal degeneration, such as retinitis pigmentosa, might be the cause.

Failing Vision in an Eye That Looks Normal

When the Fundus Is Normal

Often a patient will present with a reduction of vision in one or both eyes and yet the eyes themselves look quite 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 a routine school eye test. The next step is to decide whether the fundus is also normal, but before dilating the pupil to allow fundus examination, it is important to check the pupil reactions and to eliminate the possibility of refractive error. Once the glasses have been checked and the fundus examined, the presence of a normal fundus narrows the field down considerably. The likely diagnosis depends on the age of the patient. Infants with visual deterioration might require an examination under anaesthesia to exclude the possibility of a rare inherited retinal degeneration or other retinal disease. Other children, particularly those in the 9–12-year age group, must first be suspected of some emotional upset, perhaps due to domestic upheaval or stress at school. This can make them reluctant to read the test type. Sometimes such children discover that exercising their own power of accommodation produces blurring of vision and they might 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 Chapter 14 on squint. When, for any reason, one retina fails to receive a clear and correctly orientated image for a period of months or years during the time of visual development, 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 eight years. Young adults who present with unilateral visual loss and normal fundi could, of course, have amblyopia of disuse and the condition can 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 and normal fundi might give the history of a stroke and are found to have a homonymous haemianopic defect of the visual fields caused by 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 carefully before such a diagnosis is made.

When the Fundus Is Abnormal

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 glaucoma and retinal detachment. It is an unfortunate fact that the commonest cause of visual loss in the elderly is usually untreatable at the present time. It is known as age-related macular degeneration and forms part of the sensory deprivation, which is an increasing scourge in elderly people. These diseases are limited to the eye itself, but disease elsewhere in the body can often first present as a visual problem. 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, are easily missed. Intracranial tumours can present in an insidious manner, in particular the pituitary adenoma, and the diagnosis might 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 age-related macular degeneration, but 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 with a central retinal vein thrombosis in spite of the dramatic haemorrhagic fundus appearance. 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 can affect the eyesight, but there are very few proven oculotoxic drugs still on the market. One important example is chloroquine. When a dose of 100 g in one year is exceeded, there is a risk of retinotoxicity, which might not be reversible. Although age-related macular degeneration is normally seen in the over-60s, the same problem may occur in younger people often with a recognised inheritance pattern. A completely different condition can also affect the macular region of young adults, known as central serous retinopathy. This tends to resolve spontaneously after a few weeks, although treatment by laser coagulation is occasionally needed. Unilateral progressive visual loss in young people can also be caused by posterior uveitis, which is the same as choroiditis. The known causes and management of this condition will be discussed in Chapter 18.

The more common causes of failing vision in a normal-looking eye are summarised in Table 8.1.

Table 8.1. 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 haemianopia	Macular degeneration Central vein thrombosis Chronic glaucoma Cataract Vitreous haemorrhage Temporal arteritis

Treatable Causes of Failing Vision

Nobody can deny that the practice of ophthalmology is highly effective. Many eye diseases can be cured or arrested, and it is possible to restore the sight fully from total blindness. Many of the commoner causes of blindness, especially in the third world, are treatable. The most important treatable cause of visual failure in the UK is cataract, and, of course, no patient should be allowed to go blind from this cause, although this does occasionally happen (Figure 8.1). Retinal detachment is less common than cataract but it provides a situation where the sight could 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 before cataract surgery does not usually affect the outcome of the operation. Acute glaucoma is another instance where the sight could be lost but restored by prompt treatment. 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 can be equally effective.

It is easy to overlook the value of antibiotics in saving sight. Before their introduction, many more eyes had to be removed following injury and 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, the treatment of diabetic retinopathy has been greatly advanced by the combined effect of laser coagulation and scrupulous control of diabetes. In the past, about one-half of patients with the proliferative type of retinopathy would be expected to go blind over five years and many of these were young people at the height of their

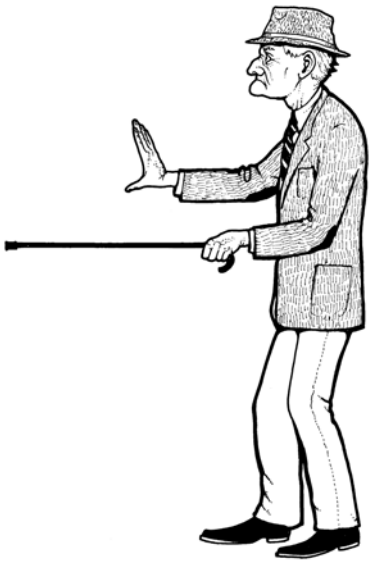


Figure 8.1. The family thought it was just old age. 🕮

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 treated effectively with systemic steroids. Amblyopia of disuse has already been mentioned; the treatment is undoubtedly effective in some cases but the results are disappointing if the diagnosis is made when the child is too old or when there is poor patient co-operation.

Untreatable Causes of Failing Vision

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, 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 inherited retinal degenerations, such as retinitis pigmentosa, although useful information is beginning to appear about the biochemistry and genetics of these conditions. Age-related macular degeneration tends to run a progressive course in spite of any attempts at treatment, and although most patients do not become completely blind, it accounts for 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 can account for visual deterioration in myopes who have otherwise undergone successful cataract or retinal 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. One must be careful here before dismissing the patient as untreatable because on rare occasions a contusion injury to the eye or orbit can result in a haemorrhage into the sheath of the optic nerve. Some degree of visual recovery can sometimes occur in these patients and it has been claimed that recovery might be helped by surgically opening the nerve sheath. There is one odd exception to this dramatic form of blindness that can follow optic nerve insult: visual loss due to optic neuritis. Patients with retrobulbar neuritis (optic neuritis) nearly always recover their vision again, whether or not they receive treatment. The explanation is that the visual loss is caused by pressure from oedema rather than to damage to the nerve fibres themselves. 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 haemianopic field defect that can follow a cerebrovascular accident.

Malignant tumours of the eye come into this category of untreatable causes of visual failure but in fact serious attempts are now being made to treat them with radiotherapy in specialised units and the prognosis appears to be improving in some cases.

9

Headache

Headache must be one of the commonest symptoms, and few specialities escape from the diagnostic problems that it can present. We must begin with the realisation that more or less everyone suffers from headache at some time or other. 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. One must, of course, be extremely cautious about not accepting symptoms at their face value, and certainly cerebral tumours have been overlooked for this reason. If the psychogenic headache is the commonest, then the headache caused by raised intracranial pressure and a space-occupying lesion must be the most important. Between these two, the whole spectrum of causes must be considered. It is essential, therefore, to memorise a permanent checklist in order that obvious causes are not omitted.

History

Often the history is the total disease in the absence of any physical signs and it is 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 could be important: raised intracranial pressure has the reputation of causing an early morning headache, which is described as bursting or throbbing and can 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 might 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 pressure of the cerebrospinal fluid is raised, the patient usually experiences a bursting pain, which can interrupt sleep or appear in the early morning. It tends to be intermittent and is made worse by coughing or lying down. It can also, of course, be accompanied by

papilloedema and vomiting, and another important symptom is blurring and transient obscurations 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 caused by a space-occupying lesion, signs of focal brain damage can also be present.

Blood Vessels

A variety of diseases involving the blood vessels can cause headache. The commonest is probably migraine. Classical migraine is thought to be caused by 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 can 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 disturbance can take the form of a hemianopic scotoma or, rarely, of a formed hallucination but, whatever their nature, they tend to last for about 10–20 min and are followed by a headache that is centred above the eye and is described as a boring pain. The headache lasts for any time between 1 h and 24 h and then disperses. The patient might experience nausea and vomiting as the attack ends. Migraine can 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 might appear by itself or the migraine attack might be accompanied by gastrointestinal symptoms or by ophthalmoplegia. The attack might be preceded by oliguria and fluid retention and be followed by a diuresis. Rarely, a permanent hemianopic scotoma or ophthalmoplegia can result from an attack of migraine, but in these circumstances the original diagnosis must be reviewed carefully. Of some importance is the fact that a history of migraine increases the risk of developing normal tension glaucoma two- or fourfold. Interestingly, migraine is one of the few risk factors for this condition.

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 by blurring of vision. Any adults with headaches should have their blood pressure measured. Another form of headache associated with abnormality of the blood vessels is that caused by 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 might be other signs of a space-occupying lesion at the apex of the orbit, for example a cranial nerve palsy or a bruit heard with the stethoscope. In the case of elderly patients, the possibility of giant cell 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 (Figure 9.1). The lumen of the affected vessels becomes occluded. Affected patients are usually over the age of 70 years and complain of tenderness of the scalp, especially over the temporal arteries, which can be seen and felt to be inflamed, and typically no pulse can be felt in them. The headache is made particularly bad by brushing the hair and other systemic symptoms include jaw claudication, weight loss and malaise. The

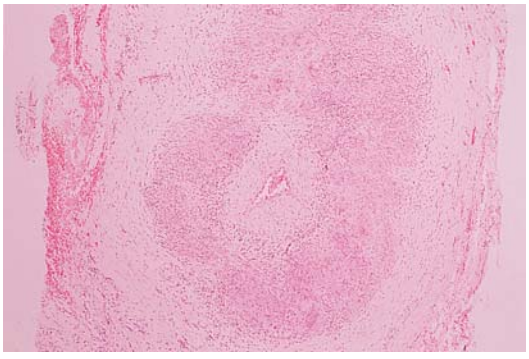


Figure 9.1. Cross-section of the temporal artery from patient with temporal arteritis. The artery is almost occluded. Note the large number of giant cells (with acknowledgement to Dr J. Lowe). 🕮

importance of this type of headache rests on the fact that the eye is involved in about 60% of cases and the patient can suddenly go blind in one eye and then a short time later go blind in the other. Patients with giant cell arteritis invariably have a raised erythrocyte sedimentation rate (ESR) and C-reactive protein level (typically the ESR is found to be above 70 mm/h). A temporal artery biopsy is helpful in specific situations to assist with the diagnosis, particularly in those with indeterminate clinical findings. When giant cell arteritis is first considered as a diagnosis, it is advisable to start treatment with systemic steroids without delay, before awaiting confirmatory diagnosis on histology. Steroid treatment is effective in preventing blindness and is required usually for a 12–18-month period. Once instituted, the response to treatment and the side effects should be very carefully monitored, preferably in co-operation with a general physician with regular measurement of the ESR and other inflammatory markers. Other less common vascular causes of headache include intracranial angioma and subarachnoid or subdural haemorrhage.

Blood

Changes in the blood itself can 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 might also complain of headache. Hypoglycaemia is another recognised cause; here, the symptoms occur after strenuous exercise or insulin excess in a diabetic patient.

Nerves

In many respects, cluster 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 can be repeated several times over a few weeks, followed by a period of remission for several months. The pain is described as being severe and unilateral. There is conjunctival congestion and constriction of the pupil on the affected side, and the attack can last from minutes to hours. Tenderness over the affected 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 meiosis and ptosis. 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 severe pain is triggered by touching a part of the cheek or by chewing and swallowing. The pain is so severe that the patient can become suicidal, and surgical division of the trigeminal nerve at the level of the Gasserian ganglion has been a method of treatment.

Postherpetic neuralgia is an extremely debilitating form of headache experienced by elderly people after an attack of trigeminal herpes zoster. The pain seems to be more severe in the elderly and it can 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 can also help, together with the application of local heat or vibration massage. Fortunately, the prompt treatment of the original attack of herpes zoster at primary care level with systemic acyclovir does seem to be reducing the incidence of this troublesome condition.

Bones

In Paget's disease of bone, 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 might show optic atrophy, and close inspection of the fundi can reveal the curious appearance of wavy lines known as angioid streaks. Oxycephaly is a congenital defect of the skull caused by premature closure of the sutures; patients sometimes complain of headache, as well as visual loss because of 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 can 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 proteins and the serum for abnormal immunoglobulins. Disease of the cervical

vertebrae is another cause of headache, because of the effect of spasm of the neck muscles. Relief of the pain by neck manipulation has been claimed, but the exact diagnosis must be made before embarking on such treatment.

Meninges

It is presumed that the pain and headache that 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 can 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, 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 at the time of examination. Patients with narrow-angle glaucoma are long-sighted – therefore, beware the middle-aged, long-sighted patient with evening headaches and blurring of vision. Chronic open-angle glaucoma 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 headaches following ocular trauma or eye surgery, that there might be secondary glaucoma. This type of glaucoma often responds well to treatment but if ignored, can lead rapidly to 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, this being a particularly important symptom following cataract surgery.

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 might have been deliberately avoiding the use of glasses and it might 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 can be accompanied by headache, which is sometimes delayed until the morning after prolonged reading. An otherwise normal person aged 45 years should be suspected of having presbyopic headaches. Uncorrected myopes do not usually complain of headaches. If the spectacle prescription is incorrect for any reason, a sensitive person can experience headache, but it is surprising how some people will tolerate an incorrect spectacle lens without complaint. Ocular muscle imbalance is an uncommon cause of headache but it is an important one because it can be corrected with considerable relief to the patient. Usually, the patient shows a significant 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 can be achieved by incorporating a prism into 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.

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 a 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 can 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 can cause similar problems. The temporomandibular joint is a recognised source of referred pain over the side of the face, and malfunctioning of the joint can result from incorrect jaw alignment or poorly fitting dentures.

Drugs

Overindulgence 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 or recognise excessive drinking. It might seem strange that such a patient should ever seek a doctor's opinion about headache, but alcoholics do sometimes seek an ophthalmological opinion for their symptoms without relating them to alcohol intake, 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 might have to be borne in mind.

Posttraumatic Headache

Nearly all patients who have suffered a significant head injury complain of headaches. The pain can remain severe for many months and in the worst case can last a few years. Usually, no obvious explanation can be found apart from the original injury. The severity of the headache can sometimes appear to be related to clinical depression following the injury but other causes of headache, such as ocular muscle imbalance or raised intracranial pressure, need to be excluded.

10

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. For this reason, some of the likely emergency requirements are considered here.

Types of Contact Lens

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 original type of moulded scleral contact lenses are still occasionally used, they have been largely replaced by the modern rigid and soft lenses, which are much smaller and thinner and hence cause less interference with corneal physiology. Rigid lenses are made from gas-permeable plastics and have generally replaced the early "hard" lenses, which were impermeable to oxygen. In 1960, the hydrophilic soft 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 as good as the rigid lens, especially when the patient has high degrees of astigmatism. Several different materials have now been used in the production of soft lenses, although the basic material used is hydroxyethylmethacrylate. The different types of soft lenses differ in their ability to take up water and transmit oxygen. Lenses are now being made that can be worn for long periods without needing to be removed and cleaned. Similarly, disposable and "planned-replacement" contact lenses are now widely available. Care should be taken that such lenses are used under professional care.

Soft contact lenses tend to absorb and adsorb material from the tear film. It is particularly important to ensure that a patient is not wearing a soft lens before fluorescein dye is instilled into the eye.

Side Effects

In general, soft contact lenses have more side effects than rigid lenses in the long term. 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 can result from clumsy handling of the lens or leaving a rigid 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 that can ensue when an abrasion becomes recurrent, the indications for wearing the lenses in the first place should be reconsidered.

The risk of infection by lens contamination or secondary to corneal abrasions is increased. Recently, *Acanthamoeba* keratitis has been described. This disease occurs more often in contact lens wearers.

Another sequel to wearing contact lenses, either rigid or soft, is the appearance of chronic inflammatory changes in the conjunctiva, often characterised by a papillary conjunctivitis. The resulting irritation and redness of the eyes can persist for some weeks after the wearing of the contact lenses ceases. Unfortunately, these symptoms can appear after wearing lenses successfully for some years and they tend to recur in spite of renewing or modifying the lenses. Some patients who tolerate contact lenses well can develop corneal changes after some years. Peripheral vascularisation can become evident and in neglected cases, there could be band degeneration of the cornea. Some contact-lens wearers complain of recurrent blurring of their vision and this could be due to an ill-fitting lens producing corneal epithelial oedema or simply to the excessive accumulation of mucus on the lens (Figure 10.1).

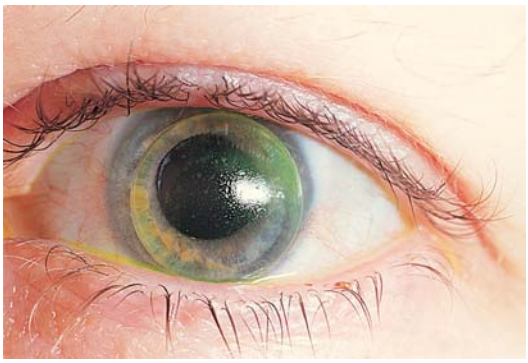


Figure 10.1. Hard contact lens with lipid deposits (with acknowledgement to Professor M. Rubinstein).

Indications

These can be considered as either cosmetic or therapeutic.

Cosmetic

There are obvious cosmetic advantages for the wearer of contact lenses, especially the teenager. However, 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. There are numerous and varied cleaning and disinfection systems on the market. Contact lenses might 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. Multifocal contact lenses are available but have limited success. Some patients tolerate being corrected in one eye for distance vision and in the other for reading with contact lenses. Care should be taken in this situation when assessing the visual acuity because the eye corrected for near vision will be blurred for distance.

Therapeutic

There are instances when the contact lens can result in much better vision than spectacles, for example in patients with high degrees of corneal astigmatism that are 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 11) by reducing the image size on the retina to such an extent that the two eyes can once again be used together. If eye drops are being regularly instilled into the eyes, soft contact lenses can 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.

11

Cataract

"Cataract" means an opacity of the lens and it is the commonest potentially blinding condition that confronts the eye surgeon. This is not to say that every person with cataract is liable to go blind. Many patients have relatively slight lens opacities that progress slowly. Fortunately, the results of surgery are good, a satisfactory improvement of vision being obtained in over 90% of cases. It is usually possible to forewarn the patients 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 might be necessary to explain that opacities in the lens are extremely common in elderly people. It is only when the opaque lens fibres begin to interfere with the vision that the term "cataract" is used. 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 fluid that 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 maintained 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 anteroposterior 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 produced by relaxation of the lens but contraction of the ciliary muscle, gradually becomes 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 the 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-40s, it becomes necessary to hold a book further from one's 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 that 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, 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 epithelial cells at 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 end of the 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" and 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 that runs in the embryonic eye from the optic disc to the vascular tunic of the lens, which is present at that stage (Figure 11.1).

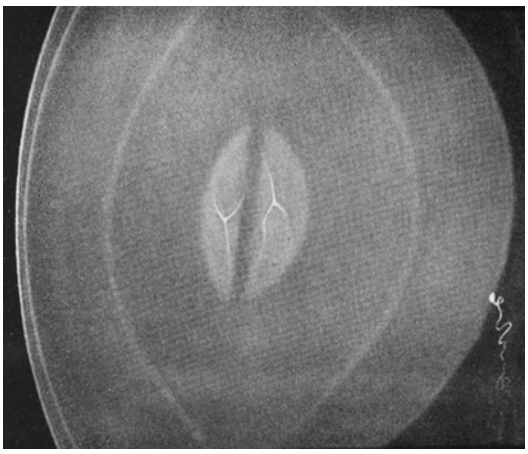


Figure 11.1. Cross-section of a child's lens: aqueous on left, vitreous on right. Note the hyaloid remnant and the "Y" sutures (with acknowledgement to M. L. Berliner, 1949). 🕮

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 might 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.

Age

The majority of cataracts are associated with the ageing process, and some of the biochemical changes in the lens fibres are now being understood. We know that certain families are more susceptible to age-related 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 might 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 impairment. 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 junior doctor working in an eye hospital must be impressed by the number of diabetics with cataracts who pass through his or her hands, and might be forgiven for deducing that diabetes is a common cause of cataract. To see the situation in perspective, one must realise that both cataracts and diabetes are common diseases of the elderly and 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 lens opacities at an earlier age. A special type of cataract is seen in young diabetics and in these cases, the lens can become rapidly opaque in a few months. Fortunately, this is not common, usually occurring in insulin-dependent (type 1) patients who 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 Causes

Cataract can be secondary to disease in the eye or disease 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 can sometimes restore a considerable amount of vision, at least for a time.

Secondary to Disease Elsewhere

It might 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 might present to the eye surgeon in their late 50s. Dysfunction of the parathyroid glands is a rare cause of cataract and Down's syndrome is a more common association.

Trauma

Contusion

A direct blow on the eye, if it is severe enough, can cause the lens to become opaque. An injury from a squash ball is a typical example of the type of force required. Sometimes the appearance of the cataract might 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 can be difficult to prove in the absence of any other signs of previous contusion. It seems unlikely that a cataract would form unless there had 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 wound of the eye bears a much higher risk of cataract formation. If the perforating object (e.g., a broken beer glass) passes through the cornea without touching the lens, the lens is usually 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 can also involve splitting of the lens capsule, with spilling out of the lens fibres into the anterior chamber. The series of events following 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 will usually gradually become absorbed, in the absence of treatment, over a period of about a month. This leaves behind the lens capsule and often a clear pupil. In spite of this, the patient cannot see clearly because most of the refractive power of the eye is lost. This has serious optical consequences and the need for an artificial intraocular lens. When the lens capsule of an adult is ruptured, a similar inflammatory reaction ensues, but there tends to be more fibrosis, and a white plaque of fibrous tissue could remain to obstruct the pupil. 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 can show a higher incidence of cataract. In practice, this is not easy to confirm. In spite of public misapprehension, ultraviolet light probably does not cause cataract either, because the shorter wavelengths fail to penetrate the globe. These shorter wavelengths beyond the blue end of the visible spectrum can

produce a dramatic superficial burn of the cornea, which usually heals in about 48 h. This injury, which is typified by "snow blindness" and "welders' flash", will be discussed in Chapter 15. Prolonged doses of infrared rays can produce cataract; this used to be seen occasionally in glassblowers and steel workers, but the wearing of goggles has now more or less eliminated this. X-rays and gamma rays can also produce cataracts, as was witnessed by the mass of reports that followed the explosion of the atomic bombs at Nagasaki and Hiroshima in Japan. Radiation cataract is now seen following whole-body radiation for leukaemia but the risk is only significant when therapeutic doses of X-rays are used.

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 can result from maternal rubella infection during the first trimester of pregnancy. The association of deafness, congenital heart lesions 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 might be overlooked, especially if they are not severe. Sometimes the cataracts can be slight at birth and gradually progress subsequently, or sometimes they can 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 meiotics, including pilocarpine. Much of our knowledge of drug-induced cataracts is based on former animal experiments. The potential danger of new drugs causing cataract was shown in the 1930s 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 might suspect that other pathology, such as macular degeneration, could 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 if carefully tested. It is a curious fact that when the cataract is unilateral, the patient can 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 noticed for the first time and interpreted as a sudden event. The history in cataract cases might be further confused by a natural tendency for patients to project their symptoms into the spectacles, and several pairs might 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 can also result from uncontrolled diabetes. If we imagine an elderly patient who requires reading glasses (for presbyopia) in the normal way but no glasses for viewing distant objects, the onset of index myopia will produce blurring of distance vision, but also the patient will discover to his or her surprise that it is possible to read again without glasses. In the same way, the hypermetropic patient will become less hypermetropic and find that it is possible to see again 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 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 be reproduced with the ophthalmoscope light. Monocular diplopia is sometimes regarded as a rather

suspect symptom, the suggestion being that if a patient continues to see double, even when one eye is closed, then he or she might not be giving an 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 might 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 might be within the requirements laid down by law (seeing a number plate at 20.5 m) but only when the patient is tested in the absence 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 might 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.

Rarely, cataracts become hypermature; that is to say, the lens enlarges in the eye and this in turn can lead to secondary glaucoma and pain in the eye. Urgent surgery might be needed under these circumstances. In its late stages, a cataract matures and becomes white, so that exceptionally a patient might complain of a white spot in the middle of the pupil.

Signs

Reduced Visual Acuity

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

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 often seen as black spokes against the red reflex (Figure 11.2). 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 age-related lens opacities are wedge shaped, pointing towards the centre of the pupil. At the same time, the central nucleus of the lens can take on a yellowish-brown colour, the appearance being termed "lens sclerosis", and ultimately, the lens can become nearly black in some 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

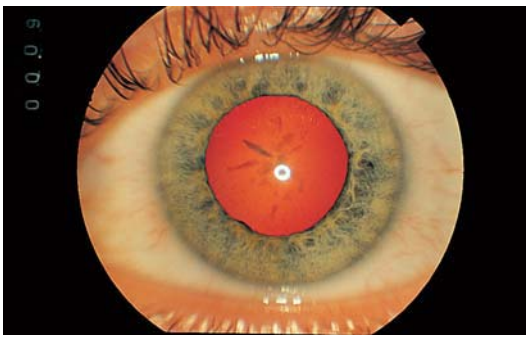


Figure 11.2. Opaque areas in the lens can be seen clearly against the red reflex.

the observer can see in, the patient can see out. If there is an obvious discrepancy between the clarity of the fundus and the visual acuity of the patient, some other pathology might be suspected. Sometimes the patient might not have performed too well on subjective testing and such an error should be apparent when the fundus is viewed. Some types of cataract can be misleading in this respect and this applies particularly to those seen in highly myopic patients. Here, there is sometimes a preponderance of nuclear sclerosis, which simply causes distortion of the fundus while the disc and macula can be seen quite clearly.

Findings on Slit-lamp Microscopy

A 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 can be seen as an early sign of senile cataract. Cataracts secondary to uveitis or to drugs might 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 can appear in a wide range of curious shapes and sizes, and earlier in the last 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 can sometimes be misleading. Congenital cataracts are usually quite easily identified by their morphology, as are some traumatic cataracts. When a unilateral cataract appears many years after a mild contusion injury, it can be difficult to distinguish this from an age-related one.

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 might lead one to suspect age-related macular 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 or her 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 seen directly. A number of other more sophisticated tests are available, for example ultrasonography, electroretinography and measurement of the visually evoked potential. Sometimes, at least an area of the peripheral retina can 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

At the present time, there is no effective medical treatment for cataract in spite of a number of claims over the years. A recent report has suggested that oral aspirin can delay the progress of cataract in female diabetics. Although this might be expected to have some effect on theoretical grounds, any benefit is probably marginal. Occasionally, patients claim that their cataracts seem to have cleared, but such fluctuation in density of the lens opacities has not been demonstrated in a scientific manner. Cataracts associated with galactosaemia are thought to clear under the influence of prompt treatment of the underlying problem.

Cataract is, therefore, essentially 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 practiced since pre-Christian times, and developments in recent years have made it safe and effective in a large proportion of cases. The operation entails removal of all the opaque lens fibres from within the lens capsule and replacing them with a clear plastic lens.

In the early part of the last century the technical side of cataract surgery necessitated waiting for the cataract to become "ripe". Nowadays no such waiting is needed and it is theoretically possible to remove a clear lens. The decision to operate is based on whether the patient will see better afterwards. Modern cataract surgery can restore the vision in a remarkable way and patients often say that they have not seen so well for many years. Indeed, many patients have quite reasonable vision without glasses but this cannot be guaranteed and, because the plastic lens implant gives a fixed focus, glasses will inevitably be needed for some distances. Probably the worst thing that can happen after the operation is infection leading to endophthalmitis and loss of the sight of the eye. Although this only occurs in about one out of a thousand cases, the patient contemplating cataract surgery needs to be aware of the possibility. Before the operation, it is now a routine to measure the length of the eye and the corneal curvature. Knowing these two measurements, one can assess the strength of lens implant that is needed. When deciding on the strength of implant, it is necessary to consider the other eye. The aim is usually to make the two eyes optically similar because patients find it difficult to tolerate two different eyes.

When to Operate

Even though the decision to operate on a cataract must be made by the ophthalmic surgeon, optometrists and the nonspecialist general practitioner need to understand the reasoning behind this decision. Elderly patients tend to forget what they have been told in the clinic and might not, for example, understand why cataract surgery is being delayed when macular degeneration is the main cause of visual loss. An operation is usually not required if the patient has not noticed any problem, although sometimes the patient can deny the problem through some unexpressed fear. The requirements of the patient need to be considered; those of the chairbound arthritic 80-year-old subject who can still read small print quite easily are different from the younger business person who needs to be able to see a car number plate at 20.5 m in order to drive. The visual acuity by itself is not always a reliable guide. Some patients who have marked glare might need surgery with a visual acuity of 6/9, whereas others with less visual demands might be quite happy with a vision of 6/12 or 6/18. Early surgery might be needed to keep a joiner or bus driver at work for which good binocular vision is needed.

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. The general health of the patient must be taken into account and this can influence one's decision in unexpected ways. Occasionally, one is presented with a patient who has difficulty with balancing, perhaps as a result of Ménière's disease or some other cause. The patient asks for cataract surgery in the hope that this will cure the problem. Unless the cataracts are advanced, the result might be disappointing. Sometimes cataract surgery is requested in a nearly blind, demented patient on the grounds that the dementia will improve with improvement of the vision. Although this occasionally happens, often the patient's mental state is made worse even though the sight is better. This raises some interesting ethical problems for the surgeon and relatives.

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 might permit a visual acuity of 6/12 or 6/18 and yet the child could be able to read small print by exercising the large amount of available focusing power. Such a child could undergo normal schooling, and cataract surgery might never be required. A complete cataract in both eyes demands early surgery and this can be undertaken during the first few months of life. There is a high risk that one eye could become amblyopic in these young patients, even after cataract surgery.

Traumatic Cataract

This is usually a unilateral problem in a younger patient and sometimes the nature of the damage to the eye prevents the insertion of an intraocular lens. The patient could be left with no lens in the eye, a situation known as aphakia. Vision can be restored by a strong convex spectacle lens but the difference between the

two eyes makes it impossible to wear glasses. This is partly because everything looks much bigger with the corrected aphakic eye; the image on the retina is abnormally large. By wearing a contact lens on the cornea, the optical problems might be solved, but it is an unfortunate fact that patients with traumatic cataracts usually have working conditions that are unsuited to the wearing of a contact lens.

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 practiced for 3000 years. The earliest method for dealing with cataract was known as couching. This entailed pushing the lens back into the vitreous, where it was allowed to sink back into the fundus of the eye. Although this undoubtedly proved a simple and satisfactory procedure in some instances, there was 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 eighteenth 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 claimed 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 eyelid. The use of local anaesthesia was introduced at the end of the nineteenth century and at the same time, attempts were made to suture the cornea back into position. By the beginning of the twentieth 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. 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 twentieth century. It involved removing the complete lens within its capsule and, by this means, avoided subsequent operations to open up residual opaque posterior capsule.

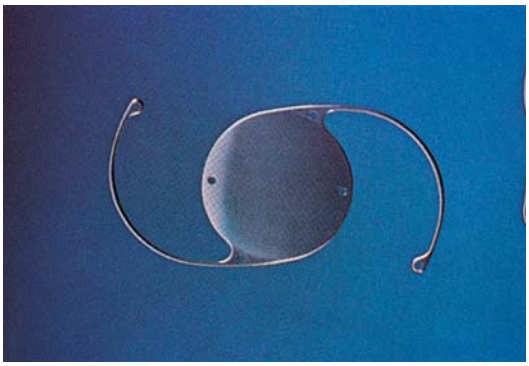


Figure 11.3. A typical plastic intraocular implant. There are different designs to suit different surgical techniques. 🕮

Perhaps the most dramatic change in cataract surgery has occurred in the latter half of the twentieth century, with the introduction of intraocular acrylic lens implants. Initially, they were mostly employed with intracapsular surgery, but a new technique for extracapsular surgery was then developed and found to be successful with implants. Many different types and designs of intraocular lens have been used over the years. Figure 11.3 shows a commonly used type of lens implant. The trend is now towards smaller incision surgery and the use of foldable or injectable implants, which unfold into position as they are being inserted into the eye. An important and widely used technique is phakoemulsification. Here, the opaque lens nucleus is removed through a complex cannula, which breaks up the lens matter ultrasonically before sucking it from the eye (Figures 11.4, 11.5 and 11.6).

Figure 11.4. Type of probe used for phakoextraction of the opaque lens nucleus. 🕮

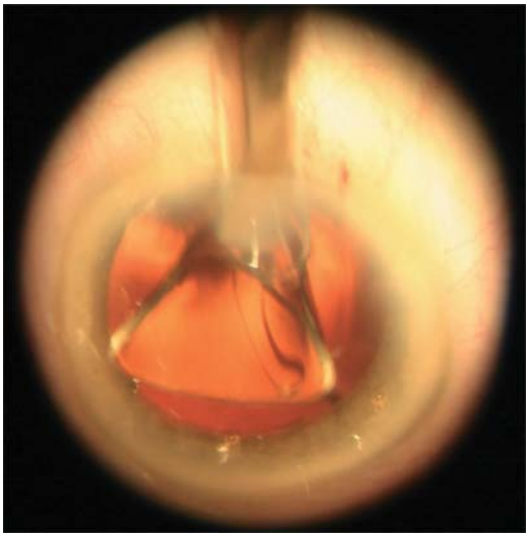


Figure. 11.5. Injection of intraocular lens implant through small incision.

Time Spent in Hospital

Many cataract operations are now done under local anaesthesia as day cases. General anaesthesia is preferred in younger patients and especially where there is a risk of straining or moving during the procedure as, for example, when the patient is deaf. An overnight stay is needed after a general anaesthetic in many cases. The elderly patient living alone with no relatives is also usually kept overnight in hospital but the trend is towards more and more day-case work, dictated partly by economic reasons, but also by safer surgery.

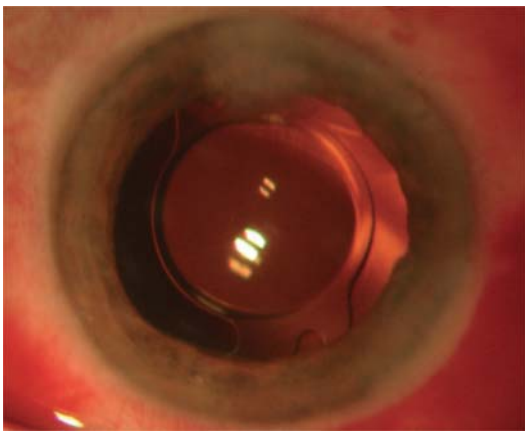


Figure 11.6. Intraocular lens implant within capsular bag.

Convalescence

It is a fair generalisation to say that an eye requires about four to six weeks for full healing to take place following a cataract operation. On the other hand, most of the healing takes place in the first two weeks. It is usual for patients to return to work after two weeks. After phakoemulsification, glasses can be prescribed at this point but after larger incision surgery the prescription of new glasses is usually done after a month. The visual recovery is undoubtedly quicker after small incision surgery but the ultimate visual result is probably no better than when a larger incision is used. Most hospitals provide a "hand-out" of do's and don'ts for the patients. The important thing is for the patient to avoid rubbing the eye and to seek immediate medical advice if the eye becomes painful, because this can indicate infection, which requires immediate treatment to prevent blindness. Following routine cataract surgery, it is usual to instill antibiotic drops combined with a steroid (usually in one bottle) four times daily for three to four weeks.

Infection is the rare but dreaded complication and this is usually heralded by pain, redness, discharge and deterioration of vision. The infection might be acquired from the patient's own commensal eyelid flora or from contamination at the time of surgery. The commonest types of bacterial infection are streptococcal and staphylococcal species. About 10–20% of patients develop opacification of the posterior lens capsule behind the implant after months or years. This is simply cured by making an opening in the capsule with a special type of laser. This is a day-case procedure, which requires no anaesthetic and takes two or three minutes. When corneal sutures have been used, these can sometimes need to be removed and this can also be done on a "while-you-wait" basis in the outpatient department.

Summary

At primary care level, it is important to be able to diagnose cataract but also to understand the benefits and risks of cataract surgery in order to

Figure 11.7. An elderly person cannot read without glasses unless he or she is myopic. Myopia in the elderly can be caused by cataract. ("Rembrandt's mother", with acknowledgement to Rijksmuseum-Stichting.) 🕮

be able to give the patient advice as to when the cataract is bad enough to need an operation. An understanding of the meaning of aphakia and the optical consequences of an implant are also useful. Most patients who present with cataracts are diagnosed as having age-related 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 symptoms of cataract is helped by understanding the meaning of index myopia.

Figure 11.7 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 this lady's eyes must be abnormal. She might have inherited myopia, allowing her to see near objects without the need for a presbyopic lens, but the myopia could also be index myopia, which in turn could be caused by early cataract formation. Another cause of index myopia could be uncontrolled diabetes.

12

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 is quite different. Glaucoma might be defined as a "pathological rise in the intraocular pressure sufficient enough to damage vision". This is to distinguish the normal elevation of intraocular pressure seen in otherwise normal individuals. Here, we 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 25–28 mmHg. The pattern of distribution fits a Gaussian curve, so that the majority of subjects have a pressure 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 low pressures remarkably well, but when the pressure is abnormally high, the circulation of blood through the eye becomes jeopardised and serious damage can ensue. For clinical purposes, an upper level of 21 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 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 by active secretion and ultrafiltration. A continuous flow is maintained through the pupil, where it reaches the angle of the anterior chamber.

On reaching the angle of the anterior chamber, aqueous passes through a grill known as the trabecular meshwork and then reaches a circular canal embedded in the sclera known as Schlemm's canal. This canal runs as 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 route 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 are not fully understood. It would appear that the production of aqueous is an active secretion, whereas the drainage is more passive, although changing the tone of the ciliary muscle can alter the rate of drainage. In normal subjects, the intraocular pressure does not differ in the two eyes by more than about 3 mmHg. Wider differences can 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 could become exaggerated as the first sign of glaucoma.

Measurement of Intraocular Pressure

The gold-standard method of intraocular pressure measurement is Goldmann applanation tonometry. The Goldmann 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, 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, 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 instill 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 photoelectric method. This air-puff tonometer is less accurate than applanation, but it is useful for screening, although abnormal results should be confirmed by Goldmann tonometry.

Clinical Types of Glaucoma

It has been mentioned above that the word "glaucoma" refers to a group of diseases. For clinical purposes, these can be subdivided into five types:

1. Primary open-angle glaucoma.
2. Normal pressure glaucoma.
3. Acute angle-closure glaucoma.
4. Secondary glaucoma.
5. Congenital glaucoma.

Primary Open-angle Glaucoma

The first important point to note about this disease is that it is 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 years are selected. This leads to the third point, which is that the incidence increases with age, being rare under the age of 40 years. This insidious, potentially blinding disease affects 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.

Primary open-angle glaucoma occurs more commonly in high myopes and diabetics; patients with Fuchs' corneal endothelial dystrophy and retinitis pigmentosa also have a higher incidence. Glaucoma is commoner in different racial groups. For example, individuals of African descent, especially those from West Africa and the Caribbean, carry a significantly greater risk of glaucoma.

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 juxtacanalicular trabecular meshwork, with endothelial thickening and oedema in the lining of Schlemm's canal. 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 the process may take five years. Younger eyes survive a raised pressure rather better than older eyes, which could already have circulatory problems. Few eyes can withstand a pressure of 50 mmHg for more than a week or two or a pressure of 35 mmHg for more than a few months.

Primary 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.

Symptoms

Most patients with chronic glaucoma have no symptoms. That is to say, the disease is insidious and is only detected at a routine eye examination, either by an optometrist or ophthalmologist, before the patient notices any visual loss. Occasionally, younger patients notice a defect in their visual field but this is unusual. Unfortunately, the peripheral loss of visual field can 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 that accounts for the lack of symptoms. Such a rise in intraocular pressure impairs the circulation of the optic disc, and the nerve 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 12.1 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. Notching of the neuroretinal rim of the optic disc tissue, especially in the inferotemporal and superotemporal region, is common. The edge of the optic disc cup corresponds to the bend in the blood vessels as they cross the disc surface. In some eyes the area of pallor can correspond to the cup, while in others the cup is larger than

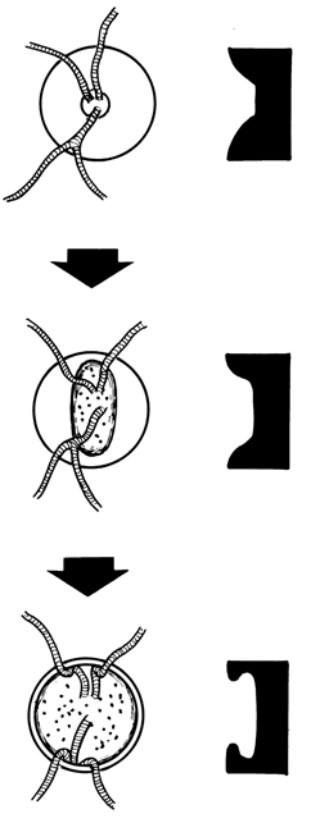


Figure 12.1. The effect of glaucoma on the optic disc.

the area of pallor. It is particularly useful to observe the way in which the vessels enter and leave the nerve head (Figure 12.2). A flame-shaped haemorrhage at the disc margin can be seen. Localised loss of retinal nerve fibres can be observed, especially with a red-free light. Diagnostic instrumentation, such as the GDx nerve fibre layer analyser, is capable of measuring the thickness of the retinal nerve fibre layer in microns, and offers an adjunctive objective measure for diagnosing and monitoring glaucoma (Figure 12.3).

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 (Figure 12.4). 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 could be well preserved and the patient can 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.

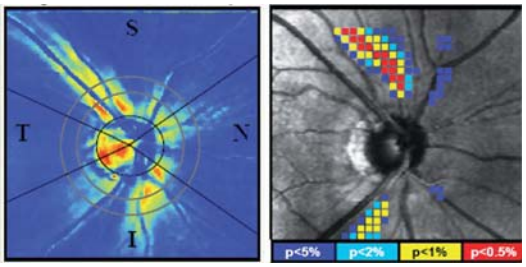


Figure 12.3. GDx nerve fibre scan result.

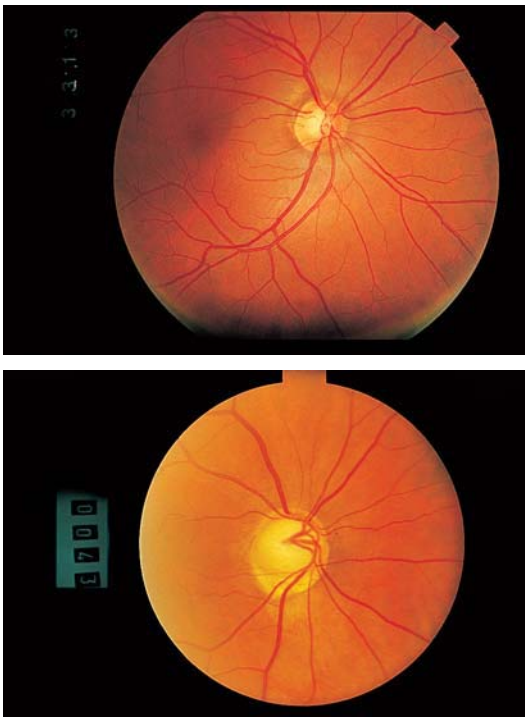


Figure 12.2. **a** Glaucomatous cupping of the disc early cupping; **b** advanced cupping.

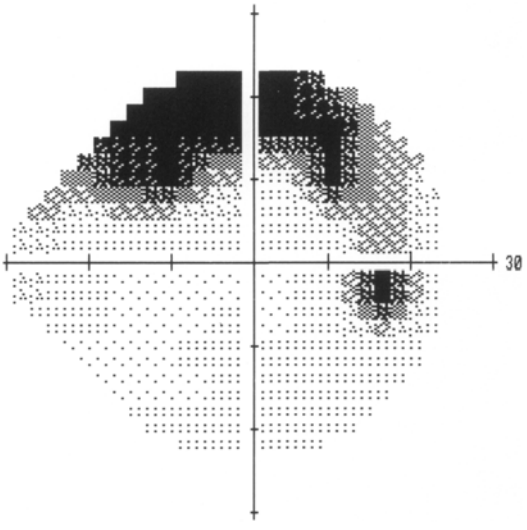


Figure 12.4. Superior arcuate visual field defect, right eye.

Treatment

For many years, the mainstay of treatment for primary open-angle glaucoma has been the use of miotic drops. The miotic of choice was pilocarpine, starting with a 1% solution and increasing to 4% if needed. Subsequently, beta-blockers, for example, timolol, levubunolol (Betagan) and betaxolol (Betoptic) replaced pilocarpine, and now prostaglandin analogues, for example, latanoprost (Xalatan) have largely replaced beta-blockers as first-line medications (Table 12.1). In practice, these medications are often used in combination.

Pilocarpine itself is effective in reducing 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 glaucoma falls to within the normal range. After about 4 h, 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, although patients are genuinely anxious to preserve their eyesight. Compliance with glaucoma medication is a major problem when medications are taken more than once daily, and is a relatively common reason for disease progression.

Timolol and other beta-blockers are effective over a 12-h period and need to be instilled only twice daily. As an ocular hypotensive agent, these are probably not quite as effective as pilocarpine, but many cases of chronic glaucoma are now satisfactorily controlled by them and furthermore, the drug may be used in combination with pilocarpine. Beta-blockers have the further advantage that they do not cause any miosis. The main side effects of beta-blockers are bronchospasm, reduced cardiac contractility and bradycardia. They are, therefore, contraindicated in patients with chronic obstructive airway disease, heart block, hypotension and bradycardia.

The cholinergic drugs (such as pilocarpine) and the anticholinesterase drugs (such as echothiopate iodide) 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 powerful and they tend to cause chronic dilatation of the conjunctival vessels in some patients, as well

Table 12.1. Topical glaucoma medication.

Drug type	Examples	Mechanism of action
ß-Blockers	Timolol Betaxolol Levubunolol Carteolol	Reduce aqueous production
Cholinergics	Parasympathomimetics: Pilocarpine Anticholinesterases: Phospholine iodide	Increase aqueous outflow through trabecular meshwork
Adrenergic agonists	Adrenaline and prodrug (Dipivefrine)	Decrease aqueous production and increase uveoscleral outflow
α_2-Agonist	Brimonidine	
Carbonic anhydrase inhibitors	Dorzolamide	Reduce aqueous production
Prostaglandins	Latanoprost (prostaglandin 2α)	Increased uveoscleral outflow

as the deposition of pigment in the conjunctiva and subconjunctival fibrosis.

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 well sustained, but it is a potent drug for reducing intraocular pressure. If a normal subject takes a 250–500 mg tablet of the drug, the eye becomes soft after about an hour. Most patients taking acetazolamide experience paresthesiae of the hands and feet and some complain of gastric symptoms. Occasionally, patients become lethargic or even confused. Young patients, particularly young males, could suffer renal colic. 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.

Newer glaucoma medications include latanoprost, dorzolamide and brimonidine. Latanoprost is a prostaglandin analogue, which produces its intraocular pressure-lowering effect through increased uveoscleral outflow. The main side effects are slight conjunctival congestion (hyperaemia) and increased iris pigmentation in some patients with mixed coloured irides. Prostaglandin analogues are licensed as first-line medication for glaucoma and have superseded beta-blockers in effectiveness and tolerability. Other prostaglandin-related medications include bimatoprost (Lumigan) and travoprost (Travatan), which have similar mechanisms of action to latanoprost.

Dorzolamide (Trusopt) and brinzolamide (Azopt) are topically administered carbonic anhydrase inhibitors. Their pressure-lowering effect is inferior to that of timolol, but they are useful adjunctive medications.

Brimonidine (Alphagan) is an α_2-adrenergic agonist, which decreases aqueous production and also increases the uveoscleral outflow. It has a pressure-lowering effect comparable with that of timolol. It has the advantage of not having any effect on the respiratory system. It can, therefore, be used in patients with obstructive airway disease.

If the intraocular pressure remains uncontrolled by safe medical treatment and there is evidence of continued loss of visual field, surgical treatment is indicated. A large number of operations have been devised for the management of primary open-angle glaucoma and most of these entail allowing the aqueous to drain subconjunctivally through an artificial opening made in the sclera. The commonest operation performed currently is known as a "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 sutured back into position. Aqueous drains out around the edge of this scleral flap into the subconjunctiva (Figure 12.5). Although most of these operations can reduce the intraocular pressure effectively and often for many years, they all tend to increase the rate of formation of cataract. This and the risk of postoperative endophthalmitis are the main reasons why surgery is usually not considered the first line of treatment in chronic open-angle glaucoma by most ophthalmologists. Often, such surgery is augmented by the use of antifibrotic agents peroperatively, such as 5-fluorouracil and mitomycin C. These agents inhibit fibroblast activity, and increase the success rate of surgery, but carry potential side effects and need to be used cautiously.

In some patients, laser treatment known as "cyclodiode" is applied externally to the eye to lower intraocular pressure by ablating part of the ciliary body (this area produces the aqueous humour). Such treatment, however, is irreversible, and although easier to perform than conventional glaucoma surgery, is generally reserved for patients with advanced uncontrolled glaucoma.

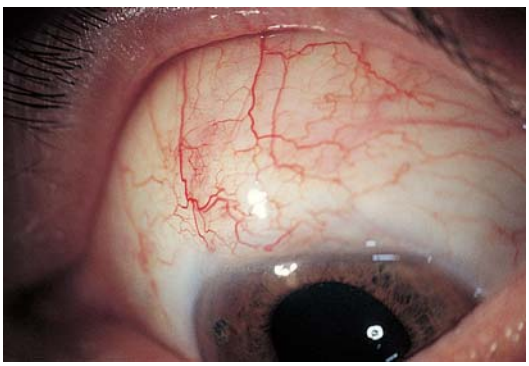


Figure 12.5. Trabeculectomy bleb.

Normal-pressure Glaucoma

This condition is similar to primary open-angle glaucoma except that the intraocular pressure is within normal limits (i.e., 21 mmHg or less at the initial and subsequent visits). The condition is probably caused by low perfusion pressure at the optic nerve head so that the nerve head is susceptible to damage at normal intraocular pressure.

Certain conditions that can mimic normal pressure glaucoma include compressive lesions of the optic nerve and chiasma, carotid ischemia and congenital optic disc anomalies.

Treatment of normal-pressure glaucoma aims to reduce intraocular pressure to 12 mmHg or less.

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 glaucoma require much time and attention. Initially, the nature of the disease must be explained and patients 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.

The initial treatment is a single topical agent – usually a prostaglandin analogue. The second-line treatments of choice are beta-blockers, brimonidine or dorzolamide. Commonly, patients with glaucoma require several different medications to control intraocular pressure effectively.

Increasingly, optometrists are providing specialist care for patients with glaucoma. Some optometrists work alongside hospital consultant-led teams, others work in the community to "refine referrals" and reduce the number of false-positive referrals to a specialist glaucoma clinic.

Acute Angle-closure Glaucoma

This condition is less common than chronic open-angle glaucoma, making up 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 that is liable to develop acute glaucoma: this is a small hypermetropic eye with a shallow anterior chamber. One rarely meets a myope with acute glaucoma in Caucasians (in Asian populations, however, angle closure and myopia more often coexist).

Pathogenesis and Natural History

Eyes that are predisposed to develop closed-angle glaucoma generally have a shallow anterior chamber and are often hypermetropic. There is forward bowing of the iris, which is more evident in these individuals, and the corneal diameter is slightly smaller than in normal eyes. Another factor is the gradual, but slight, increase in size of the lens, which takes place with ageing. Raised intraocular pressure in angle closure is caused by occlusion of the angle by the iris root and it can 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 could be left with chronic angle closure and a picture similar to that of chronic open-angle glaucoma. About half the patients with closed-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 treatment 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 years. 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 mmHg 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. Because it is evening, the patient observes that streetlights 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 might seek attention from the family doctor. Patients present as healthy people with evening headaches associated with blurring of the vision and they are wearing moderately thick convex lenses in their spectacles. Subacute glaucoma is easily missed, partly because it is rare among the large number of sufferers from headache. If attention is not sought at this stage or if the diagnosis is missed, one evening the acute attack develops.

The acute attack. After a number of subacute attacks, an irreversible turn of events can 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 might be called in the following morning, is confronted with a patient who is nauseated and vomiting and at first sight, an acute abdominal problem might 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 (Figure 12.6).

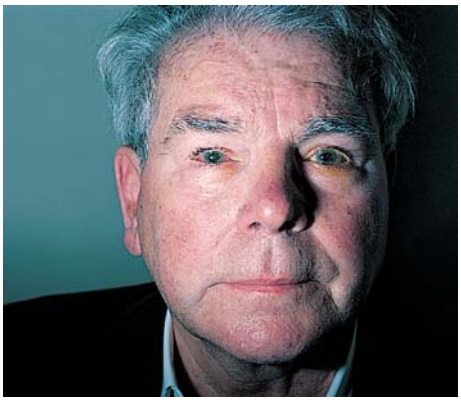


Figure 12.6. Acute angle-closure glaucoma. 🕮

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 that 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 luster 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 might 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 just deep to 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 can provide useful evidence of a previous attack that has resolved spontaneously.

Measurement of the intraocular pressure at this point could reveal a reading of 70 mmHg or more. Gentle palpation of the globe is 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 misleading and the method cannot be used to detect smaller rises in intraocular pressure with any degree of reliability (Table 12.2).

Examination of the other eye will reveal a shallow anterior chamber. Shining a focused

Table 12.2. Signs of acute glaucoma.

- Corneal oedema with resulting poor visual acuity
- Shallow anterior chamber
- Ciliary injection
- Semidilated oval pupil (caused by iris ischaemia)
- Tenderness of globe
- Hard eye

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 can assess the depth of the anterior chamber. 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 instill 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 (Figure 12.7). This instrument is a contact 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 might be necessary to apply a drop of hypertonic glycerol to the cornea to clear the oedema before applying the gonioscope.

The sooner closed-angle glaucoma is diagnosed and treated, the better are the results of treatment. Unfortunately, it is in the early subacute stage of the disease that the diagnosis can 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 can 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 narrow- or closed-angle glaucoma. Only in unsuspected cases of subacute narrow-angle glaucoma is there a real risk of precipitating an acute attack.

Figure 12.7. Preparing for gonioscopy. 🕮

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 regimen would be the application of pilocarpine 4% every minute for 5 min, then every 5 min for an hour, followed by instillation every hour. This treatment is supported by an injection of acetazolamide. If the renal function is unimpaired, acetazolamide can be given intravenously (usually 500 mg) followed by an oral dose of 250 mg four times a day. Topical beta-blockers and/or alpha-agonists, for example apraclonidine (Iopidine), and reduction of inflammation and iris congestion by topical steroids can help achieve a quicker lowering of intraocular pressure. In many cases, these measures relieve the acute attack within hours. However, some patients can require an intravenous infusion of mannitol. 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% four times a day in order to prevent a second disaster.

Once the intraocular pressure has been controlled, the cure is maintained by performing a peripheral iridotomy or iridectomy. This allows the bulging iris bombe to sink backwards like a punctured ship's sail and is a sure means of preventing further acute attacks. Usually, the fellow eye is at risk of a similar problem and is lasered at the same time. 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 might not be adequate and it might 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 develops cataracts in later years. The prognosis in adequately treated narrow-angle glaucoma is, therefore, good, but in the absence of treatment the result is disastrous.

The treatment of narrow-angle glaucoma has undergone a small revolution over the past few years. This is because a new generation of lasers has appeared, which make it possible to perforate the iris quite simply. The yttrium–aluminum–garnet (YAG) laser has replaced surgical iridectomy in most cases. A special contact lens is used to focus the laser on the peripheral iris, and one or two full-thickness openings in the peripheral iris are created. Following such laser treatment, topical steroids and pupil dilatation are given to minimise the effects of uveitis. Occasionally, trabeculectomy surgery is performed if intraocular pressures remain persistently high despite other treatments.

Secondary Glaucoma

The intraocular pressure can 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 typically appears approximately three months after the onset of the condition. The prompt appearance of this painful complication has given it the name of "hundred-day glaucoma". This type of glaucoma is usually difficult to control and even surgical measures can prove ineffective. A typical feature is the appearance of a vascular membrane over the anterior surface of the iris and sometimes 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 can 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 that develops is also resistant to medical treatment. Panretinal laser photocoagulation, when applied early, causes regression of the rubeosis. 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 can 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 topical beta-blockers, for example timolol and levubunolol, might also be required. The type of secondary glaucoma that develops after the iridocyclitis of herpes zoster infections can be particularly insidious. The intraocular pressure can 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.

In iridocyclitis, glaucoma can also be caused by pupil block (inability of aqueous to pass from the posterior to anterior chamber) because of posterior synechiae (adhesions between the iris and lens). Treatment is YAG laser iridotomy.

Secondary to Tumours

Malignant melanoma of the choroid and retinoblastoma can 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 can precipitate a rise in intraocular pressure in a number of different ways. Sometimes, especially in children, bleeding can 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 can occur on the second or third day after the injury, turning a slight event into a very serious problem. On other occasions, a contusion injury might cause splitting or recession of the angle, which is associated with glaucoma. The iridocyclitis that 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 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 a well-recognised phenomenon and "steroid responders" can be identified by measuring the intraocular pressure before and after instilling a drop of steroid. The less potent steroids, hydrocortisone and prednisolone, are less likely to cause this problem and other steroids have been manufactured that have less effect on intraocular pressure, but the anti-inflammatory strength is significantly weaker. The use of systemic steroids can be associated with glaucoma; asthmatics who use steroid inhalers frequently are at a significantly greater risk of developing glaucoma.

The possibility of inducing an attack of acute glaucoma by drugs has already been mentioned.

Secondary to Abnormalities in the Lens

A cataractous lens can become hypermature and swell up, pushing the iris diaphragm forward and obstructing the angle of the anterior chamber. This is referred to as phacomorphic glaucoma. Removing the lens relieves the situation. 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 or Developmental Glaucoma

These glaucomas occur in eyes in which an anomaly present at birth produces an intraocular pressure rise.

This type of glaucoma is extremely rare and it is often, though not always, inherited. This means that the affected child might be brought to the ophthalmologist by the parents because they are aware of the condition in the family. Children could 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.

In primary developmental glaucoma, the glaucoma is caused by a 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" (Figure 12.8).

Figure 12.8. Congenital glaucoma: note the enlarged left cornea (with acknowledgement to Mr R. Gregson).

Other 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 that covers the trabecular meshwork (goniotomy). The other (or secondary) developmental glaucomas include the rubella syndrome, aniridia, mesodermal dysgenesis, Peter's anomaly and the phacomatoses, where the intraocular pressure rise is associated with other ocular and systemic developmental anomalies.

13

Retinal Detachment

Detachment of the retina signifies an inward separation of the sensory part of the retina from the retinal pigment epithelium (RPE). There is an accumulation of fluid in the space between the neural retina and the RPE known as subretinal fluid (Figure 13.1). 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 that can be treated effectively and often dramatically by surgery. Second, retinal detachment can on occasions be the first sign of malignant disease in the eye. Finally, nowadays the condition can often be prevented by prophylaxis in predisposed eyes.

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 a week. It can be seen, therefore, that a doctor in general practice might see a case once in every two or three years, especially if we consider that some retinal detachment patients go directly to eye casualty departments without seeking nonspecialist advice. Although children are sometimes affected, the incidence increases with age and reaches a maximum in the 50–60-year age group. There is a smaller peak in the mid-20s to 30s owing to traumatic detachments in young males.

Certain groups of people are especially liable to develop detachment of the retina: severely shortsighted patients have been shown to have an incidence as high as 3.5% and about 1% of aphakic patients (see Chapter 11) have detachments.

In just under one-quarter of cases, if there is no intervention, 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 that existed during the early development of the eye, as described previously (Chapter 2). 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

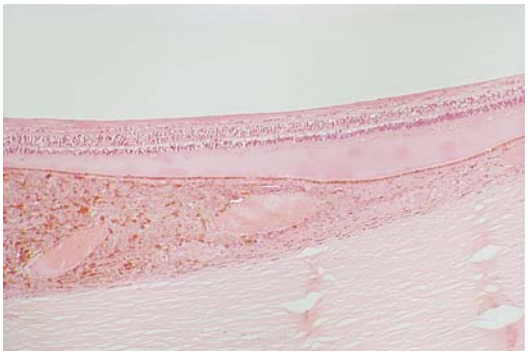


Figure 13.1. Histology of retinal detachment showing the location of subretinal fluid. This eye has an underlying choroidal melanoma. 🕮

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 RPE 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 RPE, the retina can no longer function and the sight is lost in the detached area. Both RPE 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 foveal region is supplied mainly by the choroid. When the retina is detached, the central retinal artery remains intact and continues to supply it because it is also detached with it. The outer half of the retina is deprived of nourishment, being separated from the RPE and choroid. Eventually degenerative changes appear, the fovea 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 can occur over as long a period as one or even two years.

Classification

Detachment of the retina can be classified as follows: rhegmatogenous retinal detachment, tractional retinal detachment, and exudative retinal detachment.

Rhegmatogenous Retinal Detachment

This is the most common form of retinal detachment, caused by the recruitment of fluid from the vitreous cavity to the subretinal space via a full-thickness discontinuity (a retinal "break") in the sensory retina.

Retinal "breaks" can be further subdivided into "tears", which are secondary to dynamic vitreoretinal traction, and "holes", which are the result of focal retinal degeneration (see below).

Tractional Retinal Detachment

This form of retinal detachment develops as a result of tractional forces within the vitreous gel pulling on the retina, causing the retina to be tented up from the RPE. The pure form of tractional retinal detachment is different from rhegmatogenous retinal detachment in that there are no retinal breaks. Examples of tractional retinal detachment include proliferative diabetic retinopathy and vitreomacular traction syndrome.

Exudative Retinal Detachment

This group of retinal detachments also occurs in the absence of retinal breaks. The fluid gains access to the subretinal space through an abnormal choroidal circulation (e.g., from a choroidal malignant melanoma) or, rarely, secondary to inflammation of the RPE or deeper layers of the eye (e.g., scleritis).

Rhegmatogenous Retinal Detachment

The Presence of Breaks in Retinal Detachment

It was noticed as long ago as 1853, only a short time after the invention of the ophthalmoscope, that many detached retinae have minute full-thickness discontinuities (breaks) in them, but it was not until the 1920s that the full significance of these breaks as the basic cause of the detachment became realised. The breaks can

be single or multiple and are more commonly situated in the anterior or more peripheral part of the retina. In order to understand how these breaks occur, it is necessary to understand something of retinal degeneration and vitreous changes.

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 peripheral parts of the retina supplied by the distal part of the circulation. Peripheral retinal degenerations are more commonly seen in myopic eyes, especially in association with Marfan's and Ehlers–Danlos syndromes and Stickler's disease (see reading list).

Different types of degeneration have been described and named and certain types are recognised as being the precursors to formation of retinal breaks. The most important degenerations are lattice degeneration and retinal tufts. Lattice degenerations consist of localised areas of thinning in the peripheral retina. Progressive thinning of the retina within areas of lattice degeneration can eventually lead to formation of retinal "holes".

In addition, both lattice degenerations and retinal tufts also represent areas with abnormally strong adhesions between the vitreous and the retina. The presence of exaggerated vitreoretinal adhesions can result in the formation of retinal "tears" within areas of lattice degeneration and retinal tufts during posterior vitreous detachment (see below).

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 attachment, swings back to its original position again. The vitreous is usually perfectly transparent but 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 (vitreous floaters). 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.

Posterior Vitreous Detachment

Vitreous floaters are commonplace and tend to increase in number as the years pass. But the vitreous undergoes a more dramatic change with age. Often in the late 50s, it becomes more fluid and collapses from above, separating 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. This then is the process known as posterior vitreous detachment (PVD).

When this happens, the patient might 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. On the other hand, it is also common to find a detached vitreous in an elderly person's eye in the absence of any symptoms.

Retinal Breaks Formation

In the majority of eyes the vitreous separates "cleanly" from the retina during PVD. Such "uncomplicated" PVD is common and is usually of no pathological significance. Unfortunately, on rare occasions, the collapsing vitreous causes a retinal "tear" to form at a point of abnormally strong adhesion between vitreous and retina, for example within an area of lattice degeneration or retinal tufts. There might even be an associated vitreous haemorrhage, when the PVD causes the avulsion of a peripheral retinal blood vessel.

Mechanism of Rhegmatogenous Retinal Detachment

Once a retinal tear forms as a result of abnormal vitreous traction following PVD, the fluid

from within the vitreous cavity can gain access to the subretinal space through the retinal tear. The progressive accumulation of fluid in the subretinal space eventually causes the retina to separate from the underlying RPE, similar to wallpaper being stripped off a wall. This inward separation of the retina from the RPE through the recruitment of fluid via a retinal break is the basis for "rhegmatogenous" retinal detachment, which is the most common form of retinal detachment.

Rhegmatogenous Retinal Detachment Associated with Trauma

Most rhegmatogenous retinal detachments occur as a result of spontaneous PVD-induced retinal breaks. However, retinal tears can also occur as a result of trauma. 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 or the superior nasal 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.

Signs and Symptoms of Retinal Tear and Retinal Detachment

Let us now consider a typical patient, possibly a myope in the mid-50s, either male or female, who suddenly experiences the symptoms of "flashes and floaters", sometimes spontaneously or sometimes after making a sudden head movement. 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 min. 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 can 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 these symptoms 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 a 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.

Shadow

Once a retinal tear has appeared, the patient might seek medical attention, and effective treatment of the tear can ensue. Unfortunately, some patients do not seek attention, or, if they do, the symptoms might be disregarded. Indeed, in time the symptoms might become less, but

after a variable period between days and years, a black shadow is seen encroaching from the peripheral field. This can appear to wobble. If the detachment is above, the shadow encroaches from below and it might seem to improve spontaneously with bedrest, being at first better in the morning. Loss of central vision or visual blurring occurs when the fovea is involved by the detachment, or the visual axis is obstructed by a bullous detachment. Inspection of the fundus at this stage shows that fluid seeps through the retinal break, raising up the surrounding retina like a blister in the paintwork of a car. A shallow detachment of the retina can be difficult to detect but the affected area tends to look slightly grey and, most importantly, 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 corrugated and the retinal vessels look darker than in flat retina. The tear in the retina shines out red as one views the RPE 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.

Tractional Retinal Detachment

In tractional retinal detachment, the retina can be pulled away by the contraction of fibrous bands in the vitreous. Photopsiae and floaters are usually absent but a slowly progressive visual field defect is noticeable. The detached retina is usually concave and immobile.

Advanced proliferative diabetic retinopathy can be complicated by tractional retinal detachment of the retina when a contracting band tents up the retina by direct traction. Not infrequently such a diabetic patient experiences further sudden loss of vision in the eye, when the traction exerted by the contracting vitreous pulls a hole in the area of tractional retinal detachment, resulting in a combined rhegmatogenous and tractional retinal detachment.

Exudative Retinal Detachment

In such detachments, there are no photopsiae but floaters can occur from associated vitritis or vitreous haemorrhage. A visual field defect is usual. Exudative detachments are usually convex shaped and associated with shifting fluid.

A malignant melanoma of the choroid might 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 can become difficult. It is important to avoid performing retinal 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 can be confirmed by transilluminating the eye to reveal the tumour.

Retinal detachments secondary to inflammatory exudates are not common. One example is Harada's disease, which is the constellation of exudative uveitis with retinal detachment, patchy depigmentation of the skin, meningitis and deafness. Its cause is unknown. Exudative detachments do not require surgery but treatment of the underlying cause.

Management of Rhegmatogenous Retinal Detachment

Prophylaxis

Retinal tears without significant subretinal fluid can be sealed by means of light coagulation. A powerful light beam from a laser is directed at the surrounds of the tear (Figure 13.2). This produces blanching of the retina around the edges of the hole and, after some days, migration and proliferation of pigment cells occurs from the RPE into the neuroretina and the blanched area becomes pigmented. A bond is formed across the potential space and a retinal detachment is prevented. This procedure can be carried out, with the aid of a contact lens, in a few minutes.

A wider and more diffuse area of chorioretinal bonding can be achieved by cryopexy,

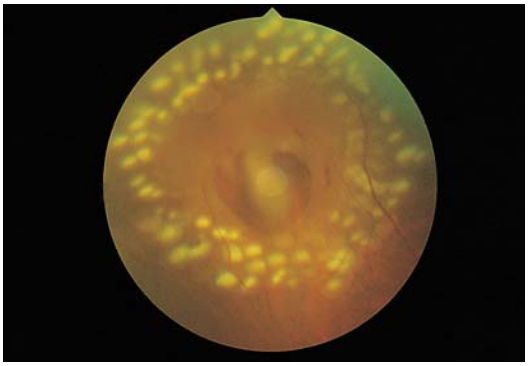


Figure 13.2. Laser photocoagulation of retinal tear (with acknowledgement to Mr R. Gregson).

which entails freezing from the outside. Cryopexy is occasionally necessary if the retinal hole is peripheral, or when there is limited blanching of the retina from laser photocoagulation because of the presence of vitreous haemorrhage. 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 (as occurs after photocoagulation) develops following this treatment, but it tends to be uncomfortable for the patient and local or general anaesthesia is required.

Retinal Surgery

In the early part of the twentieth century, it was generally accepted that there was no known effective treatment for retinal detachment. It was realised that a period 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 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 (Figure 13.3). 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 that 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 retinal reattachment surgery is carried out using either the cryobuckle or vitrectomy technique.

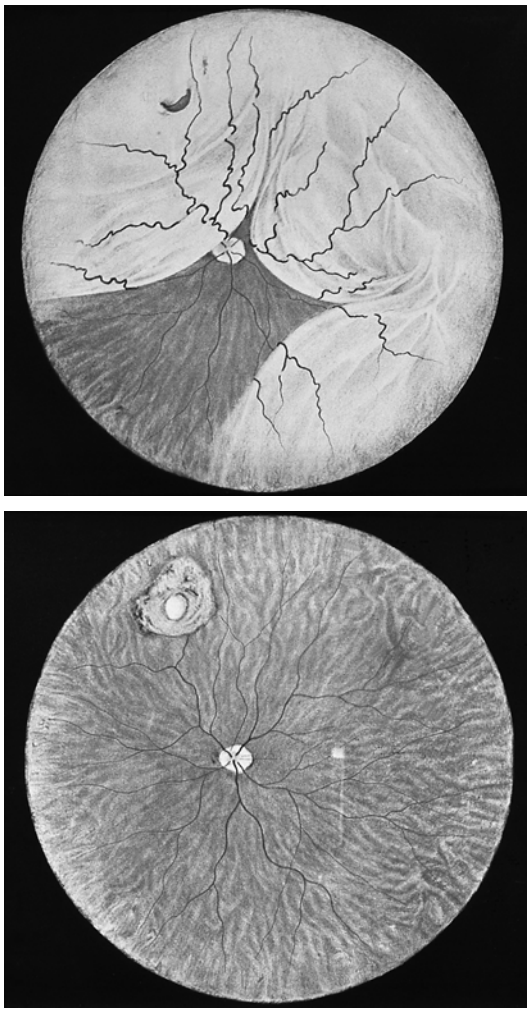


Figure 13.3. Retinal detachment **a** before and **b** after treatment. (After Gonin).

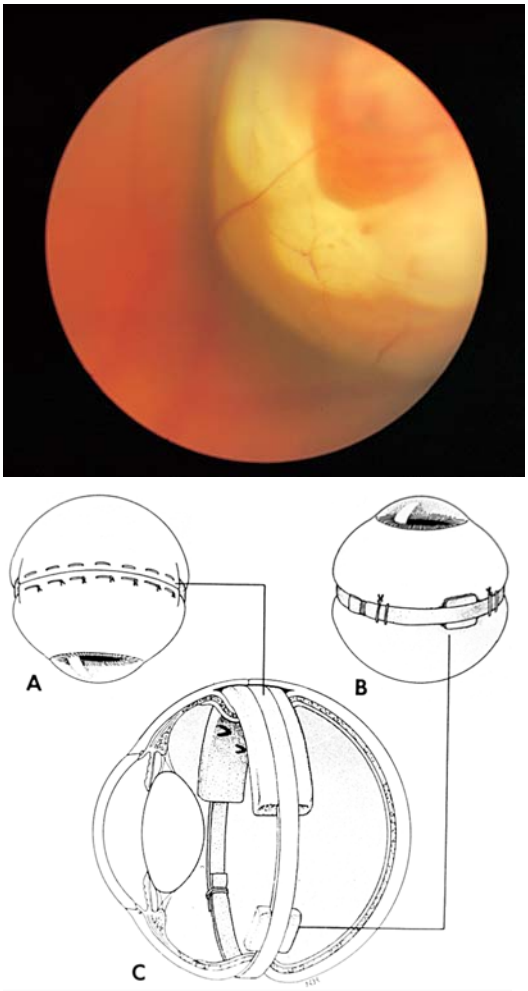


Figure 13.4. a Retinal detachment surgery: retinal tear surrounded by cryopexy and covered by indent. **b** Retinal detachment surgery: indent and encirclement band (with acknowledgement to Professor D. Archer).

Cryobuckle

This 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 (Figure 13.4). This is combined with cryopexy to the break. It is often necessary to drain off the subretinal fluid and inject air or gas into the vitreous. In more difficult cases, the eye can be encircled with a silicone strap to provide all-round support to a retina with extensive degenerative changes.

Vitrectomy

The detached retina can also be reattached from within the vitreous cavity. This involves the use of fine-calibre instruments inserted through the pars plana into the vitreous cavity. A light probe is used to illuminate the operative field, while a "vitrectomy cutter" is used to remove the vitreous, hence relieving the abnormal vitreous adhesions that produced the retinal tear in the first instance (Figure 13.5). The detached retina is "pushed back" into place from within and temporarily supported by an internal tamponade agent (air, gas or silicone oil) while the retina heals. The retinal breaks are identified and treated by either laser photocoagulation or cryopexy at the same time. Vitrectomy can also be combined with a silicone strap encirclement if further support of the peripheral retina is needed.

Historically, vitrectomy is reserved for the more difficult and complex cases of rhegmatogenous retinal detachment, where multiple tears and posteriorly located tears are present, or as a "salvage" operation following failed cryobuckle. With advances in instruments, vitrectomy is increasingly being used as the primary operation for the repair of most acute PVD-related rhegmatogenous retinal detachments, regardless of the complexity of the detachment.

Prognosis

The retina can now be successfully reattached by one operation in about 85% of cases. Of the successful cases, those in which the macular region was affected by the retinal detachment

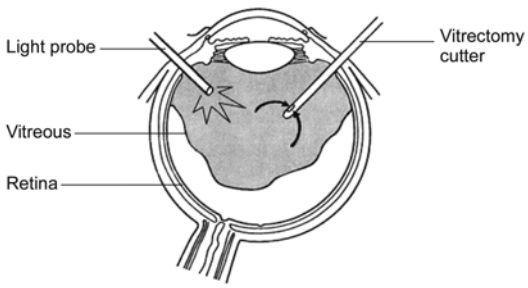


Figure 13.5. Vitrectomy.

do not achieve a full restoration of their central vision, although usually the peripheral field recovers. The degree of recovery of central vision in such macula-detached cases depends largely on the duration of the macula detachment before surgery. Even when the retina has been detached for two years, it is still possible to restore useful navigational vision.

The main cause of failure of modern retinal reattachment surgery is proliferative vitreoretinopathy. This is characterised by excessive "scarring" following initial retinal reattachment surgery, with the formation of fibrous tractional membrane within the eye, resulting in recurrent detachment of the retina.

When retinal surgery has failed, further surgery might 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, it is helpful to the patient if he is warned about this before the treatment is started.

14

Squint

The word "squint" refers to a failure of the visual axes 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 worker notices a deterioration in the ability to perform fine tasks and the elderly notice that they pour tea into the saucer rather than the cup. In time, depth perception might improve and the patient adapts to the defect to some extent; children can 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 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 are 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 might fail. The ability to use the eyes together is called binocular vision. It can be measured and graded by presenting each eye separately, but simultaneously, with a series of images. The instrument used to do this is called a synoptophore (Figure 14.1).

1. *Simultaneous macular perception* is said to be present if the subject can see two dissimilar images that are presented simultaneously to each eye, for example a triangle to one eye and 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, for example a picture of a house to one eye and a picture of a chimney to the other, and the whole picture is maintained as one as the eyes converge. The range of fusion can be measured in degrees.
3. *Stereopsis*, the third grade of binocular vision, is present if, when 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 fine degrees of impairment of binocular function need to be measured.

This ability of ours to put together the images from each eye and make a single picture in our minds seems to develop during the early years of life and furthermore, its development seems to depend on visual input. Below the age of eight

Figure 14.1. The synoptophore. An instrument for measuring the angle of deviation of a squint and the ability of the eyes to work together. 🕮

years, any misalignment of the eyes that disturbs binocular vision can permanently damage this function.

If the alignment of the eyes is disturbed for any reason during childhood, the child might at first, as one might expect, notice double vision but 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. Suppression is a temporary switching off of one eye when the other is in use, whereas amblyopia of disuse is a permanent impairment of vision, which could affect the career prospects of the patient. Amblyopia of disuse can also occur if the sight of one eye is defective as the 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 eight years. Covering one eye of a baby could lead to permanent impairment of the vision of that eye, as well as impairment of the ability to use the eyes together. An adult can have one eye covered for many months or even 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

In lay terms, the word squint can just mean screwing up the eyes but here we are referring to a deviation of one eye from the line of sight. This might be present all the time or just when the patient is tired. It is important to notice whether the eye movements are normal. For example, if there is weakness of one lateral rectus muscle, the affected eye will not turn outward and the angle of the squint will be much greater when looking to that side. Most childhood squints are not associated with weakness of one or more extraocular muscles so that the angle of the squint is the same in all directions of gaze. The deviation of the squint can be horizontal or sometimes vertical or the eyes could be convergent or divergent.

Squint in Childhood

During the first few weeks of life the eyes might seem to wander about aimlessly with limited ability to fix. Between the ages of two and six months, fixation becomes steadier even though the fovea is not fully developed, and by the age of six months convergence on a near object can 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 these early months of life. If the eyes are definitely squinting at the age of six months, urgent referral to an ophthalmologist is indicated. Before this or when there is some doubt, referral to an orthoptic screening service can be considered. These have been set up in many parts of the country. Orthoptists might be regarded as "physiotherapists of the eyes" and they are trained to examine the eye movements in great detail. We need to detect squints early in children for the following reasons:

1. The squint could be caused by serious underlying intracranial or intraocular disease.
2. The squint can result in amblyopia, which is more effectively treated, the younger the child.
3. The cosmetic effect of a squint is an important consideration.

Amblyopia of Disuse

A special word is needed about this curious condition, which accounts for unilateral impairment of vision in over 2% of the population. Any eye casualty officer is familiar with the patient with a foreign body on the cornea of one eye and the other eye being amblyopic. ("How can I drive home with this patch on, doctor?") The words "lazy eye" are sometimes used but in lay terms this can also mean squint.

The eye suffering from amblyopia of disuse shows certain features:

- Impaired Snellen visual acuity but usually able to decipher vertical lines of letters better than horizontal ones.
- Normal fundus.
- Small residual squint or, if not, the affected eye relatively hypermetropic.
- An indefinite central scotoma, which is difficult to assess by routine visual field testing.
- History of poor vision in one eye since childhood.

The diagnosis of amblyopia can be by exclusion but it must never be reached without a careful examination of the eyes. In recent years, there has been a considerable research interest in this subject and there appear to be nerve conduction anomalies in the occipital cortex, which can be induced by visual deprivation.

Causes of Squint in Childhood

- Refractive error – hypermetropia, myopia.
- Opaque media – corneal opacities, cataract, uveitis.
- Disease of retina or optic nerve – retinoblastoma, optic atrophy.
- Congenital or acquired weakness of extraocular muscles.
- 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 the object at once. A given amount of accommodation must, therefore, be associated with an equivalent amount of convergence. In hypermetropic subjects this relationship is disturbed. In order to overcome hypermetropia, the eyes must accommodate and sometimes this excessive focusing induces an excess of convergence, hence causing a squint. This type of accommodative squint can be fully corrected by wearing spectacles: when the glasses are on, the eyes are straight; and when they are off, one eye turns in. More often, the squint is only partially accommodative and is improved, but not eliminated, by wearing glasses. The convergent squint associated with hypermetropia is the commonest type of childhood squint.

Opaque Media

Congenital cataract can occasionally present as a squint. In a similar manner, a corneal opacity, as might result from herpes simplex keratitis or injury, can cause a squint to appear. 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

Sixth, third or fourth cranial nerve palsies are sometimes seen after head injuries and the surgeon must always bear in mind the possibility of a sixth or other cranial nerve palsy being associated with raised intracranial pressure. Myasthenia gravis is extremely rare in children but it can present as a squint. In some cases of squint there is a degree of facial asymmetry. These patients might also have "asymmetrical eyes", one being myopic or hypermetropic relative to the other. Sometimes there is no refractive error but there might be an asymmetry of

the insertions of the extraocular muscles as a possible cause of squint. There is a group of conditions, known as musculofascial anomalies, in which there is marked limitation of the eye movements from birth in certain directions. They are accompanied by abnormal eye movements, such as retraction of the globe and narrowing of the palpebral fissures on lateral gaze.

Overaction of muscles can cause a squint. This is seen in school children sometimes with a background of domestic or other stress. The eyes tend to overconverge and overaccommodate, especially when being examined.

Abnormalities of Facial Skeleton

This is not a common cause but it should be kept in mind.

Diagnosis

History

When faced with a case of suspected squint, certain aspects of the history can be 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 might be no true squint but merely the appearance of one. The mother herself is usually the best witness. Unfortunately, some children have a facial configuration that 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 in order to avoid sending unnecessary referrals to the local eye unit (Figure 14.2). Childhood squints often show a dominant pattern of inheritance and the family history provides a useful diagnostic indicator. From the point of view of prognosis, it is useful to find out whether the squint 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 that might have caused or initiated 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 technique 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 eyes 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 seen easily. 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 reflections is then noted carefully. The more mobile the child, the less time there is to observe this. If there is a squint, the reflections will be positioned asymmetrically in the pupil. If the patient has a left convergent squint, the reflection from the left cornea is displaced outward towards the pupil margin. A rough assessment of the angle of the squint can be made at this stage by noting the abnormal

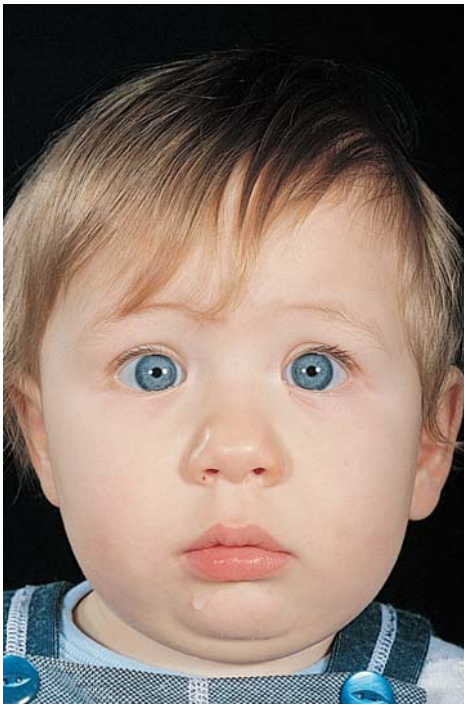


Figure 14.2. Pseudosquint. The configuration of the eyelids gives the appearance of a squint but the corneal reflexes show that this is not the case. 🕮

position of the reflection. One of the difficulties experienced at this point is because of the continuous movement of the child's eyes, which makes 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 reflections have been examined, the cover test can be performed. Once again the reflection of light from each eye is noted, but this time one of the eyes is smartly covered, either with the back of the hand or a card. If the fixating eye is covered, a movement of the nonfixing eye to take up fixation can then be observed (Figure 14.3). After some practice, it is possible to detect even slight movements of this kind. The result of the test can be misleading if the nonfixing eye is too weak to take up fixation, and quite often, an assessment of the vision of the nonfixing eye can be made at this stage.

If, having performed this first stage of the cover test, no deviation can be detected, the cover can be quickly swapped from one eye to the other and any movement of the covered eye 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 must be repeated with the patient looking at 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 might 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.

After the cover test has been performed, it is necessary to test the ocular movements to determine whether there is any muscle weakness. At this stage, it is usual to instil a mydriatic and cycloplegic drop (e.g., cyclopentolate 1% or 0.5%) in order to obtain a measure of the refractive error, by retinoscopy, when the eyes are completely at rest. Next, the optic fundi are 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.

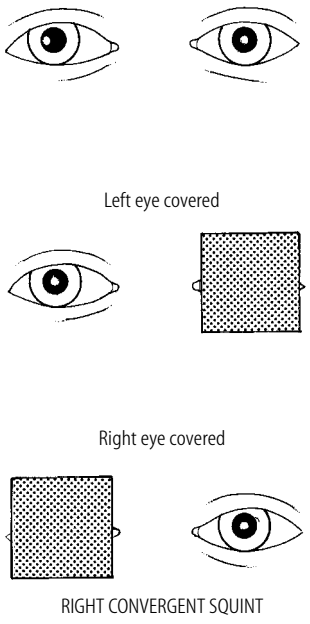


Figure 14.3. The cover test.

Management of Squint in Childhood

Glasses

Any significant 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 can be prescribed in a child as young as six to nine 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 bedtime, a previous squint might appear to become even worse and the parents should be warned about this possible rebound effect.

Orthoptic Follow-up

The orthoptic department forms an integral and important part of the modern eye unit. It is run and manned by orthoptists who carry out the careful measurement of visual acuity with and without glasses and the measurement of eye

movements and binocular function. Once the patient has been seen for the initial visit, follow up in the orthoptic department is arranged and 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 seven or eight years, 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. Sometimes atropine eye drops are used as an alternative to patching one eye. Orthoptic exercises can 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 ask if an operation can be carried out as a substitute for wearing glasses. Unfortunately, surgery to correct refractive error is not yet at a stage where it can be applied to children with squints. Squint surgery involves moving the muscle insertions or shortening the muscles and from the cosmetic point of view is highly effective. The adjustment of the muscles is measured in millimetres to correspond with the angle of the squint in degrees. Sometimes two or more operations are needed because of occasionally unpredictable results, but from the cosmetic point of view, nobody need suffer the indignity of a squint, even though a series of operations might be needed. 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 common problem, but only a small proportion of these cases 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 early surgery.

The aim of treatment for a child with squint is to make the eyes look straight, to make each eye see normally and to achieve good binocular vision. Unfortunately, all too often, the first one of these aims alone is achieved in spite of modern methods of treatment. The fault might lie partly in late referral or difficulty with patient co-operation but better methods of treatment are needed.

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 these muscles.

Anatomy of the Extraocular Muscles

The extraocular muscles can be divided into three groups: the horizontal recti, the vertical recti and the obliques.

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 mm or 8 mm behind the limbus above and below the globe. The action of these muscles depends on the initial position of the eye. For example, 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 anteroposterior axis through the globe. The important thing to realise is that the action of these muscles depends on the position of the eye (Figure 14.4).

The Obliques

These are also vertical yoke muscles but they run on a different line to the vertical recti. The superior oblique depresses the adducted eye (makes

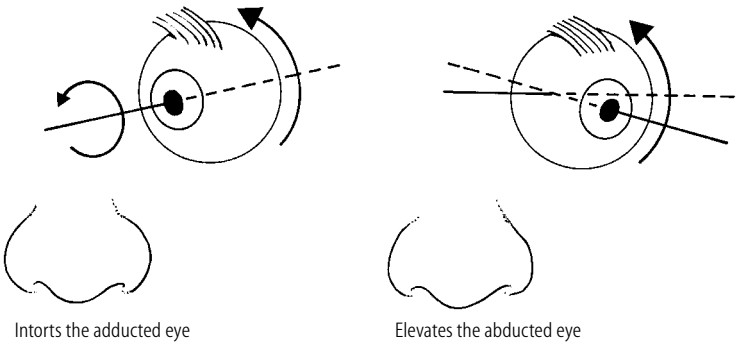


Figure 14.4. Primary and secondary actions of the superior rectus muscle. 🕮

the eye go down when it is turned in) and the inferior oblique elevates the adducted eye.

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 because of the defective action of the 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 or turn outwards. A right convergent squint is seen and the patient experiences double vision, which is worse when looking to the right. There might be a head turn to the right.

When a patient has a third cranial nerve palsy on the right side, 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 limited.

Causes of Adult Squint

Adults who present with a squint usually have a well-defined ocular muscle palsy. This can be caused by a pathological process at any point from the brain, through the nerve to the muscle. This will be discussed elsewhere but 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 concomitant squints neglected from childhood. 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 can be due solely to impaired vision in one eye.

Diagnosis

In contrast to the situation with children, who usually present with concomitant squint associated with hypermetropia, 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. In the latter case, the patient tends to complain more of blurred vision and confusion.

A carefully taken history can 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 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 eye 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.

Maddox Rod

The Maddox wing measures the deviation at reading distance and the Maddox rod is a similar device to measure the deviation when viewing a distant object. A special optical 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 using either coloured filters or a mirror. The system is arranged so that a screen is viewed with one eye and the end of a pointer with the other. The patient is told to place the pointer on various points on the screen. If the eyes are not straight, the pointer is placed away from the correct position. A map of the incorrect positions is made (Figure 14.5). The shape of the map is diagnostic of particular ocular muscle problems and serial records can be helpful in assessing progress.

Treatment

Many cases of adult squint recover spontaneously within a period of three to six months. Once the cause of the squint has been investigated, the immediate treatment entails eliminating the diplopia by occluding one or other eye. This can be conveniently achieved by applying adhesive tape to the spectacle lens. If the angle of the squint is sufficiently small, it might 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 nine months or more, 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 diplopia because of 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.

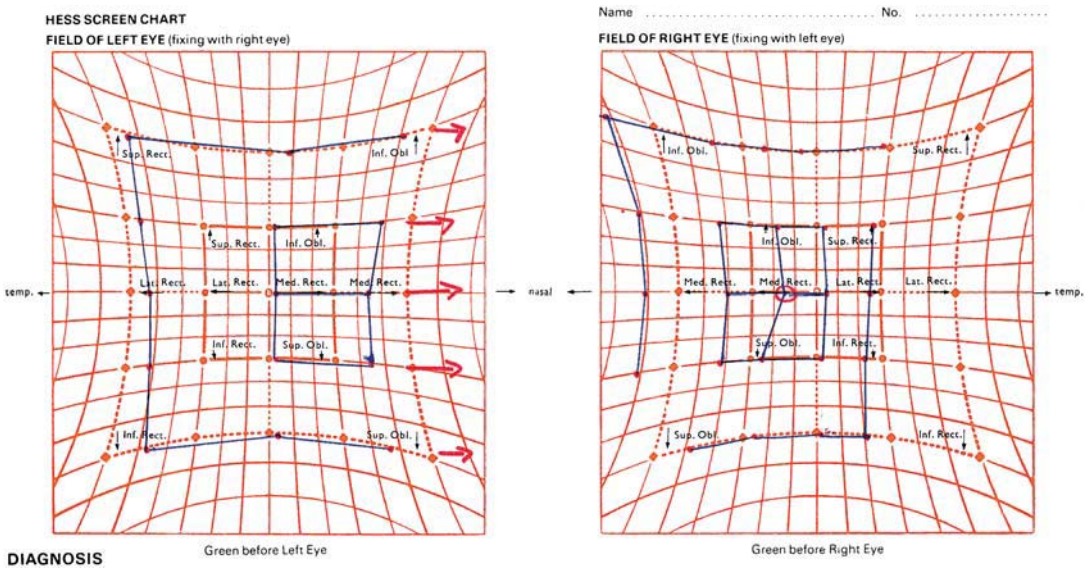


Figure 14.5. Hess chart depicting a right lateral rectus palsy. 🕮

Ocular Muscle Imbalance

Mild latent squints can sometimes go undetected until a period of stress or perhaps excessive reading precipitates symptoms of eyestrain and headache. The effort to maintain both eyes in line causes the symptoms. The latent deviation could be inward or outward but because most people's eyes tend to assume a slightly divergent position when completely at rest, a degree of latent divergence (exophoria) is almost the rule and of no significance. Vertical muscle imbalance is less well tolerated and even a slight deviation can 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 can produce dramatic relief, but we must always remember that the appearance of an ocular muscle imbalance might be the first indication of more serious disease. A small vertical deviation can be the first sign of a tumour of the lacrimal gland or thyrotoxic eye disease and a wide range of investigations might be needed before one can be satisfied with the excellent but sometimes deceptive results of symptomatic treatment.

15

Tumours of the Eye and Adnexae

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.

Choroidal Melanoma

The most common primary intraocular tumour is the malignant melanoma of the choroid. In white people, the tumour has an incidence of one in 2500 and the average age at presentation is 50 years. The incidence rises with age with a peak at 70 years. However, it is important to appreciate that no age is exempt because choroidal melanomas have been reported in children as young as three years. It is extremely rare in black people. It differs from melanoma of the skin in that it grows more slowly and metastasises late. Most choroidal melanomas are thought to originate from choroidal nevi, which are present in up to 10% of the population. At first, it is seen as a raised pigmented oval area, which can be anywhere in the fundus (Figure 15.1). It is usually brown in colour although it can be amelanotic (or greyish). As the tumour enlarges there might be an associated exudative retinal detachment or, less often, secondary glaucoma. Other associated features might include choroidal haemorrhage and serial photography might be needed to confirm the growth. The usual presentation is with decreased vision or a visual field defect. Diagnosis is confirmed with careful clinical examination, including indirect ophthalmoscopy and slit-lamp biomicroscopy (contact lens or volk lens examination), fluorescein angiography, ultrasonography and transvitreal fine-needle aspiration in equivocal cases. The most common site for metastases is the liver, so abdominal ultrasound, serum liver function tests, and chest X-ray should be performed at regular intervals. The appearance of liver metastases can be delayed for several years and can occur even if the eye has been removed, signifying micrometastases at the time of presentation. Approximately 40% of patients develop liver metastases within ten years of the initial diagnosis, while the estimated five-year mortality rate for treated medium-size melanomas is between 15% and 23%. The differential diagnosis of choroidal melanoma includes retinal detachment, metastatic choroidal tumours, wet macular degeneration, large choroidal nevi, choroidal haemangioma or choroidal effusion. Historically, treatment involved enucleation (removal of the globe); today, however, many alternative eye-sparing treatments are available, partly dependent on the size and local spread of the tumour. Options include radiotherapy (external plaque, proton beam or helium ion),

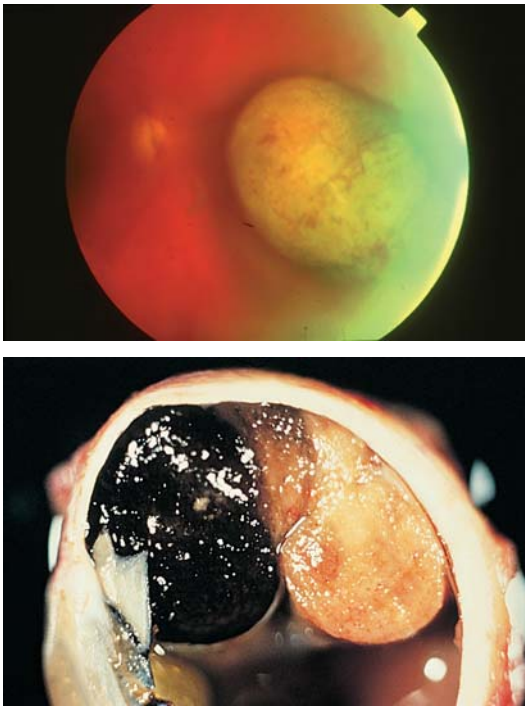


Figure 15.1. Choroidal melanoma poorly pigmented (amelanotic) melanoma **a** fundus photograph. **b** Bisected eye showing pigmented and nonpigmented portions of melanoma in same eye (with acknowledgement to Mr A. Foss).

laser photocoagulation for small lesions, local resection and transpupillary thermotherapy. Untreated, the tumour can extend into the orbit and provide an unpleasant problem for the patient.

Choroidal Metastases

These make up the most common intraocular tumours in adults. Although lesions can be demonstrated in at least 1–2.5% of patients with carcinomas, many cases remain asymptomatic unless the macula is involved. In males, the most common primary tumour is found in the lung and in females, it is the breast. The metastatic tumours are usually treated with external beam radiotherapy.

Retinoblastoma

This is a rare tumour of childhood, which arises not from the choroid but, as its name suggests, from the retina. It is, however, the commonest primary intraocular tumour in children, with an incidence of one in 15,000 live births. It shows certain rather strange and unusual features. It is not usually present from birth, but occurs most frequently in infancy to age three years (although it can occur in older patients); it is either inherited as an autosomal dominant trait or can be sporadic in nature. Approximately 40% of cases are considered to be inherited. In one-third of inherited cases it appears in both eyes. A change in the RB1 gene on chromosome 13 is found in the inherited cases. Initially, it can 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 might be in the extreme periphery of the fundus. A larger tumour can present as a white mass in the pupil ("leucocoria") and such an appearance in infancy demands immediate referral to an ophthalmologist (Figure 15.2). Other presenting features include strabismus, secondary glaucoma, proptosis or intraocular inflammatory signs. Computed tomography (CT) scanning and ultrasound might show a calcified intraocular mass. Extension tends to occur locally along the optic nerve and enucleation is often life saving. Until recently, enucleation was the treatment of choice and cure rates of 90–95% were achieved. Nowadays, eye-sparing therapy is preferred, in an attempt at avoiding the physical and psychological trauma involved in enucleating a young child. Alternative treatment options include initial systemic tumour chemoreduction with carboplatin-based regimens, followed by external beam radiotherapy, plaque radiotherapy, cryotherapy or laser photocoagulation. 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 iris tumour usually presents as a solitary iris nodule, which might or might not be pigmented. It can cause distortion of the pupil, which can be an early warning sign. Other features that can point to the diagnosis are localised lens opacity, iris neovascularisation and elevation of intraocular pressure. Melanoma of the iris is extremely slow growing and probably much less malignant than choroidal melanoma,

Figure 15.2. Retinoblastoma: leucocoria.

with a survival rate of at least 95% at five years. Treatment is usually in the form of a sector or total iridectomy.

The Eyelids

Benign Tumours

Meibomian Cysts (Chalazion)

This is the commonest eyelid lump in all ages. It is caused by blockage of the meibomian gland orifice such that the secretions accumulate. A granulomatous inflammation is set up, which results in a painless, round, firm, slowly growing lump in the tarsal plate (Figure 15.3). The cyst can become infected, when it becomes red hot and painful. Treatment is by incision and curettage.

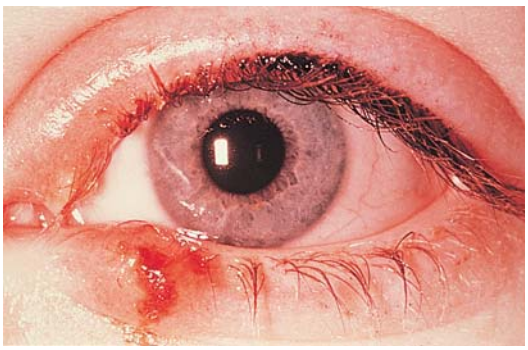


Figure 15.3. Chalazion.

Molluscum Contagiosum

This is caused by a viral infection and is most commonly seen in children. The lesions consist of several pale, waxy, umbilicated nodules on the eyelids and face. Similar lesions can be located on the trunk. The eyelid lesions shed viral particles, which produce a chronic conjunctivitis and less often superficial keratitis. The lesions might disappear in about six months, but can need curettage or cautery.

Papilloma

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

Seborrhoeic Keratosis

This is common in the elderly and consists of slow-growing, sessile, greasy lesions of the eyelid. They are usually brown and friable.

Senile Keratosis

Senile keratosis consists of multiple, flat, scaly lesions, which can occasionally undergo transformation into a squamous cell carcinoma.

Xanthelasma

These are slightly elevated lesions consisting of lipid deposits usually on the medial aspect of the eyelids. They can be associated with hyperlipidaemia, especially in the younger patient.

Keratoacanthoma

This is an example of a lesion that grows rapidly, too rapidly for a neoplasm, over a period of a few weeks and then resolves spontaneously (Figure 15.4). It usually starts as a red papule, which grows quickly into a nodule with a keratin-filled crater. The lesion can resemble a basal cell carcinoma. Small lumps on the eyelids should be removed and biopsied. Larger lumps can be biopsied by taking a small segment from them before 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.

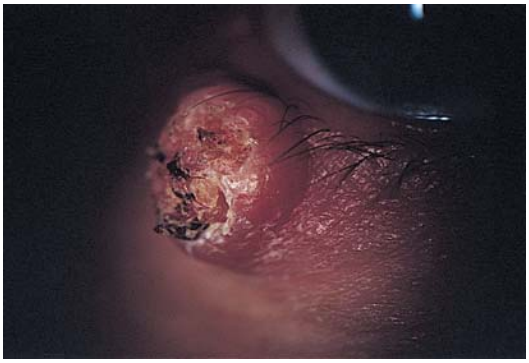


Figure 15.4. Keratoacanthoma (with acknowledgement to Mr A. Sadiq).

Kaposi Sarcoma

This is a well-known association with acquired immune deficiency syndrome (AIDS). The lesions consist of purple nodules on the eyelid and similar lesions in the lower conjunctival fornix composed of proliferating endothelial and spindle-shaped cells. Inflammatory cells might also be present with vascular channels without endothelial cell lining. Human herpes virus 8 is thought to be important in the pathogenesis of these lesions.

Benign Vascular Tumours of the Eyelids

These fall into three types: capillary haemangioma of the newborn (strawberry naevus), cavernous haemangioma and telangiectatic haemangioma.

Capillary Haemangioma of the Newborn (Strawberry Naevus)

This is usually seen before the age of six months, and nearly all examples regress spontaneously, usually in few months and by the age of five years. Tumours appear as red, slightly raised marks on the skin. Even 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 that occludes the eye, causing amblyopia. Larger tumours can produce orbital enlargement. If treatment is required, intralesional steroid injections have proved beneficial.

Cavernous Haemangioma

These tumours lie more deeply in the skin and appear as a bluish swelling in the lid, which expands when the child cries. These lesions can also disappear spontaneously or, if persistent, they can be treated by freezing.

Telangiectatic Haemangioma

Also known as the port wine stain or naevus flammeus, 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 discolouration in the skin (Figure 15.5). 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. There can be hypertrophy of the affected area of the face, leading to asymmetry.

Malignant Tumours of the Eyelids

Basal Cell Carcinoma

This is the most common malignant tumour of the eyelid in adults (80–90% of cases). Patho-

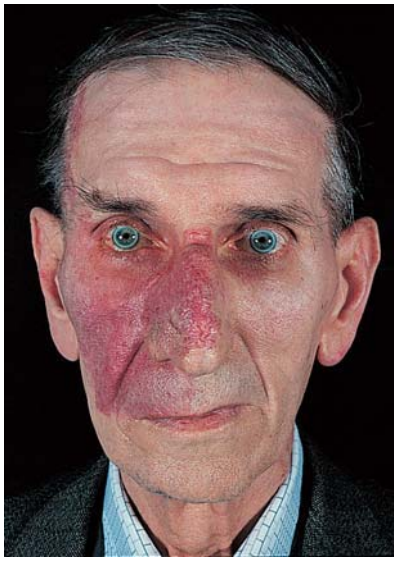


Figure 15.5. Port wine stain (naevus flammeus).

genesis is related to exposure to ultraviolet light, hence it most frequently involves the lower lid and medial canthus. The tumour begins as a small insignificant nodule, which turns into a small crater-like lesion with a slightly raised, pearly-coloured edge with fine dilated blood vessels on its surface (Figure 15.6). Although the tumour rarely metastasises, it is locally invasive, and, therefore, early diagnosis and treatment is important. In the early stages, it is a simple matter to remove the lesion and confirm the diagnosis by biopsy, but if left the tumour tends to spread into surrounding structures and into the underlying bone and orbit (Figure 15.7). Treatment depends on the size, extent and location of the tumour. Usually, surgical excision with wide margins is the technique of choice, either by a simple excisional biopsy or by the more complex Mohs' procedure. The more extensive, neglected basal cell carcinomata are treated by radical surgery, cryotherapy or palliative radiotherapy.

Squamous Cell Carcinoma

Squamous cell carcinoma is the second most common malignant eyelid lesion and constitutes 5–10% of cases. It occurs most commonly in the elderly and is related to sunlight exposure. The tumour can initially resemble a basal cell carcinoma, although the edges are usually not rolled. Spread tends to occur to the local lymph nodes (preauricular for the upper lid and submandibular for the lower lid). Treatment is similar to a basal cell carcinoma.

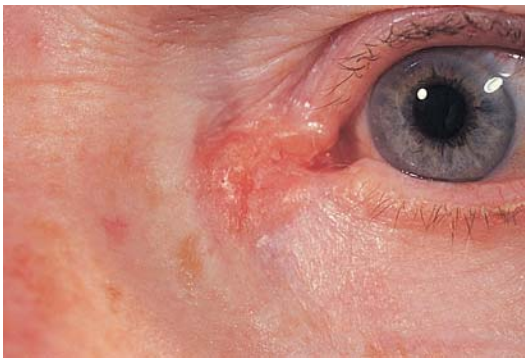
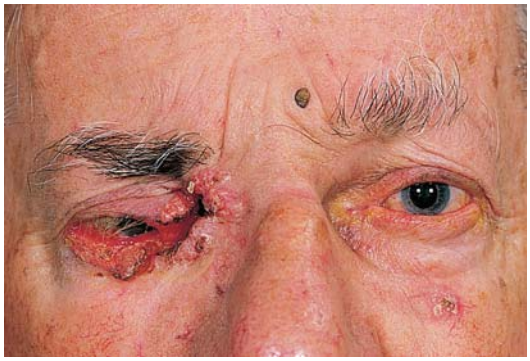


Figure 15.6. Early basal cell carcinoma of medial canthus. 🕮



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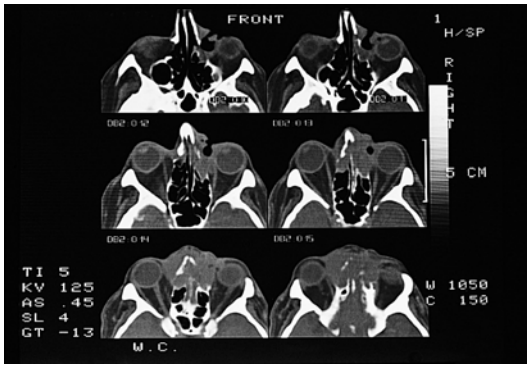

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Figure 15.7. Extensive basal cell carcinoma involving the orbit and extending across the nose to the opposite side. **a** Clinical photograph; **b** computerised tomography scan. 🕮

Sebaceous Gland Carcinoma

This uncommon tumour constitutes 1–3% of malignant eyelid tumours (higher in Asians). It arises from the meibomian glands in the tarsal plate. It appears as a discrete, firm nodule, which often presents as a "recurrent chalazion", thereby delaying diagnosis. Treatment involves wide excision with or without radiotherapy. Mortality ranges from 6% to 30%, depending on site, size, symptom duration and histological classification.

Melanoma of the Eyelid

Malignant melanoma of the eyelids is similar to malignant melanoma elsewhere, appearing as a raised, often shiny, black lump. It metastasises at an early stage and the prognosis does not seem to be altered by excision.

The Conjunctiva

Benign Lesions

Benign Pigmented Lesions of the Conjunctiva

Conjunctival epithelial melanosis occurs in approximately 90% of black people and 10% of white people, and is noticeable in early life. The lesions are flat, brownish patches scattered throughout the conjunctiva, but might be more noticeable at the limbus (Figure 15.8). Usually, they do not grow. Other pigmented lesions, for example the benign naevus, require closer attention and specialist evaluation.

Nonpigmented Lesions

Pingueculum is a common mass lesion of the conjunctiva. It is seen as a yellowish nodule usually on the medial interpalpebral fissure. It is a fibrovascular degeneration and is seen in all climates.

Pterygium is a growth of abnormal fibrovascular tissue extending from the conjunctiva over the cornea (Figure 15.9). It is thought to result from to chronic irritation from dust and solar radiation. It is more common in hot climates and individuals who work out of doors. Recurrent inflammation of the pterygium is often self-limiting but responds to a short course of topical steroids. If it extends over the visual axis of the cornea it can cause visual impairment and, therefore, surgical excision might be required, although regrowth occurs in a large proportion of patients.

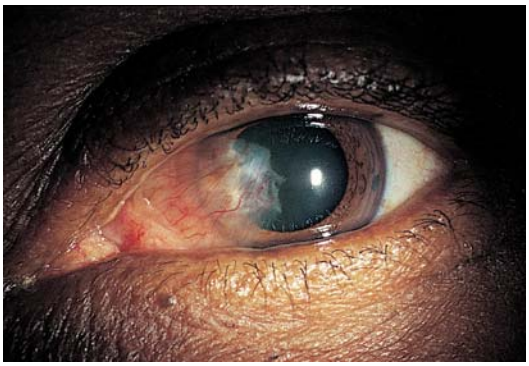


Figure 15.9. Pterygium. 🕮

Malignant Lesions

Melanoma of the Conjunctiva

Malignant melanomata can occur on the conjunctiva (Figure 15.10) 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 benign lesions should be excised and biopsied if they become irritable or sometimes simply on cosmetic grounds, but they rarely become malignant. The treatment of conjunctival malignant melanoma involves wide surgical excision with adjuvant cryotherapy or radiotherapy. The five-year survival rate is approximately 85%.

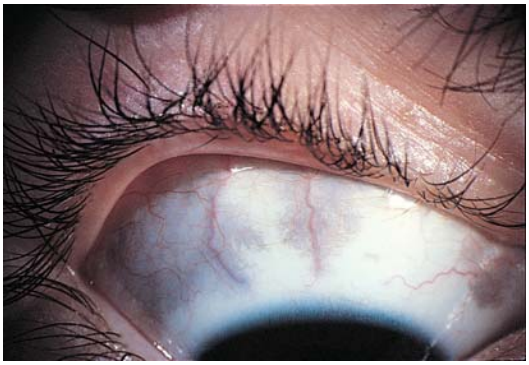


Figure 15.8. Conjunctival melanosis. 🕮

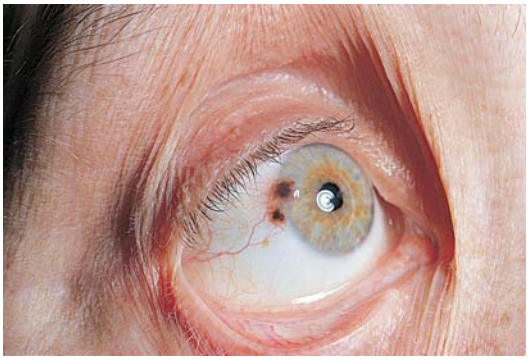


Figure 15.10. Melanoma of conjunctiva. 🕮

The Orbit (see Table 15.1)

Lacrimal Gland and Sac Tumours

Lacrimal gland tumours can either be inflammatory, mixed cell tumours or adenocarcinomas. They present with proptosis or a mass in the outer part of the eyelid superotemporal orbit. Lacrimal sac tumours are less common and present with sac swelling. Benign lesions and infections need to be excluded.

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 can occupy a cavity in the bone and a computed tomography (CT) scan is advisable when this is suspected. Rupture of the cyst can lead to profound orbital inflammation. Excision on cosmetic grounds and for diagnosis is usually indicated.

Cavernous Haemangioma

This is the commonest primary neoplasm of the orbit in adults. It is benign. It is unusual for surgery to be necessary in such cases. It is usually located within the muscle cone, and gives rise to axial proptosis.

Glioma of the Optic Nerve

This rare tumour causes progressive proptosis and optic atrophy but it can be slow growing. There is an association with Von Recklinghausen's disease (neurofibromatosis type 1) and the presence of pigmented patches in the skin should make one suspect this. Treatment by surgical resection and/or radiotherapy is indicated if intracranial spread is documented.

Table 15.1. Primary orbital tumours.

Vascular	Capillary haemangioma
	Cavernous haemangioma
	Lymphangioma
Neural	Optic nerve glioma
	Meningioma
	Neurofibromatoma
Lacrimal gland	
Lymphoproliferative	
Rhabdomyosarcoma	
Histiocytosis	

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 combining radiotherapy and chemotherapy. The tumour is thought to arise from striated muscle and the histological diagnosis is confirmed by finding striation in the tumour cells. It is usually located in the superonasal orbit.

Metastatic Tumours and Tumours from Neighbouring Sites

A wide variety of tumours can invade the orbit and produce proptosis and often diplopia. Lymphoma is one example. It can 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. In children, orbital metastases arise most commonly from neuroblastoma and Ewing's sarcoma. In the adult, the commonest primary sites are bronchus, breast, prostate and kidneys.

"Pseudotumour" (Idiopathic Orbital Inflammatory Disease)

This is an inflammatory swelling in the orbit of unknown cause, which can present with pain, proptosis and diplopia. A mass might be palpable in the orbit and biopsy reveals nonspecific inflammatory tissue consisting mainly of lymphocytes. Diagnosis can eventually be made by exclusion of other causes of proptosis. In severe cases, a course of systemic steroids and/or radiotherapy is usually effective.

Exophthalmos and Proptosis

Both these terms mean forward protrusion of the eyes but traditionally exophthalmos refers to the bilateral globe 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 (see Table 15.2)

When one eye seems to bulge forward, the doctor might have a serious problem on his hands and the following likely causes should be considered:

- *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 (orbital floor). These need to be distinguished from a true proptosis.
- *Thyrotoxicosis*. This is the commonest cause of unilateral or bilateral proptosis; diagnosis is achieved from the history, examination and tests of thyroid function (Figure 15.11).
- *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 can expand after bending down or crying. Ultrasound and CT scanning can confirm the diagnosis. Occasionally, angiography might be required.
- *Pseudotumour*. Biopsy should be carried out if possible, and other causes excluded.
- *Mucocele of sinuses*. Diagnose by X-ray or CT scan.
- *Lymphoproliferative disease*. A biopsy, full blood count and sternal marrow puncture should be carried out.

Table 15.2. Causes of proptosis.

• Endocrine
• Vascular abnormalities
• Inflammatory disorders
• Primary orbital tumours
• Metastases

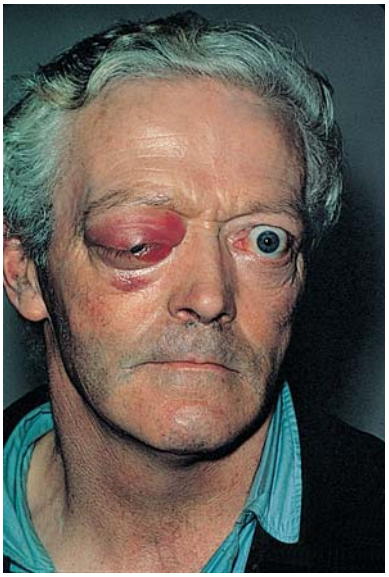


Figure 15.11. Proptosis: dysthyroid disease. 🕮

- *Others*. There are a large number of possible but rare causes of 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 accurate (especially for the novice) but best results are achieved by ensuring that they are made by the same person, using the same instrument on each occasion for a given patient.

Once thyroid disease and trauma have been excluded, the patient would require further investigations including systemic examination, full blood picture, orbital ultrasound, CT scan, magnetic resonance imaging (MRI) scan, possibly carotid angiography and sometimes orbital biopsy.

16

Ocular Trauma

The fact 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, might be overlooked. Sometimes, the eyelids might be so swollen that it is difficult to examine the eyes and a serious perforating injury could 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 even though the problem might have at first seemed slight.

Injuries to the Globe

Contusion

The eye casualty officer comes to recognise a familiar pattern of contusion, the effect of squash ball injuries and blows from flying objects in industry or after criminal assault. Injuries from industrial causes have now become quite uncommon thanks to better control by means of protective clothing and proper guarding of machinery. As a result, sporting injuries have become more evident, although here increasing public concern has also led to some improvement. Notable instances of good control are the use of protective guards in ice hockey and cricket. The surrounding orbital margin provides good protection to the eyes from footballs and even tennis and cricket balls, but the rare golf ball contusion injury usually leads to loss of the sight of the eye. Squash balls and especially shuttle cocks have earned a bad reputation for inflicting contusion injuries and, from the economic point of view, leading to loss of time at work and hospital expenses.

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 might 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, if necessary using an eye speculum under general anaesthesia. In the primary care situation, one must be very careful not to apply more than gentle pressure to the eyelids in case the globe of the eye has been perforated and when there is doubt, referral to the eye department is advisable. The important clinical features of contusion injury are best considered by looking at the anatomical parts of the eye.

Cornea

The most common injury to the cornea is from the corneal foreign body and this has already

been described in Chapter 5. Almost as common is the corneal abrasion. It is odd how this is so often caused by the edge of a newspaper, a comb or a child's fingernail. Abrasions from the leaves of plants or twigs need special attention because of the type of infection that can occur (fungal), but any abrasion can lead to the condition known as recurrent abrasion. Here, the patient experiences a sharp pain in the eye in the early morning usually on waking, sometimes many months after the initial injury. It is thought that the lid margin adheres to the area of weakened healed corneal epithelium during sleep. The diagnosis is easily missed if the patient has forgotten about the original injury and if the cornea is not examined carefully with the slit-lamp biomicroscope. This problem of recurrence is a reason to treat these abrasions with some care and to provide the patient with a lubricating ointment to be used at night for some time after the original injury has healed. Sometimes, recurrent abrasion results from a rare inherited disorder of the corneal epithelium.

When a patient presents with a corneal abrasion, the eyelids are often swollen perhaps from rubbing and the distress and agitation can be considerable. Examination may be impossible without first instilling a drop of local anaesthetic. These drops should never be continued as treatment because they could seriously delay the healing of the cornea.

Anterior Chamber

A small bleed into the anterior chamber of the eye is seen as a fluid level of blood inferiorly ("hyphaema") (Figure 16.1). This is a sign of potential problems because of the risk of secondary bleeding after two or three days. This risk is especially serious in children and the complication can lead to secondary glaucoma and at worst, the loss of the eye. The parents need to be warned about this if there is a hyphaema. Treatment is by strict rest with little or no head movement to avoid further bleeding and regular measurement of the intraocular pressure.

Iris

When confronted by a flying missile, the normal reaction is to attempt to close the eyelids and to rotate the eyes upward. This is the reason why the commonest point of impact is the lower temporal part of the eye and it is in this region of the iris that one is most likely to see peripheral iris tears ("iridodialysis").

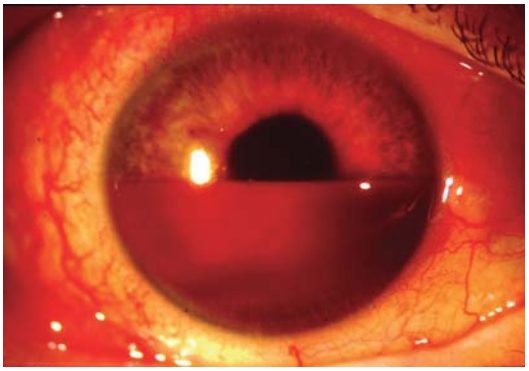


Figure 16.1. Hyphaema showing anterior chamber half filled with blood.

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 (Figure 16.2).

Contusion can result not in a tear of the iris root, but in a tangential splitting of the iris and ciliary body from the sclera producing recession of the angle of the anterior chamber; the appearance is often associated with secondary glaucoma, sometimes many years after the injury and is identified using the special contact lens known as the gonioscope.

A sudden impact on the eye can also produce microscopic radial tears in the pupillary

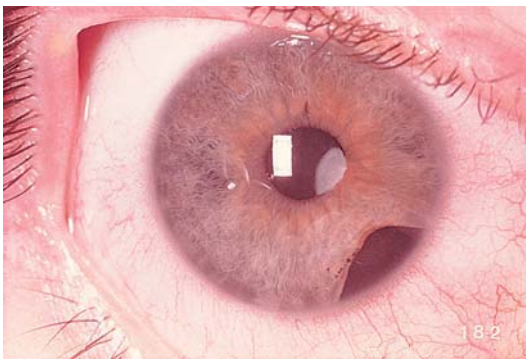


Figure 16.2. Iridodialysis or splitting of the iris root in lower temporal quadrant. A sure sign of previous contusion. 🕮

sphincter of the iris. This could be a subtle microscopic sign of previous injury when no other signs are present, or the damage might be more severe, resulting in persistent dilatation of the pupil (traumatic mydriasis). Unless the eye is examined, this widening of the pupil after injury can be mistaken for a third cranial nerve palsy.

Lens

Any severe contusion of the eye is liable to cause cataract, but the lens might not become opaque for many years after the injury. The lens can also become subluxated (slightly displaced because of partial rupture of the zonular ligament) or even dislocated either anteriorly into the anterior chamber or posteriorly into the vitreous.

Vitreous

The vitreous can become displaced from its attachments around the processes of the ciliary body or around the optic disc after a contusion injury if it has not already undergone this change as part of the normal ageing process. The patient might be aware of something floating in front of the vision. More extensive floating black spots can indicate a vitreous haemorrhage caused by excessive vitreous traction on a retinal blood vessel. Although such haemorrhages usually clear completely in time, they tend to accompany more serious damage to the retina, which can only be fully revealed once clearing has taken place.

Retina

Bruising and oedema of the retina are seen as grey areas with scattered haemorrhages. The macular region is susceptible to oedema after contusion injuries, causing permanent damage to the reading vision. Just as tears can occur to the peripheral iris, so a similar problem is seen in the peripheral retina. These crescent-shaped retinal dialyses are also most common in the lower temporal quadrant and their importance lies in the fact that they may lead to a detachment of the retina unless the tear is sealed by laser treatment. Any significant contusion injury of the eye requires a careful inspection of the peripheral retina.

Choroid

Tears in the choroid following contusion 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 (Figure 16.3). They are also potential sites for choroidal neovascularisation.

Optic Nerve

A variable degree of optic atrophy can become apparent a few weeks after a contusion injury. Blunt injuries to the eye can cause bleeding into the optic nerve sheath or tearing of the tiny pial blood vessels that supply the nerve, both resulting in complete, irreversible loss of vision on the affected side. Attempts have been made to relieve the situation by emergency decompression of the optic nerve, nerve sheath fenestration, use of hyperbaric oxygen and high-dose steroids. No treatment has shown a clear benefit except optic nerve decompression in specific circumstances.

Perforation

As soon as the globe of the eye is penetrated there is a serious risk of infection. The vitreous is an excellent culture medium and in the

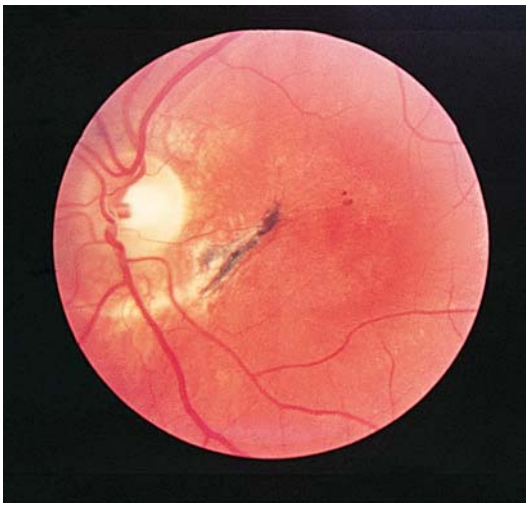


Figure 16.3. Healed choroidal tear. Another sign of previous injury. 🕮

pre-antibiotic era, eyes were totally lost within two or three days as a result of this. *A perforating wound of the eye must, therefore, be considered a surgical emergency.* Perforating injuries are seen in children from scissor blades, screwdrivers, darts and other more bizarre objects. In adults, there has been a dramatic fall in the incidence of such injuries since the introduction of compulsory seat belts but "do-it-yourself" accidents and assaults still take their toll. Following such an injury it is important to consider the possibility of an intraocular foreign body, especially when there is a history of using a hammer and chisel.

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 can be obtained by careful suturing under general anaesthesia using the operating microscope. If the lens has been damaged, early cataract surgery might be needed and deeper penetration can result in the need for retinal detachment surgery.

On admission or in the casualty department, the patient is given tetanus prophylaxis and both systemic and local antibiotics. If early surgery under general anaesthesia is likely to be needed, it is better for the patient not to eat or drink to avoid delays in hospital. If it becomes clear that the injury is a serious one, it is better to warn the patient at an early stage about the possible risk of losing the sight of the eye or even the need to replace it with an artificial one.

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 eye protection. These injuries might seem slight at first and sometimes patients do 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 can cause immediate infection, or at a later date the deposition of ferrous salts, in a process known as siderosis. This can eventually lead to blindness of the eye. Other metals also tend to give reactions, particularly copper and for this reason the metallic fragment should be removed (Figure 16.4). This is achieved either by using intravitreous forceps under microscopic control or using a magnet. The exact surgical technique is planned beforehand once the foreign body has been accurately localised in the eye. Airgun pellets cause particularly severe eye injuries and the eye is often lost because of the extensive disruption at the time of the injury. Some intraocular foreign bodies, such as glass particles or some alloys, might be tolerated quite well and a decision could have to be made as to whether observation is preferable in the first instance. This especially applies when the sight of the eye remains good. When a foreign body is not to be removed immediately, many ophthalmologists would insert intravitreal antibiotics as a prophylactic measure against endophthalmitis. When a foreign body is found lying deeply in the cornea, its removal can 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.

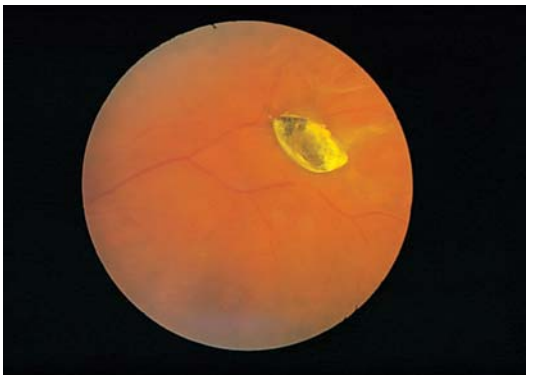


Figure 16.4. A small metallic foreign body lying on the retina. 🕮

Sympathetic Ophthalmia

This rare complication of perforation is more common in children. The injured eye remains markedly inflamed and the wound might have been cleaned inadequately or too late. Over a period of two weeks to several months or even years a particular type of inflammatory response begins in the uvea and subsequently a similar reaction occurs in the other eye. The inflammation in both eyes can be so severe as to cause blindness. The condition does, however,

respond well to steroid treatment and it is extremely rare. Occasionally, one sees patients who have an artificial eye complaining of transient blurring of the vision of their remaining eye. They need to be examined carefully for signs of uveitis.

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 eyelids 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. When more than this is lost or when it has been 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 using the operating microscope and the fine suture material available in an eye department (Figure 16.5). An untidy repair can result in a permanently watering eye because of kinking of the eyelid. This interferes with the proper moistening of the cornea during blinking or when asleep. Special attention must be paid when the medial part of the eyelid has been torn, as this contains the lacrimal canaliculus. Again, unless repair is carried out using an accurate technique under general anaesthesia in theatre, the risk of a permanently watering eye is increased.

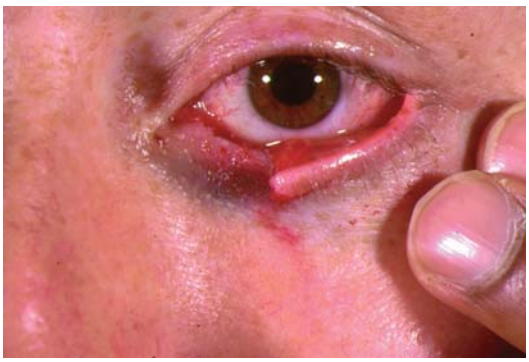


Figure 16.5. Full thickness lower lid laceration.

Contusion of the eyelids, otherwise known as a black eye, is of course a common problem, especially on Saturday nights in a general casualty department. 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. It is unusual to find damage to the eyes after Saturday night fist-fights, unless a weapon was involved. Broken beer glasses produce devastating injuries to the eyes as well as to the eyelids.

Injuries to the Orbit

Blows on the side of the cheek and across one or other eye occur in fights, industrial accidents and road traffic accidents. The most common type 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 of the maxillary sinus. The inferior rectus muscle becomes tethered in the wound so that there is mechanical limitation of upward movement. The infraorbital nerve, which traverses the orbital floor, can 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 of the eye itself often shows evidence of contusion. A considerable improvement from the functional and cosmetic point of view can be obtained by positioning a plastic or Teflon implant in the floor of the orbit after freeing the prolapsed tissue.

Fractures of the skull that extend into the orbit can 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 can be affected by optic nerve damage. A blow on the eye can result in sudden blindness with at first no other evidence of injury (apart from an afferent pupillary defect), but subsequently, the optic disc becomes pale and atrophic after two or three weeks.

Radiational Injuries

The eyes might be exposed to a wide range of electromagnetic radiation from the shorter wavelength ultraviolet rays through the wavelengths of visible light to the longer infrared waves, X-rays and microwaves. X-rays pass straight through the eye without being focused by the optical media and, in large enough doses, can cause generalised damage. It is 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, destruction of the secretory cells within the lacrimal glands and retinal neovascularisation. As one might expect, visible light does not normally damage the eyes, although an intense light source can be absorbed by the pigment epithelium behind the retina and converted to heat, producing a macular burn. After eclipses of the sun, there are usually a number of patients who arrive in the casualty departments of eye hospitals with macular oedema and sometimes serious permanent impairment of visual acuity. Sun gazing, with consequent retinal damage, has been reported after taking lysergic acid diethylamide (LSD).

The laser beam provides a source of intense light, which is used widely in ophthalmology as a deliberate means of producing gentle burns in the retina or making holes in the lens capsule after cataract surgery. However, uncontrolled use of lasers can cause blinding foveal burns as the subject tends to look directly at the beam momentarily, until they realise what it is. Ultraviolet rays, which are shorter than visible light, do not normally penetrate the eye but in large enough doses produce burns of the eyelids and cornea. On the skin this is seen as erythaema 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 welder's 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 ultraviolet injury show a delayed effect, the symptoms appearing 2 h or 3 h after exposure and lasting for about 48 h. There is usually severe pain and photophobia so that it might 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, infrared 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 because of 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 would not be expected to be dangerous in this respect. Concern is quite often expressed in the press or elsewhere 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. Someone not used to working with a visual display unit who is suddenly made to spend several hours a day in front of one might experience eyestrain, especially if incorrect spectacles are worn.

Chemical Injuries

These are quite common but usually not severe enough to warrant hospital attention. In industrial premises there is now nearly always a first-aid post with facilities to wash out the eyes. Plain water or a salt solution is the best fluid to use and valuable time may be lost if washing is delayed in order to search for a specific antidote. More severe burns can result from the catalysts used in the manufacture of plastics or from alkalis, such as caustic soda. Alkalis penetrate the eye rapidly as they saponify lipids within cell membranes, aiding passage, and can quickly reach the posterior segment. Acid burns as from exploding car batteries are quite commonly seen in large casualty departments but are usually less severe as acids tend to coagulate corneal proteins, thereby slowing penetration.

17

Testing Visual Acuity

Measurement of visual acuity is the most important part of the ocular assessment performed by the doctor and yet it is surprising how often the nonspecialist omits it in examination. 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 in the injured eye. Simple measurement of visual acuity is of limited value without a knowledge of the spectacle correction or whether the patient is wearing the appropriate spectacles. The best corrected visual acuity (i.e., with lenses in place) therefore needs to be recorded for each eye. This corrected visual acuity can also be estimated with a pinhole held in front of the eye. The effect of the pinhole is to eliminate the effect of refraction by the cornea and the lens on the extremely thin beam of light produced by the pinhole.

Measuring the visual acuity means measuring the function of the macula, which is of course only a small part of the whole retina. A patient might 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 unable to read. This state of affairs is seen in patients with age-related macular degeneration and can be compared with the situation in which a patient has grossly constricted visual fields but normal macular function, as is sometimes seen in retinitis pigmentosa or advanced primary open-angle glaucoma. Here, the patient appears to be blind, being unable to find his way about, but he might surprise the ophthalmologist by reading the visual acuity chart 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 close together (resolution). 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 that 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 11 μm 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 if they are separated by 1 mm when placed 10 m away. Two such points would be separated by 2 μm on the retina. This might be surprising considering that a spot of light casts a minimum size of image of 11 μm because of 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 1.5 μm.

In the clinic, the distance 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 from the eye. The single large letter at the top of this chart is designed to be just discernible to a normal-

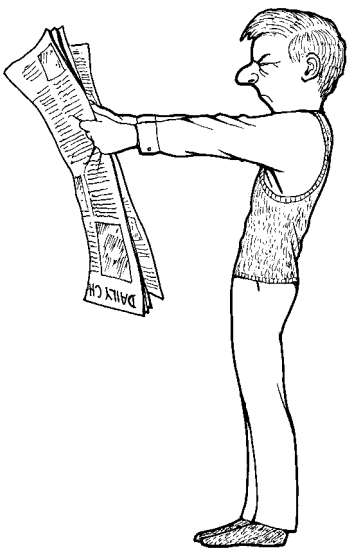
sighted person at a range of 60 m. If the patient's vision is so poor that only this and no smaller letter can be seen at 6 m, the vision is recorded as the fraction "6/60". The normal-sighted person who can read the chart down to the smaller letters designed to be discerned at 6 m is recorded as having a visual acuity of 6/6. The normal range of vision extends between 6/4 and 6/9, depending on the patient's age. In some European countries, the visual acuity is expressed as a decimal instead of a fraction. Therefore, 6/60 would be expressed as 0.1. In the USA, metres are replaced by feet, so 6/6 becomes 20/20. This is where the term "twenty twenty" vision originates from, meaning clear or near-perfect vision. Recently, a new type of visual acuity chart has entered use in the clinic and in research studies. It is called the LogMAR chart and differs from the conventional Snellen chart (Figure 3.1) by having five letters on each line rather than the "pyramid" shape of the Snellen chart. There are also smaller differences in type size between lines. Some of the advantages of using this new chart are that the measurement of poor visual acuity is more accurate as more larger letters are included and small changes in acuity are easier to detect (easier to detect disease progression or treatment success).

The near visual acuity is also measured using a standard range of reading types in the style of newsprint 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 years (Figure 17.1). 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 can be worse than the vision of each eye tested separately (e.g., in cases of macular disease causing distortion).

A number of other tests have been developed to measure visual acuity in the nonliterate patient. 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 18 months or two years, the optokinetic drum might 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 then 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



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

Figure 17.1. Reading glasses in presbyopia. 🕮

shy children or elderly patients can sometimes be misleading.

Other ways of measuring visual acuity have been developed. One is to assess the patient's 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 (contrast sensitivity) can be measured. This type of test has certain theoretical advantages over standard methods but it is not widely used clinically as a routine. Finally, the electrical potentials generated by the retina and optic nerve can be measured to give an estimate of visual acuity when the eye is presented with targets of varying size and contrast. This method is useful in infants and in the assessment of adults with nonorganic visual loss.

Measuring for Spectacles

If a patient has not been tested recently for spectacles, not only can the measurement of visual acuity be inaccurate, but the symptoms might be caused by 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 accurate method because some patients' observations as to the clarity of letters can be unreliable. Furthermore, a healthy young person might see quite clearly with a wide range of lenses simply by exercising the ciliary muscle (i.e., accommodation). 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 lenses (retinoscopy). 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 instill 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 be measured.

Subjective Refraction

Here, considerable skill is also needed because many patients become quite tense when being tested in this way and might not initially give accurate answers. Lenses both stronger and weaker than the expected requirement are placed in the trial frames 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 min or 15 min 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, or when the patient overaccommodates. One further way of assessing 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 checkerboard. When fine checks are viewed, interposing different lenses can modify the response. This method is of great interest but it is still not 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 function in the centre of the visual field only and it is possible to have advanced loss of peripheral 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 3 together with various other measurements of different aspects of vision.

18

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 can 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 that 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) can 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 can often be extremely painful. Photophobia is usual and often pain on focusing on near objects is a feature. The age incidence is wide but anterior uveitis is commonly seen in the third and fourth decades of life, and every eye casualty officer becomes 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 h and then resolves on treatment in two or three weeks; however, it may last as long as six weeks. A further exacerbation might occur during this period and there is a strong tendency towards recurrence after a few months or several years 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 anterior chamber of the eye itself. The pupil is small because the iris 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 might be pos-

sible 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 (Figure 18.1). Normally, of course, the aqueous is crystal clear even when examined with the slit-lamp biomicroscope.

The presence of an occasional cell in the aqueous can be normal, especially if the pupil has been dilated with mydriatic eye drops, but suspicion should be raised if more than three or four cells are seen. In fact, the early diagnosis of anterior uveitis can entail 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"s (Figure 18.2). The microscopic appearance of the KP is determined by the type of cells. If a granulomatous type of inflammatory reaction is taking place, involving epithelioid cells and macrophages, the KP might 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 nongranulomatous, a fine dusting of the posterior surface of the cornea could be evident. KPs tend to become absorbed but they can remain more permanently as pigmented spots on the endothelium.

Figure 18.2. Keratic precipitates.

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 because parts of the iris remain stuck to the pupil giving it an irregular appearance. In severe cases of anterior uveitis, pus can collect in the anterior chamber to the extent that a fluid level can be seen where the layer of pus has formed inferiorly. This is known as hypopyon – literally, "pus below" (Figure 18.3). A hypopyon is an indication of severe disease in the eye and the patient

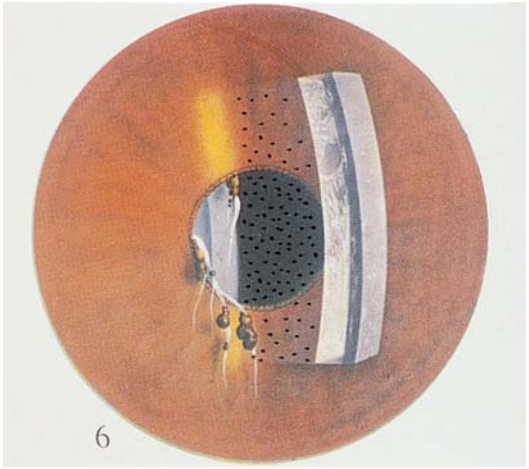


Figure 18.1. Flare.

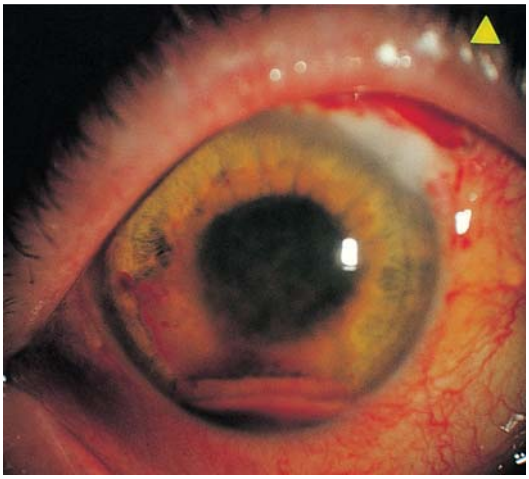


Figure 18.3. Hypopyon. In addition, there are red blood cells and fibrinous exudate in the anterior chamber (with acknowledgement to Professor H. Dua).

should preferably be treated in hospital as an inpatient. Hypopyon 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 hypopyon uveitis, and ulceration of the mouth and genitalia. A hypopyon is occasionally seen following cataract surgery and in such cases can be infective or noninfective in origin. It is fortunately a rare complication of modern cataract surgery and the use of intraocular acrylic lenses.

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 can cause serious problems and a careful check on the intraocular pressure must be maintained. The rise in intraocular pressure might be due to direct obstruction of the aqueous outflow 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 forward, giving the appearance known as iris bombe. Secondary glaucoma can also result from the use of topical steroids in predisposed individuals. Cataract is a further serious complication, which can appear after repeated attacks of anterior uveitis. It nearly always first affects the posterior subcapsular zone of the lens and, unfortunately, interferes with the vision at an early stage. Cataracts can also result from long-term use of topical or systemic steroids.

Causes

For the majority of patients who present to eye outpatient 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 might 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 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 high on the list of suspected causes but this would appear to be a less common cause nowadays. Herpes simplex and zoster can also cause anterior uveitis. Septic foci in adjacent structures, such as dental sepsis or sinusitis, have also been under suspicion but these are now thought to be relatively unimportant. In the case of the elderly, the onset of anterior uveitis can 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 hypermature cataract. Three other types specific of anterior uveitis must be mentioned at this stage.

Sympathetic Ophthalmia

This is a rare but dramatic response of the uvea in both eyes to trauma. The significance of the condition rests in the fact that although the trauma has only affected one eye, the inflammatory reaction occurs in both. It can follow perforating injuries, especially when uveal tissue has become adherent to the wound edges. Occasionally it can occur following intraocular surgery. The injured eye, which is referred to as the "exciting eye", remains severely inflamed and, after an interval of between two weeks and several years, the uninjured eye ("sympathising eye") becomes affected. The inflammation in the sympathising eye usually starts in the region of the ciliary body and spreads anteriorly and posteriorly. It is granulomatous. Careful wound toilet and repair of the injured eye can probably prevent many cases, as can also removal of

blind, painful and inflamed eyes within the critical two-week period following injury.

Heterochromic Iridocyclitis

This type of anterior uveitis presents in 20–40 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; posterior synechiae do not develop. The inflammation does not usually respond at all to treatment. Cataracts and chronic glaucoma occur commonly. The condition has been mimicked by denervating the sympathetic supply of the eyes in experimental animals and it seems possible that there might be a neurological cause, unrelated and distinct from other types of uveitis.

Pars Planitis (Intermediate Uveitis)

This refers to a low-grade inflammatory response, which is seen in young adults. It affects both eyes in up to 80% of cases, although the severity can be asymmetrical. 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. A mild-to-moderate peripheral retinal phlebitis can occur. The condition runs a chronic course and occasionally can be complicated by cataract, cystoid macular oedema and tractional retinal detachment. The cause is unknown in the majority of cases, although there is a known association with sarcoidosis.

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 especially taking into account any previous chest or joint disease. An X-ray of the chest, and a blood count, including measurement of the erythrocyte sedimentation rate (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 might lead one to suspect the possibility of venereal disease. In the case of some infective types of anterior uveitis, the diagnosis is usually 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. By contrast, anterior uveitis can be an important clue to the diagnosis of a venereal disease.

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 can 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 cyclopentolate drops can 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 a specialist ophthalmologist and, when the condition is affecting both eyes, it might be preferable to admit the patient to 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 can lead to intraocular infection. The development of anterior uveitis, weeks, and occasionally 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, the disease might 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 then to the vitreous. When this happens the vision becomes markedly blurred, even when the original focus is remote from the macula region. Alternatively, the inflammation might originate from the retina and spread to involve the choroid and vitreous subsequently. Choroiditis at the macula itself usually leads to permanent loss of central vision.

Signs

In its early stages, choroiditis can be seen as a grey or yellowish raised area, which can be discrete or multiple and anywhere in the fundus. A cellular reaction could appear in the overlying vitreous, seen as localised misting with the ophthalmoscope, and eventually the whole vitreous can 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. Retinitis manifests as an indistinct white cloudy area. When a patch of choroiditis heals, the margins become pigmented and a white patch of bare sclera remains (Figure 18.4). This is 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 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. Retinal vasculitis can occur. A predominantly arteriolar inflammation can indicate a viral cause, whereas venous involvement is more common with other aetiologies. Optic nerve inflammation or oedema can also occur.

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.

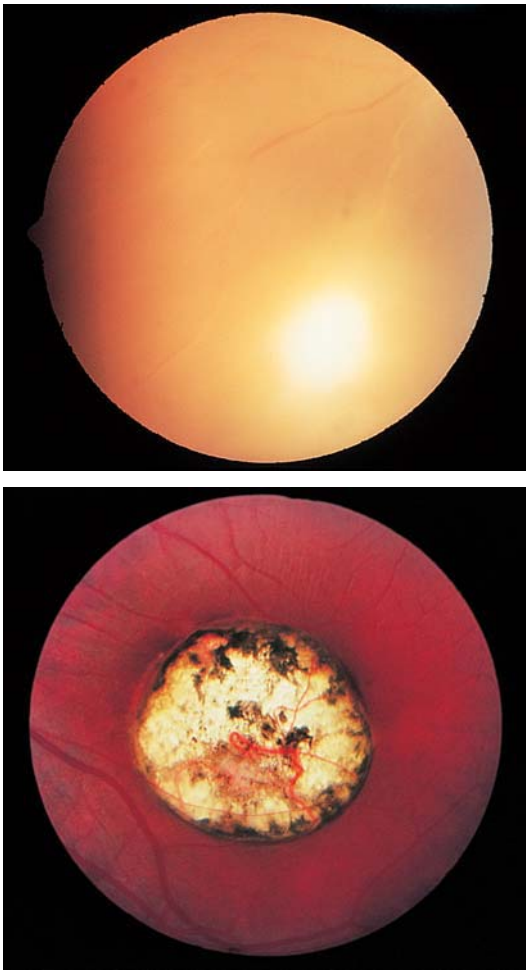


Figure 18.4. Chorioretinitis: **a** active with hazy vitreous; **b** inactive scar.

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 can be infected. In the adult with the acquired infection, systemic symptoms are usually mild. Similarly, the ocular symptoms of acquired toxoplasmosis can be mild. However, a severe form of acquired ocular toxoplasmosis has been recognised. In such cases, there has been contact with wild cats in stables. In the case of infected pregnant mothers, the child *in utero*

could be infected by the more severe congenital form of the disease. The organism enters the brain as well as the eyes and can cause mental deficiency and epilepsy. A characteristic type of calcification is seen on skull X-ray or computed tomography (CT) scan. In the eye, a focal type of choroiditis often affects both eyes and this is usually at the posterior pole in the macular region. Histologically, the toxoplasma organism is found in the eye lesions. The diagnosis can be confirmed by sending some blood for serological tests. Four such tests are currently in use clinically: the toxoplasma dye test, indirect fluorescent antibody test, haemaglutination test and enzyme-linked immunosorbent assay (ELISA). These tests 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 can be less easy in acquired toxoplasmosis, where evidence of systemic involvement can 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. The most specific of these tests is the ELISA.

All the currently available antitoxoplasma treatments have potentially serious side effects. Therefore, not all active toxoplasma retinochoroiditis lesions require treatment. Such treatment is required only if an active lesion involves or threatens the fovea and/or optic nerve. Treatment is also required when there is severe vitritis.

A combination of pyrimethamine and sulfadiazine has been recommended, but such treatment can cause a serious fall in the white cell count. An alternative antimicrobial treatment is clindamycin. This needs to be given with a sulfonamide in order to reduce the risk of colitis. Other antitoxoplasma agents include atovaquone. It is generally accepted that systemic steroids have some beneficial effect and can help to clear the vitreous more rapidly, but this treatment should be given only with antimicrobial cover. Steroids on their own will produce exacerbation or progression of the chorioretinitis. In fact, the majority of cases resolve spontaneously, leaving more or less chorioretinal scarring at the macular region. Recurrences are fairly common, with or without treatment, and the fresh choroidal inflammation tends to arise at the edge of a previous scar.

Toxocariasis

Toxocariasis is caused by *Toxocara cati* (from cats) or *T. canis* (from dogs). This nematode has been found in the enucleated eyes of young patients with a severe type of chorioretinitis. It is a unilateral disease found in children who are in close contact with puppies or eat dirt (through faecal contamination). The vitreous tends to be filled with a white mass of inflammatory cells so that the presence of a tumour might be suspected (e.g., retinoblastoma). Endophthalmitis tends to develop in these cases and the sight of one eye might be completely lost. During the acute stage, the peripheral blood can show an eosinophilia. Treatment is unsatisfactory and includes a combination of antihelminthic agent taken by mouth (thiobendazole or diethylcarbamazine) and steroids.

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 tuberculosis. The relationship seems less likely now that tuberculosis has been almost eliminated from the population. However, this diagnosis must not be forgotten especially in the immunosuppressed patient and those with recalcitrant or atypical uveitis, as there is currently a slight re-emergence of the disease. Choroidal tubercles are a well-described entity: these raised yellowish granulomatous foci were used as a diagnostic feature of miliary tuberculosis and, occasionally, chronic pulmonary tuberculosis. They are usually seen in extremely ill patients and the yellowish tubercles become pigmented as they heal. Treatment is as for systemic tuberculosis.

Sarcoidosis

The eye is frequently involved in sarcoidosis. Involvement usually takes the form of an anterior or posterior uveitis. The choroiditis is more often peripheral and accompanied by

inflammatory changes in the retinal veins. Sheathing of the veins can be seen and the vision might be impaired by macular oedema. The inflammatory changes might be similar to those seen in pars planitis. When the diagnosis is suspected, the conjunctiva and skin should be searched for possible nodules, which can be biopsied, and an X-ray of the chest can reveal enlargement of the hilar lymph nodes. The management of the ophthalmological problem might 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.

Presumed Ocular Histoplasmosis

Histoplasmosis is a fungal infection (causative agent *Histoplasma capsulatum*). Infection with this organism occurs throughout the world but is more common in the Mississippi Valley and does not occur in the UK. A severe pneumonitis can occur but most cases are asymptomatic.

Presumed ocular histoplasmosis is not seen in patients with active histoplasmosis. The evidence for infection in the originally described cases was necessarily circumstantial – hence the expression "presumed ocular histoplasmosis". The syndrome consists of a certain type of haemorrhagic macular lesion (choroidal neovascularisation) combined with discrete foci of peripheral choroiditis and peripapillary scars.

Syphilis

Syphilis is a chronic infection caused by *Treponema pallidum*. Iridocyclitis occurs in patients with secondary acquired syphilis. It is a bilateral disease in which the iris vessels are particularly engorged. Chorioretinitis can be either multifocal or diffuse and involves the mid periphery and peripapillary area. In the healed phase, perivascular bone spicule pigmentation could be seen similar to that observed in retinitis pigmentosa.

In congenital syphilis, other possible features occur such as deafness and corneal scarring from previous interstitial keratitis. The scattered pigmentation in the fundus might 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 *T. pallidum* immobilisation test and the fluorescent treponemal antibody test are the most sensitive and specific.

Behçet's Disease

Behçet's disease is a multisystem disease associated with HLA–B5. It was originally thought to occur only in the Mediterranean and Japan, where it is most common. It is characterised by an obliterative vasculitis. The clinical syndrome consists of oral and genital ulceration in combination with recurrent uveitis and skin lesions. The uveitis consists of recurrent bilateral nongranulomatous anterior and/or posterior uveitis. Central nervous system involvement occurs as a serious form of the disease.

Other Causes

A wide variety of infective agents have been shown to cause posterior uveitis on rare occasions. The leprosy bacillus 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, meningo-encephalitis, uveitis and exudative retinal detachments.

The Role of Autoimmunity in Uveitis

Although it has been recognised for a long time that bacterial and viral infection can 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. 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 a

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 (HLA–B27). It has been suggested that HLA might 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 that 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

Increased 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. Immunosuppressive agents, such as cyclosporin A, tacrolimus, azathioprine, and cyclophosphamide, are now sometimes used to supplement or replace steroids in difficult cases. 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. Large doses of systemic steroids are best administered on an inpatient basis, especially if the sight is threatened. This has the added advantage of allowing a more detailed pretreatment examination and investigations, and often the opinion of a general physician or immunologist can be valuable at this stage. Secondary glaucoma might also need to be treated and immunosuppressive agents can be administered to resistant cases. When posterior uveitis keeps recurring at the edge of previous healed foci, laser coagulation has been used in selected patients with toxoplasma retinochoroiditis. 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 can prove reversible with intensive antibiotic treatment. When endophthalmitis and panophthalmitis are not properly and aggressively treated, the sight is usually lost permanently and after months or years the whole 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 can become selectively inflamed, either diffusely or in localised nodules. In the case of episcleritis, 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 can be gritty but not painful. Episcleritis is seen from time to time in the casualty department and the patient might be otherwise perfectly fit and well. Such cases tend to recur and some develop signs of dermatological disease. The condition responds to local steroids, but systemic aspirin can also prove effective. Scleritis is less common and more closely linked with rheumatoid arthritis and other collagen diseases. The eye is red (diffuse or localised) and painful. In severe cases, the sclera can become eroded with prolapse of uveal tissue. Topical treatment is of no benefit. The condition responds to systemic anti-inflammatory agents, particularly oral flurbiprofen (Froben), which can be supplemented with systemic steroids and/or immunosuppressants.

19

The Ageing Eye

Although the eye and its supporting structures undergo a number of well-defined changes with age, the distinction between these involutional 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 instill drops into their eyes, either prescribed for them or as self-medication. It is important that adequate advice is received. Advising the elderly is often time consuming and might 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 age-related macular degeneration (AMD). The first can be cured, the second arrested or prevented, while the third generally tends to run a progressive course and treatment is unsatisfactory at present, although significant progress has been made recently. Attempts to measure the incidence of these problems have produced a wide range of figures. 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 AMD, whereas 5% or less have chronic open-angle glaucoma. Visual impairment owing to glaucoma is more prevalent and occurs at an earlier age in blacks than in whites. Although there is an unexpectedly high incidence of cataract in patients with chronic simple glaucoma, the association of macular 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 can ride level with or slightly above the corneal margin, in an elderly subject the upper lid might cross a significant part of the upper cornea. An area of white can 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 might 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. This loss of focusing ability is also contributed to by the progressive loss of ciliary muscle tone. A child might be able to observe details of an object held 5 cm from the eye, but as a result of hardening of the lens and weakening of the ciliary muscle, 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 years if the eyes are otherwise normal, and the phenomenon is called presbyopia. Some degree of opacity of the lens fibres is 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-year 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 can cause troublesome symptoms, for example flashing lights and floaters, 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 13. The appearance of the fundus also shows gradual changes; the retinal arterioles become straighter and narrower, as also do the venules. Colloid bodies or drusen are more commonly seen because of 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 somewhat paler and a degree of optic atrophy is accepted by many clinicians as a senile change unrelated to disease.

Eye Disease in the Elderly

The prevalence of blindness increases with age. The prevalence and causes of blindness also vary from one community to another depending on the age structure of the population and environmental conditions. In England and Wales (1980), the prevalence of blindness was found to be nine per 100,000 children under five years of age and 2324 per 100,000 individuals above 75 years.

A recent survey in the USA 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 (65–75 years), the incidence of AMD is 9.6% for males and 6.9% for females. Both these conditions are, therefore, common and they demand time and medical expertise, both at the primary care level and in hospital.

With increasing longevity throughout the world, especially in the developing countries, there will be a continuing increase in the number of blind people, especially those suffering from diseases related to age, such as cataracts, glaucoma and macular degeneration.

Age-related Macular Degeneration

AMD is the commonest cause of incurable blindness in the elderly in western countries. It is a bilateral disease in which visual loss in the first eye usually occurs at about 65 years of age. The second eye is involved at the rate of approximately 10% per annum and accounts for half of all registered visual impairments in the UK.

There are two main types of AMD: "dry" or atrophic, and "wet" or neovascular. Blindness is usually associated with the wet form of AMD, and among the eyes with severe visual loss, 80–90% of cases are because of wet AMD, while 10–20% are because of the dry form.

Older patients with macular degeneration complain of blurring of their vision and inability

to read. Younger or more observant patients notice that straight edges might look kinked. Usually one eye is considerably more affected than the other, although both eyes can be affected simultaneously. 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. The wet form occurs more commonly in Caucasians and about one-third of the patients give a family history of similar problems. Several preventable factors, including smoking, systemic hypertension, cardiovascular disease and low antioxidant intake, are associated with increased risk of AMD.

In the early stages of dry AMD, inspection of the fundus shows spots of pigment in the macular region. Drusen are also often seen (Figure 19.1). These are small round yellowish spots, often scattered over the posterior pole. Unfortunately, the word "drusen" has been used rather loosely in ophthalmology to refer to two or three types of swelling seen in the fundus. It is used to describe the 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.

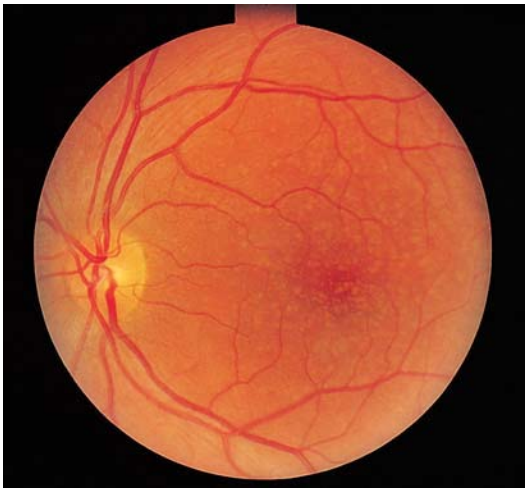


Figure 19.1. Drusen.

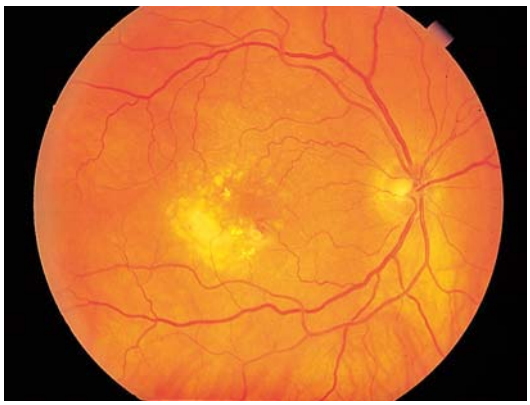


Figure 19.2. Dry macular degeneration.

Under the microscope, colloid bodies are seen as a degenerative change in Bruch's membrane. Drusen can have varying degrees of hyperpigmentation. Most eyes with drusen maintain good vision, but a significant number will develop progressive atrophy of the retinal pigment epithelium (RPE) and choriocapillaris. This is inevitably associated with photoreceptor loss (Figure 19.2). There is usually a moderate loss of vision. This atrophic change in the RPE, choroid and photoreceptors is referred to as "dry" AMD. This is because there is no leakage of fluid or bleeding into the retina or subretinal space.

In the "wet type" of macular degeneration a fan of new vessels arises from the choroid – choroidal neovascularisation (CNV). 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 can be either under the RPE or subretinal (Figure 19.3). A sudden loss of central vision might be experienced as the result of such an episode. Subsequently, "healing" of the leaking vascular complex results in scar tissue formation, which further destroys the central vision permanently.

The terms "classic" and "occult" describe the different patterns of CNV leakage on fluorescein angiography.

Management

No effective treatment is available for dry AMD. There is an increasing vogue for administering

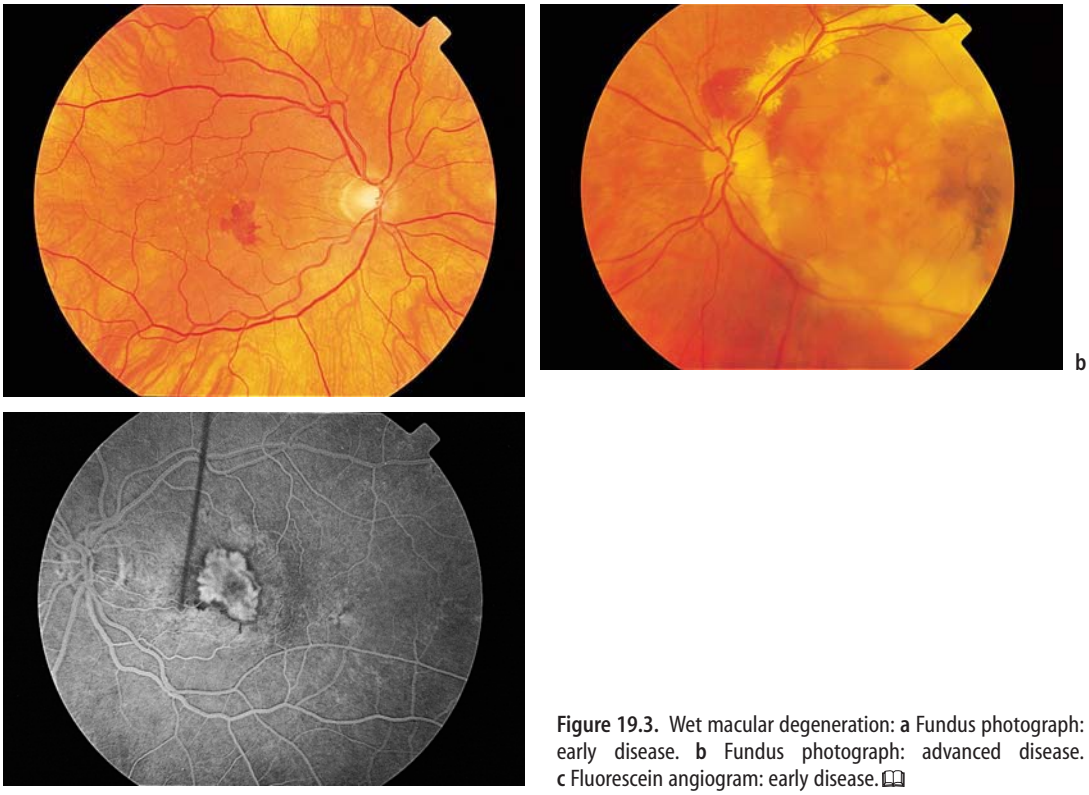


Figure 19.3. Wet macular degeneration: **a** Fundus photograph: early disease. **b** Fundus photograph: advanced disease. **c** Fluorescein angiogram: early disease.

vitamins A, C and E, selenium, copper, zinc, zeaxanthin, carotenoids and lutein preparations to patients. These have been shown to protect against progression of dry AMD to more advanced stages of the disease in high-risk patients. They are thought to reduce the damaging effects of light on the retina through their reducing and free-radical scavenging actions.

Some types/stages of wet AMD are treatable. Currently, there are two clinically proven treatments for wet AMD, although the treatment for some eyes is still unsatisfactory.

Controlled trials of the effect of laser photocoagulation of the choroidal new vessels have shown that this treatment is useful in extrafoveal CNV (i.e., when the leakage is not directly under the fovea). Laser photocoagulation ablates the CNV. It is important that those cases that are likely to benefit from treatment are first identified quickly. At the present time, this entails photography of the fundus and fluorescein angiography, and infrared angiography with indocyanine green. Often patients present at the stage when new vessels have already advanced across the macular region to the subfoveal area or where the fovea has already been permanently damaged by haemorrhage or exudate, making effective laser treatment impossible. Only about 10–20% of cases of CNV are eligible for laser photocoagulation. Another limitation of laser treatment is the high rate of recurrence of the CNV within a short time following treatment.

The second proven treatment is photodynamic therapy (PDT) with verteporfin (Visudyne). PDT specifically targets the CNV complex for damage by low-energy laser, but avoids damage to the unaffected tissue, including the photoreceptors. This treatment aims to preserve vision.

Apart from photocoagulation and PDT, there are other treatment modalities currently under investigation. These include radiotherapy, thermal thermotherapy and drugs including triamcinolone, anercortave, and vascular endothelial growth factor (VEGF) aptamers or

antagonists, which are delivered via injections into the vitreous.

Practical measures can be taken in the management of these patients to alleviate their handicap. Telescopic lenses might be needed for reading or watching television and full 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 within one to three 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. Cataract surgery is required only if vision is sufficiently reduced so far as to interfere with the patient's normal lifestyle. The contraindications for cataract surgery are few and even in extreme old age the patient can benefit. Surgery might 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 eye drops and the domestic chores during the postoperative period might require some attention but is not a contraindication. About one-third of the population aged over 70 years suffers from a 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 quickly, 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 might be coincidental AMD, except if the cataract is of the posterior subcapsular type.

Glaucoma

The various types of glaucoma have also been considered already, and the reader would realise 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 and optometrist 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 years is thought to suffer from chronic simple glaucoma and the figure could rise to as high as 30% in those over 75 years. In most instances, the treatment is simple but requires the co-operation and understanding of the patient. The treatment is preventative of further visual loss rather than curative. Chronic simple glaucoma is best managed in 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. More recently, the care of glaucoma patients is being shared between hospital units and selected (trained) optometrists in the community.

Deformities of the Eyelids

Both entropion and ectropion are common in the elderly and a complaint of soreness and irritation in the eyes as well as watering 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 can be intermittent and not present at the time of examination, but usually under these circumstances there is a tell-tale slight inversion of the lid, which is made apparent by comparing the two sides. Ectropion is nearly always an obvious deformity because of the easy visibility of the reddened and everted conjunctiva, but slight degrees of ectropion are less obvious. The lower punctum alone can be slightly everted, causing a watering eye, and the symptoms might be relieved by applying retropunctal cautery to the conjunctiva. Both ectropion and entropion respond well to lid surgery and there is no

reason why geriatric patients should put up with the continued discomfort and irritation when a complete cure is readily available. These lid deformities can recur sometimes and require further lid surgery, but careful surgery in the first instance should largely prevent this.

Temporal Arteritis

This condition, also known as giant cell arteritis, seen only in the elderly, can rapidly cause total blindness unless it is treated in time. The disease is more common than was originally supposed but it is rare under the age of 50 years. 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 might not be altered. In early disease, 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 might be painful. Often there is low-grade fever and there can be aches and pains in the muscles and joints, as well as other evidence of ischaemia in the brain and heart. Scalp ulceration and jaw claudication can occur. The blurring of vision is caused by ischaemia of the optic nerve head or occasionally central retinal artery occlusion. The diagnosis rests largely on finding a raised erythrocyte sedimentation rate (ESR), elevated C-reactive protein levels 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. Polymyalgia rheumatica is a syndrome consisting of muscle pain and stiffness affecting mainly the proximal muscles without cranial symptoms.

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 (Figure 19.4). Once the disease is suspected, a biopsy is essential and this should be done without delay. Treatment can be commenced immediately, sometimes even before biopsy. However, it is advisable that the lag between starting treatment and biopsy is as short as possible (preferably less than two weeks). The symptoms disappear rapidly after administering systemic steroids, initially in a high dose (e.g., 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 (on average 18 months).

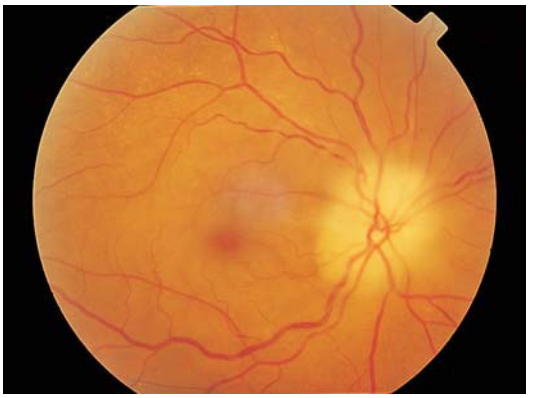


Figure 19.4. Giant cell arteritis: ischaemic optic neuropathy.

Temporal arteritis is recognised as a self-limiting condition. About one-quarter of all patients are liable to become blind unless adequate treatment is administered and in some instances, extraocular muscle palsies causing diplopia and ptosis can confuse the diagnosis. For simplicity, one might summarise the disease by saying it causes headache in patients aged over 70 years with an ESR over 70 and 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 might 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 well defined and can cut through the point of fixation. Fortunately, the central 2° or 3° of the visual field are often spared. When there is so-called macular sparing, the visual acuity as measured by the Snellen chart can be normal. Patients tend to complain of difficulty in reading if the right homonymous field is affected rather than the left, and although they might 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 might have normal fundi and visual acuity of 6/6 and yet be unable to read the newspaper. The picture can be further complicated by true dyslexia and the patient might admit to being able to see the paper and yet be unable to make any sense of it. Dyslexia might 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 might fail to notice it in himself. It is unusual for a homonymous hemianopia to show any signs of recovery, but once the patients understand the nature of the handicap they can learn to adapt to it to a surprising degree.

20

The Child's Eye

How the Normal Features Differ from Those in an Adult

At birth the eye is large, reaching adult size at about the age of two years. One might expect that before the eye reaches its adult size, it would be long-sighted, being too 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. None the less, more than three-quarters of children aged under four years are slightly hypermetropic. The slight change of refractive error that 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 but tends to appear between the ages of six and nine years and gradually increases over subsequent years. The rate of increase of myopia is maximal during the growing years and this can often be a cause of parental concern.

The iris of the newborn infant has a slate-grey colour because of the absence of stromal pigmentation. The normal adult colouration does not develop fully until after the first year. The pupil reacts to light at birth but the reaction can be sluggish and it might not dilate 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, that is the spot of reflected light from the fovea, is absent or ill-defined until the infant is four to six months old. By six months the movement of the eyes should be well co-ordinated, and referral to an ophthalmologist is needed if a squint is suspected. Once children learn to identify letters, at the age of four or five years, the Snellen chart can be employed to measure visual acuity, which by this age is normally 6/9 or 6/6. The Stycar test can be used for three- to four-year olds or sometimes younger children and a similar level of visual acuity is seen as soon as the child is able to co-operate 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.

How to Examine a Child's Eye

The general examination of the eye has been considered already, but in the case of the child, certain aspects require special consideration. Before the age of three or four years, it might not be possible to obtain an accurate measure of the visual acuity, but certain other methods that 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 can also be a helpful guide, for example the response to a smile or the recognition of a face. 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 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 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 co-operation. The cover test can only be performed well under such conditions and once this has been done the pupils and anterior part of the eye 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 the mother's arms, this can be beneficial because it is a simple matter to raise one eyelid and peer in without waking the patient. In the case of children between the ages of three and six years, fundus examination can be more easily achieved by sitting down and asking the standing patient to 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 might 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 casualty situation, which occurs from time to time, is when a child is brought in distressed 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

In an ideal world, all children's eyes would be examined at birth by a specialist and again at six months to exclude congenital abnormalities and amblyopia. This is rarely achieved, although most children in the UK are examined by a nonspecialist at these points. Most children are also screened routinely in school at the age of six years, and any with suspected poor vision are referred for more detailed examination. A further examination is often conducted at the age of nine or ten years and again in the early teens. The commonest defect to be found is refractive error, that is simply a need for glasses without any other problem. The ophthalmological screening is usually performed by a health visitor in the preschool years and a school nurse for older children. Screening tends to include measurement of visual acuity alone but checking any available family history of eye problems would be helpful. When there is a difference in the visual acuity of each eye, the screener should suspect the possibility of a treatable medical condition rather than just a refractive error. A test of colour vision should also be included in the screening programme for older children and this can be conveniently done using the Ishihara plates. It is worth remembering that colour blindness affects 8% of men and 0.4% of women and it might have important implications on the choice of a job. It is also equally important to realise that colour blindness can vary considerably in degree and

can often be so mild as to cause only minimal inconvenience to the sufferer.

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 causing purulent discharge can be treated effectively by the use of antibiotic drops. Although these should clear the unpleasant discharge, the eye continues to water as long as the tear duct is blocked. The mother can be shown how to massage the tear sac. This manoeuvre causes mucopurulent material to be expressed from the lower punctum when there is a blockage and can be used as a diagnostic test. If carried out regularly, this helps to relieve the obstruction. In most cases, spontaneous relief of the obstruction occurs, but if this does not occur after about six to nine months, probing and syringing of the lacrimal passageway under general anaesthesia is an effective procedure, which can be done as a day case. It is important to remember that a watering eye can be caused by excessive production of tears as well as inadequate drainage, and in a child, a corneal foreign body or even congenital glaucoma might 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. This 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 can give the misleading impression that a squint is present. Severe epicanthus can be repaired by a plastic procedure on the eyelids.

Ptosis

Congenital drooping of the eyelid can be unilateral or bilateral and sometimes shows a dominant inheritance pattern. The ptosis can be associated with other lid deformities. Referral for surgery is indicated if there is significant head tilt and especially if the lid covers the visual axis. See Chapter 5 for more information about eyelid deformities.

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 might simply occur because the child cannot see (sensory nystagmus) or they might be caused by an abnormality of the normal control of eye movements (motor nystagmus). It is important to distinguish congenital nystagmus from acquired nystagmus because of 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 other causes of visual impairment in both eyes. A special kind of retinal degeneration known as Leber's amaurosis can present as congenital nystagmus. The condition resembles retinitis pigmentosa, being a progressive degeneration of the rods and cones, and occurs at a young age. It tends to lead to near blindness at school age. Patients with congenital nystagmus usually need to be examined under general anaesthesia, and electroretinography (a technique that can detect retinal degenerations at an early stage) 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 such cases show recessive inheritance. Other abnormalities might 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 the right or left. The distance vision is usually impaired to the extent that the patient might never be able to read 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 one or two years. Like other forms of congenital nystagmus, it can be associated with head nodding.

Albinism

The lack of pigmentation might be limited to the eye, ocular albinism, or it might be generalised. The typical albino has pale pink skin and white hair, eyebrows and eyelashes. There is often 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 can be partial or complete. Albinos need strong glasses to correct their refractive error, which is usually myopic astigmatism. Dark glasses are also usually required because of photophobia. Tinted contact lenses can sometimes be helpful.

Structural Abnormalities of the Globe

There are many different developmental abnormalities of the globe but most of these are fortunately rare. Coloboma refers to a failure of fusion of the foetal cleft of the optic cup in the embryo. Coloboma of the iris is seen as a keyhole-shaped pupil and the defect can extend into the choroid, so that the vision might be impaired. Inspection of the fundus reveals an oval white area extending inferiorly from the optic disc. Children can be born without an eye (anophthalmos) or with an abnormally small eye (microphthalmos). It is always important to find out the full extent of this type of abnormality and if the mother has noticed something amiss in the child's eye, referral to a paediatric ophthalmologist is required without delay. Often a careful discussion of the prognosis with both parents is needed.

Aniridia

Aniridia (congenital absence of the iris) can be inherited as a dominant trait and can be associated with congenital glaucoma. The lens can be subluxated or dislocated from birth. This might be suspected if the iris is seen to be tremulous. This strange wobbling movement of the iris used to be seen in the old days after cataract surgery without an implant, but it is now still seen after injuries to the eye and signifies serious damage. Congenital subluxation of the lens is seen as part of Marfan's syndrome (congenital heart disease, tall stature, long fingers, high arched palate). Congenital glaucoma has already been discussed in the chapter on glaucoma; it can 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"). This enlargement with raised pressure does not occur in adults.

Congenital Cataract

The lens can be partially or completely opaque at birth. Congenital cataract is often inherited and can be seen appearing in a dominant manner together with a number of other congenital abnormalities elsewhere in the body. The condition might also be acquired *in utero*, the best known example of this being the cataract caused by rubella infection during the first trimester of pregnancy: remember the triad of congenital heart disease, cataract and deafness in this respect. 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. The lens fibres are laid down from the outside of the lens throughout life. If the opaque lens fibres are laid down *in utero*, this opaque region can remain in the centre of the lens. Only when the cataract is thick does it present as a white appearance in the pupil and often it is difficult to detect it. It is important to examine the red reflex and see whether the darker opaque lens fibres show up. The surgeon has to decide whether the vision of the child has been significantly affected and unless the cataracts are dense it might be better to wait until the school years approach in order to obtain a more accurate measure of the vision. Sometimes the vision can turn out to be surprisingly good with apparently dense cataracts. The surgical technique is similar to that for cataract surgery in the adult. Before the introduction of lens implants, the risk of developing a retinal detachment in later life was high in

these patients. When the cataract is unilateral, this presents a special case because the affected eye tends to be amblyopic, thus preventing a useful surgical result.

Other Eye Conditions in Childhood

Abnormalities of Refraction

Nowadays children whose vision is impaired because they need a pair of glasses are usually discovered by routine school testing of their visual acuity. They might also present to the doctor because the parents have noticed them screwing up their eyes or blinking excessively when doing their homework. Some children can tolerate quite high degrees of hypermetropia without losing visual acuity simply by exercising their accommodation, and unless there appears to be a risk of amblyopia or squint, glasses might not be needed. 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 five and 14 years, 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 the main features. All cases of squint require full ophthalmological examination because the condition can be associated with treatable eye disease, most commonly amblyopia of disuse. 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 together (binocular vision). In general, the earlier in life that treatment is started, the better the prognosis.

Amblyopia of Disuse

This 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 because of 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 ("lazy eye") but it is also used to refer to loss of sight caused by drugs. Amblyopia of disuse is 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 in early childhood or, more specifically, below the age of eight years. Amblyopia of disuse, therefore, arises as the result of a squint or a one-sided anomaly of refraction, or it can 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 can rapidly lead to amblyopia. Once a clear image has been produced on the retina, either by the wearing of spectacles or other treatment, the vision in the weak eye can 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 eight years it is unlikely that any significant improvement can be achieved by this treatment and, by the same token, it is unlikely that amblyopia will appear after the age of eight years. An adult could suffer total occlusion of one eye for several months without experiencing any visual loss in the occluded eye.

Leucocoria

This term means "white pupil" and it is an important sign in childhood. There are a number of conditions that can 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, opaque nerve fibres in the retina, retinopathy of prematurity, endophthalmitis, some rare congenital abnormalities of the retina and vitreous and, not common but most important, retinoblastoma.

Retinopathy of Prematurity

In the early 1940s, premature infants with breathing difficulties began to be treated with oxygen, and 12 years elapsed before it was

realized that the retinopathy seen in premature children was caused by this treatment. During the course of 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 vessels 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 can rapidly become blind, although some are minimally affected. The management of the condition now involves screening of those children at risk and monitoring of blood oxygen levels. When the condition occurs, treatment with cryotherapy to the peripheral retina has been shown to be beneficial. Now that children are being born at an earlier and earlier stage, it seems that extreme prematurity runs the risk of blindness from this cause even in the absence of supplementary oxygen.

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. It has also been shown that chlamydial infection of the genital tract can lead to the same problem, as can infection by the herpes simplex virus. The blindness that 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 because of the fact that midwives are trained to screen for the condition. Bacterial conjunctivitis usually occurs between the second and fifth day after birth, whereas chlamydial infection tends to occur a little later, between the sixth and tenth day. Purulent or mucopurulent discharge is evident and the eyelids can 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 penicillin drops every hour. 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 history of infection in the parents needs to be explored and managed by a genitourinary specialist.

Uveitis

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

Optic Atrophy

One must be rather wary about the diagnosis of optic atrophy in young children because the optic discs tend to look rather pale in normal individuals. 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 can be listed as follows:

- Causes of optic atrophy without systemic disease include:
  - hereditary optic atrophy
  - drug toxicity.
- Causes of optic atrophy with systemic disease include:
  - glioma of chiasm and craniopharyngioma
  - post meningitic
  - post traumatic after head injury
  - hydrocephalus
  - cerebral palsy
  - disorders of lipid metabolism.

Juvenile Macular Degeneration

This is a rare cause of progressive visual loss in children, the diagnosis being made perhaps

once in a lifetime at primary care level. For this reason, the diagnosis can easily be missed, especially as the patient finds difficulty in reading but no difficulty in walking around. Some cases show dominant inheritance and so the family history can be important.

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 might not become evident until later life. Often, examination of the eye reveals the diagnosis. In Von Recklinghausen's neurofibromatosis, multiple neuro-fibromata are seen on the skin, and the eyelids may be enlarged and distorted. Gliomata can develop in the optic nerves and scattered pigment "cafe au lait" patches are seen in the skin. Brown nodules can be seen on the iris. 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 can leak and expand and lead to detachment of the retina. Similar tumours can be present intracranially.

21

Systemic Disease and the Eye

Diabetes

Diabetes mellitus affects 1–2% of the UK population. The disease is more prevalent in other countries. Diabetic retinopathy is the commonest cause of legal blindness in patients between the age of 20 and 65 years such that about 1000 people are registered blind from diabetes per year in the UK. The management of diabetic eye disease has improved greatly over the past 20 years so that much of the 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 (noninfective) disease that gives rise to blindness. Many diabetics remain free of eye problems, but a diabetic is 25 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, because the incidence of diabetic retinopathy is most related to the duration of diabetes. Other risk factors are listed in Table 21.1.

Diabetic retinopathy is extremely rare under the age of ten 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 can 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 third and sixth cranial nerve palsies than nondiabetics of the same age group. Sometimes isolated third nerve palsy can be painful and the pupil is spared. A fasting blood sugar might be required in patients presenting with isolated third nerve palsies. Hypertension and arteriosclerosis need exclusion.

Cornea and Conjunctiva

Some diabetics have microcirculatory changes, for example conjunctival vascular irregularity and dilatation. Corneal ulcers in diabetics can prove particularly troublesome. Minor trauma

Table 21.1. Risk factors for diabetic retinopathy. 🕮

- Age
- Duration of diabetes
- Smoking
- Hypertension
- Poor diabetic control
- Hyperlipidaemia
- Renal impairment
- Pregnancy

to the cornea can lead to the formation of indolent chronically nonhealing or infected ulcers, which respond slowly to intensive treatment with local antibiotics. Inadequate treatment can 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 diabetics after cataract surgery when there is a severe plastic reaction. It is important that such cases are treated adequately to prevent the development of posterior synechiae, which will make subsequent fundal examination difficult. Many surgeons consider it advisable, therefore, to use mydriatic drops (cyclopentolate) after cataract surgery in diabetics.

Iris

The iris itself often shows degenerative changes in longstanding diabetics. The pupil can react sluggishly and fail to dilate 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 can 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 biomicroscope or a magnifying lens soon reveals the minute irregular blood vessels on its surface. The appearance is known as "rubeosis iridis" or neovascularisation of the iris (Figure 21.1). Neovascular glaucoma occurs once the rubeosis involves the anterior chamber angle. If left untreated, few eyes with rubeosis iridis retain useful sight. The iris should be examined carefully before pupillary dilation.

Lens

It was mentioned in an earlier chapter that diabetics tend to develop senile cataracts at an earlier stage than normal. Once developed, cataracts also progress more quickly in diabetics compared with nondiabetics. In addition, a rapidly advancing type of cataract is seen in young poorly controlled patients. This is a true diabetic cataract. This cataract is bilateral and consists of snowflake posterior or anterior opacities, matures rapidly and is similar to the rare cataract seen in starvation from whatever cause. 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 might change in response to a rise in the blood sugar level. This results from increased hydration of the lens in patients with high uncontrolled blood sugar levels. Undiscovered diabetics quite often become short-sighted because of this so-called index myopia. They can 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 can improve with treatment. In some instances index myopia proves irreversible, being the first sign of cataract formation.

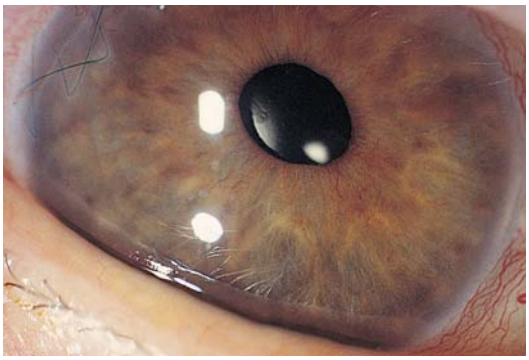


Figure 21.1. Rubeosis iridis. 🕮

Retina and Vitreous

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

Diabetic retinopathy is essentially a small vessel disease affecting the retinal precapillary arterioles, capillaries and venules. The larger vessels can be involved. The vascular disease can take the form of vascular leakage or closure, with resultant ischaemia, or both.

Background Retinopathy

There are usually no ophthalmic symptoms initially, but inspection of the fundi of most diabetics who have had the disease for ten years or more reveals, at first, a few microaneurysms. They are often on the temporal side of the macula but often scattered over the posterior pole of the fundus (Figure 21.2). These might come and go over months and the overall picture could be unchanged for several years. The vision is not affected unless the microaneurysms are clustered round the macular region and leak fluid, resulting in macular oedema. Exudates are also seen and these tend to form rings around areas of diseased vessels, although only one part of the ring might be present at any given point. These are yellowish-white deposits with well-defined edges, which are the result of precipitation of leaked lipoproteins from diseased blood vessels. Capillary dilatation is a more subtle sign of diabetic retinopathy. Haemorrhages, which can be small ("dot") or large blot, result from the venous end of capillaries and are in the deep retina. Flame-shaped haemorrhages can also occur in the nerve fibre layer. "Cotton-wool" spots represent axoplasmic accumulation adjacent to microinfarction of the retinal nerve fibre layer. They

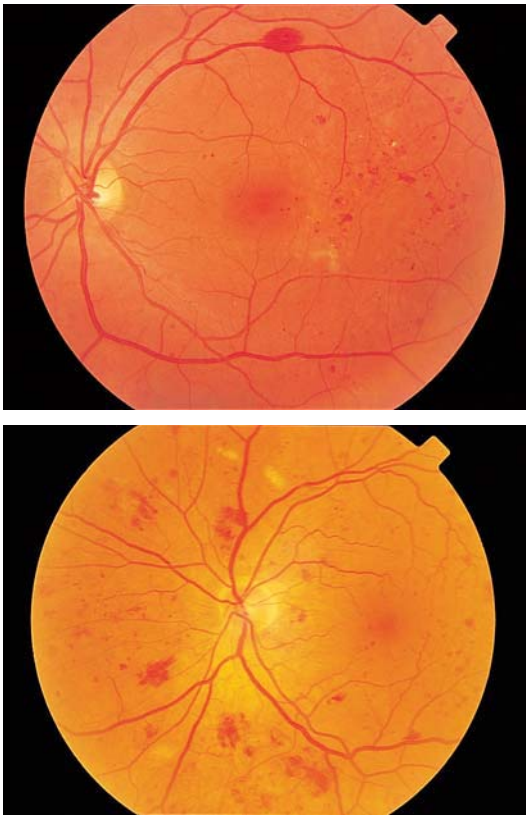


Figure 21.2. Background diabetic retinopathy. **a** Early: microaneurysms and haemorrhages. **b** Severe: extensive haemorrhages, cotton-wool spots and venous dilatation. 🕮

are greyish-white with poorly defined fluffy edges. 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 (pericytes) (Figure 21.3).

The Preproliferative Stage

Proliferative retinopathy is typically seen in poorly controlled diabetics (usually type I diabetes). The situation can become bad quickly and it is important to be able to recognise the warning signs, which occur before proliferation. There are three of them: a large number of dark blot haemorrhages, irregular calibre and dilatation of the retinal veins (beading) and finally, the presence of intraretinal microvascular abnormalities. These warning signs can herald

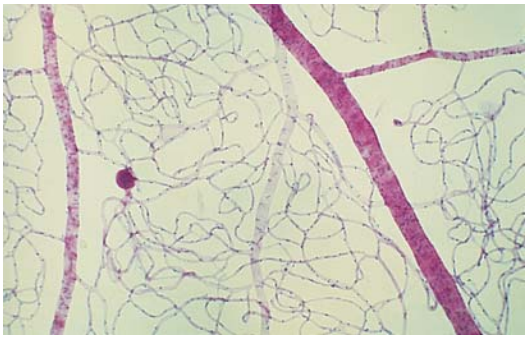


Figure 21.3. Trypsin digest of retina showing microaneursyms and loss of some mural cells.

the appearance of the retinal or optic discs new vessels, which should not be confused with normal disc capillaries or with widened collateral vessels. Approximately 50% of eyes with preproliferative changes will progress to proliferative disease within one year.

Proliferative Retinopathy

Proliferative diabetic retinopathy occurs in 5% of all diabetics. Younger-onset diabetics have an increased risk of the disease after 30 years. Until recently 50–70% of cases of proliferative diabetic retinopathy became blind within five years.

Proliferative diabetic retinopathy is characterised by the development of new blood vessels (neovascularisation) on the optic nerve head or the retina (Figure 21.4). These occur as a response to retinal ischaemia. These new vessels can appear as small tufts, which ramify irregularly. They might be flat initially but enlarge and move forward into the vitreous cavity as they grow. Once the new vessels form and grow, there is increased risk of an acute pre retinal or vitreous haemorrhage. This is a significant threat to vision because the vitreous haemorrhages can become recurrent or dense, preventing any meaningful examination and treatment. Retinal fibrosis, traction retinal detachment and neovascular glaucoma can occur at a later stage.

It is important to appreciate that proliferative retinopathy can be quite severe before the patient notices anything and the situation might have to be explained carefully to him or her.

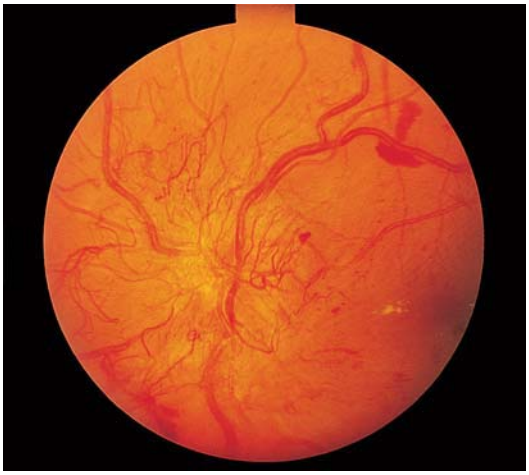


Figure 21.4. Proliferative diabetic retinopathy.

Diabetic Maculopathy

This is the commonest cause of visual impairment in diabetics. It occurs more commonly in type II diabetics. Three types of maculopathy are known and these can occur in isolation or in combination with each other. The three types are:

- Focal caused by focal leakage from a microaneurysm or dilated capillaries and surrounding exudates are seen (Figure 21.5).
- Diffuse oedema caused by diffuse leakage from dilated capillaries at the posterior

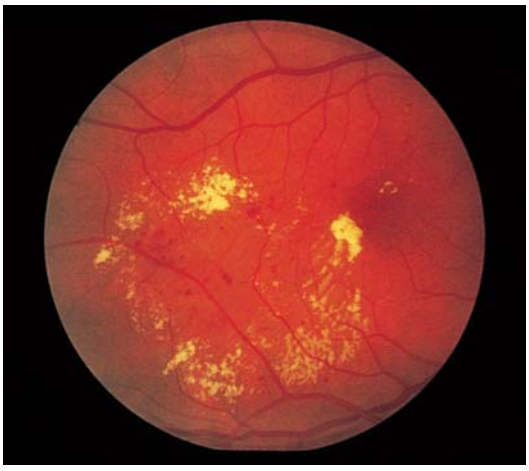


Figure 21.5. Focal maculopathy.

pole of the eye. Retinal oedema is diffuse and can be associated with microaneurysms and few haemorrhages but exudates are absent (Figure 21.6).

- Ischaemic maculopathy is caused by closure of the perifoveal and surrounding vascular network. In addition to diffuse oedema, several dark haemorrhages might be present (Figure 21.7a). Fluorescein angiography might be required to confirm the ischaemia and determine its severity (Figure 21.7b).

Treatment

Control of Diabetes

This aspect of treatment might seem self-evident but in the past the value of careful control has not always been fully recognised. Some patents have the impression that eye problems develop anyway if the diabetes has been present long enough. Nothing could be further from the truth. Control of the diabetic state needs to be sustained. Control or elimination of the known risk factors (see Table 21.1) is also important in reducing the severity of diabetic retinopathy.

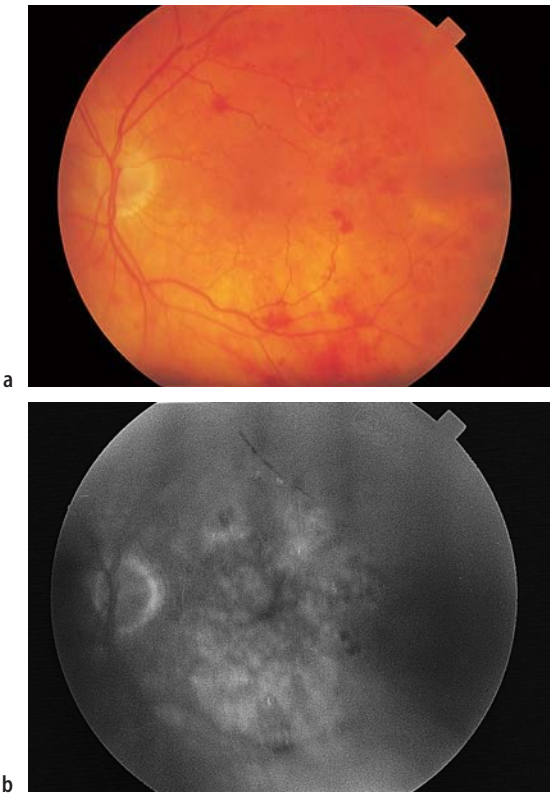


Figure 21.6. Macular oedema: **a** colour photograph; **b** fluorescein angiogram of eye in **a** showing diffuse and cystoid oedema.

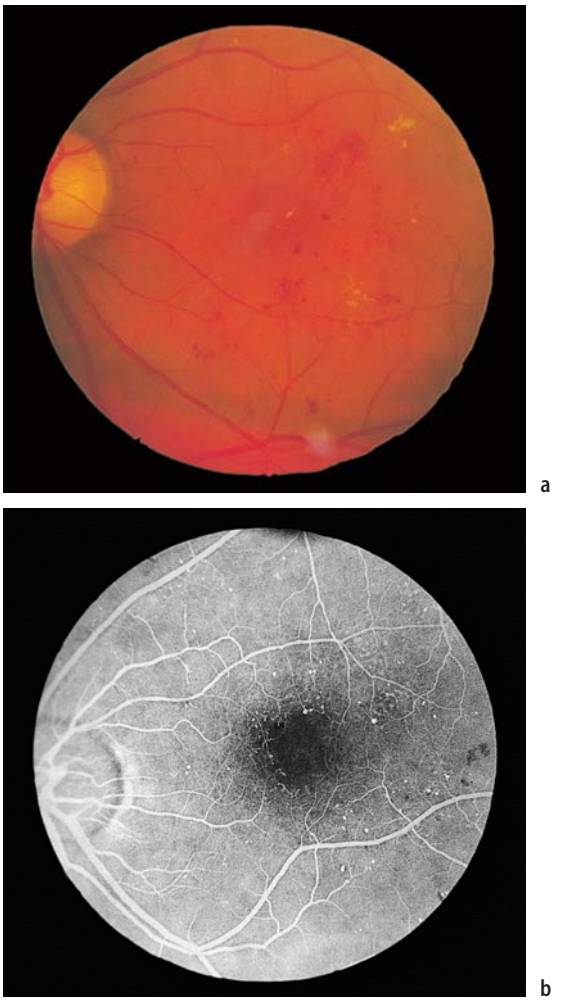


Figure 21.7. Ischaemic maculopathy: **a** colour photograph; **b** fluorescein angiogram.

Laser Photocoagulation

The use of a focused light beam to cauterise the retina has been practiced for several years and the value of this treatment has been confirmed by extensive clinical trials for both proliferative disease and some types of maculopathy. The exact mode of action is not known but it has

been suggested that the photocoagulation of ischaemic areas prevents the release of some, as yet unidentified, vasoformative factor in proliferative disease. The treatment must be applied promptly in the early proliferative stage or sometimes before. About 2500–3000 burns (of 500 µm spot size) are needed in an eye with proliferative retinopathy. This might require several treatment sessions (Figure 21.8). The laser treatment of focal and diffuse maculopathy involves application of small number of burns (of 100–200 µm spot size) to the leaking area, avoiding the fovea. Ischaemic maculopathy generally is less amenable to laser treatment.

Glaucoma Surgery

Drainage surgery might be needed if neovascular glaucoma is not controlled by medical means. Rubeosis iridis initially requires panretinal laser photocoagulation. Chronic simple glaucoma can also be more common in diabetics. Drainage surgery in these cases is less successful than in nondiabetics. Special measures need to be taken to avoid failure.

Vitreo-retinal 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, and relieve traction retinal detachment. Vitrectomy for vitreous haemorrhage tends to be performed sooner these days because of the relative safety of the technique. It can be combined with intraoperative laser photocoagulation.

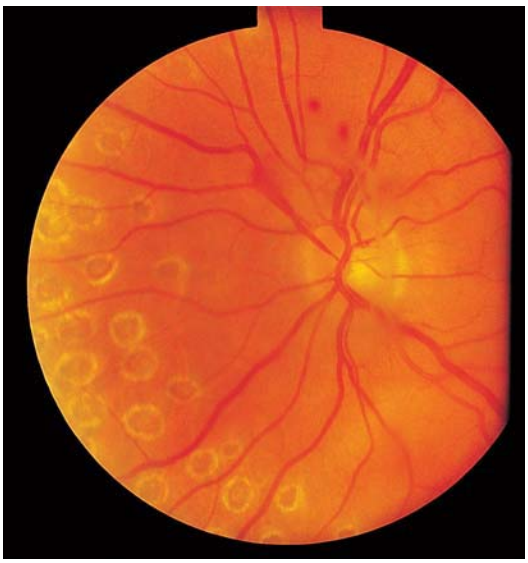


Figure 21.8. Panretinal laser photocoagulation in proliferative diabetic retinopathy.

Prognosis

A better understanding of diabetic retinopathy has resulted from the use of more accurate methods of investigation, especially fluorescein angiography, and 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 and modern technology have 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. Patient education is vital in order to maintain continuing improvement in visual prognosis for diabetics. After 20 years, 75% of diabetics will develop some form of retinopathy. About 70% of patients with proliferative retinopathy will progress to blindness if untreated in five years.

Thyroid Eye Disease

Dysthyroid eye disease is an autoimmune disease in which the manifestations can be notable in the hyperthyroid, euthyroid or hypothyroid phase. Although the ophthalmic features of thyroid disease are often diagnosed in the hyperthyroid phase, a significant number of patients may be euthyroid (i.e., have no other evidence of thyroid disease) or less often hypothyroid at the time of detection of the eye changes. Thus, the ophthalmic disease might precede, be coincidental or follow the systemic manifestations.

Grave's disease is a term used to describe the most common form of hyperthyroidism that has an autoimmune basis. Hyperthyroidism can arise from other conditions, for example thyroid tumour or pituitary dysfunction. It usually affects women between 20 and 45 years. Usually it is characterised by goitre, infiltrative

Table 21.2. The 13 possible eye signs of thyroid disease.

- Proptosis
- Raised intraocular pressure when looking up
- Lid lag
- Lid retraction
- Lid swelling
- Chemosis
- Conjunctival congestion
- Double vision
- Exposure keratitis
- Corneal ulceration
- Optic disc swelling
- Impaired visual acuity
- Constriction of visual field

ophthalmopathy, thyroid acropathy (clubbing) and pretibial myxoedema. When these ophthalmic changes occur in isolation, the condition is described as ophthalmic Graves' disease.

The systemic features of hyperthyroidism include weight loss, high pulse rate, poor tolerance of warm weather and fine tremor. The eye signs of thyroid disease are eyelid retraction and lid lag, puffiness of the eyelids, chemosis, proptosis, exposure keratitis, double vision from muscle involvement and optic neuropathy (Tables 21.2 and 21.3).

- *Lid retraction*. Eyelid drawn up slightly, more on one side than the other. Reveals white sclera above corneoscleral junction (Figure 21.9).
- *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 can be present (Figure 21.10).
- *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. When severe, the conjunctiva overhangs the lower lid margin.

Table 21.3. Routine tests for thyrotoxicosis.

- Serum thyroxine (T4)
- Thyroid autoantibodies
- T3 assay

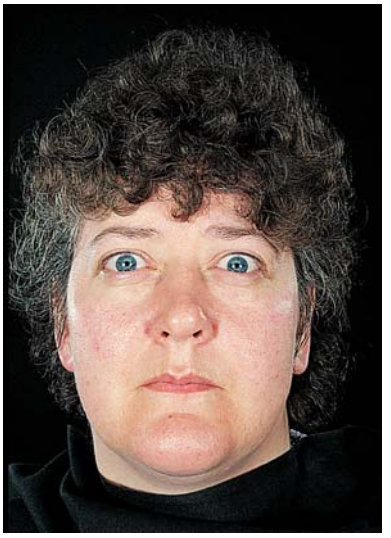


Figure 21.9. Dysthyroid eye disease: eyelid retraction.

- *Proptosis*. Lid retraction can 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. Dysthyroid disease is the commonest cause of unilateral or bilateral proptosis. Forward protrusion of the globe can lead to severe exposure keratitis demanding urgent attention.
- *Exposure keratitis*. Punctate staining with fluorescein across the lower part of the cornea is characteristic and caused by inadequate closure of the retracted upper lid.

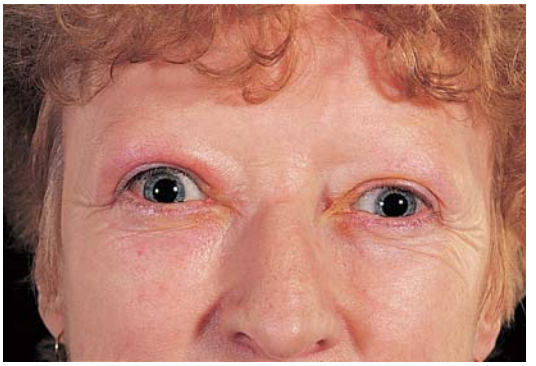


Figure 21.10. Dysthyroid eye disease: lid oedema.

- *Limitation of extraocular muscle action*. The muscles become infiltrated and thickened, producing a characteristic appearance on computed tomography (CT) scan, which helps to distinguish this form from other causes of diplopia. The main restriction of movement is due to infiltration, then subsequently tethering of the inferior recti with limitation of upwards gaze. The resulting pressure on the globe can cause the intraocular pressure to rise on looking up and this has been used as a diagnostic test. The other extraocular muscles are involved less frequently.
- *Optic nerve compression*. This condition occurs in only 5% of cases of thyroid eye disease. However, it is important because of the seriousness of the condition. It is caused by the increased pressure within the orbit, where enlargement of the extraocular muscle causes crowding of the orbital apex with subsequent compression of the optic nerve. The first sign can be swelling of the optic disc, followed by optic atrophy. It is, therefore, vitally important to monitor the visual acuity and central visual field in these cases.

Management

Reassurance is all that might be required in the mild forms of the disease. In some cases, treatment is usually limited to that of the exposure keratitis. Ocular lubrication with artificial tear drops, and 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 can also be improved by the use of guanethidine eye drops.

If there is visual deterioration (from optic nerve compression or significant proptosis), 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 might be necessary to continue with a maintenance dose for many months. Some ophthalmologists might use other immunosuppressive agents, such as azathioprine, or orbital radiotherapy in severe cases of proptosis and/or optic nerve compression. If there is no response between 24 h and 48 h, surgical decompression of the orbits is required. If double vision persists beyond the acute stage, extra-ocular muscle surgery can be helpful and operations have also been designed to deal with lid retraction.

Hypertension

Although the effects of raised blood pressure on the appearance of the fundus of the eye were recognised in the nineteenth 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 because of natural ageing. It is now accepted that the exact cause of the raised blood pressure does not by itself influence the fundus appearance. However, the appearance of the retinal vessels and associated changes serve as a good guide to the severity of the disease and urgency of treatment.

The Effect of Age on the Retinal Blood Vessels

In older patients the retinal arteries are seen to be narrower and straighter and the veins are also narrower than in younger patients. The term "retinal arteriosclerosis" is used to describe these changes.

The Effects 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 because of spasm of the vessel walls. This hypertonicity 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 can be distended. Occasional flame haemorrhages, cotton-wool spots and exudates might indicate more severe vascular damage but do not necessarily lead to "malignant" hypertension (Figure 21.11).

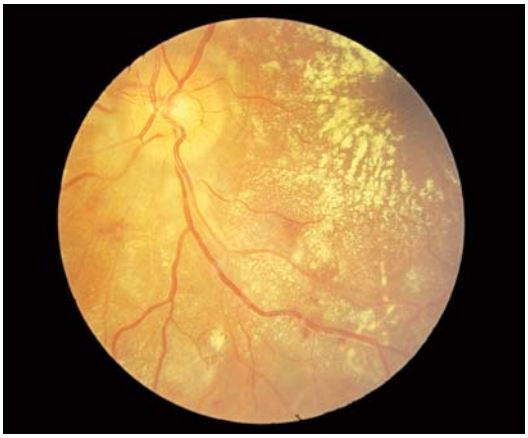


Figure 21.11. Hypertensive retinopathy: haemorrhages, cotton-wool spots, exudates, vascular calibre changes.

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 might be present in more severe cases. The cotton-wool spots of hypertension reflect ischaemic damage to the nerve fibre layer caused by obstruction of the retinal precapillary arterioles. Exudates are caused by abnormal vascular permeability.

"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 might be only slightly reduced unless there is significant macular oedema and there might be some enlargement of the blind spot and 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 110–120 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 from other causes. 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.

Other Associated Vascular Changes

Retinal Vascular Occlusion

This is more common in hypertensive patients compared with normotensives. The most frequent occurrence is the central retinal vein occlusion (CRVO), although branch retinal vein occlusions can occur at arteriovenous crossings. The fundus appearance in CRVO is dramatic with numerous scattered haemorrhages and swelling of the optic disc, and the patient experiences sudden blurring of vision in one eye (Figure 21.12). This can be compared with occlusion of the central retinal artery, which is less common and in which the prognosis is uniformly worse. Here, the fundus appears pale and the arteries are narrowed. There is a cherry-red spot at the macula.

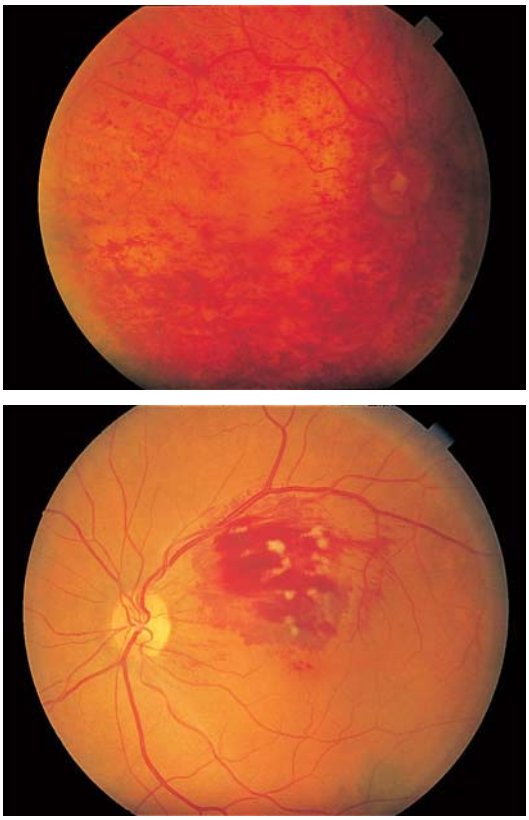


Figure 21.12. **a** Central retinal vein occlusion and **b** macular branch retinal vein occlusion.

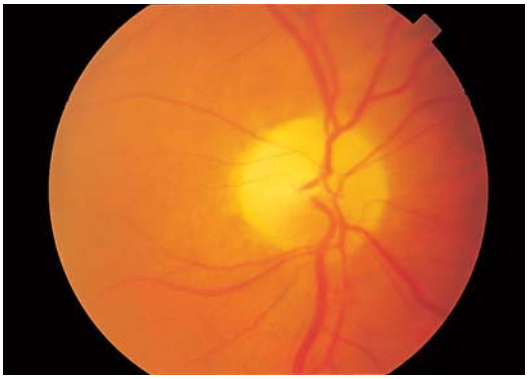


Figure 21.13. Anterior ischaemic optic neuropathy. The superior part of the disc is pale.

Emboli

Cholesterol emboli can be seen in the retinal arteries, sometimes in association with arterial occlusion. These usually arise from atheromatous plaques in the carotid artery. Calcified emboli can be seen in association with diseased heart valves and platelet or fibrin emboli can 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 retinal artery occlusion, giving an altitudinal defect of the visual field. This appearance should suggest the possibility of temporal arteritis and an erythrocyte sedimentation rate (ESR) and a temporal artery biopsy should be considered as urgent investigations (Figure 21.13).

However, there is a group known as "non-arteritic" or idiopathic anterior ischaemic optic neuropathy (AION), which occurs in otherwise healthy individuals between 45 and 65 years of age. About one-third of these patients develop bilateral disease. In these patients, retinal arterial occlusion is absent. There is no known treatment for nonarteritic AION but giant cell arteritis needs to be excluded.

Anaemia

When the haemoglobin concentration in the blood is abnormally low, this becomes apparent in the conjunctiva and ocular fundus. The conjunctiva, similar to oral mucosa, is pale. 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 can be misleading. In severe cases, small haemorrhages are usually seen, mainly around the optic disc. The haemorrhages tend to be flame-shaped but a special feature of anaemic retinopathy is the presence of white areas in the centre of some of the haemorrhages. The haemorrhages might be due to associated low platelet counts. In pernicious anaemia, retinal haemorrhages and bilateral optic neuropathy that manifests as centrocaecal scotomas are seen. In severe cases, the optic nerves are atrophic. Anaemia secondary to blood loss can give rise to ocular hypoperfusion, which leads to anterior ischaemic optic neuropathy. 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

All ocular tissue can be involved in leukaemia. The eye changes can occur at any time during the course of leukaemia, or they can make up the presenting features of the disease. These changes are more common in the acute leukaemias than in the chronic types.

Two groups of ophthalmic manifestations are recognised in leukaemias. The first group consists of leukaemic infiltration of ocular structures, for example retinal and preretinal infiltrates or anterior chamber and iris deposits. All of these are quite uncommon. The second group of manifestations is considered to be secondary to the haematological changes, for example thrombocytopenia, increased blood viscosity and highly increased leucocyte count. These changes include subconjunctival haemorrhages and intraretinal haemorrhages, including white centred ones, cotton-wool spots, "slow flow retinopathy" (Figure 21.14) and retinal venous occlusions (especially CRVO).

Less common manifestations include choroidal infiltrations, and retinal and optic disc neovascularisations. Apart from eye changes, the vision can be impaired by

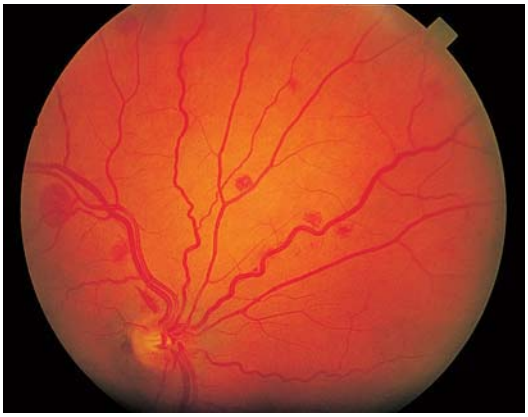


Figure 21.14. The fundus in leukaemia. Note dilated veins and haemorrhages.

leukaemic infiltrates elsewhere in the visual pathway (leading to field defects).

Ocular disease can also occur as complications of treatment of the leukaemia, for example opportunistic infections such as herpes zoster, graft-versus-host reactions and intraocular haemorrhage.

Sickle-cell Disease

This condition is mentioned separately because of the severe and devastating effect it can have on the vision. The sickle-cell haemoglobinopathies are inherited and result from the affected person having one or more abnormal haemoglobins as 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. The abnormal haemoglobins occur either in combination with normal haemoglobins resulting in AS (sickle-cell trait) or in association with each other: SS (sickle-cell anaemia or disease) or SC (sickle-cell haemoglobin C disease) and S thal (thalassaemia). Individuals with cell trait usually lead a normal life and do not have any systemic or ocular complications. The red blood cells in patients with sickle-cell (SS, SC, S thal) disease adopt abnormal shapes under hypoxia and acidosis. These abnormal red cells are less deformable compared with normal, leading to occlusion of the small retinal blood vessels especially in the retinal periphery.

Sickle-cell retinopathy can be divided into two types: (1) nonproliferative and (2) proliferative. In nonproliferative sickle retinopathy there is increased venous tortuosity, peripheral chorioretinal atrophy, peripheral retinal haemorrhages, peripheral haemosiderin deposits, which appear refractile, and peripheral arterial occlusion. These lesions are usually asymptomatic. When central retinal arterial or venous occlusion, macular arteriolar occlusion or choroidal ischaemia occurs, there is significant visual deficit.

When significant ischaemia is present, retinal neovascularisation occurs. This is generally in the retinal periphery. Such peripheral neovascularisation can respond to laser photocoagulation or cryotherapy of the retina. Occasionally vitrectomy is required.

Onchocerciasis

Onchocerciasis, commonly known as river blindness, is caused by the filaria *Onchocerca volvulus*. The name "river blindness" is derived from the occurrence of the disease in focal areas along rivers and streams where the blackfly (*Similium*) breeds in fast-flowing water. The blackfly can travel several kilometres and does not respect international borders.

The disease is characterised by a few adult worms encased in nodules and the invasion of the body by microfilaria produced by the adult worms. It is endemic in equatorial Africa – West and Central – and Central and South America. It is estimated that there are about half a million blind people because of onchocerciasis.

The adult worm has a lifespan of 15–30 years. The microfilaria is sucked up by the blackfly when it takes its blood meal. Subsequently, division within the blackfly gives rise to latter stages of the larva, which are re-injected into the skin of the next victim of the blackfly's bite. The microfilariae migrate under and through the skin and may mature in about one year. Newly produced microfilariae migrate to the eye through the skin or blood.

Clinical manifestations of onchocerciasis can be divided into extraocular and ocular manifestations.

Extraocular Features

Skin involvement is in the form of pruritis – a maculopapular rash, which can be associated with hypo- or hyperpigmentation, dermal and epidermal atrophy or "onchodermatitis".

There might be subcutaneous nodules, which are firm, round masses in the dermis and subcutaneous tissue, especially close to joints in the head and shoulder.

Ocular Features

Intraocular microfilariae can be seen in the anterior chamber. Dead microfilariae are usually seen in the cornea (especially peripherally).

Other ocular features are punctate keratitis and sclerosing keratitis; anterior uveitis, usually of the nongranulomatous type with loss of the pigment frill, and posterior synechiae are common. Secondary cataract and glaucoma can develop.

Chorioretinitis of the chronic nongranulomatous type can occur, with secondary degenerative changes in the retinal pigment epithelium (RPE) neuroretina and the choriocapillaries. There might be granular atrophy of the RPE, subretinal fibrosis, retinal arteriolar attenuation and vasculitis. Optic atrophy and neuropathy are not uncommon.

Diagnosis is confirmed by skin snip and the Mazzoti test, which depends on a Herxheimer reaction to a single dose of diethylcarbamazine. Care is required with this test because the reaction could be severe.

Management

One method is by vector control. An international (World Health Organisation) programme, the onchocerciasis control programme, has been successful in reducing the endemicity of the disease in the Volta river basin.

Chemotherapy of infected patients now uses Ivermectin, which in a single dose rids the patient of microfilariae for one to two years. This medication needs to be repeated over several years in mass administration projects.

Diethylcarbamazine is the older treatment for the microfilariae but is more toxic and requires to be taken over a two- to three-week period. Adult worms can only be killed by suramin, or removed surgically.

Acquired Immune Deficiency Syndrome (AIDS)

Acquired immune deficiency syndrome (AIDS) refers to the final stages of infection by the human immunodeficiency virus (HIV). The earlier stages of the disease are often asymptomatic (Table 21.4).

In western countries, AIDS commonly affects homosexuals, haemophiliacs, and intravenous drug abusers, although there is now a significant heterosexual and paediatric pool of patients. In Africa, it is generally a heterosexual disease, and a significant paediatric population is also known. Transmission is through sexual intercourse, parenteral or transplacental routes.

Ocular features occur in 75% of patients with AIDS. The major ocular complications of AIDS occur later in the disease and can be predicted by CD4 T-cell levels. At CD4 level $>200 \times 10^6$/L common ocular complications are toxoplasmosis and herpes zoster ophthalmicus and retinitis, while at CD4 levels $<50 \times 10^6$/L cytomegalovirus (CMV) retinitis is common.

AIDS microangiopathy (noninfectious) occurs in about 50% of patients (in both developing and western countries). It consists of microaneurysms, telangiectasia, cotton-wool spots and a few retinal haemorrhages. Retinal peripheral perivascular sheathing may sometimes occur in the absence of intraocular infections.

Other ocular involvement of AIDS includes infections with opportunistic and

Table 21.4. Classification of human immunodeficiency virus (HIV) infection (Centers for Disease Control, Atlanta, 1992).

Group I	Acute infection: asymptomatic with seroconversion
Group II	Asymptomatic carrier
Group III	Generalised, persistent lymphadenopathies; usually good state of general health
Group IV AIDS	
Sub-groups	(A) Constitutional (cachexia, fever, etc.). (B) Neurological. (C) Infections diagnostic of AIDS. (D) Malignancies. (E) Others, e.g. CD4 count $<200 \times 10^6$/L.

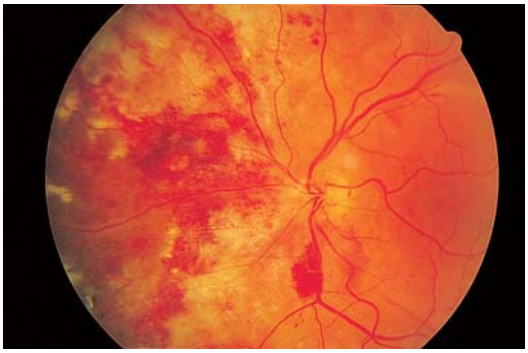


Figure 21.15. Cytomegalovirus retinitis in acquired immune deficiency syndrome (AIDS).

nonopportunistic organisms (e.g., CMV, cryptococcus and *molluscum contagiosum*) (Figures 21.15 and 21.16). Neoplasms of the conjunctiva, lids and orbit, and neurophthalmic complications are other features.

In western countries, the commonest ophthalmic complication of AIDS is CMV retinitis, while in developing countries (such as Africa), CMV is not a major problem. Herpes zoster ophthalmicus and conjunctival carcinoma are common in AIDS patients in Africa and AIDS patients die of other complications, for example tuberculosis. Therefore, short-term survival from AIDS itself is a problem in developing countries, while in western countries quality of life for the longer term is the main problem.

Treatment with the highly active antiretroviral therapy (HAART) regimen leads to significant elevation of CD4 T-cell levels such that the ocular complications, especially opportunistic infections, are less commonly encountered.

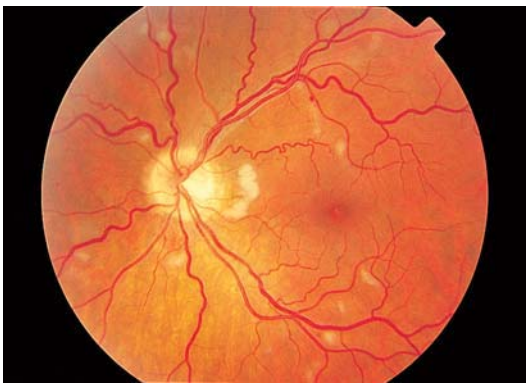


Figure 21.16. Human immunodeficiency virus retinopathy.

Ophthalmological Signs of AIDS

1. Noninfectious retinopathy:
   (a) cotton-wool spots
   (b) retinal haemorrhages
   (c) microvascular changes.
2. Opportunistic infections:
   (a) Involvement of posterior segment:
      - CMV retinitis
      - acute retinal necrosis (herpes simplex, herpes zoster)
      - toxoplasmic chorioretinitis
      - *Pneumocystis carinii* choroiditis
      - tuberculous choroiditis
      - endophthalmitis caused by *Candida albicans* – usually intravenous drug users
      - cryptococcus chorioretinitis
      - syphilitic retinitis

   (b) Involvement of anterior segment:
      - chronic keratitis and keratouveitis caused by herpes zoster and herpes simplex
      - keratoconjunctivitis caused by CMV, microsporum and gonococcus
      - corneal ulcer caused by *Candida albicans*, and bacteria (*Pseudomonas aeruginosa, Staphylococcus aureus*, and *Staphylococcus epidermidis*)
      - syphilitic and toxoplasmic iridocyclitis
      - conjunctivitis caused by CMV, herpes zoster and herpes simplex
      - bacterial conjunctivitis.
3. Neoplasms:
   (a) conjunctival, palpebral and orbital Kaposi's sarcoma
   (b) intraocular lymphoma
   (c) other neoplasms:
      - conjunctival squamous carcinoma
      - palpebral and orbital lymphoma.
4. Neuro-ophthalmological signs:
   (a) Involvement of cranial nerves:

- internuclear ophthalmoplegia
- third, fourth and sixth cranial nerve palsies
- retrobulbar neuritis and papillitis

(b) Homonymous haemianopia

(c) AIDS–dementia complex with cortical blindness.

5. Other signs:

(a) Conjunctiva:
- nonspecific conjunctivitis
- keratoconjunctivitis sicca
- nonspecific conjunctiva microvascular changes in the inferior perilimbal bulbar region (haemorrhages, microaneurysms, column fragmentation, dilatation and irregular vessel diameter)
- bacterial conjunctivitis

(b) Cornea:
- nonspecific punctate keratitis

(c) Sclera:
- necrotising scleritis.

(d) Retina:
- talc-induced retinopathy (only intravenous drug users)

(e) Eyelids:
- herpes zoster ophthalmicus
- palpebral *molluscum contagiosum*
- palpebral cryptococcosis

(f) Orbit:
- orbital apex granuloma
- orbital pseudotumour
- orbital infiltration by *Aspergillus*, *Pneumocystis carinii*
- orbital cellulitis

(g) Visual and refraction defects:
- night blindness because of vitamin A and E malabsorption
- progression of myopia
- decreased accommodation

(h) Acute closed-angle (bilateral) glaucoma caused by choroidal effusion.

22

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 a subspecialty. Retrobulbar neuritis, for example, is a condition that 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 can require urgent medical 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 nonseeing 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 or contour, the vessel entry, the central cup and the presence or absence of haemorrhages.

Colour

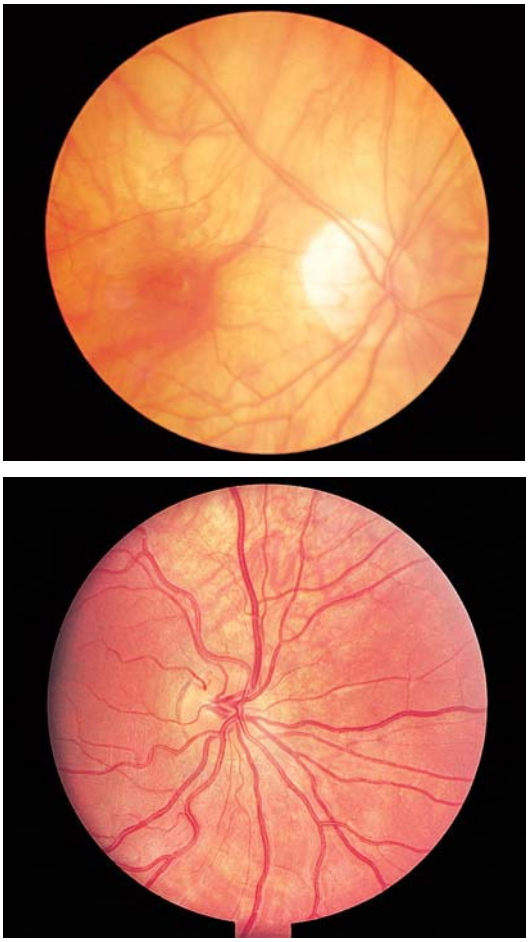
The disc is pink but often slightly paler on the temporal side. That of the neonate might be deceptively pale and some elderly discs appear atrophic without evidence of disease. Pallor of the disc is caused by loss of nerve tissue and small blood vessels of the surface of the disc. In severe optic atrophic cupping, there is exposure of the underlying sclera. The myopic disc is relatively pale, whereas the hypermetropic disc is pinker than normal (Figure 22.1).

Margins

These are better defined in myopic than in hypermetropic subjects. In hypermetropes the edges of the disc can appear raised, sometimes resembling papilloedema. It is common to see a crescent of pigment on the temporal side of the disc. Frequently, an area of chorioretinal atrophy is present at the disc margin in myopes and can give rise to difficulty in deciding where the true disc margin is.

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 spontaneously in 80% of the population if examined carefully. In the other 20% of normal individuals, venous pulsation at the disc can be induced by gentle pressure on the globe.



a

b

Figure 22.1. Normal optic disc in **a** myope and **b** hypermetrope.

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 one-third (or less) of the total disc diameter in normal subjects. The ratio between the vertical diameter of the cup and the total disc diameter is known as the cup-to-disc ratio. Thus, the normal cup-to-disc ratio is <0.3.

Haemorrhages

Haemorrhages are never seen on or adjacent to normal discs. If present, they warrant further investigation.

Congenital Disc Anomalies

A number of minor congenital abnormalities are seen on the disc. In an astigmatic eye, the disc is often oval. The central cup might be filled in by "drusen" – small hyaline deposits, which can be found on the surface or buried in the substance of the disc. This appearance can mimic papilloedema. Alternatively, the central cup might be hollowed out further by a congenital pit in the disc. Myelinated retinal nerve fibres are recognised by their strikingly white appearance, which obscures any underlying vessels, and their fluffy margin (see Figure 22.3). The central cup can be filled in by persistent remnants of the hyaloid artery (Bergmeister's papilla), which runs in the embryo from disc to lens. Some of these and other congenital abnormalities of the disc can be associated with visual field defects that 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 and the cup of disc tends to be larger 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 difficult 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 can be mimicked by optic atrophy because of chiasmal compression. When optic atrophy follows swelling of the optic disc, there is more gliosis than when it is "primary", that is, caused by disease in the nerve

itself. Gliosis makes the appearance of the disc more grey or yellowish-grey than white and the cribriform markings often seen in optic atrophy might not be evident.

The following are the important causes of optic atrophy:

- Glaucoma.
- Vascular. Following obstruction of the central retinal artery or vein, giant cell arteritis and nonarteritic anterior ischaemic optic neuropathy.
- Following disease in the optic nerve, for example optic neuritis, or compression of the nerve by an aneurysm or tumour (Figure 22.2).
- Following papilloedema. The disc can become atrophic as a direct result of the chronic swelling, irrespective of its cause.
- 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 toward blindness, the discs become atrophic. Optic atrophy might also appear in certain families without any other apparent pathology, for example Leber's hereditary optic neuropathy and autosomal dominant optic atrophy. It is also seen in the rare but distressing cerebroretinal degeneration, which presents with progressive blindness, epilepsy and dementia.

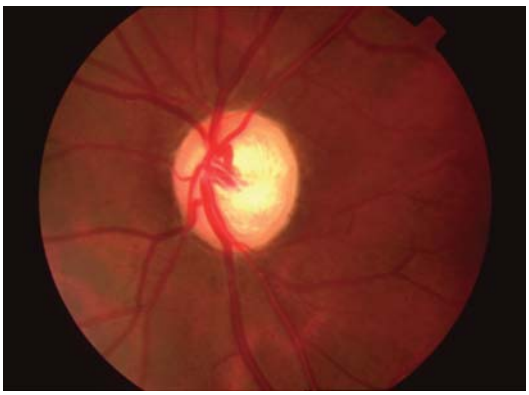


Figure 22.2. Optic atrophy caused by pituitary compression of the optic nerve.🕮

- 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 resulting from excessive smoking of coarse tobacco, usually in a pipe and often in association with a high ethyl alcohol intake. Reversal can be achieved by abstention in the early phases of the disease. Other toxic agents include ethambutol, isoniazid, digitalis and lead.
- Trauma. The optic nerve can be damaged by indirect injury if bleeding occurs into the dural sheath. This can 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 could be blind but the fundus normal. The pupil reaction to direct light is impaired from the time of the injury. Such an injury can result in complete and permanent blindness in the affected eye but a degree of recovery is achieved in a small proportion of cases, if decompression of the nerve sheath is undertaken early.

Swelling of the Optic Disc

This is a serious sign because it could be caused by 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 might 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 (Figure 22.3).

Vascular

The disc can 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 anterior ischaemic optic neuropathy. In the latter instance, swelling of the disc occurs in

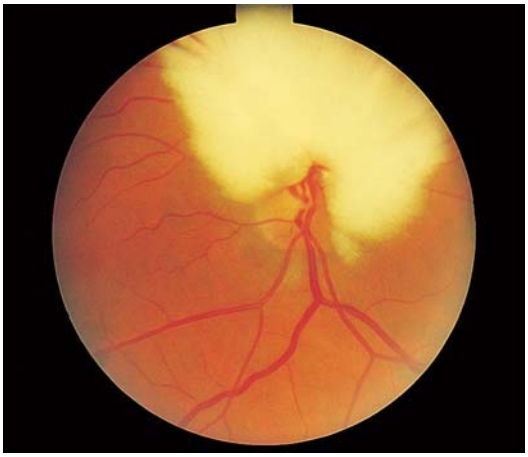


Figure 22.3. Myelinated nerve fibres. 🕮

association with arterial disease and one must take pains to exclude temporal arteritis in the elderly.

Postoperative

Swelling of the disc is not uncommon in the immediate postoperative period after intraocular surgery. It is caused by ocular hypotony. It can persist for longer periods if the intraocular pressure remains low. It is not usually regarded to be of serious significance, because the swelling regresses following normalisation of the intraocular pressure.

True Papilloedema

Papilloedema is swelling of the optic discs because of increased intracranial pressure. Every doctor must be aware of the triad of headache, papilloedema and vomiting as an important feature of raised intracranial pressure. The optic disc might be markedly swollen and haemorrhages are present around it, but not usually in the peripheral fundus (Figure 22.4). In chronic papilloedema, the disc is paler and haemorrhages might be few or absent. Although these patients might 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 noninflammatory swelling of the disc, which results from raised intracranial pressure. The most common causes of raised intracranial pressure are cerebral tumours, hydrocephalus idiopathic (benign) intracranial hypertension, subdural haematoma, malignant hypertension and cerebral abscess.

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 can also be of great diagnostic help in difficult cases when abnormal disc leakage occurs.

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 (retrobulbar neuritis).

Other Causes

Chronic intraocular inflammation, such as anterior, intermediate or posterior uveitis, can be complicated by disc swelling. Severe diabetic eye disease can sometimes be marked by disc swelling (diabetic papillopathy). In severe cases of thyroid orbitopathy, the orbital congestion

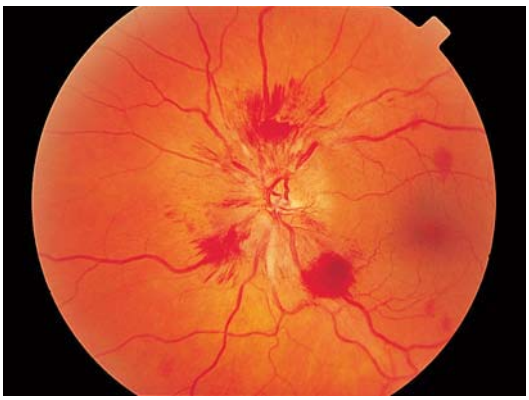


Figure 22.4. Papilloedema. 🕮

can cause disc swelling (dysthyroid optic neuropathy). In both instances, the doctor should be warned that serious consequences might ensue unless prompt treatment is applied. Infiltration of the disc by leukaemia, lymphoma or chronic granulomata (as in sarcoidosis) can also cause disc swelling.

Multiple Sclerosis

This common and important neurological disease can often present initially as an eye problem and its proper management requires careful co-ordination at the primary care level. It is important to realise that multiple sclerosis should not be diagnosed after one single attack of optic or retrobulbar neuritis because this could cause unnecessary alarm about something that might never happen. Studies have shown that between 45% and 80% of patients with optic neuritis will develop multiple sclerosis after 15 years of follow-up. Furthermore, optic neuritis has causes other than multiple sclerosis. 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 demyelination plaques are detectable on magnetic resonance imaging scans of the brain. The optic nerve between globe and chiasm is commonly involved at an early stage and there might 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

Optic or 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 severe cases, the sight of the affected eye can be lost completely. On examination, a relative afferent pupil defect on the affected side might be the only objective evidence of disease. It is essential to test the pupil before dilating it with eye drops. The fundus is often normal initially (retrobulbar neuritis), although there can be slight swelling of the optic disc (optic neuritis). After two or three weeks the optic disc starts to become pale. The visual prognosis is generally good. Most patients make a complete or nearly complete recovery after 6–12 weeks. The attack is unilateral in 90% of cases, although there is a risk that the other eye can be affected at a later date and recurrent attacks in one or both eyes can 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 advisable to make the diagnosis in retrospect. The patient might give a history of visual loss in one eye, which has recovered, and at a later date, presents with other nonocular signs and symptoms of demyelinating disease. If it can be confirmed that the patient has had a previous attack of optic neuritis, this can help in the confirmation of the diagnosis of disseminated sclerosis. Under these circumstances, the pallor of the disc can be helpful, but careful assessment of the colour vision, visual acuity and measurement of the visually evoked potential can provide conclusive evidence. At the time of the acute attack, testing the visual field might reveal a central scotoma. The size of this defect diminishes as healing occurs, often leaving a small residual defect between blind spot and central area.

Corticosteroids administered systemically can speed up recovery of vision. However, the final visual outcome is unchanged by such treatment.

Nystagmus

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

Internuclear Ophthalmoplegia

Whereas double vision is a common symptom in multiple sclerosis, it is unusual to see an

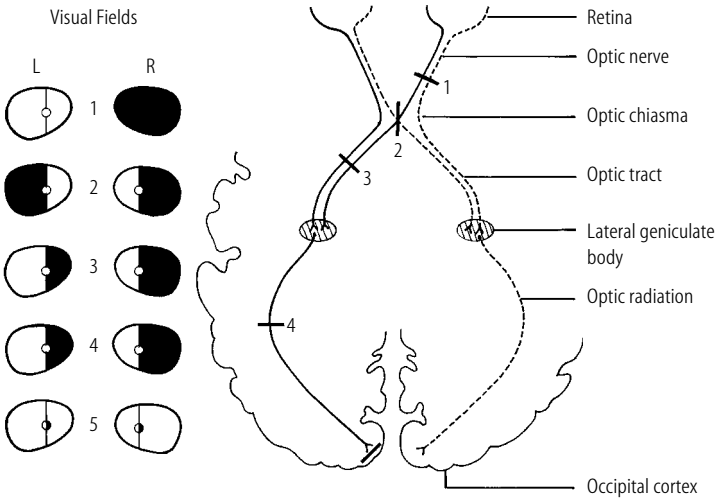


Figure 22.5. The visual pathway.

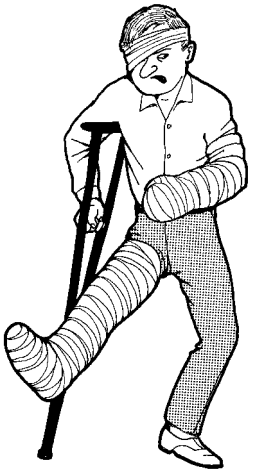
obvious defect of the ocular movements. Sometimes it can be seen that one eye fails to turn inwards when the patient is asked to look to the opposite side, and yet when the patient is made to converge the eyes on a near object, the medial rectus moves normally. This failure of the muscle action with certain co-ordinated eye movements only (i.e., limitation of adduction), while the opposite abducting eye shows nystagmus, is termed "internuclear ophthalmoplegia". It is characteristic of multiple sclerosis when seen in young people (when the internuclear ophthalmoplegia is usually bilateral and is caused by a demyelinating lesion in the pons) but usually has a vascular cause in the elderly (when it is usually unilateral).

Other Features

Other types of ocular muscle dysfunction, 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, especially in the periphery (peripheral retinal vasculitis).

Defects in the Visual Fields

The pattern of a visual field defect gives useful localising information for lesions in the visual pathway. The right half of each retina is linked by nerves to the right occipital cortex and the splitting of nerve fibres from each half occurs at the chiasm. For this reason, 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 (Figure 22.5). Cortical lesions tend to be more congruous. That is to say, the blind areas on each side tend to be similar in shape and size. Cortical lesions also show better preservation of central vision ("macular 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 because of ocular disease are relatively common as, for example, those seen in the elderly with glaucoma. Care must be taken to interpret field defects with this possibility in mind. Notice from the diagram in Figure 22.5 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 might be unaware of a severe visual field defect, especially in hemianopia, providing that the macula is spared (Figure 22.6).



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

Figure 22.6. The effects of hemianopia.

Miosis refers to a small pupil, mydriasis to a large pupil (big word, big pupil). The pupil grows smaller with age, as does reactivity. In young children the pupils are relatively large and sometimes anxious parents bring up their children because they are concerned about this. During sleep, the pupils become small. 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 light pupil reaction, but as a general rule, the pupils remain of equal size. It should be apparent from Figure 22.7 that the patient with cortical blindness (lesion within the occipital cortex) might have a normal pupil reaction. We must also remember that a pupil might not react to light because it is mechanically bound down to the lens by adhesions (posterior synechiae). When both maculae are damaged by senile macular degeneration, the

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 travelling within the third cranial nerve. The afferent stimulus is conveyed along the optic nerves and decussates at the optic chiasm and continues as the optic tract. The specific pupillomotor nerve fibres leave the optic tract without synapsing in the lateral geniculate nucleus and pass to the pretectal nucleus of the midbrain, where they synapse with interneurons. The interneurons project to both Edinger–Westphal nuclei (part of the third cranial nerve nucleus). The pupillomotor fibres then travel within the third cranial nerve to the pupil constrictor muscles of the ipsilateral eye via the ciliary ganglion (Figure 22.7).

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.

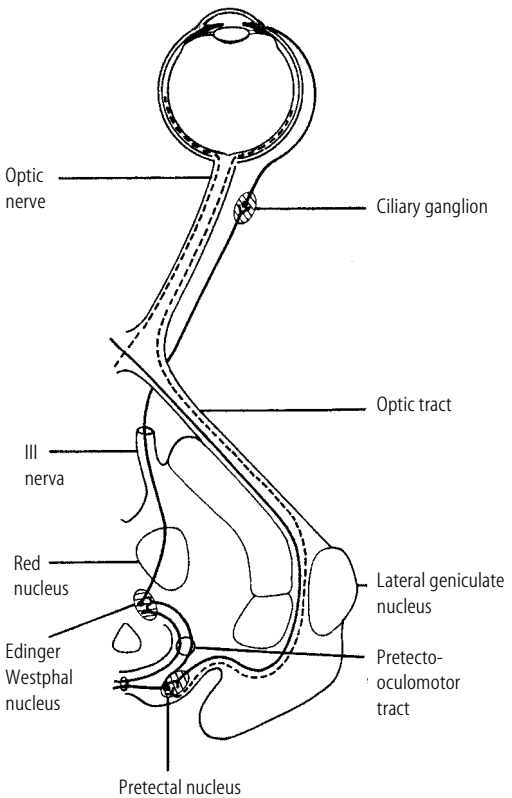


Figure 22.7. The pupillary pathway.

pupils can be slightly wider than normal and might show sluggish reactions. A relative afferent pupil defect (also known as a Marcus Gunn pupil) implies optic nerve or severe retinal disease.

The Abnormally Dilated Pupil

The most common reason for unilateral mydriasis is drugs in the form of locally administered eye drops, either prescribed by an ophthalmic department or obtained from a friend's medicine cabinet. The next most common cause is probably the Adie's pupil, a condition that is more common in young female patients. The affected pupil is usually dilated and contracts slowly in response to direct and indirect stimulation. In bright light, the pupil might constrict slowly on the affected side and take some time to dilate in the dark. The vision might also be blurred, particularly at near fixation because of the effect of the disease process on the ciliary muscle (necessary for accommodation). The pupillary constriction to near fixation is tonic and prolonged and worm-like. When this tonic pupil reaction is combined with absent tendon jerks in the limbs, it is known as Holmes–Adie syndrome. When the vision is blurred and the pupil widely dilated, the symptoms can 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. It is thought to have an underlying viral aetiology.

Acute narrow-angle glaucoma can occasionally present in this manner and confusion can arise if the eye is not red; however, closer examination of the eye should make the diagnosis obvious. Because 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 can be a serious sign of raised intracranial pressure after head injury. One pupil might be wider than the other as a congenital abnormality (congenital anisocoria).

The Abnormally Constricted Pupil

Again, drugs are a common cause. Miotic drops are still encountered in the treatment of 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 because of paralysis of the smooth muscle in the eyelids (Müller's muscle), loss of sweating over the affected side of the forehead, a slight reduction of the intraocular pressure and enophthalmus (sunken globe). Horner's syndrome can be caused by a wide diversity of lesions anywhere along the sympathetic pathway. While a Pancoast's apical lung tumour is classically associated with Horner's syndrome, it is quite often noted in the elderly as an isolated finding and investigation fails to reveal a cause. The Argyll Robertson (AR) pupil is a rare but famous example of the miosed 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. Visual acuity is normal in such patients.

Double Vision

Double vision (diplopia) can 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 a cataract or corneal disease. Some patients say that they can see double when they mean that the vision is blurred. A clear distinction must, therefore, be made. Binocular 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 can 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 distance vision or whether there is horizontal or vertical displacement of the second image, then a possible cause can be suspected even before examining the patient.

Assessment of Eye Movements in Diplopia

The complaint of double vision suggests that the separate eyes are not both fixed on the point of regard. The eye that is "off line" sees the object of regard but it appears displaced. This failure of the eyes to work together is because of malfunction of one or a group of eye muscles or the neurological mechanisms that 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 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 (Figure 22.8). 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.

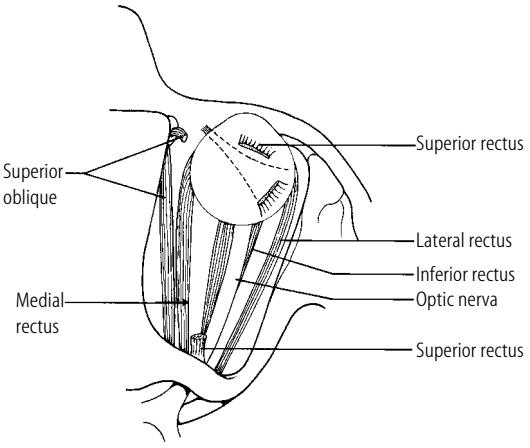


Figure 22.8. The extraocular muscles.

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 rapidly by suppressing the image from one eye, and under the age of eight years the suppression can lead to 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 of Diplopia

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 can 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.

Sixth Cranial Nerve Palsy

The affected eye is converged because of a weakness of the lateral rectus muscle. It occurs most commonly as an isolated microvascular episode in hypertensive elderly patients and heals spontaneously in three to six months. Elderly diabetics are also more prone to sixth cranial nerve

palsies. In young patients, the possibility of multiple sclerosis or even raised intracranial pressure must be borne in mind.

Fourth Cranial Nerve Palsy

The eye fails to look down when it is turned in and might be turned slightly up when the other eye is looking straight ahead. Trauma (a blow over the head) is an important cause in younger patients but a full investigation for an intracranial space-occupying lesion is usually needed.

Third 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 a posterior communicating aneurysm should also be considered, particularly if it is associated with pain. Other causes include demyelination, diabetes, microvascular occlusion and herpes zoster infection. Recovery of nerve function particularly after compressive lesions can lead to a phenomenon known as aberrant regeneration. This can manifest as atypical pupil or lid responses on attempted eye movement.

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. When the inflammation has settled, the infiltrating cells are replaced by fibrous tissue, further restricting muscle action.

Myasthenia Gravis

This disease presents sometimes with diplopia with or preceded by ptosis, which becomes worse as the day goes by. Any one extraocular muscle or group of muscles can be affected. The symptoms and signs show a transient improvement seconds after the intravenous injection of edrophonium chloride (Tensilon). Diagnosis can be confirmed by high serum titres of acetylcholine receptor antibodies. Approximately 10% of cases are associated with a thymoma, which can become malignant. A chest X-ray is, therefore, mandatory in any patient suspected of having myasthenia gravis. Treatment is with an anticholinesterase such as pyridostigmine.

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. There might be a relative enophthalmos. Surgical intervention to repair the orbital wall defect might be required if the patient suffers from prolonged diplopia or marked enophthalmus.

23

Genetics and the Eye

Many eye diseases are inherited or have familial clustering. It is, therefore, always advisable to enquire about the family history when interviewing a patient with an ophthalmological complaint. Some types of inherited eye disease lead to blindness and relatives of patients with such conditions often seek advice concerning their risk of developing the disease. Patients might also consult with a view to prenatal testing, particularly if the disease leads to blindness at a young age.

Recent advances in molecular biology have led to a dramatic increase in our understanding of eye diseases. The discovery and the unravelling of the role of numerous ocular disease genes has also helped in our understanding of normal eye development and functioning. Because of the advances made in ophthalmic molecular genetics, we are now able to refer to an inherited ocular condition not only by the mode of inheritance, but also to denote the abnormal chromosome, the abnormal gene's position on the chromosome and its nucleotide sequence. To date, over 150 different gene defects have been described for retinal conditions alone (www.sph.uth.tmc.edu/retnet/home.htm). For many disorders, we also now know the role the abnormal gene plays in the pathogenesis of the disease, either because it leads to the production of an abnormally functioning protein or because the gene defect leads to the abnormal regulation of nearby or distant genes. Once the abnormal gene product (protein) associated with a disease can be identified, then drugs can be designed specifically, either to suppress its production or to replace the lost function.

Examples of eye disease that have been mapped out to different chromosomes are shown in Table 23.1.

Several methods are used in molecular biology to link disease to particular gene loci. Work usually starts by finding and classifying the disease in question in a large family or series of families. Next, the disease chromosome is sought (unless the inheritance pattern is X-linked, then this step can be omitted) and then the position of the gene in question is gradually narrowed down (by the use of linkage analysis followed by chromosome walking). This usually produces a region of the chromosome on which a number of candidate genes are found. Sequencing of these genes and comparison with normal individuals or animal models usually allows the disease gene to be identified (this can be a very time-consuming operation). Once the sequence of the gene is known, this can be compared on computer databases with similar known genes and the putative structure and function of the disease gene and its product can be determined. The potential of the latter has been greatly improved by the project to sequence the entire human genome.

Eye screening in selected patients at risk of inherited disease might detect important life-threatening conditions, for example familial adenomatous polyposis, retinoblastoma, Marfan's syndrome, neurofibromatosis and von Hippel Lindau disease.

Table 23.1. Chromosome mapping for common eye diseases 🕮

Chromosome	Eye disease
1	Leber's congenital amaurosis, Stargardt's disease, open-angle glaucoma (type 1A), congenital cataract, retinitis pigmentosa
2	Congenital cataract, iris coloboma aniridia type 1, AR retinitis pigmentosa, congenital glaucoma
3	Usher's syndrome, AD retinitis pigmentosa
5	Treacher Collins mandibulofacial dysostosis
7p	Goldenhar's syndrome
11	Aniridia type 2 (sporadic aniridia/Wilms tumour), Best's disease
12	Stickler's syndrome, congenital cataract
13q	Retinoblastoma
17	Neurofibromatosis type 1 (NF1; Von Recklinghausen's disease)
22q12	Neurofibromatosis type 2 (NF2)
X Chromosome	Ocular albinism Juvenile retinoschisis Norrie's disease Choroideremia Retinitis pigmentosa
(Xq 28)	Colour blindness – blue cone, red cone, green cone

Basic 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 basic knowledge of the mechanism of genetic transmission.

The nucleus of each cell in the body contains 46 chromosomes arranged as 23 pairs. The twenty-third pair comprises the sex chromosomes (the remainder being known as autosomes). These sex chromosomes 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 can be similar (homozygous) or different (heterozygous). If different, one can exert an overriding influence and is said to be dominant. The gene that is overridden is said to be recessive.

Genetic disorders can be divided into three broad groups:

- abnormalities of chromosomes – numerical or structural
- abnormalities of individual genes, which are transmitted to offspring
- abnormalities involving the interplay of multiple genes and the environment.

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 can 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. There are also some other terms that are important when describing genetic abnormalities: penetrance refers to the proportion of individuals who carry the gene and who express the disease, while expressivity refers to the clinical spectrum of severity of a particular genetic condition. The four important patterns of inheritance are:

- autosomal recessive
- autosomal dominant
- sex-linked recessive
- mitochondrial inheritance.

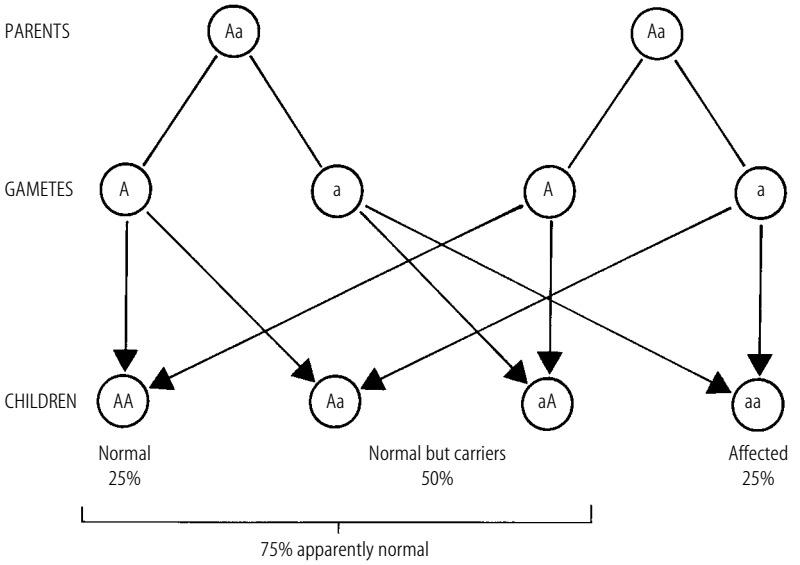


Figure 23.1. Recessive inheritance.

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", 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 (Figure 23.1). If a patient has recessively inherited disease, his or her parents are likely to be normal but there might 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, all the offspring will be affected. If one spouse is a carrier and the other has the disease, there is a risk that 50% of the offspring would be carriers and 50% would be affected. When a carrier marries a normal individual, 50% of the offspring are carriers. These expected findings could be calculated quite easily using the type of diagram shown in Figure 23.1. Common diseases inherited in this manner include sickle-cell disease and cystic fibrosis.

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 can, therefore, have 50% of affected children when married to a normal spouse. Of course, if both spouses carry the abnormal dominant gene, all the offspring will be affected. Dominant inheritance can only be shown with certainty if three successive generations show the disease and if about 5% of individuals are affected. Also, one sex should not be affected more than the other (Figure 23.2). Examples of this type of inheritance are hereditary retinoblastoma and Marfan's disease.

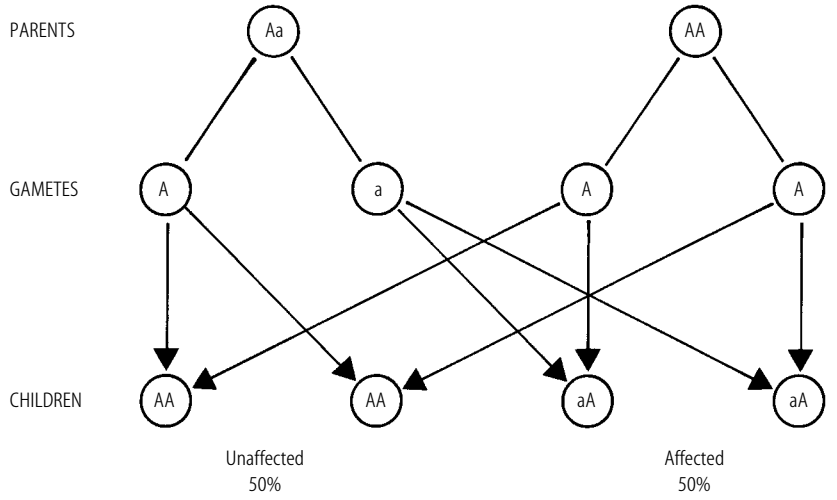


Figure 23.2. Dominant inheritance.

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 can 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 can also show this pattern in some families. When inheritance is 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 (Figure 23.3).

This description of the three important modes of inheritance should make it apparent

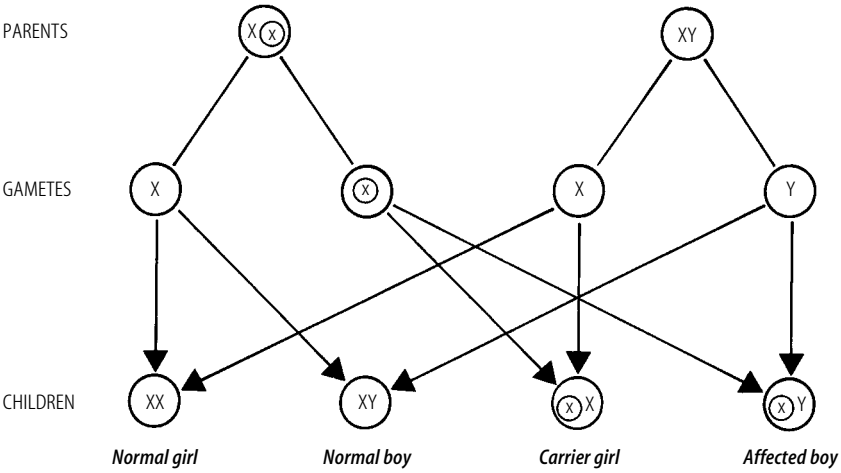


Figure 23.3. X-linked inheritance.

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, can be inherited in different ways in different families. In most large centres, there are now genetic clinics in which time is devoted specifically to the investigation of families and also to the detection of carriers.

Mitochondrial Inheritance

Mitochondria are the only organelles of the cell besides the nucleus that contain their own DNA. They also have their own machinery for synthesising RNA and proteins. Instead of individual chromosomes, mitochondria contain circular DNA similar to bacteria (from which they are thought to be derived). Mitochondrial DNA contains 37 genes, predominately encoding the enzymes necessary for the respiratory chain. All mitochondria in the zygote are derived from the ovum; therefore, a mother carrying a mitochondrial DNA mutation will pass it on to all of her children (maternal inheritance) but only her daughters will pass it on to their children. Mitochondrial DNA mutations are usually manifest clinically in tissues with a high metabolic demand, for example brain, nerve, retina, muscle and renal tubule. Examples of ophthalmic diseases caused by mitochondrial DNA mutations include Leber's hereditary optic neuropathy, chronic progressive external ophthalmoplegia, maternally inherited diabetes and deafness, and Kearns–Sayre syndrome.

Chromosomal Abnormalities

Microscopic studies of the chromosomes themselves have revealed that abnormal numbers of chromosomes can be produced by a fault at the moment of fertilisation. These might be caused by changes in numbers or structure of chromosomes. Numerical chromosomal changes include the absence of a chromosome (monosomy), as in Turner's syndrome, or an additional chromosome (trisomy), as in Down's syndrome. Cytogenetic studies have shown that patients with Down's syndrome have an additional 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 disease in which there are known to be abnormalities of the chromosome, 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.

Structural abnormalities occur when recombination or reconstitution in an altered form follows chromosomal breaks. Such changes can be in the form of deletions, duplication inversions, translocations or isochromosomes.

Multifactorial Diseases

These are disorders that arise from an interplay of genetic and environmental influences. The genetic contribution is made up of at least two abnormal genes acting in concert to express a "dosage-related" type effect, which is significantly influenced by several environmental factors. This leads to variable phenotypic expression. Examples include diabetes mellitus, some malignancies and perhaps age-related macular degeneration.

24

Drugs and the Eye

It is possible to achieve a high concentration of many drugs in the eye by applying them as eye drops. In this way, a high local concentration can be reached with minimal risk of systemic side effects. However, the systemic side effects of drops cannot be discounted, particularly in susceptible individuals. For example, timolol drops can precipitate asthma and slow the pulse rate in elderly patients, and pilocarpine drops can cause sweating and nausea. The action of local medications can be prolonged by incorporating them in an ointment, but for most purposes drops are supplied in 5 ml or 10 ml containers. After the container has been opened, it should not be kept for longer than one month. In order to avoid undue stinging, drops can be buffered to near the pH of tears and they contain a preservative, such as benzalconium chloride. It must be borne in mind that patients who develop an allergic reaction to drops might be reacting to the preservative. Single-application containers are also used, which do not contain a preservative but are expensive.

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 to flush out foreign bodies or irritant chemicals. Fresh mains tap water is an adequate substitute.

One of the major drawbacks of using eye drops is that, although high local concentrations of the drug are achieved in the anterior segment of the eye, little if any drug penetrates to the posterior segment. Drops are, therefore, of little use in treating diseases of the vitreous and retina. One way of delivering a drug to the posterior segment is to give it systemically. An example of this is the use of systemic prednisolone for posterior uveitis. This treatment method has the drawback of systemic side effects. This can be reduced by delivering the drug to the posterior segment by local injection either directly into the vitreous, along the orbital floor, within the sub-Tenon's space or in the subconjunctival space.

Treatment of Infection

Chloramphenicol is rarely used as a systemic drug nowadays, but it has been useful for many years in the form of eye drops. It remains a drug of choice in the UK for superficial eye infections. Other broad-spectrum antibiotics in use include gentamycin, framycetin, tobramycin and neomycin, as well as ciprofloxacin and ofloxacin. 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 intravitreal administration might be needed if the infection is intraocular. A number of antiviral drugs are now available, but acyclovir in the form of Zovirax ointment is the most widely used treatment of herpes simplex keratitis. The use of systemic acyclovir and famcyclovir for herpes zoster ophthalmicus has made a great impact on the severity of ocular complications.

Drops That Widen the Pupil

The pupils can be dilated either by local blockade of the parasympathetic pathway or by local stimulation of the sympathetic pathway.

Parasympathetic Antagonists

Routine mydriasis to allow examination of the fundus is best achieved by tropicamide 0.5% or 1% drops because the effect lasts for only about 3 h. Cyclopentolate 1% (0.5% in babies) can last for 24 h, but because of its cycloplegic effect (blockade of accommodation) is preferable for the examination of children's eyes when refraction is also needed. Dilating the pupil runs the risk of inducing an attack of acute narrow-angle glaucoma in a predisposed individual. Because the vision could remain blurred, driving should be avoided within the first 6–8 h after mydriasis. Atropine in drop form is a long-acting mydriatic, which is used when it is necessary to prevent or break down adhesions between iris and lens in acute iritis (posterior synechiae). It is also used in the treatment of amblyopia in children. Its effect lasts for about seven days. Allergic reactions are quite common and occasionally systemic absorption can cause central nervous system symptoms of atropine toxicity.

Sympathetic Agonist

Phenylephrine (5 or 10% drops) is a sympathomimetic amine. It is used with a parasympathetic antagonist when extremely wide pupil dilation is required (e.g., for intraocular surgery or for peripheral retinal examination). There are reports of severe acute hypertension after use of 10% drops.

Drops That Constrict the Pupil

In the past, meiotics have been widely used for the treatment of chronic open-angle glaucoma. Pilocarpine is available in 1%, 2%, 3% or 4% solutions. Although it is effective in reducing the intra-ocular pressure, the side effects of dimming of vision and accommodation spasm can be disabling and mean that this treatment has largely been superceded. Pilocarpine is still used in the treatment of acute glaucoma attacks to constrict the pupil and open up the closed drainage angle. Sometimes it is necessary to constrict the pupil rapidly during the course of intraocular surgery and this is achieved by instilling acetylcholine directly into the anterior chamber. Strong meiotics run the risk of causing retinal detachment in susceptible individuals. Meiotics have been used to reverse the effect of mydriatic drops used for fundus examination, but this practice is no longer recommended as a routine because it is unnecessary and the symptoms of meiosis may make matters worse.

Drugs in the Treatment of Open-angle Glaucoma

There has been a small revolution involving the type of eye drops used for the treatment of glaucoma in recent times. For years, the mainstay of treatment was pilocarpine and the topical beta-blockers, for example timolol, but the potential systemic side effects of these drugs have led to the introduction of other novel types of ocular hypotensive agents. In general, these new agents can be divided into alpha$_2$-adrenergic agonists, carbonic anhydrase inhibitors and prostaglandin analogues.

The production of aqueous humour can be reduced by either blockade of the beta-receptors on the ciliary body epithelial cells (i.e., with a beta-blocker) or by agonism of the alpha$_2$-receptors. Brimonidine and apraclonidine are both alpha$_2$-receptor agonists and show good efficacy compared with timolol. A significant number of patients, however, do develop an allergy to these agents and this has limited their widespread use. Acetazolamide was introduced as a diuretic many years ago; although not a very good diuretic, it has proved to be a potent ocular hypotensive when given orally. Again because of side effects its use has been restricted to short-term treatment. In 1995, dorzolamide was introduced and more recently, brinzolamide has become available. These are also carbonic anhydrase inhibitors but they are available in drop form and are able to penetrate the cornea. Their ocular hypotensive effects are generally not as great as topical beta-blockers but they are useful as adjuvant agents. It has recently been discovered that a second aqueous outflow route

exists in the eye – the uveoscleral route. It is known that certain prostaglandins increase the flow of aqueous via this route and a number of topical prostaglandin $F_{2\alpha}$ analogues are now available. Latanoprost, travoprost and bimatoprost have all been shown to as effective as topical beta-blockers with minimal side effects.

All these medications have the problem of compliance. Elderly patients may forget to instill drops on a regular basis. In some cases, even instillation of three different glaucoma drops fails to control the intraocular pressure. In these instances, the only sure way of lowering the pressure is by glaucoma drainage surgery.

Drugs in the Treatment of Acute Angle-closure Glaucoma

Angle-closure glaucoma is a surgical problem. Once the acute attack has been aborted by the use of intensive pilocarpine drops and Diamox, a small hole is made in the iris with the Yttrium–Aluminium–Garnet (YAG) laser to allow redirection of the flow of aqueous. In many patients this provides a permanent cure. Beta-blockers may also be used during the acute stage and more recently the alpha$_2$-agonist apraclonidine has been shown to be a useful adjunct.

Drugs in the Treatment of Allergic Eye Disease

With the increasing incidence of atopy, the treatment of allergic eye disease has gained in importance in recent years. Treatments are designed to interfere with either the type 1 (immunoglobulin E [IgE]-mediated) or type 4 (delayed) hypersensitivity response, both of which are thought to be important in disease pathogenesis. For mild disease, initial treatment should involve antigen avoidance (if known) and frequent use of artificial tears (hypromellose) to wash away antigens from the ocular and conjunctival surface. Treatment of more severe disease involves the use of systemic or topical antihistamines (levocabastine, emedastine and azelastine), which are helpful for relief of symptoms, and topical mast cell stabilisers (sodium cromoglicate, nedocromil sodium and lodoxamide), which are useful in disease prevention if used regularly. The treatment of severe (sight-threatening) disease involves the use of courses of topical and occasionally oral steroids.

Local Anaesthesia in Ophthalmology

Proxymetacaine (Ophthaine) is a useful short-acting anaesthetic drop that is comfortable to instill and so is particularly useful when examining children. Amethocaine and benoxinate are also widely used but are longer-acting and sting quite markedly. Local anaesthetic drops should not be used as pain relievers on a long-term basis because the anaesthetized cornea becomes ulcerated and severe infection of the eye can occur. Lignocaine (1% or 2%) with or without adrenaline is injected into the eyelids for lid surgery. Local anaesthesia for intraocular surgery is obtained by topical drops, sub-Tenon's injection, periorbital injection (outside the cone of extraocular muscles) or sometimes retrobulbar injection (within the muscle cone) of lignocaine. For a longer effect, this is sometimes combined with marcaine.

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 can take up and store the preservative from some kinds of drop. The preservative benzalkonium chloride is especially liable to be absorbed onto a contact lens. When it is essential that drops are administered to a patient wearing contact lenses, it is often possible to prescribe in the form of single-dose containers that do not contain a preservative.

Artificial Tears

Artificial tears provide one of a number of measures that are used to treat tear deficiency. Other measures include occlusion of the lacrimal

puncta or the use of mucolytic agents. The first step is to make the diagnosis. Once a deficiency of tears has been confirmed, the mainstay of treatment is hypromellose. Adsorptive polymers of acrylic acid can also give symptomatic relief. Polyvinyl alcohol is another compound present in a number of tear substitutes. Recently, a new agent, sodium hyaluronate (0.1%) has been shown to improve symptom relief and improve the ocular surface abnormalities in cases of severe dry eye. By their nature, tear substitutes tend to adhere to the surface of the eye and in the conjunctival sac. For this reason, their prolonged use is liable to give rise to preservative reactions. Preservative-free preparations are often preferable. Some patients with a severe dry eye problem might need to instill the drops every hour or even more frequently.

Anti-inflammatory Drugs 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. It must be remembered that systemic steroids give the patient a sense of well-being, which might give a false impression of the real benefit obtained. Furthermore, systemic steroids can have serious and life-threatening side effects, such as vertebral collapse through osteoporosis and perforated gastric ulcer (Figure 24.1).

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 two-fold: first, local steroids enhance the multiplication of viruses, especially herpes simplex; and second, they can cause glaucoma in certain predisposed individuals. In such individuals, the instillation of one drop of steroid can cause a temporary rise of intraocular pressure. The most potent steroid in this respect is dexamethasone, followed by betamethasone, prednisolone and hydrocortisone. It has been claimed that rimexolone, clobetasone and fluorometholone are relatively safe in this respect.

Recently, a number of nonsteroidal anti-inflammatory drugs (NSAIDs) have been made available in topical form (diclofenac [Voltarol Ophtha], ketorolac [Acular] and flurbiprofen [Ocufen]) to reduce our dependence on topical steroids. They have been shown to be of use in the treatment of postcataract surgery inflammation and in reducing the pain after excimer laser surgery and corneal abrasions.

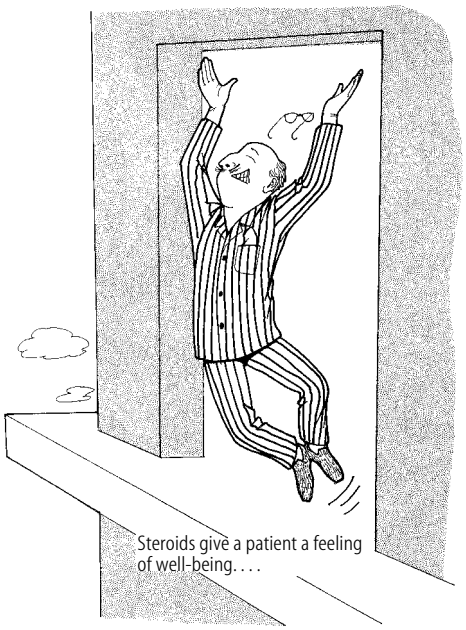


Figure 24.1. There might be a false impression of the real benefit obtained. 🕮

Anti-angiogenic Drugs and the Eye

Uncontrolled angiogenesis (growth of new blood vessels) is a common finding in many potentially blinding conditions, such as proliferative diabetic retinopathy, central retinal vein occlusion, wet age-related macular degeneration (ARMD) and retinopathy of prematurity. Inhibiting their growth offers us the hope of dramatically reducing the number of patients going blind each year. It is thought that the angiogenic response is caused by elevated levels of a cytokine called vascular endothelial growth factor (VEGF) produced by abnormal or ischaemic cells within the eye. Attempts to reduce the levels of VEGF and hence turn off the angiogenic drive have involved intravitreal

injections of anti-VEGF antibodies or oligonucleotide aptamers, which bind VEGF, or the intravitreal/sub-Tenon's injection of an anti-angiogenic steroid (triamcinolone). All of these treatments are showing great promise in clinical trials. An alternative mechanism of treatment is the destruction of preformed new vessels. Recently, a new type of treatment for wet ARMD has seen the use of a light-activated dye, injected intravenously, which preferentially locates in the choroidal neovascular membrane (photodynamic therapy). Activation of the dye by light of a specific wavelength causes thrombosis and destruction of blood vessels harbouring the dye. Treatment of patients with one particular subtype of wet ARMD (classic with no occult blood vessels) has shown stabilisation of vision in 60–70% of cases.

Damage to the Eyes by Drugs Administered Systemically

There are a number of drugs, which if given in excessive doses, can lead to severe visual handicap and blindness. Some of these are still available on prescription. Chloroquine and hydroxychloroquine in excessive doses can lead to pigmentary degeneration of the retina and blindness. Certain antipsychotic drugs can also cause fundus pigmentation in excessive doses; melleril and chlorpromazine have been incriminated in this respect in the past. Recently, a number of cases of uveitis have been reported in patients using bisphosphonates for the treatment and prevention of osteoporosis. Interestingly, sudden visual loss has been reported in a number of patients taking the oral anti-inflammatory COX-2 inhibitors (celecoxib and rofecoxib). The vision has returned to normal upon cessation of treatment.

Apart from causing glaucoma in some patients, systemic steroids are thought to increase the rate of formation of cataracts. Ethambutol and isoniazid can cause optic atrophy. Sometimes excessive doses of quinine are taken as an abortifacient and as the patients regain consciousness they are found to be blind from quinine toxicity. Methyl alcohol is toxic to the ganglion cells of the retina and blindness is a hazard of meths drinkers. It sometimes contaminates crudely prepared alcoholic beverages leading to unexpected loss of vision. The list of drugs with ocular side effects is large and the reader should consult a specialised textbook for more information. Nowadays, disasters and indeed lawsuits should be avoidable if the drug literature is checked before prescribing an unfamiliar drug.

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Blindness

Blindness marks the failure or inefficacy of ophthalmological treatment. Once a patient becomes permanently blind, he or she might be lost from the care of the ophthalmologist. This means that the ophthalmologist might not have personal experience of the size of the problem and might 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.

In the UK, the major problem is among the elderly where visual loss is often combined with defective hearing. Sensory deprivation is thus a major scourge at the present time; the problem is undoubtedly going to be much worse as the proportion of elderly people increases.

Definition

In the UK, 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". When a patient's vision falls below this level, registration as a blind person can be considered. This is a voluntary process, which allows the patient access to the social services for the visually handicapped, as well as certain tax concessions. Registration is usually initiated in the hospital clinic. Some patients are referred by their general practitioners or social workers for registration by the ophthalmic specialist. A special form is completed and copies go to the patient, the general practitioner, the social services department and the Office of Population and Censuses.

Certain guidelines are laid down when considering blind registration; the visual acuity should be less than 3/60 in the worse eye but if the field of vision is constricted, the visual acuity might be better than this. Patients whose vision is not bad enough for blind registration but none the less have significant visual handicap can have their name placed on the partially sighted register. In these patients the binocular vision should normally be worse than 6/18. Patients sometimes erroneously claim the benefits of the partially sighted because they have only one eye, even though the remaining 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, 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 can 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.

Benefits for the Visually Handicapped

There is no blind pension in the UK but those registered blind have a special income tax allowance and some exemptions from deductions from income support. Blind persons can have parking concessions and a free National Health Service (NHS) sight test, as well as rail cards and bus passes. Disability living allowance can be available for blind people under the age of 65 years but for the over-65s, only those who are both blind and deaf can qualify. Those seeking these concessions should consult an expert in the field. There are a number of voluntary organisations that run clubs, social centres and supply various other aids and benefits. For example, the Royal National Institute for the Blind (RNIB) provides a comprehensive range of services including the popular talking-book service. It also supplies regular funds for research into the causes of blindness.

The system of registration applies equally to children. In this instance, registration calls attention to the need for special educational requirements. These can include a specialist resource teacher, low visual aids, and other special supplies and equipment. If necessary, special schooling might need to be considered.

Standards of Vision for Various Occupations

The standards for various occupations can vary from year to year and are more or less exacting, depending on the occupation. In the UK, in order to drive a private motor vehicle, one must be able, in good daylight, to read a number plate with glasses or contact lenses at 67 feet or 20.5 m. A full binocular field of vision is also now required. This must extend at least 120° horizontally and 20° above and below. The field is measured by perimetry using a standard target. It is assumed that any healthy person applying to drive has a normal field of vision but if the driver has any eye condition that might lead to visual handicap, he or she must declare it. The driver and vehicle licensing centre might then ask for a report from an ophthalmologist or an optometrist. Double vision is a bar to driving, if it cannot be corrected by prisms in the glasses or the wearing of an eye patch.

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. Of the male population, 8% suffers from some form of congenital colour blindness. This is usually in the form of "red–green blindness". Inheritance of this type of defect is sex-linked so that unaffected female carriers pass the gene to 50% of their sons. The screening of school children for colour blindness is now widely practiced because of the occupational implications. The Ishihara test is the simplest and the best test for congenital colour blindness. Occupations that 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 of Blindness

In England and Wales, the prevalence of blindness in 1980 for children under five years was 9:100,000. This figure increased to 2324:100,000 for adults over 75 years. In the western world, blindness in children is largely because of inherited genetic disease and birth trauma. In adults aged 20–60 years, the major causes are diseases of the retina, including diabetic retinopathy and optic atrophy. Over the age of 60 years, macular degeneration, glaucoma and cataract are the important problems.

In Africa and Asia, the causes of blindness are rather different; many children become blind from corneal scarring associated with vitamin A deficiency and measles. Cataract is the most important cause in adults but in certain areas, for example southern Sudan, onchocerciasis and trachoma are still a serious problem.

It is apparent that the problems of blindness in Europe and North America are different from those in poorer parts of the world where much could still be done by improving standards of 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 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.

Many different electronic devices have been tried but by and large these are only useful to younger patients who can make full use of them. Scanning systems are now available which, when moved across the page, can read out the page. Most blind patients are unable to afford this type of aid. Many of these devices rely on the patient's hearing to identify an audible warning signal, but most blind people prefer to use their undistracted sense of hearing as an important clue to their whereabouts.

Guide dogs are specially trained by the Guide Dogs for the Blind Society 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 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 and newspapers are now very popular among blind and partially sighted people of all ages. The Talking-Book Service provides a comprehensive library for the use of the visually disabled.

There are numerous other gadgets that 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 pad with large numbers on it can be helpful. Other ingenious devices range from relief maps that can be felt by the blind person, to a telephone that speaks back through the earpiece the digit that has just been pressed. Research has also been carried out on aids that signal the position of objects by means of electrical stimuli to the skin and even by means of implanted electrodes in the visual cortex.

One important advance has been voice synthesis by computers. Many current models have this facility, so that the user can hear emails, and programmes are available to allow printing in Braille. In spite of these advances, the elderly visually handicapped patient can benefit most from someone who is prepared to give the time to read out letters or books. Some voluntary local societies can provide this service.

When the patient has a visual acuity of better than 6/60, much can be achieved by the use of optical magnification. An ordinary hand magnifying glass is the simplest and can often be the most effective form of assistance. If this is not adequate and the patient has been a keen reader, a telescopic lens can be fitted to a spectacle frame with advantage. These multi-lens systems are known as low visual aids and hence the popular "LVA" clinics in eye departments for the testing and provision of these items. 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 presented on a television screen.

The well-being of a blind or partially sighted person can be greatly enhanced by relatively simple social measures. Advice in the home about the use of gas or electricity can 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 can 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 directed onto the page. The distance of the bulb from the page is as important as the wattage of the bulb.

Artificial Eyes

These can be made of glass or plastic molded 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 can indicate that the socket is becoming infected. This, in turn, might be because of roughening of the artificial eye with wear. Under these circumstances, arrangements should be made for the prosthesis to be replaced or polished. It should always be borne in mind that a patient with an artificial eye might have had the eye removed because it contained a malignant tumour, in which case one must consider the possibility of local or systemic spread of the tumour. A well-made artificial eye is almost undetectable to the untrained eye but normal movements of the eye can be restricted. Nowadays, the use of orbital prostheses deep to the conjunctiva and attached to the eye muscles gives greatly increased movement. After many years and after renewing the artificial eye on several occasions, the eye can appear to sink downwards.

Surgical removal of an eye (enucleation) is considered in the following circumstances:

- when the eye is blind and painful
- when the eye contains a malignant tumour
- when the eye is nearly blind and sympathetic ophthalmitis is a risk following a perforating injury.

Before having an eye removed, the patient must be made fully aware of all the advantages and disadvantages. A general anaesthetic is needed and the patient remains in hospital for one to two nights after the operation. It is common practice to fit the socket with a transparent plastic "shell" for a few weeks until the artificial eye is fitted.

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