The Eye (Globe), Eyelids and Associated Structures: Part I

27

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27.1 Applied Anatomy and Physiology

27.1.1 The Eye (Globe)

The globe lies in within the orbit. It is surrounded by fat, but separated from it by a membranous sac, the Tenon's capsule. This is attached to the corneoscleral junction anteriorly (at the limbus), and to the optic nerve posteriorly. The eyeball is not a simple sphere but can be visualised as the result of fusing a smaller dome (the cornea) onto a larger sphere (the scleral segment). The cornea is at the front of the eye and forms about one-sixth of the eye volume. In health it is transparent so that light can pass through. Overall, the globe has a diameter of about 2.5 cm. Normally, these dimensions are relatively constant, varying among normal individuals by only a few millimetres.

Embryologically the eye develops from four different sources, commencing around day 22 of development.

1. the neuroectoderm of the forebrain. The optic nerve, retina and the posterior layer of the iris all come from the neuroectoderm of the forebrain vesicle.
2. the ectoderm of the head. This becomes the lens and the epithelium covering the anterior aspect of the cornea.
3. the neural crest. These cells migrate and form the choroid, sclera and corneal epithelium)
4. the surrounding mesoderm. This is 'sandwiched' in between the neuroectoderm and the surface ectoderm. It gives rise to fibrovascular elements of the eye.

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M. Perry (ed.), *Diseases and Injuries to the Head, Face and Neck*,
https://doi.org/10.1007/978-3-030-53099-0_27

Optic 'grooves' initially appear in the developing forebrain. These grow forward and invaginate to form hollow "optic vesicles" or "cups". As these come into contact with the surface ectoderm, the ectoderm undergoes a process of invagination and becomes the lens placode. This ultimately becomes detached from the surface ectoderm to form the lens. Further invagination of the optic vesicle forms a double walled "optic cup". This consists of an outer pigmented layer and an inner neural layer. Anteriorly, the lens enters its opening. On the under surface of the optic cup and optic stalk a grove develops (the choroidal groove or fissure). This encloses the adjacent vascular mesenchyme and hyaloid vessels (the hyaloid artery is branch of the ophthlamic artery). With closure of this fissure (at around week 7), the axons of the ganglion cells from the retina and the vessels are contained within the optic stalk, now the optic nerve. The hyaloid vessels later become central artery and vein of retina. Thus, the optic nerve is an outgrowth of the brain (diencephalon) and not a peripheral-type cranial nerve. As such, like the spinal cord, (1) it is myelinated by oligodendrocytes, (2) it is not capable of regeneration following injury and (3) it is surrounded by the meninges and subarachnoid space (containing CSF) (Figs. 27.1, 27.2, and 27.3).

Several congenital anomalies can occur during development. These include (1) anophthalmos (complete failure of development), (2) synophthalmos (the eyes become partially fused), (3) microphthalmos (development of a tiny eye) and (4) cylopia (fusion of two eyes in the midline). In cylopia the nose is usually absent, but if a rudimentary appendage develops above the single eye, this is known as 'proboscis'. Microphthalmia is usually associated with intrauterine infections from toxoplasma, rubella virus, cytomegalovirus, and herpes simplex virus. Nanophthalmos, is a subtype of microphthalmia where the globe is small (20 mm in an adult) but has no gross structural abnormality. Persistence of the hyaloid vessels is seen in 3% of full term infants and 95% of premature infants. This may be recognised as "Mittendorf's dot", located at the posterior lens capsule or as "Bergmeister's

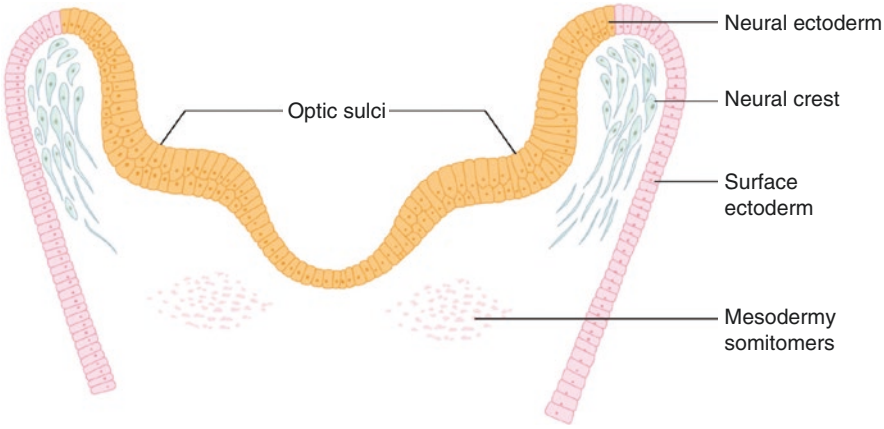


Fig. 27.1 Embryonic development: day 22

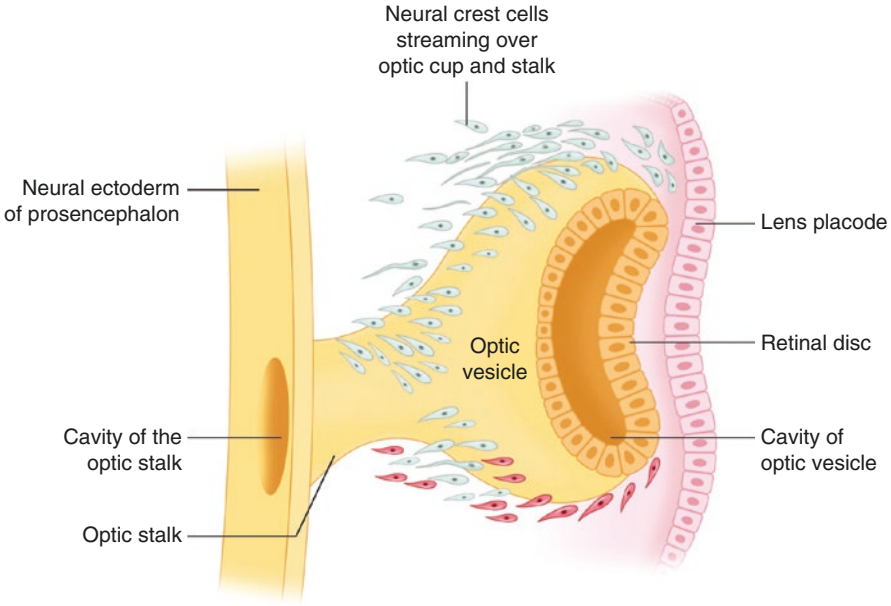


Fig. 27.2 Embryonic development: day 27

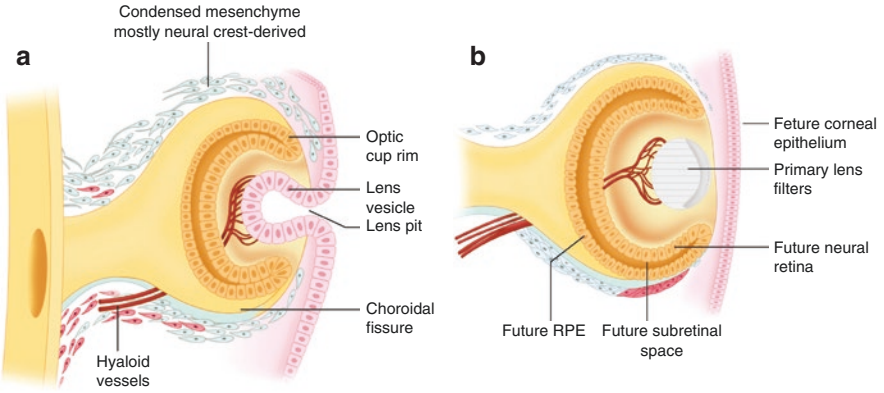


Fig. 27.3 Embryonic development: (**a**) day 29 (**b**) day 37

papilla", located at the optic disc. Eyes may also be abnormally small or large because of acquired diseases—these discussed later. Phthisis is the condition where the globe becomes small and shrunken often in association with thickened sclera, usually as an end-stage result of severe ocular disease such as endophthalmitis and trauma (Figs. 27.4 and 27.5).

Hypoplasia (septo-optic dysplasia) is one of the milder forms of developmental disorders that can involve brain and orbit and is associated with incomplete

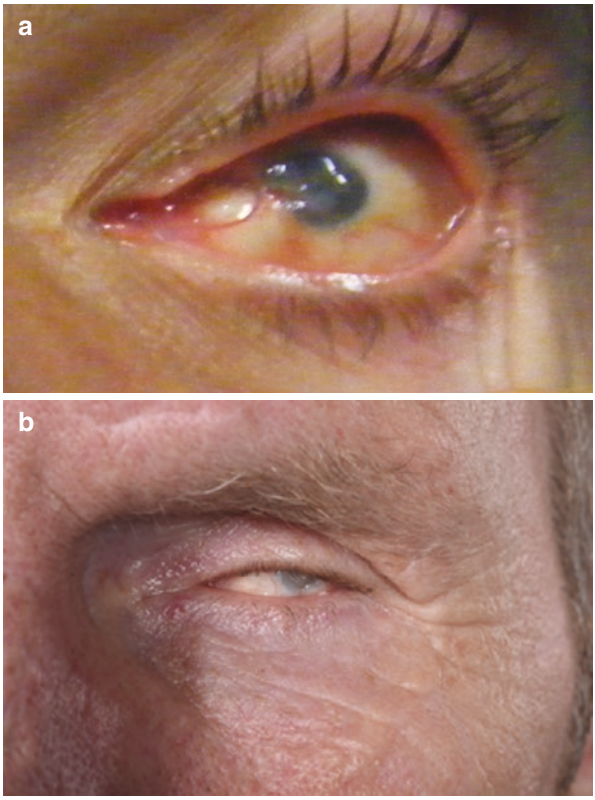


Fig. 27.4 (**a**, **b**) Pthiscal eyes following infection

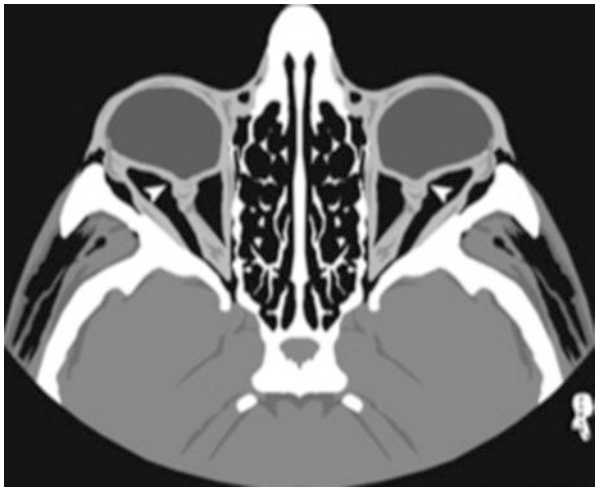


Fig. 27.5 Axial orbital CT illustration shows bilateral, abnormal, tilted insertion of the optic nerves into the orbit, a typical sign of "tilted disc syndrome" (arrowheads)

induction of the neural axis during the first trimester. It is also known as de Morsier syndrome. Aplasia of the optic nerve and eye are rare. This can occur sporadically and is usually due to failure of the optic vesicle to form. Absence of a globe then results in concomitant orbital hypoplasia and facial deformity. Leber's hereditary optic neuropathy can also eventually leads to bilateral optic nerve atrophy. With mild optic nerve hypoplasia the vision can be near normal. However with smaller and paler optic nerves, visual impairment may be severe. Children with bilateral disease may also develop nystagmus and there may also be pituitary dysfunction (Figs. 27.6, 27.7, and 27.8).

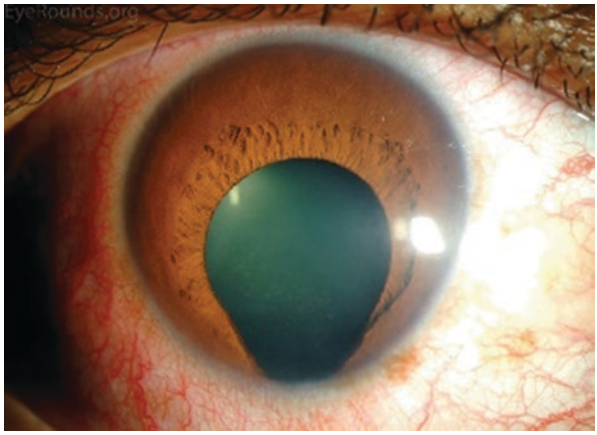


Fig. 27.6 Iris coloboma

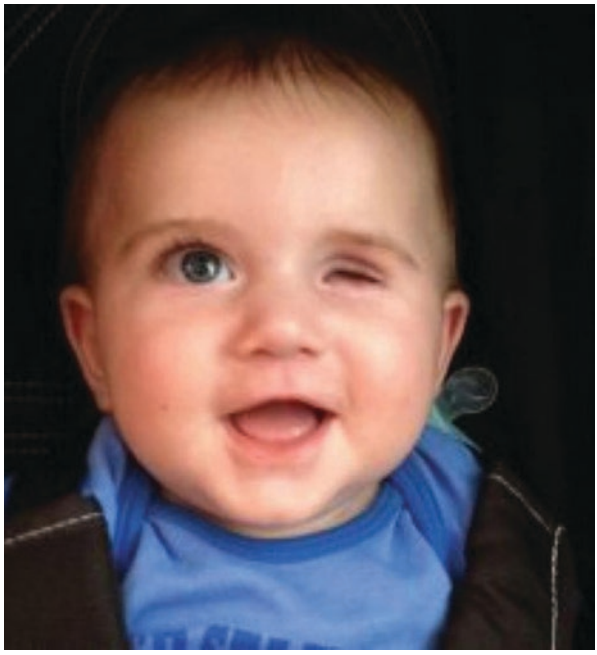


Fig. 27.7 Primary anophthalmia

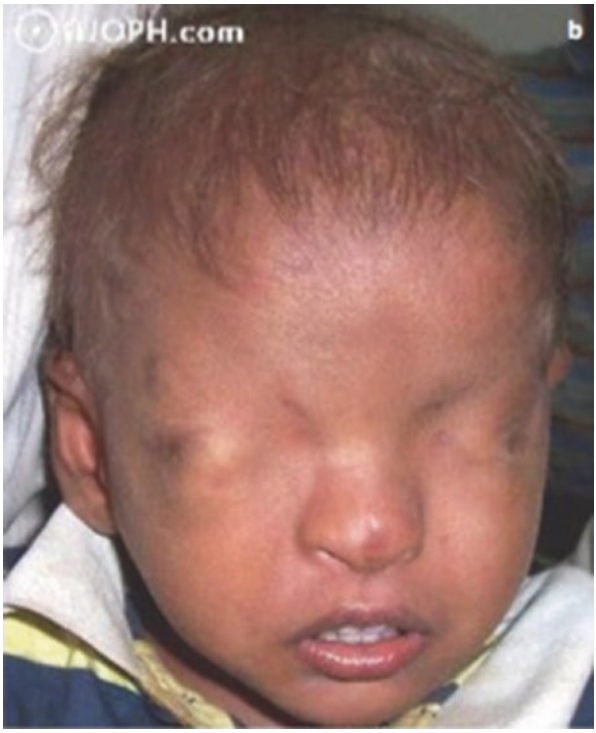


Fig. 27.8 Microphthalmia with cryptophthalmos

The wall of the adult eye is made up of several layers. These enclose the clear aqueous humour and lens (in the anterior chamber) and the vitreous body posteriorly.

1. Anteriorly, a conjunctival layer covers the sclera and Tenons layer, attaching at the limbus. This is reflected from the sclera onto the undersurface of the eyelids to form the tarsal conjunctiva.
2. Tenon's Capsule (fascia bulbi) is a fascial sheath that envelops the eye and separates it from the orbital fat. It separates the conjunctiva from the sclera and fuses with the sclera just behind the corneoscleral junction. It also passes backwards as a sheath around the recti muscles. Posteriorly, it is perforated by the optic nerve and its sheath and by the ciliary nerves and vessels.
3. A structural layer is composed of collagen-elastic tissue. This layer comprises the transparent cornea and the sclera. The junction between the cornea and sclera is called the limbus. The extra-ocular muscles attach to the sclera further back, while the optic nerve leaves the globe posteriorly. The cornea is about 0.5 mm thick centrally and thicker peripherally.
4. A middle or vascular pigmented layer (the uvea). This contains the main blood supply to the eye and consists of the pigmented choroid posteriorly and the cili-

ary body and iris anteriorly. The choroid nourishes the outer two-thirds of the retina and regulates the temperature of the globe.
5. An innermost layer—the retina. This lines the choroid. It is a laminated structure with photoreceptor cells on its deep surface.

The cornea, iris and lens of the eye form the anterior chamber. The crystalline lens lies within this chamber, behind the iris, suspended by a series of fine ligaments and fibrils (the zonules). These pass between the lens and the ciliary body and are under tension. The ciliary body contains the ciliary muscle. Its action alters the shape of the lens and thus the focus of the eye. It also provides the attachment for the iris. The ciliary epithelium secretes aqueous humor and is therefore is important in maintaining ocular pressure. Between the iris and lens and the ciliary body lies the posterior chamber (distinct from the vitreous body). Both the anterior and posterior chambers are filled with aqueous humor.

27.1.1.1 The Anterior Chamber (Iridocorneal) Angle

The anterior chamber of the eye develops as a cleft in the mesenchyme between the developing iris and cornea. The posterior chamber develops from a cleft in the mesenchyme between iris and the lens. With the disappearance of an initial papillary membrane these chambers freely communicate with each other. The anterior chamber angle is the anatomical angle created by the peripheral attachments of the iris and the cornea at the corneoscleral junction. This site is important in the flow of aqueous fluid. It is lined by the Trabecular meshwork. In the sclera outside this angle, Schlemm's canal allows aqueous humor to drain from the anterior chamber into the venous system. This region is often referred to as the drainage angle. The relationship between the iris and the cornea is very important as this can significantly affect the ability of the eye to drain the aqueous. The drainage angle in a healthy eye is approximately 30°. However this varies with sex, age, anterior chamber depth and iris anatomy. In eyes where the iris and corneal endothelium are "closed" against one another, the aqueous will not drain. This results in an increase in the intraocular pressure (angle closure glaucoma) (Fig. 27.9).

27.1.1.2 Aqueous Humor (AH)

Aqueous humor (AH) is a clear fluid contained within the anterior and posterior chambers of the eye. It is formed by the ciliary epithelium on the ciliary processes of the ciliary body, by selective transfer of solutes (ions, glucose, ascorbate, amino acids and others) and water from the blood. This is continuously secreted by the epithelium. It enters first into the posterior chamber and then flows forward through the pupil into the anterior chamber. Intra-ocular pressure is normally maintained by a balance between the formation of aqueous and its drainage via the trabecular meshwork at the drainage angle. A healthy eye makes about 2 µl of aqueous a minute i.e. around 80–90 l during a lifetime. About 95% of people have an IOP between 12 and 22 mm Hg.

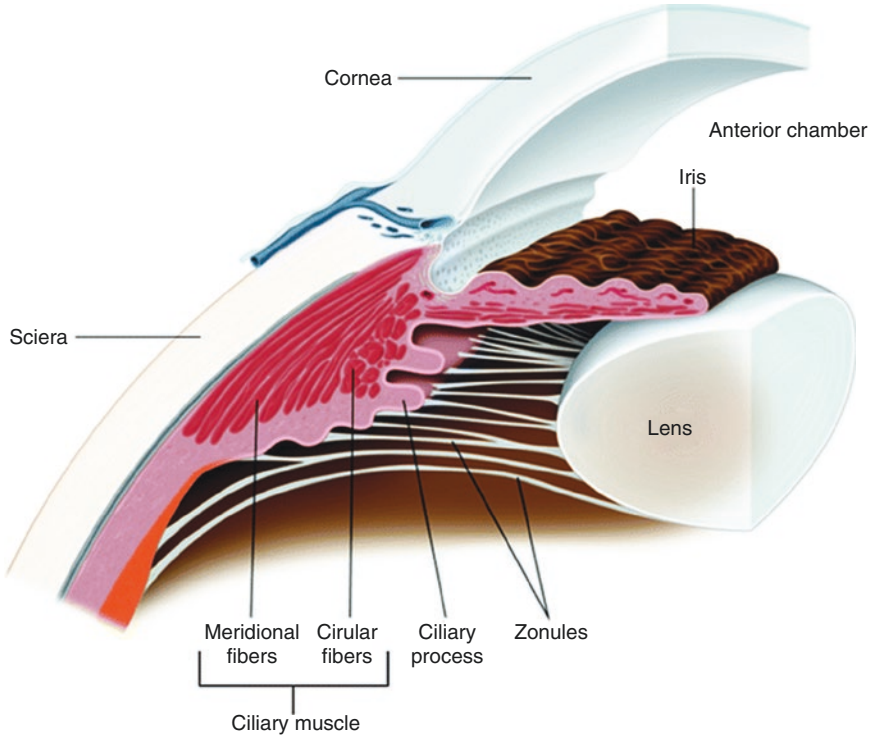


Fig. 27.9 Schematic representation of anterior chamber anatomy. *CB* Ciliary body, *S* Sclera, *SS* Scleral Spur, *SC* Schlamm canal*I*ris, *C* Comea, *TM* Tabecular Meshwork, *IP* Iris Process, *SL* Schwalbe's line, Z Zonules. Photo courtesy of AAO.org BCSC Sect. 2: fundamentals and principles of ophthalmology

27.1.1.3 Vitreous Body

Between the lens and the retina lies the vitreous body, occupying most of the posterior globe. This is a clear, transparent gelatinous mass. This initially develops from the neural crest mesoderm within the optic cup (the primary vitreous humour), but becomes replaced by a more gelatinous secondary vitreous, derived from the inner layer of the optic cup and the lens vesicle. The adult vitreous body is comprised of a vascularised mass of transparent gel-like intracellular substance. This hyaloid artery usually obliterates to form a hyaloid canal of adult eye. Persistence of the hyaloid artery can occur. This remains nonfunctional and can sometimes be seen as a 'worm-like' structure from the optic disk.

Unlike the aqueous, which is continuously replenished, the vitreous is stagnant. Therefore, any blood, cells or products of inflammation that leak into the vitreous, will remain there unless removed surgically. If these lay within the visual axis, they will be perceived as 'floaters'. Whilst these are generally harmless, any sudden onset of recurring floaters may indicate posterior vitreous detachment (PVD) or some other disorder of the eye. The vitreous abuts the retina and presses it against

the choroid, keeping it in place. If it is pulled away from the retina, this is called vitreous detachment. This can occur with increasing age and earlier in nearsighted eyes, or following injury or inflammation (uveitis).

27.1.1.4 The Conjunctiva

This is the fine translucent mucous membrane which covers and joins the anterior surface of the eyeball to the posterior surfaces of eyelids. It lines the visible white part of the globe, starting at the edge of the cornea (limbus) and passing out over the anterior surface of the sclera. It then inverts forwards, to line the inner surfaces of the eyelids. At the medial canthus the conjunctiva folds and thickens, to form the semilunar fold. Conjunctiva is subdivided into three parts depending on its location (palpebral conjunctiva, bulbar conjunctiva and conjunctival fornix). Histologically it is comprised of multiple layers (epithelial, adenoid and fibrous). These contain a variety of structures, notably glands, melanocytes, langerhans cells, mast cells and lymphoid tissue. Normal conjunctiva is thin and transparent. In the bulbar region, the underlying sclera with a fine network of episcleral and conjunctival vessels can often be seen. In the palpebral region and fornices, it looks pinkish because of underlying fibrovascular tissue. The conjunctiva may become discoloured as a result of various local and systemic diseases.

1. Red discolouration—bright red homogeneous discolouration suggests a subconjunctival haemorrhage.
2. Yellow discolouration—may occur from bile pigments in jaundice, blood pigments in malaria and yellow fever, conjunctival fat in the elder and some ethnic groups
3. Greyish discolouration—may occur from soot and mascara
4. Brownish grey discolouration—may be seen argyrosis (following prolonged application of silver nitrate).
5. Blue discolouration—is usually due to ink from pens or pseudopigmentation in patients with blue sclera and scleromalacia perforans.
6. Brown pigmentation may be seen in Addison's disease, melanocytic pigmentation, conjunctival epithelial melanosis, subepithelial melanosis and pigmented tumours (notably benign naevi and malignant melanoma) (Figs. 27.10, 27.11, 27.12, and 27.13).

27.1.2 The Orbit

This is described in detail in the chapter on the cheek and orbit. By way of summary, the orbit is a roughly pyramidal-shaped structure positioned at an angle of around 20° the midline. At its posterior apex is the optic canal, which transmits the optic nerve to the optic chiasm, tract and lateral geniculate body. The more lateral superior orbital fissure allows passage of the 'orbital' cranial nerves and vessels from the middle cranial fossa into the orbit, to innervate and supply the orbital structures. The lacrimal gland lies anteriorly, in the superolateral aspect of the orbit. Each orbit

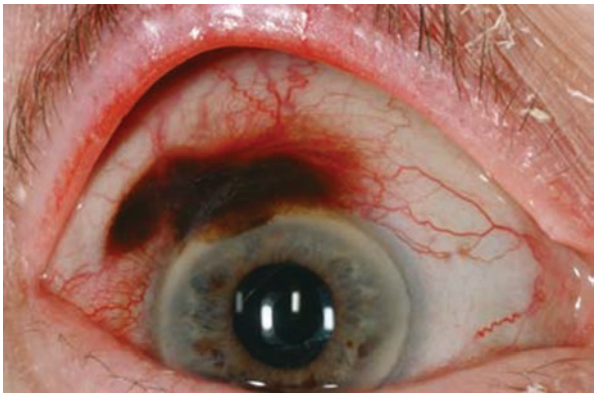


Fig. 27.10 Conjunctival malignant melanoma

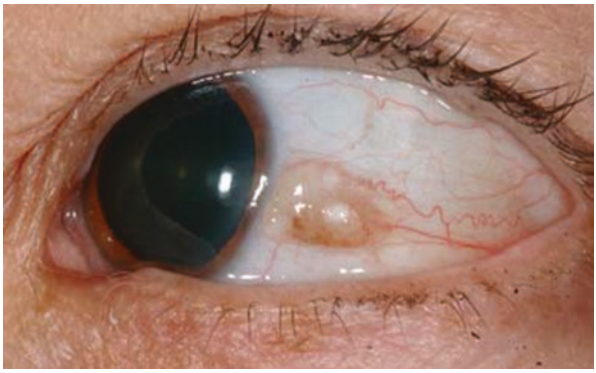


Fig. 27.11 Conjunctival nevus

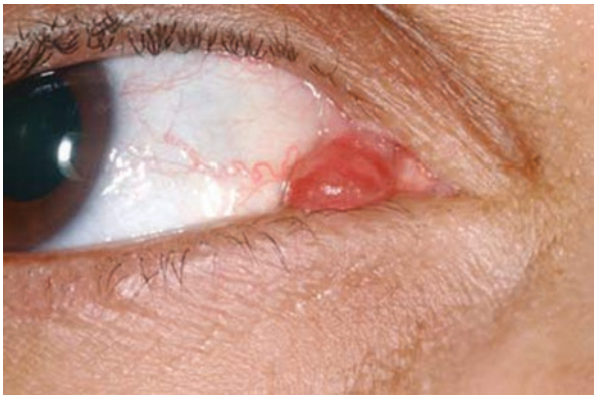


Fig. 27.12 Conjunctival papilloma

has a volume of approximately 30 ml. Together, they are aligned in such a way that their medial walls are almost parallel to each other, while their lateral walls lay at approximately 90° to each other. This is best seen on axial and coronal CT imaging.

The superior, medial and inferior walls of the orbit are extremely thin. They are composed of a number of different bones that vary considerably in thickness and

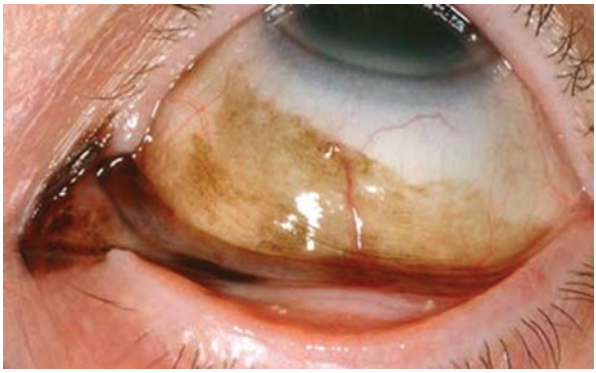


Fig. 27.13 Conjunctival primary acquired melanosis

strength. Fractures of the orbital roof should be regarded as skull base fractures. The inferior wall (floor) and medial wall are particularly delicate and prone to injury, either in isolation (blowout fractures), or in combination with injuries of the adjacent bones (zygomatic/nasoethmoid fractures). Together, they form a complex shape and familiarity with this geometry is essential in understanding orbital fractures. This is especially important at the back of the orbit, where the junction between the floor and medial wall becomes less obvious and the two merge to form a "posteromedial bulge". This is a very important area in the repair of orbital fractures. The infraorbital nerve passes forward along the orbital floor, supplying sensation to most of the cheek and ipsilateral half of the nose and upper lip.

27.1.2.1 Orbital (Retrobulbar) Contents

Attached to the eye are the four extraocular (recti) muscles and the two oblique muscles (superior and inferior obliques). These are responsible for the movements of the eyeball. The recti pass backwards and are attached to the back of the orbit by the annulus of Zinn. This encloses the optic foramen and the lower part of the superior orbital fissure (collectively referred to as the "orbital apex"). This 'conical' arrangement of the muscles effectively divides the orbital contents into "intraconal" and "extraconal" compartments. The compartments communicate with each other through the narrow gaps between the recti muscles. All the structures that enter the orbit through the optic canal and lower part of the fissure therefore lie at first within the cone of the recti. The intraconal space thus contains:

1. The optic nerve (CN II). Developmentally this should be considered as an outgrowth or extension of the brain. It is surrounded by the meninges, which are continuous with those of the brain and contains the subarachnoid space. Intracranially, the optic nerve is related to the internal carotid and ophthalmic arteries and to the hypophysis.
2. Cranial nerves—oculomotor nerve, nasociliary nerve (and its branches), abducens nerve
3. The ophthalmic artery and its branches

4. The superior ophthalmic vein
5. Fat. Any space within the orbit that is not occupied with muscle, nerve or blood vessels is filled with orbit fat. This cushions the eye, and supports and stabilises the extraocular muscles.

The extraconal space contains:

1. Extraocular muscles (Levator Palpebrae Superioris; Superior and Inferior oblique muscles). These develop from the mesoderm of somitomeres 1, 2, and 3 (preotic myotomes) that surround the optic cup.
2. Blood vessels
3. Lacrimal gland, lacrimal sac, nasolacrimal duct
4. Medial palpebral ligament and lateral palpebral ligament
5. Suspensory ligament of the eyeball
6. Trochlea of superior oblique
7. Extraocular Fat.

Throughout the two spaces, fine but tough fibrous septa divide the orbital fat into numerous tiny, interconnected fat-filled compartments. This delicate "bubble-wrap" type arrangement provides structural support for the globe and allows its free movement. In a sense, retrobulbar fat acts like a bursa, facilitating movement and providing support. Damage to the bony orbit can result in herniation of the fat (and occasionally the extra ocular muscle) with trapping of the septa. This can restrict eye movements, resulting in diplopia.

27.1.3 The Eyelids

The eyelids provide protection of the globe. Opening of the eyelids is achieved mostly by elevation of the upper lid. The lower lid remains essentially in the same position, although it will retract on when looking down. Embryologically, upper and the lower cutaneous folds containing mesoderm approach each other over the anterior surface of the developing cornea. These initially fuse, enclosing an ectodermal "conjunctival sac", but later separate to form the palpebral fissure. In many lower mammals (e.g. cats), the babies are born with closed eyelids. In humans, the eyelids remain fused until the 10th week but can sometimes remain adhered to each other until the 28th week. The mesoderm within each lid forms the tarsal plates and the connective tissue of the eyelids. The eyelashes and glands develop from the ectoderm in a similar manner to those in the skin. Thus, each eyelid has a 'sandwich' like arrangement of three layers: skin and muscle on the outside, a mucosal lining on the inside, and a 'skeletal' element in between.

1. The skin of the eyelid is extremely thin. In some places, upper lid skin can measure only 1 mm in thickness and less than 10 cell layers. This is the thinnest skin in the body. The underlying orbicularis muscle enables forced eyelid closure.

2. A tough collagenous layer (the tarsal plate) provides structural support to the eyelid. This houses the meibomian oil glands, which secrete the lipid component of the tear film. The levator palpebrae muscle is attached to the upper tarsal plate. This elevates the upper eyelid.
3. The inner epithelium (mucosa) lines the tarsal plates and blends with the palpebral conjunctiva. This forms an uninterrupted layer passing from the free edge of one eyelid, over the globe, to the free edge of the other lid. Where the conjunctiva reflects from lid onto globe, the recesses are referred to as the fornices (superior and inferior). This lining provide a smooth surface allowing free gliding movement between the lids and the globe.

The Meibomian (tarsal) glands, are sebaceous in nature. These secrete an oily substance onto the conjunctiva, which facilitates blinking. There are approximately 10–20 on the lower lid and 20–40 on the upper lid. These glands may be the site of several inflammatory processes (styes, granulomas, chalazia, or blepharitis). Medially, the lids contain the punctae, through which tears flow into the lacrimal drainage system. Arranged in multiple rows along the margins of the eyelids are the eyelashes. Next to these are the openings of the Ciliary glands of Moll and Sebaceous glands of Zeis. The eyelids carry out multiple functions:

1. Mechanical protection to the anterior globe
2. Spreading of the tear film over the conjunctiva and cornea with each blink, thereby preventing drying of the eyes

The levator muscle arises at the back of the orbit and passes forwards to attach to the tarsal plate in the upper lid. It is innervated by the third nerve. Damage to the nerve or the lid results in drooping of the eyelid—'ptosis'. A flat smooth muscle (Mullers muscle) arising from the deep surface of the levator also inserts into the tarsal plate. This is innervated by the sympathetic nervous system. If the sympathetic supply is damaged (as seen in Horner's syndrome) this can also result in ptosis. Anomalies of the eyelids include:

1. Coloboma of eyelid. This is a congenital condition in which part of the eyelid is missing, commonly seen as a small notch in the upper eyelid. Coloboma of the lower eyelid is rare.
2. Congenital ptosis can occur if the levator palpebrae superioris muscle fails to develop (Fig. 27.14)
3. Epicanthus is the development of a crescentic fold of skin passing from the upper eyelid to canthus. This is a normal feature in some races.
4. Cryptophthalmos is failure of the eyelids to separate from each other resulting in absence of the palpebral fissure. The eyeball is small and defective and covered by the skin.

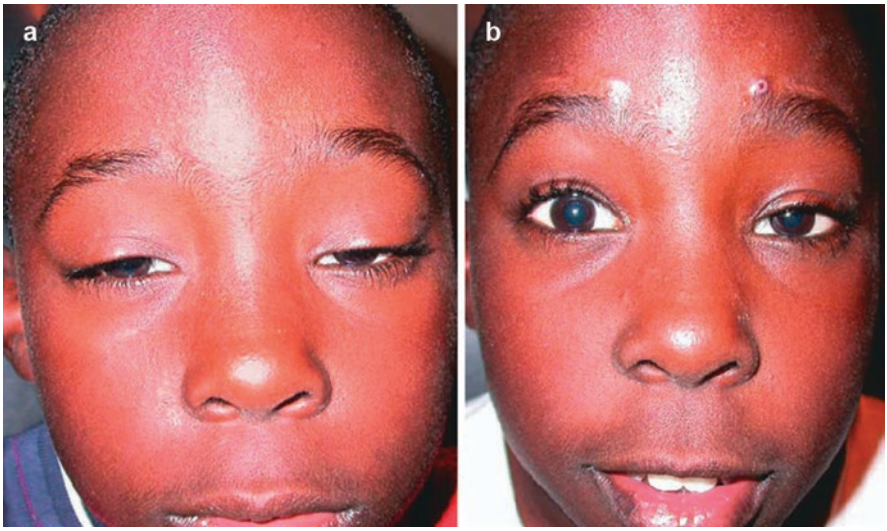


Fig. 27.14 (**a**) Congenital ptosis with poor levator function (preoperative). (**b**) Postoperative bacterial infection with recurrence of ptosis and purulent discharge from wound above left brow following bilateral ptosis repair with frontalis suspension with donor fascia lata

27.1.4 The Lacrimal System

The lacrimal gland is located in the upper, outer part of each orbit, in the lacrimal recess. This arises from multiple buds that sprout from the developing conjunctival sac. Two pouches develop—orbital and palpabral. These gradually become canalised to form the acini and the ductules of the developing gland. The ducts of lacrimal gland thus open in the conjunctival sac. In the adult, the gland is composed of two lobes—a main orbital and a smaller palpebral lobe. Its ducts empty into the upper lateral half of the superior fornix. Blinking distributes tears medially across the globe, where they reach the puncta and lacrimal drainage system. Here, the tears drain through the upper and lower puncta into the lacrimal sac via the upper and lower canaliculi. The canaliculi form a common canaliculus before entering the lacrimal sac. The nasolacrimal duct then passes from the sac into the nose, where the tears finally drain. The lacrimal gland receives innervation from the lacrimal branch of the oculomotor division of the trigeminal nerve and secretory parasympathetic fibres from the geniculate ganglion (related to the facial nerve). Anomalies of the gland include agenesis, ectopic or a nonfunctioning gland.

Development of the nasolacrimal duct begins with thickening of the ectoderm along the line of fusion between the lateral nasal and maxillary processes. This thickening separates away from the surface ectoderm and becomes buried as a solid cord. It later becomes canalised, forming the lacrimal sac and nasolacrimal duct. The nasolacrimal duct only becomes fully patent after birth. Failure of the distal part to fully canalise is a common cause of a watering, sticky eye in babies. This tends

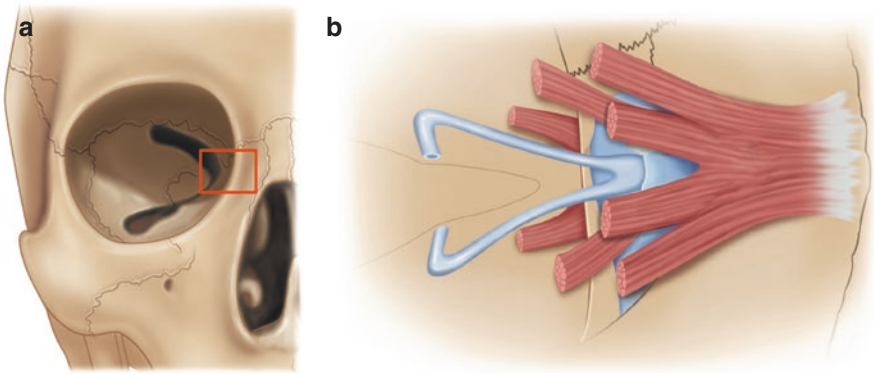


Fig. 27.15 (**a**, **b**) The canthal region (corner of the eye) is a key anatomical site both functionally and cosmetically

to resolve spontaneously by the second year of life. Cysts can also develop in any part of the lacrimal apparatus. In adults the nasolacrimal duct runs from medial corner of the eye to the inferior meatus in the lateral nasal wall. Anomalies of this system may be seen in some oblique facial clefting defects. Here, the duct is converted into an open gutter. Atresia of the nasolacrimal duct and supernumerary canaliculi can also occur. Tear drainage is an active process. Each blink of the lids helps to pump tears through the system (Figs. 27.15 and 27.16).

27.1.5 Ocular Blood Supply

The globe receives most of its blood supply from the ophthalmic artery (a branch of the internal carotid artery). This gives off the retinal artery, ciliary arteries and muscular arteries. Fine anastomoses also exist with branches of the external carotid artery. This is a clinically important point in the cosmetic industry. Injection of subcutaneous filler in the periocular area has been reported to lead to central retinal artery occlusion.

Rarely the ophthalmic artery does not arise from the internal carotid artery. The most common abnormal origin is from the middle meningeal artery by an enlargement of an anastomosis between the recurrent branch of the lacrimal artery and the orbital branch of the middle meningeal artery through the superior orbital fissure or a foramen in the greater wing of the sphenoid. This anastomosis is present during foetal life and becomes stronger when the ophthalmic artery is stenosed or not connected to the internal carotid artery. In some cases the ophthalmic artery trunk origin from the internal carotid artery may be markedly stenosed, so that the major supply to the ophthalmic artery is from the middle meningeal artery. Irrespective of its origin, all the blood to the optic nerve comes from the ophthalmic artery, making it a vulnerable structure. The artery follows a tortuous path within the orbit, crossing the optic nerve where the short posterior ciliary arteries, the long posterior ciliary

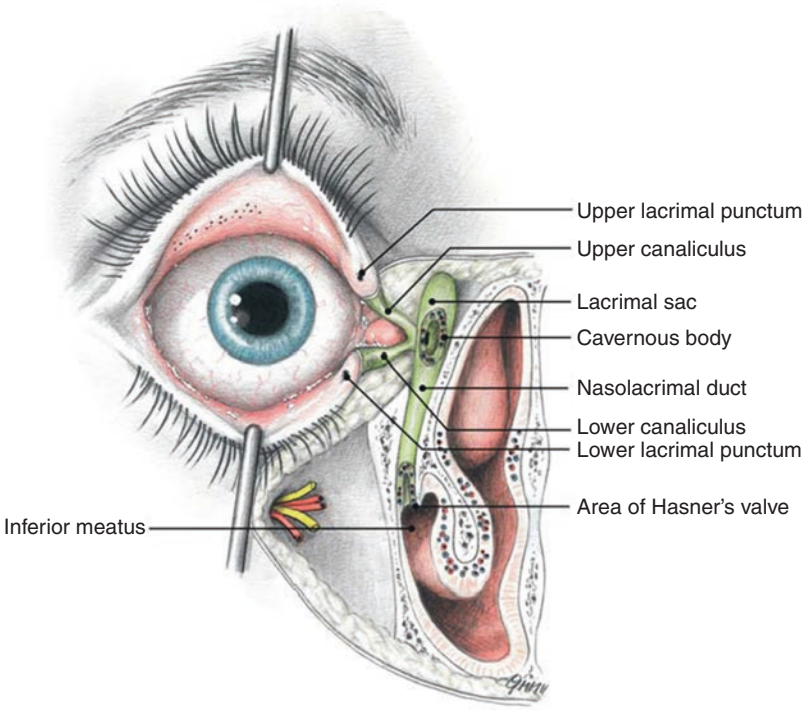


Fig. 27.16 Ocular surface and nasolacrimal ducts. The ocular bulbus with cornea and bulbar conjunctiva, as well as tarsal conjunctiva, are visible. At the medial rim of the upper and lower lid open the lacrimal puncta leading into the lacrimal sac via the upper and lower canaliculi. The lacrimal sac is situated in the orbital lacrimal fossa and proceeds into the nasolacrimal duct. The nasolacrimal duct is surrounded by a bony canal created by the maxillary and lacrimal bones and opens into the inferior meatus of the nose. Both lacrimal sac and nasolacrimal duct are surrounded by a vascular plexus comparable to a cavernous body that is connected to the cavernous system of the nose

arteries and the central retinal artery branch off. The anterior part of the optic nerve is supplied from the ciliary arteries. The choroid receives its blood supply mainly from the posterior ciliary arteries. The inner retina is supplied by arterioles from the central retinal artery. These are end-arterioles, that is, each supplies a volume of retina, with little anastomoses or overlap. Occlusion will therefore result in ischaemia of that tissue supplied by the arteriole. The capillary endothelial cells are joined by tight junctions so that the vessels are impermeable to proteins. This forms a 'blood–retinal barrier', similar to the blood-brain barrier. The capillaries of the choroid, however, are fenestrated and leaky. Breakdown of these barriers (in some vascular diseases) may thus be recognisable on fundoscopy (Fig. 27.17).

In contrast, the vascular system of the eyelids is rich and consists of numerous overlapping vessels. The arterial blood supply to the conjunctiva is derived from

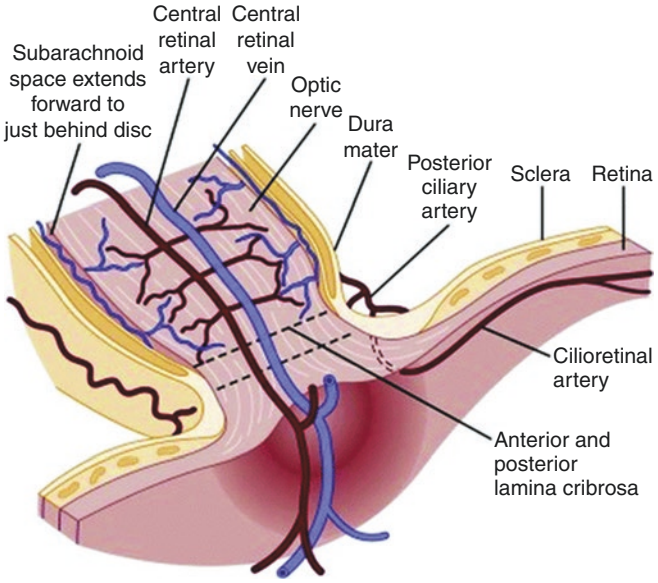


Fig. 27.17 From Balcer LJ and Prasad S, "Abnormalities of the Optic Nerve and Retina". Reprinted with permission from Springer Publishing Company

branches of the ophthalmic artery, the anterior and posterior conjunctival arteries. Blood drains from the orbit via the superior and inferior ophthalmic veins, which pass through the superior orbital fissure. Communications are present between these vessels and the facial vein and pterygoid plexuses. Of significance is the cavernous sinus and the potential for infection to spread from the face to this via the venous system. This is described in the chapter on the head.

27.1.6 The Third, Fourth and Sixth Cranial Nerves

The nuclei of the third (oculomotor) and fourth (trochlear) cranial nerves lay in the midbrain; the nuclei of sixth nerve (abducens) lays in the pons. The third nerve leaves the midbrain ventrally between the cerebral peduncles. It passes between the posterior cerebral and superior cerebellar arteries and then lateral to the posterior communicating artery. Aneurysms of this artery may therefore cause a third nerve palsy. All three nerves enter the lateral wall of the cavernous sinus and from there pass into the orbit through the superior orbital fissure. The sixth nerve has a long intracranial course. This is important because it can often be involved in various intracranial pathologies (such as base of skull fractures, invasion by nasopharyngeal tumours and raised intracranial pressure).

27.2 Ocular Related Functions

27.2.1 The Tear Film

The surface of the eye is constantly bathed by the tears, secreted mostly by the lacrimal gland. This watery secretion is supplemented by secretions from the meibomian glands. These secrete oil onto the anterior surface of the tear film during blinking, where it slows evaporation. The normal tear film is thus comprised of:

1. a thin mucin layer in contact with the globe
2. an aqueous layer
3. a surface oil layer

The tear film is about 3 μm thick. Tears not only provide a smooth interface for distortion-free refraction of light, but they also removes debris and foreign particles and have antibacterial properties. Lysozyme, lactoferrin, defensins and secretory IgA have all been found in tear fluid.

27.2.2 The Cornea

Developmentally, the cornea is derived from several sources.

1. The outer squamous epithelium is derived from the surface ectoderm
2. The lamina propria is derived from the mesoderm. This is continuous with the sclera.
3. The substantia proprea and inner corneal epithelium is derived from the neural crest cells.

Development is initiated by the lens vesicle. This induces the surface ectoderm to become transparent and avascular. The main functions of the cornea are to contain and protect the internal structures of the globe, whilst allowing light to enter. Together with the lens, it refracts and precisely focuses light onto the retina. The junction between the surrounding air and the surface of the cornea is an important refractive interface. This must be smooth and uniform. Any disruption to the cornea or this interface can significantly impair vision. The cornea is avascular and devoid of lymphatic drainage. It receives its nutrition from the surrounding peripheral capillaries and the aqueous humor which bathes its posterior surface. Some of its oxygen requirement is also provided by the ambient air. Any tightly fitting contact lens can therefore deprive the anterior cornea of oxygen and result in corneal oedema. The cornea is innervated by the ophthalmic division of the trigeminal nerve (the long ciliary nerves).

27.2.2.1 Corneal Dystrophies

These are inherited disorders in which the cells of the cornea develop with inborn defects. These undergo pathological changes over a period with the development of

a corneal haze in otherwise normal eyes. Dystrophies occur and are occasionally noticed at birth, but more usually during first or second decade and sometimes later. They are classified according to their site of development and other factors. They include Epithelial and subepithelial dystrophies, Bowman layer dystrophies, Stromal dystrophies and Descemet membrane and endothelial dystrophies.

27.3 The Aqueous Humor

This oxygenates and nourishes the cornea, lens, anterior vitreous and trabecular meshwork through diffusion. It also removes metabolic waste products. The hydrostatic pressure formed by the balance of aqueous production and its resorption establishes the IOP and helps maintain the correct alignment of the lens and cornea. Aqueous also contains anti-oxidants and of immunoglobulins.

27.3.1 The Ciliary Body and Iris

The ciliary body develops as a wedge-shaped extension from the anteriormost part of the choroid. It is formed by the adjacent mesoderm. The pigmented layer of the ciliary epithelium including the ciliary processes, is derived from the outer layer of the optic cup. This is continuous with the retinal pigment epithelium. The non-pigmented layer is an anterior extension of the neural retina, but lacks neural tissue. The ciliary epithelium over the ciliary processes later becomes secretory, producing the aqueous humor. The ciliary muscle is comprised of smooth muscle. Both it and the connective tissue of the ciliary body are derived from mesoderm. In the adult eye the ciliary body is divided into three parts: (1) the ciliary muscle, (2) the ciliary processes (pars plicata) and (3) the pars plana. The ciliary muscle is made of smooth muscle and is innervated by the parasympathetic system. It is responsible for the changes in the lens's curvature and thickness during accommodation. The ciliary processes (pars plicata) secretes the aqueous.

The iris develops from the rim of the optic cup, as an anterior extension of the two layers. These grow inward and partially covers the lens, but not fully—the central circular hole persists as the pupil. The epithelium of iris is thus derived from both layers of the optic cup. The connective tissue of the iris is derived from the mesoderm. The dilator and sphincter muscles of the iris are neuroectodermal in origin. The epithelium continues posteriorly into the double layered epithelium of the ciliary body. The iris is attached peripherally to the ciliary body. Its aperture (the pupil) can vary in size by its sphincter and dilator muscles. This controls the amount of light entering the eye. The sphincter muscle is innervated by the parasympathetic system. The smooth dilator muscle is innervated by the sympathetic system. The colour of the iris depends on its concentration and distribution of pigment-containing chromatophores. When melanin is confined to the epithelium on the posterior surface of the iris, it appears blue. With more anterior distribution the iris appears brown. Oculocutaneous albinism (OCA) is a group of autosomal recessive genetic disorders caused by a mutation in

chromosome 11q14, which encodes for the tyrosinase enzyme. This results in a disorder in which melanocytes fail to produce melanin pigment. Clinical features include pink skin, blue-grey eyes and disorders of the hair (Fig. 27.18).

Persistence of the anterior part of the vascular membrane of the lens can completely cover the pupil. This is commonly seen in premature infants. Normally the membrane undergoes atrophy, however persistence results in "atresia of the pupil". This usually requires surgical intervention. Coloboma of the iris is a defect in the inferior part of the iris. This results in an "keyhole" appearance. In some patients the defect may extend to the ciliary body and the retina. It occurs as a result of failure of the retinal fissure to close (the ventral groove formed by invagination of the optic cup and its stalk).

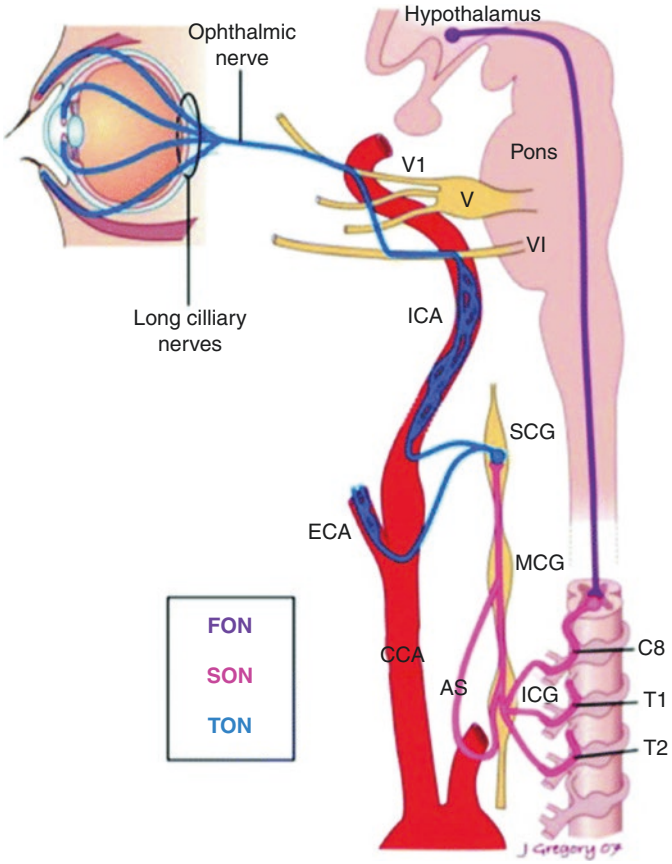


Fig. 27.18 Three-neuron sympathetic innervation of tile eye. *AS* ansa subclavia, *ECA* external carotid artery, *ICA* internal carotid artery, *ICG* inferior cervical ganglion, *MCG* middle cervical ganglion, *SCG* superior cervical ganglion, *FON* first-order neuron, *SON* second-order neuron, *TON* third-order neuron. From Reede D. Garcon E, Smoker W. Kardon R. Horner's Syndrome: Clinical and Radiographic Evaluation. Neuroimaging Clinics of North America [serial online]. January 1, 2008;18 (CraniaI Nerves):369–385. Available from: Science Direct. Ipswich. MA. Reproduced with permission from Science Direct

27.3.2 The Lens

The lens develops from a thickening of the surface ectoderm—the lens placode. This is induced by the underlying developing optic vesicle. Failure to develop results in congenital aphakia. The placode invaginates and separates from the ectoderm to form the lens vesicle. The vesicle is at first made of a single layer of cells. However, the cells of posterior wall elongate and extend into the cavity of vesicle. The posterior cells later lose their nuclei and become transparent primary lens fibres. The cells of anterior layer persist as an epithelial layer. The lens slowly grows during childhood as new lens fibres (secondary lens fibres) are added to it by the cells. The original primary lens fibres become old and harden. In the early stages of development the lens is supplied by the hyaloid artery (from the ophthalmic artery). However this eventually involutes and the lens becomes an avascular structure. It then obtains nutrition from the aqueous humor in the front and the vitreous humor at the back. A cataract is an opacity in the lens. This can be congenital or acquired. Congenital cataracts usually occur bilaterally. They are caused by a number of conditions affecting lens development, notably rubella, toxoplasmosis, Down's syndrome (trisomy 21) and galactosemia (an inborn error of metabolism). Congenital cataract can also be seen in avitaminosis and parathyroid deficiency.

In the adult eye, the lens comprises the second major refractive element of the eye, after the cornea and tear film. It is composed of an outer collagenous capsule under which is a layer of epithelial cells. This gives rise to the lens fibres which make up the bulk of the lens. The lens is supported by zonular fibres which pass between the ciliary body and its capsule. The lens grows throughout life. When young, it can easily change shape from thin to thick (during accommodation), allowing the eye to focus on close objects. This ability diminishes in most people after the age of 40—presbyopia, hence the need for reading glasses as we get older. This is important to remember when assessing visual acuity. Most people over 40 who are tested without corrective lenses will appear to have poor vision, even though it may be normal for them.

27.3.3 The Sclera, Choroid and Retina

The choroid and sclera develop from mesenchymal tissue around the optic cup. This differentiates into an inner vascular choroid and an outer fibrous sclera. The sclera continues anteriorly with the cornea, whilst posteriorly it is continuous with the dura mater surrounding the optic nerve. The choroid also continues anteriorly with the ciliary body. Fully developed sclera is formed from interwoven collagen fibres. It is of variable thickness, being thinnest just distal to the extraocular muscle insertions. Blue sclera is an asymptomatic condition characterised by a marked, generalised blue discolouration of the sclera as a result of thinning. It is typically associated with osteogenesis imperfecta. Other causes are Marfan's syndrome, Ehlers-Danlos syndrome, pseudoxanthoma elasticum, buphthalmos, high myopia and healed scleritis. Choroid is composed of arterioles, venules and a dense

capillary network. This is loosely attached to the sclera. The choroid nourishes the deeper layers of the retina. If the sclera is extremely thin, the underlying pigments of the choroid become visible. This is known as 'Blue sclera'.

The retina itself is a complex structure, comprised of multiple layers. It is derived from the walls of the optic cup. The outer layer of the cup is thin and develops into the retinal pigment epithelium (RPE). The inner layer of the cup is thick and becomes the neural layer of the retina. During early development these layers are initially separated, but this "intraretinal" space slowly disappears and the two layers come to approximate very closely. However, they do not fuse together. In detachment of the retina the neural layer separates from the pigment layer. The neural layer eventually develops into the light-sensitive zone containing modified neurones (photoreceptors—the rods and cones), bipolar neurons and ganglion cells. Axons from the ganglion cells pass into the optic stalk to eventually form the optic nerve. Photoreceptors convert light into electrical signals. Cones are stimulated by daylight and colour vision. They are concentrated at the fovea, the site which is responsible for detailed vision (such as reading fine print). Rods are responsible for night vision and form the majority of the photoreceptors in the remaining retina. Myelination of the fibres occurs following exposure of the eye to the light during the first 10 weeks after birth. In new born infants vision is initially poor. Coloboma of the retina is a defect just inferior to the optic disc. Like the iris, this occurs due to defective closure of the retinal fissure. Retinocoele results from herniation of the retina into the sclera from failure of the choroid fissure (Fig. 27.19).

27.3.4 The Optic Nerve

The optic nerve is formed by the coalescence of the axons arising from the retinal ganglion cell layer. These pass out of the eye through the cribriform plate of the

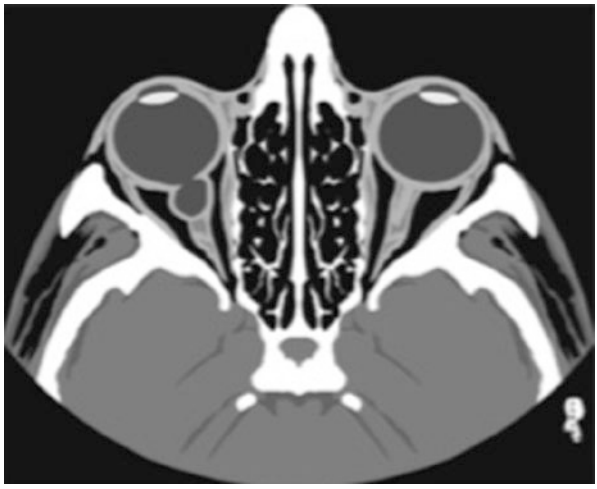


Fig. 27.19 Axial orbital CT illustration shows cystic expansion of the right optic disc (optic disc coloboma)

sclera, a sieve-like structure, at which point the nerve fibres become myelinated. The central retinal artery and vein enter the eye via the centre of the nerve. In the orbit the optic nerve is surrounded by a sheath of dura, arachnoid and pia mater. This is continuous with the layers surrounding the brain and contains cerebrospinal fluid. Because of the close proximity between the optic nerve and several large pulsating arteries (notably the carotid artery), it has been suggested that optic nerve compression by these vessels may be the pathological basis in some patients with normal tension glaucoma.

27.3.5 The Vitreous

The vitreous is a clear gel occupying the posterior two-thirds of the globe. It develops in two stages—primary and the secondary. Primary vitreous comes from the neural crest cells. Secondary vitreous is believed to arise from the inner cell layer of the optic cup. Vitreous has both a nutritive and structurally supportive role. It is composed mostly of water which contains hyaluronic acid and a few cells. The vitreous is firmly attached to the peripheral retina and around the optic disc, but is loosely attached elsewhere. Collapse of the vitreous gel (vitreous detachment) therefore puts traction on these points of attachment, which can result in a retinal tear.

27.3.6 Overview of the Visual Pathway

Light enters the globe through the cornea. It passes through the aqueous, the lens, and the vitreous to strike the retina. This stimulates the photoreceptor cells (rods and cones) in its deepest layer. Depolarisation of these cells occurs, which transmits impulses to the bipolar cells in the intermediate layer. These cells are the primary afferent neurons of the visual system. Impulses are then transmitted to ganglion cells, on the superficial layer of the retina. The axons of the ganglion cells pass across the superficial surface of the retina and converge at the posterior pole of the globe, where they pierce the sclera and form the optic nerve. Myelination of the axons occurs only outside of the globe. Axons within the globe are not myelinated. The optic nerve passes through the orbit and enters the skull via the optic canal. Both optic nerves combine at the optic chiasma, just above the sella turcica. Axons from the nasal half of each retina decussate and join the opposite optic tract, whereas axons from the temporal half of the retina continue their journey uncrossed. This arrangement has evolved because each visual field projects onto both retinas, with the right visual field projecting onto the nasal half of the right retina and the temporal half of the left retina and vice versa. Fibres from both retinas, carrying information about the right visual field, thus combine at the optic chiasma, to form the left optic tract. As a result the entire right visual field is transferred to the left hemisphere. The optic tracts leave the optic chiasma and pass dorsolaterally around the hypothalamus to synapse with cells of the lateral geniculate bodies. Cell bodies of

the lateral geniculate nucleus then give rise to the optic radiation, which passes to the primary visual cortex on the medial aspect of the occipital lobes.

The entire visual pathway therefore results in a precise topographic arrangement, enabling clear three-dimensional perception of the environment. Because of these predictable pathways, discrete lesions affecting the visual pathway can often be localised by clinical examination. For example:

1. Lesions affecting the optic nerve will result in monocular blindness.
2. Lesions of the decussating nasal fibres at the optic chiasma will result in loss of each eye's temporal visual field (bitemporal hemianopia).
3. Partial interruption of the retrochiasmal visual pathway will result in incomplete hemianoia.
4. Complete interruption of the retrochiasmal visual pathway will result in total homonymous hemianopia (loss of half of the field of view on the same side in both eyes).
5. Interruption of the pathway at the optic tract causes contralateral homonymous hemianopia.
6. If the optic radiations are interrupted within the temporal lobe, a contralateral superior quadrantanopia results.
7. Lesions of the occipital pole can result in central macular hemianopic deficits (Fig. 27.20).

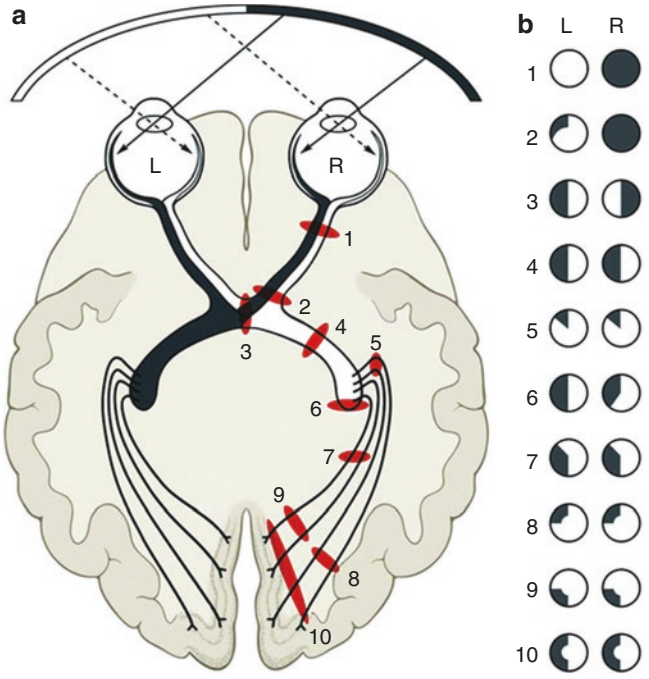


Fig. 27.20 (**a**, **b**) Visual pathways and visual field defects associated with lesions at various locations. From BCSC Neuro-Ophthalmology. Reproduced with permission from American Academy of Ophthalmology

27.4 Diplopia (Double Vision)

Diplopia is the simultaneous perception of two images of a single object. It can be horizontal (two images in the horizontal plane), vertical (one image above the other) or torsional. It is most commonly seen following trauma, but can occur secondary to other pathologies. Following trauma, diplopia is usually the result of either impaired function of the delicate extraocular muscles (EOMs), or entrapment of the orbital fat within an orbital fracture (most commonly the floor or medial wall). Other trauma related injuries include 'deeper' orbital fractures involving the superior orbital fissure. These are higher energy injuries and often associated with optic and cranial nerve, skull base and intracranial problems. Non-traumatic causes include disorders of the neuromuscular junction and cranial nerves (III, IV, and VI) following ischaemic events, inflammation or tumour infiltration, or occasionally ingestion of toxins. Physical displacement of the globe by tumour, orbital swelling or fracture displacement can also result in double vision. The list of possible causes is therefore long, representing a diverse range of mechanical, ophthalmologic, infectious, autoimmune, neurological and neoplastic disorders.

1. Muscle/fat entrapment following trauma
2. Isolated third fourth or sixth cranial nerve palsies (traumatic and non-traumatic)
3. Pathology involving the cavernous sinus
4. Strabismus
5. Cerebral tumours and brain stem lesions
6. Cataracts
7. Diabetes
8. Orbital tumours
9. Orbital infections and inflammation
10. Botulism
11. Drunkenness
12. Graves disease
13. Guillain–Barré syndrome
14. Keratoconus
15. Migraine
16. Multiple sclerosis
17. Myasthenia gravis
18. Wernicke's syndrome
19. Giant cell arteritis
20. Decompensating phoria

Most cases of diplopia are binocular, that is, due to an imbalance of action between both eyes (neurological or mechanical in origin). If one eye is covered the diplopia resolves. Monocular diplopia is less common and is double vision that is

perceived in one eye only. Such cases are usually due to a corneal problem, subluxation of the lens, or some other structural problem within the eye itself. Occasionally it can arise secondary to a lesion in the visual cortex.

27.4.1 Causes of Monocular Diplopia

Common	Uncommon
Edge effect from intra ocular lens	Cataract
Abnormal lens shape (lenticonus)	Corneal oedema
Polycoria	Astigmatism
Subluxation of crystalline lens or IOL	Corneal ectasia (e.g. keratoconus)

Most causes of double vision are painless. Neurogenic diplopia is worse when the patient looks in the direction of action of the affected muscle. In mechanical restriction there is worsening of diplopia when looking in the opposite direction of the trapped muscle. Painful diplopia is usually associated with orbital inflammation, infections and trauma. Space occupying lesions or aneurysms can cause a variety of painful isolated cranial nerve palsies, notably a posterior communicating artery aneurysm and painful third nerve palsy. Isolated painless cranial nerve palsies may be due to microvascular pathology. When examining the eyes it is therefore important to note the following:

1. Monocular or Binocular diplopia
2. Horizontal or vertical diplopia
3. Obvious restriction of eye movements
4. Signs of orbital injury or disease (proptosis, inflammation, visual acuity)
5. Abnormal pupil responses, notably signs of a 3rd cranial nerve palsy
6. Ptosis

Finally examine all the cranial nerves and look for any other neurological deficits. Diplopia as an isolated clinical problem and is unlikely to present acutely, unless following trauma. If there is obvious restriction of movement in one of the eyes CT is usually indicated to look for orbital fractures or pathology. All cases of diplopia should be discussed with an ophthalmologist, especially in children.

27.4.1.1 Third Nerve Palsy

Third nerve palsy due to a posterior communicating artery aneurysm is a potentially fatal condition and needs prompt diagnosis. Rupture of the aneurysm can result in subarachnoid and intracerebral haemorrhage (discussed in the chapter on the head). Third nerve palsy results from a complete or partial palsy of the oculomotor nerve. The nerve arises in the midbrain, passing through the subarachnoid space and over the edge of the tentorium cerebelli. It then passes near to the vessels in the circle of

Willis before entering the cavernous sinus and finally the orbit. It innervates the levator muscle of the upper eyelid, superior, medial and inferior recti, and inferior oblique muscles, as well as the parasympathetic supply for pupillary constriction and accommodation. Causes of third nerve palsy include:

1. Aneurysm—Just after the nerve leaves the midbrain it is intimately related to the posterior communicating artery. Aneurysms here may compress the nerve.
2. Raised intracranial pressure—Expanding lesions above the tentorium can result in herniation the uncus of the temporal lobe through the tentorial notch (uncal herniation). Compression the midbrain passing through the notch and the nearby oculomotor nerve results is pupillary dilatation (unopposed sympathetic action since the parasympathetic fibres in III are affected).
3. Tumours (parasellar/nasopharyngeal)
4. Trauma
5. Microvasular ischaemia—diabetes, hypertension, atherosclerosis, smoking. It is not uncommon for diabetics to suffer from an acute vasculitis of the oculomotor nerve.
6. Vasculitis
7. Basal meningitis
8. Demyelination
9. Migraine
10. Idiopathic
11. Tolosa-Hunt syndrome

Vascular or other lesions of the midbrain can thus affect the nerve. They can also affect the substantia nigra resulting in Parkinsonian symptoms (e.g. resting tremor), the red nucleus (also causing extrapyramidal symptoms), and the descending corticospinal fibres in the cerebral peduncles leading to a contralateral upper motor neuron lesions (UMNL). Benedikt's syndrome involves the nerve as it passes through the red nucleus—there is oculomotor paralysis with contralateral extrapyramidal dyskinesia. In Weber's syndrome the lesion is more ventral, also involving motor fibres in the cerebral peduncles: oculomotor paralysis is associated with contralateral UMNLs.

Patients present with horizontal and vertical diplopia.The affected eye is exotropic with hypotropia (turned out and down) and is associated with a ptosis. There is reduced adduction, elevation and depression, whilst abduction and intorsion are normal. Examination of pupils is the key to diagnosing a surgical cause—the pupils are dilated if there is a mass lesion causing compression. With medical causes the pupillary sizes and reactions are normal. Localised pain and headache and important symptoms. Painful third nerve palsy should be considered to be secondary to a surgical cause until proven otherwise. Medical third nerve palsies are usually painless. Rupture of a posterior communicating artery aneurysm and pituitary apoplexy are two important diagnoses. Pituitary apoplexy presents with headache, reduced

vision (unilateral or bilateral), diplopia and ± nausea and vomiting and an altered level of consciousness. All third nerve palsies requires urgent neuroimaging. High resolution CT angiography is now the investigation of choice. MRI /MRA are also helpful. Pupil sparing third nerve palsies need evaluation of vascular risk factors (BP, lipids, Blood glucose, ESR, CRP). Intracranial bleeds, aneurysms and tumours require immediate transfer to a neurosurgical unit. Management of the diplopia itself requires orthoptic evaluation of ocular movements. Diplopia can be relieved by prisms or occluding the affected eye. In some patients, aberrant regeneration of third nerve can occur following injury or removal of compressive lesions. These can cause bizarre ocular movements which do not fit any particular pattern.

27.4.2 Fourth Nerve Palsy

The trochlear nerve is the thinnest and most fragile of the cranial nerves. It is therefore vulnerable to trauma. Section of the nerve would result in the affected eye being turned medially. The fourth (trochlear) nerve nucleus is in the caudal midbrain. This slender nerve exits posteriorly and crosses its opposite neighbour to wrap around the brainstem. It then follows a long path to supply the contralateral superior oblique muscle. Intorsion and depression of the eye are the main functions of this muscle. Consequently a palsy makes reading and walking down stairs difficult. Causes of fourth nerve palsy include:

1. Congenital
2. Trauma
3. Microvasular ischaemia—diabetes, hypertension, atherosclerosis, smoking
4. Tumours
5. Demyelination
6. Vasculitis
7. Infections
8. Tolosa Hunt syndrome

Patients present with vertical diplopia which is worse on downgaze. The affected eye is hypertropic (deviated up), especially if the head is tilted to the side of the palsy (Bielschowsky test). Patients may adopt a characteristic head posture to compensate for the diplopia—the chin is lowered and the head turned and tilted to the opposite side. Associated cranial nerve palsies and neurological deficits should be looked for. Bilateral palsies may be more difficult to detect and more common following trauma. Unlike third nerve palsies, fourth nerve do not require routine neuroimaging. Assessment of vascular risk factors is required if the palsy is thought to be due to microvascular ischaemia. Orthoptic assessment and vertical prisms may alleviate diplopia. Patients generally manage well with head posture, prisms, and occasionally surgery.

27.4.2.1 Sixth Nerve Palsy

This involves the abducens nerve. This nerve, with a relatively low origin compared to its destination, has the longest intracranial course of any cranial nerve. It can be involved in fractures of the base of the skull or in intracranial disease. The sixth nerve nucleus is in the pons. It passes around the facial nerve nucleus to leave the brainstem anteriorly. The nerve then courses over the petrous bone towards the cavernous sinus and from there passes into the orbit. It supplies the lateral rectus muscle and abducts the eye. Causes of sixth nerve palsy include

1. Congenital
2. Microvasular ischaemia—diabetes, hypertension, atherosclerosis, smoking
3. Tumours (acoustic neuroma)
4. Raised intracranial pressure—As this rises the cerebrum may be displaced backwards and downwards, stretching this long intracranial nerve. The resulting lateral rectus palsy is a false localising sign.
5. Trauma (skull base fracture)
6. CNS Infections
7. Vasculitis
8. Demyelination
9. Cavernous sinus thrombosis
10. Caroticocavernous fistula

Patients develop horizontal diplopia, which is usually worse for distance vision and when looking to the side of the affected muscle. The affected eye is esotropic (turned in). Isolated palsies are usually ischaemic in origin, bilateral passes are usually associated with trauma or space occupying lesions. Neuroimaging is therefore required. Patching of the eye or prisms can give relieve symptoms. Recovery depends on the cause and varies over several months. Ischaemic palsies have the best prognosis.

27.5 Photophobia

Photophobia is the sensitivity of the eye to light. This is often painful. It is commonly associated with meningitis and subarachnoid haemorrhage. Causes include:

27.5.1 Ocular Conditions

- Cornea
  - corneal foreign body
  - corneal abrasion
  - infective/microbial keratitis
  - dry eye syndrome
- Conjunctivitis—viral, bacterial, allergic
- Acute anterior uveitis

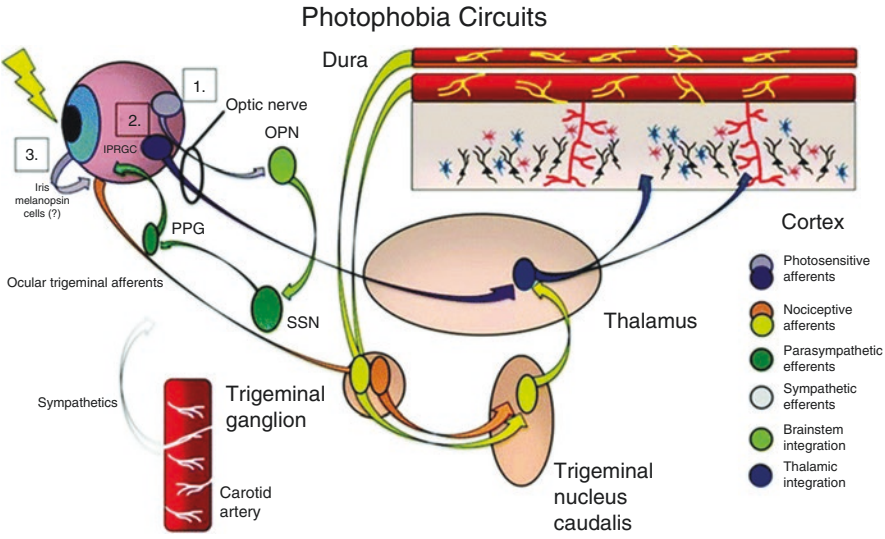


Fig. 27.21 Proposed pathway for chronic photophobia From Katz and Digre [254]. Reproduced with permission from Wolters Kluwer

27.5.2 Central Nervous System Causes

- Migraine
- Meningitis
- Subarachnoid haemorrhage

If photophobia is associated with severe headache, nausea and vomiting, neck stiffness, high fever and irritability or confusion, urgent investigations and referral are required (Fig. 27.21).

27.6 Important Considerations When Taking a History

When taking an 'ocular' history, there are a number of key points that help to triage the patient's disease or injury. Very rarely, the ocular symptoms may be the first sign of carcinoma or metastasis. Disturbances in the vision is perhaps the most single important indicator of severity and prognosis. The history and examination should focus on three main aspects—(1) assessment of the patient's visual function, (2) assessment of symptoms affecting the eye and surrounding structures and (3) determining the systemic status of the patient. Irrespective of the presenting features, in

all patients it is important to ask specifically about visual loss and ocular discomfort. The time of onset and progression of these symptoms should be noted, together with any obvious precipitants, alleviating and aggravating factors. It is also important to determine whether one or both eyes are affected and if both, which is the worse.

In some patients, symptoms may be recurrent, or prodromal episodes may have been occurring. In acute angle closure glaucoma, for example, repeated bouts of blurred vision and seeing haloes around lights may occur before the onset of pain. Temporary visual loss can occur in migraine, but also following retinal artery embolism or with raised intracranial pressure. Loss of colour perception, especially red, is common in optic neuritis, whereas difficulty seeing in low light intensity may be associated with a retinal dystrophy. These useful clues may be overlooked if a detailed history is not taken.

27.6.1 Visual Disturbances

If the patient's vision has changed, it is important to try and quantify this and get an idea of how much this has been reduced and how quickly. It can vary from blurring to total loss of vision. Sudden complete loss, or rapidly deteriorating vision requires urgent referral. Gradual loss of vision can arise from clouding of the ocular contents, such as by a cataract. Macular degeneration, Diabetic retinopathy and optic nerve compression will also result in gradual loss. Sudden visual loss implies inflammatory or ischaemic causes, or retinal detachment and neurological disease.

Blurred vision which resolves on blinking is likely to be related to debris or tears on the surface of the cornea. Unusual visual experiences e.g. floaters, photopsia (flashing lights), shadows/scotoma (abnormal blind spot), distortion of an image should be noted. If the patient uses spectacles or contact lenses, this should also be noted. With all visual disturbances try to determine:

1. Is this sudden, or gradual and longstanding?
2. Is this painful or painless?
3. Does it involve one eye or both eyes?
4. Does it involve and entire visual field or just part of one?

If these four questions can be answered a diagnosis will be made much easier.

Normally the cornea is transparent. Any condition which disrupts its anatomy or physiology can result in loss of transparency to a variable degree. Common causes include:

1. Corneal oedema
2. Drying of cornea

3. Depositions on cornea
4. Inflammation of cornea
5. Corneal degenerations
6. Dystrophies of cornea
7. Vascularisation of cornea
8. Scarring of cornea (corneal opacities).

Corneal oedema is a common finding. The normal cornea is about 80% water. This is delicately maintained by the balance between the intraocular pressure, swelling pressure of the stromal matrix and the active pumping action of the corneal endothelium and barrier function of the epithelium. Any disturbance in any of these leads to corneal oedema, with an increased water content and swelling. Causes of corneal oedema therefore include:

1. Raised intraocular pressure
2. Endothelial damage (trauma, corneal dystrophies and inflammation)
3. Epithelial (trauma, inflammation and infections).

Patients initially develop a stromal haze with reduced vision. Over time permanent oedema develops with epithelial vesicles and bullae formation (bullous keratopathy). There is then marked loss of vision, pain, discomfort and photophobia from rupture of the bullae.

27.6.2 Pain

Pain in and around the eye can be considered as (1) Gritty/foreign body type sensation. This tends to occur with corneal surface problems. (2) An ache or dull pain in the eye, (3) Headache and associated symptoms such as jaw claudication, (4) Photophobia—whilst there are several benign causes of this, meningitis and subarachnoid haemorrhage must always be considered. Assess the level of discomfort present, its onset, character and duration. Does it fluctuate, or is it relentless? Severe pain is often related to corneal damage (such as from an abrasion) due to the high number of pain receptors innervating its surface. Both trauma and infections of the cornea can result in severe pain and photophobia. Pain is also disproportionately severe in acanthamoeba keratitis.

27.6.3 Injuries

Not all injuries are necessarily obvious to the patient (or the clinician). Injury from tiny high-speed projectiles (e.g. following grinding or hammering) may not be realised to have occurred. Symptoms may also develop late. If trauma is suspected, ask about the circumstances and try to determine if it was most likely blunt or penetrating in nature. If possible determine the mechanism of injury and when it

occurred. Sudden onset of pain would suggest a penetrating or corneal injury. Ask about:

1. The circumstances—was the injury low velocity (dust, twigs, finger), or high velocity (hammering, grinding, drilling)?
2. What was the date and time of the injury?
3. Sharp or blunt injury
4. The size of the injuring object
5. Presence of chemical or organic matter
6. Was eye protection used?
7. What are the current symptoms (notably vision, pain and fever)

27.6.4 Additional Symptoms and Changes

Enquire about any discharge from the eye and its nature. Allergic conjunctivitis tends to produce a white, stringy discharge whilst in viral infections it is usually very watery. Bacterial infections produce a thicker mucopurulent discharge. If unilateral blood is noted, consider trauma, tumours and more aggressive pathology. Changes in the appearance of the globe, eyelids and surrounding tissues may indicate pathology extending into, or originating from adjacent sites. A "red eye" is a common symptom, generally caused by inflammation, infection, trauma or raised intraocular pressure. This is discussed later. Oedema, proptosis and reduced ocular movements suggest a space occupying process, commonly orbital cellulitis (Fig. 27.22).

Note any systemic symptoms such as nausea, fever, headache or a localised rash. Herpes zoster often presents with a characteristic rash in a dermatomal distribution. Hutchinson's sign (a rash involving the nose) is reported to indicate viral involvement of the nasociliary nerve which increases the likelihood of globe involvement. Acne rosacea and atopic dermatitis may be associated with blepharitis.

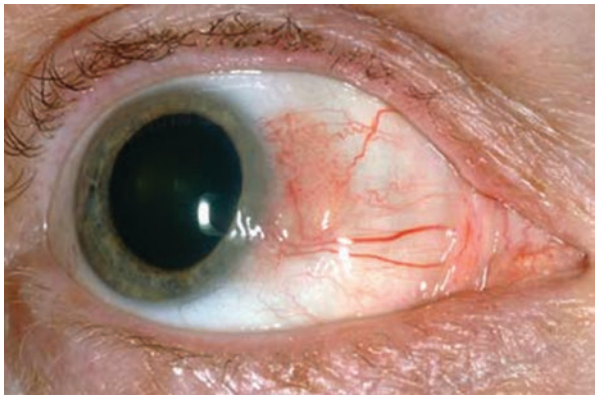


Fig. 27.22 Conjunctival squamous cell carcinoma

27.6.5 Double Vision

This has many causes, ranging from physiological to life and sight-threatening. Intermittent double vision often occurs when tired, but can be a feature of myasthenia gravis. Ask if it involves one, or both eyes. Also determine if it is acute, progressive, painful or associated with a change in the position of the globe (dystopia). Trauma, thyroid disease, cataracts, diabetes and hypertension are common underlying causes. Consider also the possibility of a space occupying lesion. The following characteristics of the diplopia may help with diagnosis:

1. Is the double vision vertical, horizontal, or a combination?
2. Does the double vision involve one or both eyes (monocular or binocular double vision)?
3. Does the double vision get worse when looking in a certain direction?
4. Is it progressive and painful?
5. Is there a history of childhood strabismus or muscle surgery?

27.6.6 Systemic Symptoms

Review systemic symptoms. Some ocular conditions (e.g. scleritis and uveitis) may be associated with symptoms of autoimmune diseases elsewhere. Ask about symptoms of hypertension, diabetes etc.

27.6.7 Past Ocular and Medical History

If there is a history of recent surgery or injury, consider the possibility of endophthalmitis or a retained foreign body. Complications of surgery may also include retinal tears, cataract and glaucoma. Note any preexisting ocular conditions such as amblyopia, squints, diabetes, glaucoma. The patient's normal vision and use of aids is important. This helps determine the severity of any visual deterioration and risks of some conditions. Nearsighted patients ("high myopes") have an increased risk for posterior vitreous detachment and retinal tears, whilst longsighted patients (hypermetropes) are at risk of acute angle closure glaucoma.

The past medical history is important as many systemic diseases can affect the eye.

1. Cardiovascular—consider hypertension, cholesterol, atrial fibrillation and risks of emboli. Subconjunctival haemorrhage may be a sign of hypertension or clotting disorder
2. Endocrine disorders such as diabetes and hyperthyroidism commonly involve the eye and orbit.

3. Autoimmune diseases, such as rheumatoid and sarcoid, can result in ocular inflammation.
4. Immunodeficient patients are at risk of severe ocular infections such as CMV, TB and toxoplasmosis.

Note any medications. Some systemic drugs (such as steroids and hydroxychloroquine) can have ocular side effects. Other drugs such as antidepressants, anticoagulants, adrenergic antagonists and antiepileptic dugs have been reported to produce ocular symptoms, especially in those with predisposed angle closure. These can result in pupillary dilation, which can precipitate acute angle-closure. Preservatives in some eye drops can result in irritation.

27.6.8 Family History

Enquire about glaucoma, macular degeneration, retinal tears or detachment, strabismus, amblyopia, hypertension, and diabetes. Unexpected visual loss in young family members should be asked about. Glaucoma is believed to have a genetic component.

27.6.9 Social History

Smoking can adversely affect macular degeneration. It predisposes to arteriosclerosis, emboli, wound infections, poor healing and optic neuropathy.

Travel abroad may have resulted in unusual infections. Also enquire about close-contacts if infection is suspected.

27.6.10 Triaging Ocular Symptoms and Signs

Not all ocular problems are sight-threatening. But to the inexperienced this is an ever present concern, particularly if patients are to be discharged home. However there are a few ‘red flag’ symptoms that are worth noting. These can often indicate significant pathology and the need for urgent referral. They include:

1. Significantly reduced or deteriorating visual acuity. This can indicate serious ocular disease. If blurriness improves with blinking, there is probably a collection on the corneal surface. Any sudden decrease in vision is significant. If it is not associated with pain or redness, consider a retinal detachment.
2. Repeated but transient visual loss. This may be a symptom of amaurosis fugax—indicative of embolism, possible carotid artery disease and risk of stroke.
3. Pain. If severe or worsening, this suggests a number of significant diseases such as keratitis, corneal ulceration, iridocyclitis or acute glaucoma. The acutely pain-

ful red eye, with blurred vision, should be regarded as an emergency. Similarly acute pain following injury or surgery needs urgent referral. Deep seated pain is significant. Pain in the eyelids and chronic pain or discomfort are generally less significant.
4. Headaches. Any history of headaches, should raise suspicion. The nature of the headache requires careful evaluation as described in the chapter on the head. Consider raised intracranial pressure, glaucoma and temporal arteritis.
5. Photophobia. This may occur in iritis, following injury to the cornea or acute glaucoma. CNS diseases should also be considered.
6. Photopsia (flashes of light). This can be a sign of a retinal tear, posterior vitreous detachment, migraine and occipital lobe infarction.
7. Distorted vision or a central blind spot may suggested macular degeneration. Coloured halos can occur when there is corneal oedema (following an acute rise in intraocular pressure—acute glaucoma).

27.7 Examining the Eye, Eyelids and Associated Structures

In most patients who present acutely, specialist equipment is not usually necessary in order to complete an adequate eye examination. Sufficient information can usually be obtained using the following:

1. A small but powerful torch
2. Visual acuity chart (Snellen or Sheridan-Gardiner)
3. Magnification—a handheld magnifying glass or simple magnification loupe will often suffice, although a slit lamp is preferable. This will help assess the anterior structures of the globe.
4. Cotton buds—for removal of foreign bodies and to evert the upper eyelid.
5. Fluorescein—drops or strips. A blue light source is also required to highlight the staining.
6. Local anaesthetic drops e. g. Amethocaine.
7. Dilating drops (Mydriatics) e.g. Tropicamide 1.0% (0.5% for neonates).
8. An ophthalmoscope to visualise the fundus.

27.7.1 External Examination

Initial general inspection of the eye, eyelids and adnexa should always be part of every examination, especially following trauma. This may reveal important clues.

1. A well defined ‘black eye’ or surgical emphysema usually indicates a fracture somewhere in the orbit.
2. Signs of an impact to the lateral brow or forehead can sometimes indicate the possibility of traumatic optic neuropathy.
3. Full thickness eyelid lacerations may be covering an underlying globe injury. Any fat prolapsing through an eyelid laceration is highly suggestive of orbital

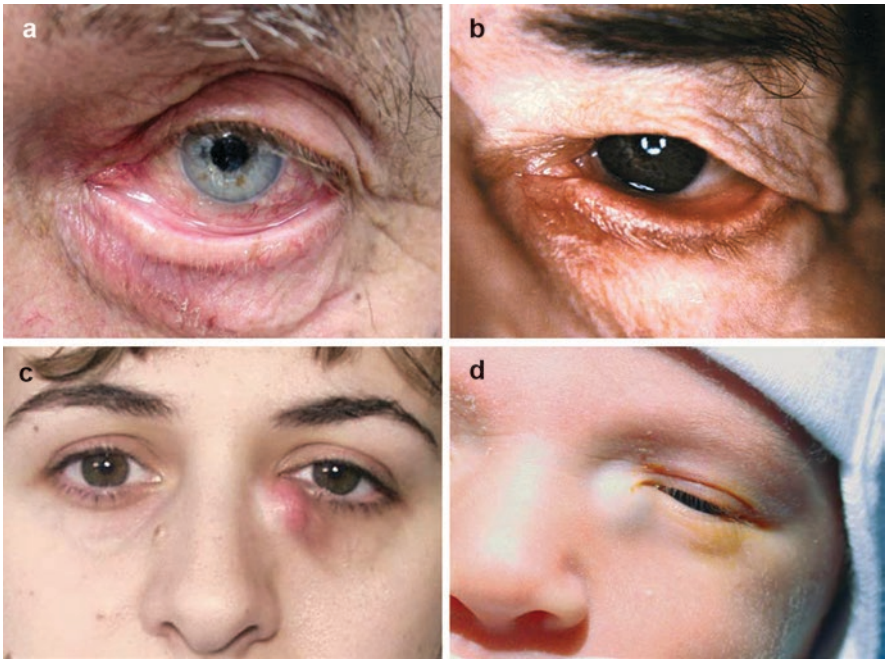


Fig. 27.23 External examination (**a**) medial ectropion (**b**) Lateral lid laxity and ectropion (**c**) Acute dacryocystitis (**d**) Mucocele

penetration, globe injury and a foreign body. A full thickness laceration medially may have affected the canaliculus and lacrimal drainage and requires ophthalmic input within 48 h. If the injuring mechanism is high velocity, intracranial injury should also be considered (Figs. 27.23, 27.24, 27.25, and 27.26).

Forced closure of the eyelids ("screwing up" the eyes), reflexively occurs when anticipating an impact. This is also associated with an upward rotation of the globe— Bell's phenomenon. Therefore, when assessing penetrating eyelid injuries it is important to carefully examine the lower limbus of the eye and lower sulcus. These will have been vulnerable at the moment of impact but later become covered by the lower eyelid when the eye returns to its normal position. The presence of bulging (proptosis) and changes in position of the globe (dystopia) should be noted. This may occur as a result of haemorrhage, infection, inflammation or tumour. Looking at the orbits from above (bird's-eye view) or below (worm's-eye view) can help in assessing for any degree of proptosis. When facing the patent directly, the reflection of any light source (pen torch, lighting, window) off the patient's corneas should be in the same position in both eyes. This means that the globes are level and looking in the same direction.

Using a pen torch, the conjunctiva, cornea, anterior chamber, and the lens should all be inspected at the bedside, although a slit lamp examination is preferred. The normal sclera is white and surrounds the iris and pupil. The sclera is covered the thin transparent conjunctiva, which reflects forwards to line the underside of the eyelids.

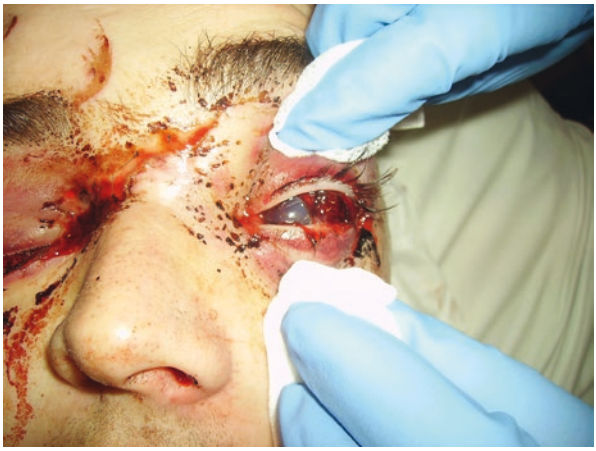


Fig. 27.24 Avoid globe pressure when opening eyelids

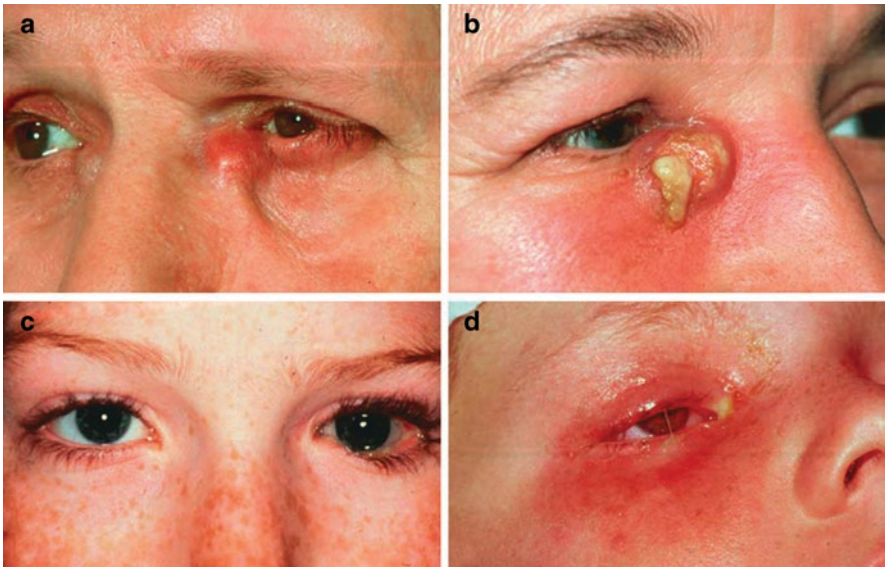


Fig. 27.25 Lacrimal sac mass (**a**) Mucocele, (**b**) acute dacryocyctitis, (**c**) and (**d**) Orbital cellulitis

Normally, it's invisible except for the fine blood vessels that run through it. When infected or inflamed, it can appear red—conjunctivitis. Severe vascular engorgement of the conjunctiva results in a 'red eye' appearance. Alternatively, the conjunctiva can appear pale if patient is very anaemic. By gently applying pressure and pulling down and away on the skin below the lower lid, the conjunctival reflection can be seen. Look for foreign bodies, tears and oedema (chemosis). Non-haemorrhagic chemosis can sometimes indicate a rupture or penetrating injury of the globe, where fluid accumulates under the conjunctiva. The cornea and lens

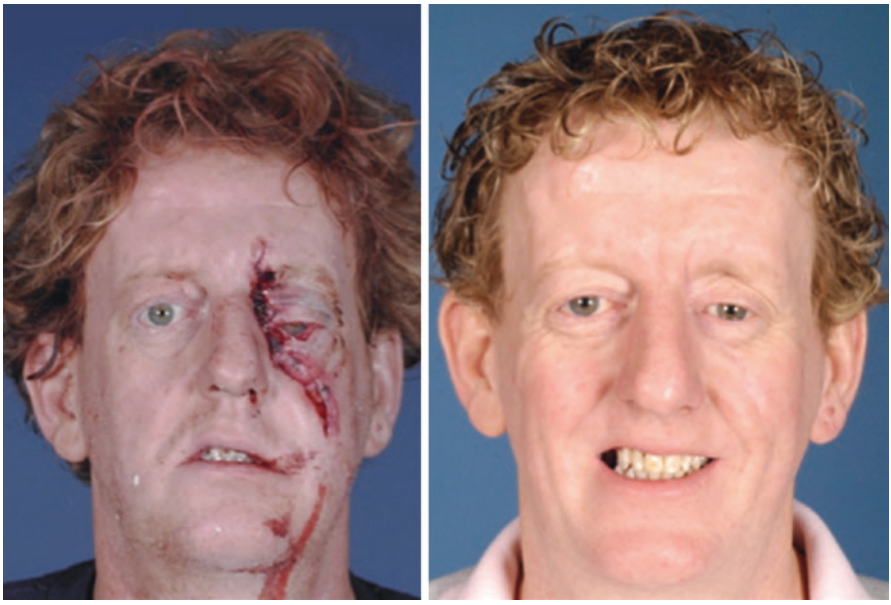


Fig. 27.26 Severe lacrimal trauma requires urgent and careful assessment

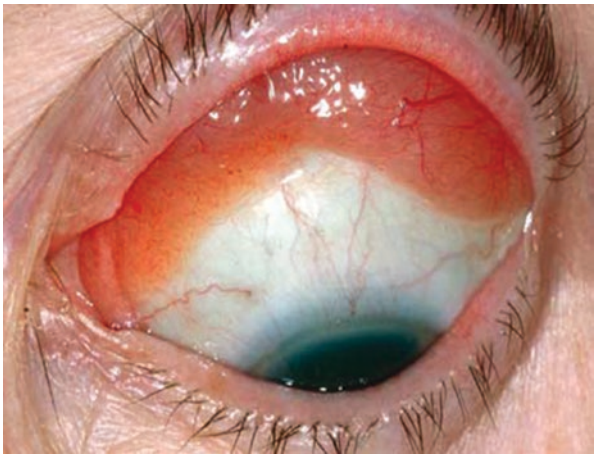
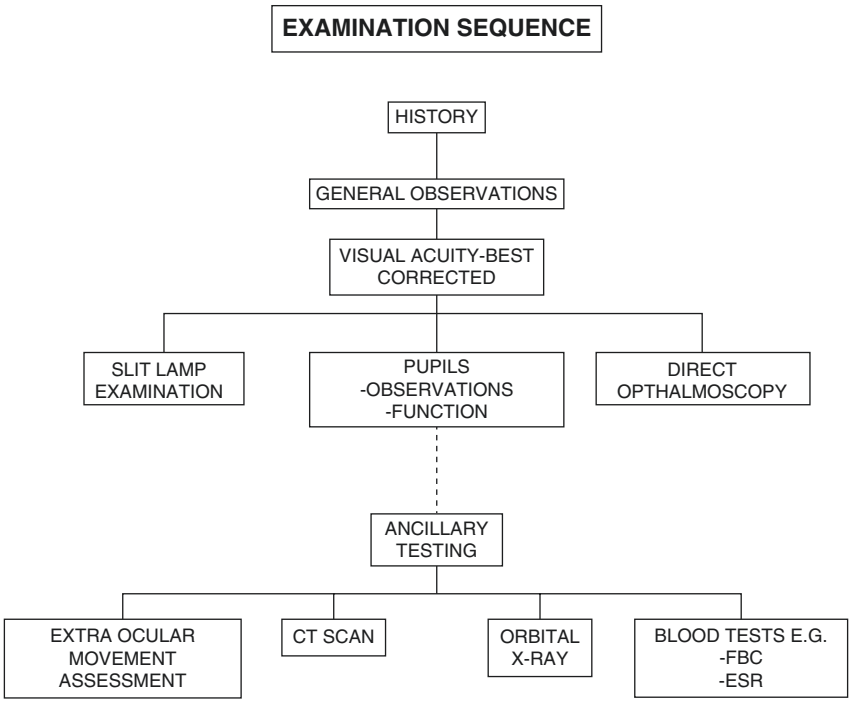


Fig. 27.27 Conjunctival lymphoma

should both appear clear, with an obvious red reflex. Clouding of the cornea may be either acute or longstanding (such as acute infection or an old scar). Clouding of the lens is usually from a cataract. In the normal eye the anterior chamber should be clear, with an appreciable depth. If the anterior chamber looks shallow or flat following an injury, globe rupture should be suspected. The presence of blood (hyphaema) or pus (hypopyon) should also be noted (Fig. 27.27).



27.7.2 Examination of the Eyelids

These should be examined with good illumination. The tissues surrounding the eyes are easily distended by oedema, suggesting injury, inflammation, cardiac or renal failure. Look first at the position of rest and the relative heights of the two eyelids, then the surface and edge (skin and lashes). Both eye lids should cover approximately the same amount of eyeball. If the difference in height between the two palpebral fissures is more than 2 mm, there may be pathology. Less than this is common and may represent normal mild asymmetry. If a difference is seen, try to determine which eyelid is abnormal (upper or lower). If the sclera above the limbus in the 12 o'clock position is visible, the upper eyelid is abnormally high (lid retraction). If the upper eyelid covers the pupil, it is abnormally low (ptosis). Lid signs of hyperthyroidism include retraction of the upper lids, giving the patient a wide-eyed, staring appearance. Lid lag is where the upper lid does not follow the eyeball exactly when the eye looks down. A drooping eyelid, called ptosis, suggests a third nerve paralysis, Homer's syndrome, or myasthenia gravis. Or it may be congenital.

Normally, the lowermost part of the iris is covered by the lower lid. No sclera should be visible. The conjunctiva lining the inner eyelid of the lower lid may be inspected by gently pulling down the lid with a finger. The conjunctiva lining the upper lid can only be observed by everting the upper lid. This may be necessary if a

superficial foreign body is suspected. Foreign bodies can often pass up under the upper eyelid and become lodged in the upper fornix. These can easily be overlooked if the upper eyelid is not everted. However never evert the upper eye lid if a penetrating injury or corneal thinning (from ulceration) is suspected. To evert an upper eyelid:

1. Instil a drop of local anaesthetic and fluorescein dye
2. Ask the patient to look down
3. With one hand, hold the eyelashes of the upper eyelid between thumb and index finger
4. With the other hand, place a cotton bud (or paper clip or other small blunt elongated object) horizontally along the lid, midway from its margin
5. Evert the eyelid over the cotton bud
6. If a foreign body is seen, gently remove it with a moistened cotton bud.
7. When finished, ask the patient to look up and the eyelid will flip back in to its normal position (Fig. 27.28)

Look also at the lower lid. Note if the edge is turned inwards (entropion) or outwards (ectropion) and any signs of inflammation. Assess the lid margins for redness,

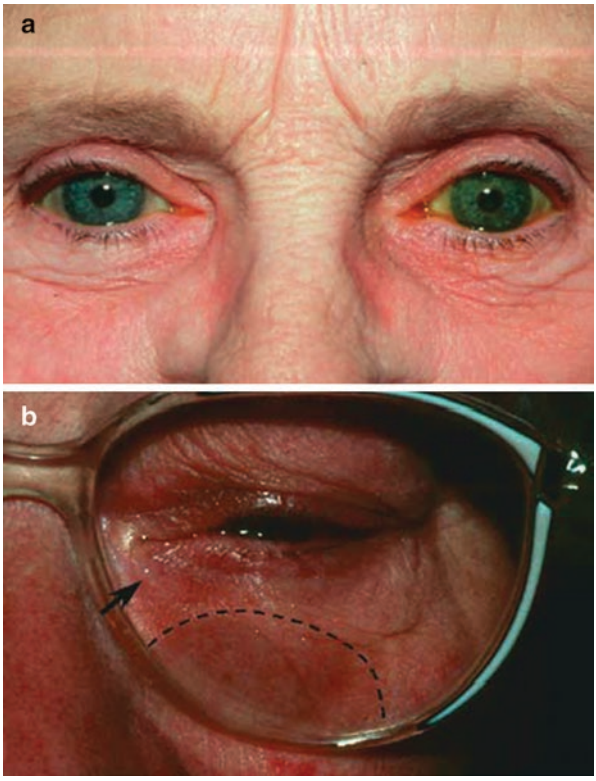


Fig. 27.28 (**a, b**) "Flow" characteristics of lacrimal drainage: (**a**) Excessive tear-lake and slow clearance of fluorescein dye in both eyes, especially the left side. (**b**) Medial spillage of tears (arrow) from excessive tear lake, with pooling between spectacle lens and cheek (outlined)

irritation and scaling—blepharitis. The lacrimal apparatus is also checked by observing for excess dryness or tearing. Gently pressing the lacrimal sac at the medial corner of the lower lid and nose will normally not express any discharge. The lid margins are often subject to localised bacterial infections (styes). These common lesions appear as classic red, swollen, tender lumps with a pussy discharge. A lump in the lid without signs of acute inflammation is likely a chalazion. Tumours of the lid margin are uncommon but do occur. A discrete waxy yellowish deposit in the medial aspect of the lid is an xanthelasma. This suggests the possibility of abnormal blood lipids.

Following injury, both eyelids should be carefully examined for lacerations and other signs of penetrating injury. Always consider also the possibility of a penetrating orbital and brain injuries—these can often be obscured by a trivial looking wound. A retained foreign body may also be present. This will necessitate the need for imaging (CT) and at the same time contraindicate MRI. When attempting to open a swollen eyelid, it is important not to press on the eye itself. If so, this can cause or exacerbate any globe injury. The direction of applied force should be upwards and downwards on the respective lids, onto the (hopefully) intact orbital rims (Fig. 27.29).

27.7.3 Visual Acuity

In the United States, normal visual acuity is designated as 20/20. This indicates that the person can see at 20 ft away the same as the average person at the same distance. In Europe the standard testing distance is usually 6 m, so normal vision is depicted as 6/6. Decreased visual acuity of 20/30, 20/40 etc. indicates that the patient can see at 20 ft what an average person can see at 30 or 40 ft, respectively. Conversely someone with 20/10 vision can see at 20 ft what the average person must be 10 ft away from to see. When measuring the visual acuity the following steps are important:

1. Test visual acuity in each eye separately.

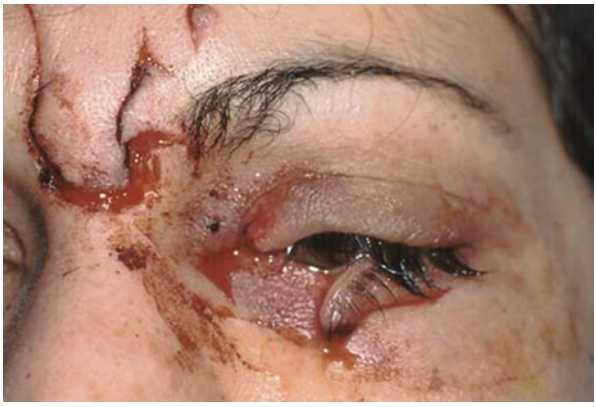


Fig. 27.29 A 46-year-old patient with large glabellar laceration, an avulsed laceration of the upper and lower eyelids, and extensive laceration of the distal canaliculi and common canalicular structures. Establishment of the integrity of the globe is the primary responsibility of the surgeon prior to reconstructing the eyelids and the lacrimal system

2. Topical anaesthetics may help if the patient has pain or blepharospasm.
3. Use a Snellen Chart or Sheridan-Gardner Chart (e.g. for children who can't read) at 6 m, with the patient wearing their spectacles or contact lenses (if normally used for distant vision). If these are unavailable, small or moderate refractive errors are overcome with the use of a "pinhole". A patient who normally wears distance spectacles and can achieve 6/24 on the Snellen chart should be able to manage 6/9 or better through a pinhole if no serious pathology is present.
4. Record the vision as a fraction. The numerator (first or upper number) is usually 6 (test performed at 6 m) and the denominator (second or lower number) corresponds to the line on the chart they could read. If the patient can only read the top line with the biggest letter (60 line), the vision is recorded as 6/60. This means that the patient was tested at 6 m and could read the line a normal person should have managed to read at 60 m.
5. Ask the patient to work their way down the chart reading successively smaller lines.
6. Record the smallest print size that the patients can clearly read
7. If the patient fails to read the top line, try to ascertain if the patient can count fingers at 1 m (counting finger vision). Failing that, ascertain if the patient can see movements of the hand in front of the eye (hand movement vision). Failing that, test the vision with a light source (light perception or no light perception vision).
8. If the Snellen Chart is not available, a mini Snellen chart can be used with the patient wearing their reading glasses.
9. I-phone apps are now available with these charts, corrected in size accordingly to be read at arms length.

The table demonstrates visual acuity measurements (from poorest vision to best). This is measured in metres, i.e. a normal person would be able to see the 6/60 letter at 60 m away, where as a patient with 6/60 vision can only read that letter from a distance of 6 m.

No Light Perception (NLP)	
Light Perception (LP)	
Hand Movements (HM)	
Counting Fingers (CF)	
6/60	
6/36	
6/24	
6/18	
6/12	Driving standard[a]
6/9	
6/6	Normal visual acuity
6/5	

[a]In the United Kingdom

Although visual acuity is probably the best single test of overall ocular integrity and function, falsely reassuring or falsely alarming results can occur. Eyelid or orbital swelling can sometimes distort and deform the eye. Together with any watering of the eye this can temporarily affect the patient's vision, resulting in a lower recorded acuity. By way of contrast, serious ocular injuries (including penetrating or peripheral retinal tears), may have no obvious effect on central visual acuity initially.

Although accurate testing is very difficult in the emergency room, it should still be attempted. This is important to establish a baseline, act as a means of communication and documentation, and for medicolegal purposes. Poor results commonly arise from lack of spectacle correction, blood or mucus in the tear film, and poor compliance due to anxiety, pain or intoxication. Despite such limitations an eye that can not see light at all is a clear indication of severe ophthalmic problem (or malingering).

In the cooperative patient, one of the most sensitive tests of optic nerve integrity is red colour saturation. Formal colour vision testing makes use of Ishihara pseudoisochromatic plates. However, if these are not available a bright red object such as a container top is shown to each eye in turn. The saturation in one eye is compared with the other. The patient is asked whether the object's colour is of equal hue and intensity in each eye. The red object may appear more dull, orange, or brown than it does with the (normal) contralateral eye. Even patients with mild colour blindness should be able to tell a difference. Test each eye separately. Colour vision defects are common in men, affecting 1 in 8 males (most commonly confusing reds and greens). Loss of colour perception can be a manifestation of optic nerve pathology such as optic nerve compression. This simple test can provide useful evidence if there is a suspicion of a relative afferent pupillary defect. If an optic nerve has suffered significant injury or is diseased, colour perception will be altered in the corresponding eye.

27.7.3.1 Visual Field Determination

Visual field testing in an awake and cooperative patient can be very useful if it is done correctly. There are three parts to the assessment.

1. Central visual field assessment. This evaluates the overall central retinal (macular) function. There are several formal methods, but a simple method is to sit a few feet from the patient, cover each of their eyes in turn and ask the patient to focus on your nose. While doing so, the patient should be able to see all the features of your face, including your ears, without any dark or blurry spots.
2. Peripheral visual field assessment. To do this, sit 2–3 ft in front of the patient. Both you and the patient cover opposing eyes (if the patient covers the right eye, you cover your left). With the eye covered use your other hand (which must be equidistant between yours and the patient's heads), to bring a wiggling finger or hat pin, in from the far periphery. Have the patient indicate when the moving finger is first visible. You and the patient should see the finger at approximately the same time (assuming your visual fields are normal of course). Test each of the four quadrants in turn

3. Double simultaneous confrontation.This is part of a complex neurologic evaluation and not usually required in the emergency setting. Visual neglect, a defect in central processing, may not be apparent using the peripheral visual field test. Double simultaneous confrontation is performed with both eyes open. The examiner presents stimuli in both the right and left visual fields at the same time. A patient who has neglect, may be able to identify a stimulus when each quadrant is tested independently, but when two stimuli are presented together will only be able to identify one. The other may appear blurry or not be seen at all.

27.7.3.2 Pupillary Assessment

Pupil size per se, has no direct relationship to optic nerve or visual function. Its size is determined by the balance of muscle stimulation between the sympathetic and parasympathetic fibres. Vision is dependent on the optic nerve. Therefore a completely blind eye can still have a normal sized pupil, and an eye with a dilated pupil may have normal vision. It is also important to remember that a coexisting head injury as well as many medications can affect the size of the pupil(s). There are three elements to pupil examination.

1. The size, shape and symmetry of the pupils should be recorded. 1 mm or more difference may be normal. An irregularly shaped or non-concentric pupil (corectopia), should raise concern about an injury to the anterior chamber. Teardrop-shaped pupils are highly suggestive of a penetrating injury. The point of the teardrop often points toward a small laceration, this is where part of the iris has herniated through the wound. The herniation then seals the wound, preventing further deflation of the globe.
2. The pupillary reaction to a bright light should then be assessed. Each pupil should be tested independently. Whilst terms like "sluggish" and "slow" are often used, they are unreliable, cannot be quantified and if used incorrectly can mislead. Following this, the light is swung from one eye to the other and back again, with delay of 1 s, to determine whether there is an afferent pupillary defect (APD, or Marcus Gunn pupil). This is best conducted in a semi-darkened room. In a normal reaction to this "swinging flashlight" or Relative Afferent Pupil Defect (RAPD) test, both pupils constrict when one is exposed to light. As the light is being moved from one eye to the other, both pupils begin to dilate, but constrict again when light stimulates the other eye. An afferent pupil defect (APD) is present if light stimulation in one eye fails to constrict either pupil (that is, no stimulus is reaching the midbrain). However, if one of the pupils constricts, then it is likely that a pupil defect (sphincter muscle tear or dilating drop in the eye) or efferent pathway defect is responsible for the abnormality.
3. The pupils are then assessed during convergence, looking from far to near. As an object gets closer to the patient's face they should constrict. Although of little value when examining the injured eye, this is important when assessing non-traumatic presentations.

Results are then documented—for example, "pupils: 4 mm, round, briskly and equally reactive to light."

27.7.3.3 Assessing Eye Movements

Patients should be able to painlessly move their eyes in all directions. When assessing eye movements move the object slowly—otherwise subtle restriction may be overlooked. Limited motility has many causes (notably fractures with muscle injury or entrapment, cranial nerve palsy, infections, tumours and in thyroid eye disease). Assess whether both eyes can work together while the patient is facing straight ahead (in the position of primary gaze). Ask the patient to look straight ahead and then follow a target to the eight positions of gaze. Ask if there is any double vision or discomfort during movement. At the same time look for any obvious restriction. If diplopia is noted it is important to determine whether it is monocular or binocular. Most problems are identified on looking up, down, left, or right. Children younger than 6 years are at risk for developing amblyopia if they have prolonged restriction and can not use their eyes together. Obvious restriction in upward movement of the globe is a useful sign for orbital floor entrapment, however diplopia on downward gaze can be associated with dysfunction of the inferior rectus from simple bruising. This is discussed further in the chapter on the cheek and orbit.

Examination of the Fundus

Examination of the retina, optic nerve, and retinal vessels can be difficult, especially through an undilated pupil. Dilating the pupil will greatly facilitate examination, but this may be contraindicated following trauma. If allowed, it is important to note this in the hospital records and advise the staff looking after the patient. Points to note when looking at the fundus include:

1. A poor red reflex indicates opacity in the intervening media (cataract, vitreous haemorrhage).
2. Leukocoria—a white reflex noted on funduscopic examination. It can be the result of intraocular tumors, detachments, or other significant pathology (se below)
3. If there is a good red reflex but no view of the retina, the posterior chamber may be filled with blood.
4. A large white or pale area in an otherwise red retina may indicate retinal ischaemia.
5. Retinal detachment may appear as large irregular, pale folds. If some parts of the retina appear to be out of focus compared to others, this suggests the retina is at different levels.
6. A swollen disc or papilloedema (Fig. 27.30)

Fundoscopy may therefore detect intraocular haemorrhage, retinal oedema/detachment and avulsion or swelling of the optic disc. However following trauma, normal appearances on fundoscopy can be misleading. This is because the optic nerve takes time to atrophy. If fundoscopy is not possible the red reflex should at least be checked and compared between each eye. Follow the red reflex and look at the disc (to find the disc follow the branching vessels towards the back of the eye).

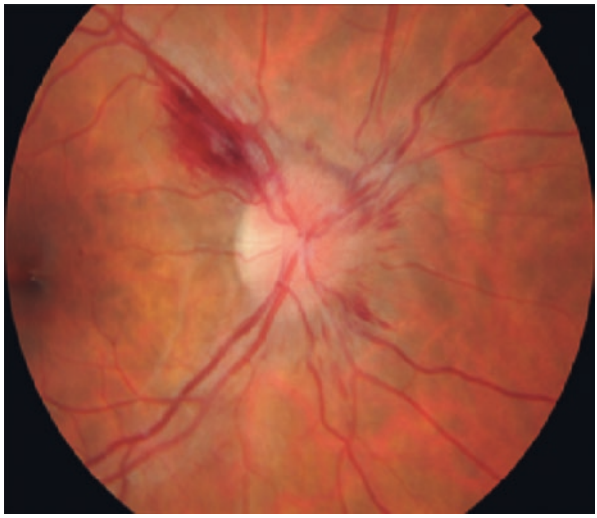


Fig. 27.30 Optic disc swelling

Assess the disc margins (distinct or blurred), rim colour (pale or hyperaemic), cup (full or empty) and its blood vessels (congested, pulsating or attenuated). Papilloedema is suspected if the disc margin is blurred, the disc appears hyperaemic, its cup is full, and the vessels are congested. Because the optic nerve is enclosed within the meninges, like the spinal cord, it is surrounded by CSF within the subarachnoid space. Any increase in CSF pressure (from raised intracranial pressure) will thus impede venous return from retina. The retinal vessels themselves are also covered by the pia mater and lie in the subarachnoid space. Fluid will therefore accumulate in the optic disc resulting in swelling—papilloedema.

Leukocoria

This is a white reflex noted which is immediately apparent on funduscopic examination. Diseases causing leukocoria include retinoblastoma, a number of non-neoplastic diseases and total retinal detachment. Imaging is usually required to distinguish between a tumour and non-neoplastic diseases. Non-neoplastic conditions causing leukocoria include Coat's disease (exudative retinitis of unknown aetiology), persistent hyperplastic primary vitreous, retinopathy of prematurity, Norrie disease, Warburg syndrome and endophthalmitis secondary to toxocara infection. Some of these condition are discussed later.

27.7.3.4 Slit-Lamp Examination

The slit lamp is a binocular microscope that provides a magnified 3D view of the ocular contents. In experienced hands it can identify many serious diseases and injuries including (1) macular degeneration, (2) detached retina, (3) cataracts, (4) injury to the cornea, (5) retinal vessel diseases. It is therefore an essential piece of equipment in most ophthalmic departments. Examination should proceed systematically, from anterior to posterior. First, take a quick second to observe the patient

as a whole for iris heterochromia and eyelid neoplasms. Begin with the external features and work towards deeper structures.

1. Eyelids—Starting with the lids closed, the lid margins and lashes should be examined lacerations, tumours, marginal blepharitis and signs of infection. Next, the patient should be asked to open his or her eyes, and the lid margin be examined for patency of the tear ducts and meibomian glands.
2. Conjunctiva—Ask the patient to look in the four directions of gaze (up, down, left, right) to enable a view of as much bulbar conjunctiva and sclera as possible; hold eyelids up or down to get an unobstructed view. Look for subconjunctival haemorrhage or vesicles and the possible presence of a pingueculum or pterygium. If blood is noted in the tear film following trauma, but there is no lid laceration, then a conjunctival laceration (or deeper injury) must have occurred. Consider a serious globe injury (Fig. 27.31).
3. Cornea—assess the cornea for gross opacification or central corneal clouding produced by hard lens wear. Look around the limbus, at the limbal vasculature and assess the degree of corneal vascularisation noting any neovascularisation. Look for arcus senilis, foreign bodies, microcysts, stromal striae and folds in the endothelium. Note any keratic precipitates (KP's) and prolapsed tissues (iris or uveal tissue. These normally appear dark). Add fluorescein and reassess.
4. Anterior chamber—In the normal eye the anterior chamber is 'dark' because it is optically empty. However, in some conditions, such as in anterior uveitis, inflammatory cells and proteins, can migrate into the chamber. These can be then be viewed. The scatter of light from these particles, similar to dust seen in sunlight, is termed the Tyndall effect. Blood and inflammatory debris can settle, forming a fluid level (hyphema and hypopyon, respectively) (Fig. 27.32).
5. Iris—The integrity of the iris may also be examined using retroillumunation. This is an indirect illumination technique that utilises light reflected from posterior surfaces to view the area of interest. The iris should be round

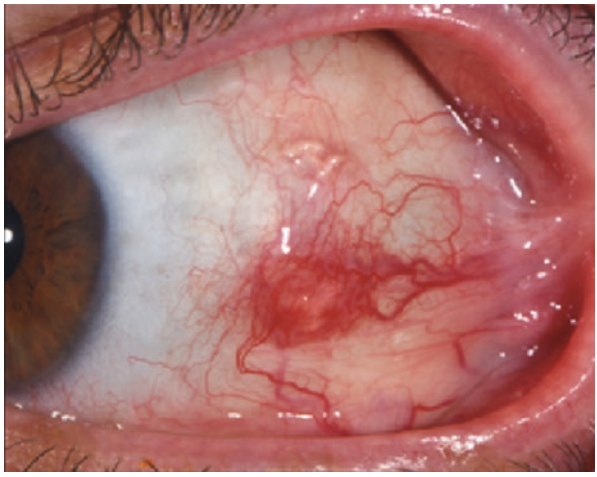


Fig. 27.31 Suture granuloma following squint surgery

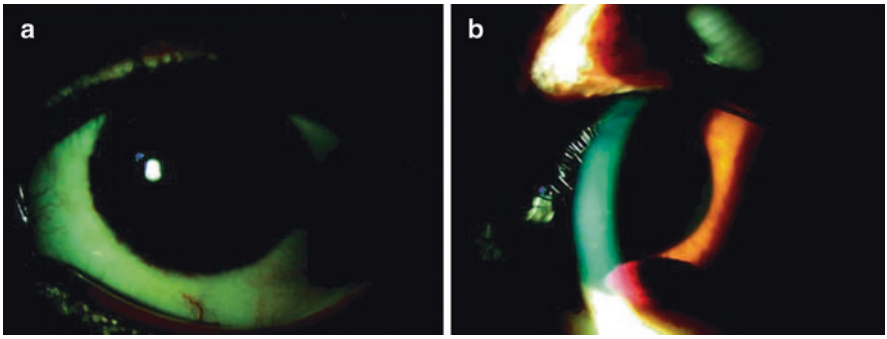


Fig. 27.32 (**a**) Anterior segment photo showing hyphema inferiorly in the anterior chamber of a sickle cell trait patient, (**b**) Slit lamp magnified view of hyphema settling inferiorly in anterior chamber. Kings County Hospital Center, Brooklyn, NY

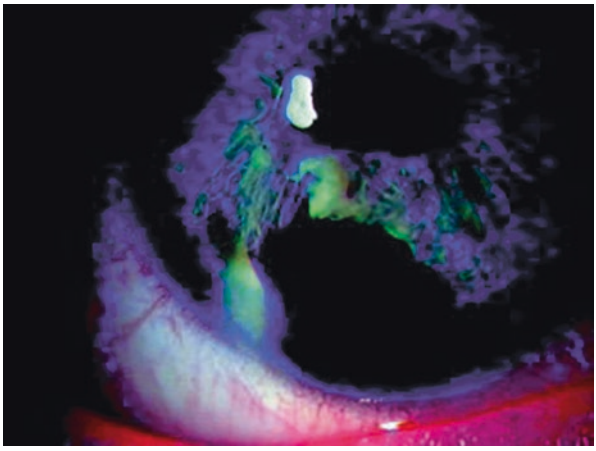


Fig. 27.33 Anterior segment photo showing inferior iridodialysis after injury with a projectile nail gun to the eye. (Photo courtesy of Minas Coroneo, MD. University of New South Wales, Sydney, Australia

and reactive to light. Any irregularity in its shape should be considered as abnormal. Look for defects in the periphery (a dialysis). This indicates a significant injury resulting in avulsion of the iris from the ciliary body (Fig. 27.33).

6. Lens—this should be clear. An opacity in the lens is known as a cataract. Other opacities can be inspected with the use of the retro-illumination technique. Posterior capsular opacification, a complication of cataract surgery can be viewed in this way, with reflection of light from the red fundus.
7. The anterior vitreous can be viewed just behind the lens. Ask the patient to look up, down and then straight ahead. This causes movement of the vitreous and may reveal floaters. It can also reveal debris released by the retinal pigment epithelium after a retinal break (Schaffer's sign), indicating the presence of a retinal tear

8. Fundoscopy—The slit lamp may also be used to view the retina. This technique uses a high-power positive lens, held in front of the patient's eye.

Staphyloma is a localised bulging of a weakened and thin cornea or sclera, which is lined by uveal tissue. This is usually visible through the thinned outer fibrous coat. Anatomically, staphyloma can be divided into anterior, intercalary, ciliary, equatorial and posterior staphyloma. Ectasia of the weakened scar tissue can occur during healing of a perforating injury, corneal ulcer progression of secondary angle closure glaucoma, scleritis or pathological myopia. Some staphyloma can be resected (Figs. 27.34 and 27.35).

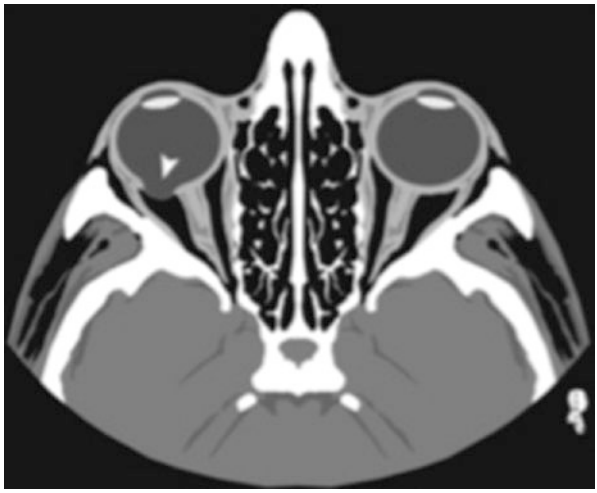


Fig. 27.34 Axial orbital CT illustration shows thinning of the scleral-uveal region of the right globe, representing a classic sign of staphyloma (arrowhead)

Fig. 27.35 Handheld slit lamp biomicroscopy

27.7.4 Examining Children

Assessing a non cooperative child, most likely in distress can be very difficult and requires a lot of patience. Try to assess with the parent holding the child instead of alone as this may make it easier. Any toys or other distractions commonly available on a paediatric ward may also help. Useful steps include:

1. Obtain a detailed history from an adult witness. If this is not available, always suspect an injury being the cause of a red or painful eye.
2. Assess the visual acuity—fixing and following objects of interest, reaching out for objects of interest, '100s and 1000s', Sheridan-Gardner test depending on the age and verbal ability of the child. Test each eye in turn if possible.
3. General observation e.g. periorbital redness or bruising
4. Test pupil responses.
5. Test for red reflexes.
6. If a globe injury is suspected, do not try to pry the eyelids open as this can exacerbate a perforating eye injury.
7. If periorbital bruising is present, especially if associated with other injuries, suspect non-accidental injury as a possible cause.
8. If the eyelid is red, tender and swollen, especially if the child is febrile, suspect periorbital cellulitis.
9. Purulent discharge in a baby in the first month of life, is suspicious of ophthalmia neonatorum. This should be investigated to rule out gonorrhoea or chlamydial infection.
10. If a white pupil or leukokoria is present (absent red reflex), consider congenital cataract and retinal abnormalities (retinoblastoma, Coat's disease, infections).
11. A white blow-out fracture should be suspected if there are signs of a sunken globe, minimal periorbital haemorrhage, restricted eye movements, disproportionate pain and an irritable child.
12. If an eyelid laceration is present, always consider the possibility of a penetrating injury however small the size of the laceration e.g. a toddler falling on a pencil and penetrating the orbit (Fig. 27.36).

Care is required when assessing the eyes in the elderly. In some patients a dilated pupil itself may precipitate ocular problems. Acute angle closure glaucoma can be precipitated by dim light, and some drugs. This should be considered in any elderly patient who develops a painful, tense, "red eye."

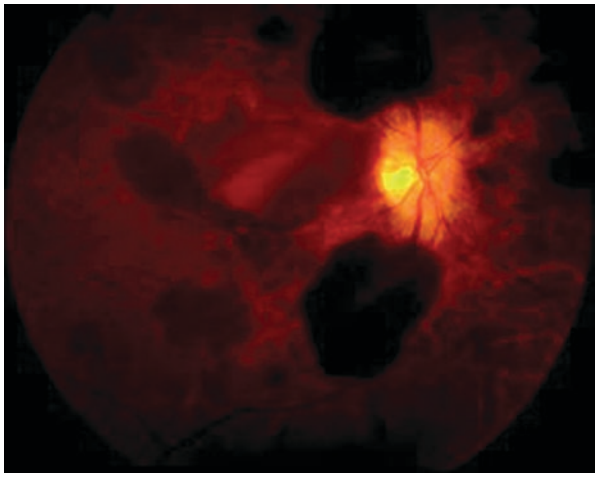


Fig. 27.36 Shaken-baby syndrome is depicted. Hemorrhages are seen at all layers of the retina. Image is courtesy of the ASRS image bank

27.8 Investigating Symptoms and Signs

Not all the following investigations are required acutely. Nevertheless they are noted here for completeness.

27.8.1 Laboratory Tests

ESR/CRP should be measured in all inflammatory conditions, notably suspected temporal arteritis. Eye swabs should be taken in suspected chlamydial or other unusual infections. Conjunctival scrapes can be useful in detecting chlamydial and fungal infections, allergy, and dysplasia, but are rarely done. Tear protein analysis measures the lysozyme contained within tears (which makes up approximately 20–40% of the total protein content).

27.8.2 Seidel Test

10% fluorescein (which appears dark orange) is applied to an injured eye, asking the patient not to blink. It is then viewed with a slit lamp using a cobalt blue light source, or Wood's light. If aqueous fluid is leaking through a corneal laceration, this will be seen running through the dye. This finding is indicative of an open globe injury. However, a negative Seidel's test does not exclude a penetrating injury—this may still occur with small or spontaneously sealing lacerations.

27.8.3 Van Herick Test

This test allows a quick, non-invasive assessment of the anterior angle. It is performed on the slit lamp. A narrow slit of light is projected onto the peripheral cornea at an angle of 60° as near as possible to the limbus. This results in a slit image on the surface of the cornea the width of which is compared with the peripheral anterior chamber depth ("black space"). A four-point scale is used to grade the probability of angle closure.

27.8.4 Smith Test

This is another optical test used to determine the anterior chamber depth using a slit-lamp. The lamp is located in the subject's temporal field at an angle of 60° and the slit-beam is projected horizontally.

27.8.5 Gonioscopy

Gonioscopy still represents the gold standard in the assessment of the anterior chamber angle. Direct visualisation of the chamber angle is undertaken using a contact lens, or mirrors and prisms to reflect light from the angle to the viewer.

27.8.6 Schirmer's Test

This measures the amount of fluid bathing the eye. This test is useful for determining the severity of dry eyes.

27.8.7 Tear Breakup Time (TBUT) Test

This measures the time it takes for tears to break up in the eye. It is determined after placing a drop of fluorescein in the fornix.

27.8.8 Visual Evoked Potential (VEP)

This tests the integrity of the visual pathway. Electrodes are placed on the patients scalp. The patient then looks at various targets and the electrical activity produced is recorded. This test may be helpful in the diagnosis of disorders that affect the optic nerve (for example MS). Flash VEP testing (VEP's recorded in response to a

flashing light source) has been shown to be correlate to visual acuity outcome in patient with traumatic optic neuropathy. This can be useful in the unconscious patient.

27.8.9 Optical Coherence Tomography (OCT) Scanning

This is a non-invasive method of viewing the retina in cross-section. Patients effectively have a cross sectional photograph taken of the retina with a camera using laser rather than visible light. OCT assesses the reflectivity of the different layers of the retina (including the nerve fibre layer) to a specific wavelength of laser light.

27.8.10 Miscellaneous Tests

- Keratometry
- Cycloplegic refraction
- Amplitude of accommodation
- Corneal topography
- Corneal pachymetry
- Scheimpflug ocular imaging
- Retinal tomography
- Scanning laser polarimetry
- Electrooculography
- Electroretinography
- Ultrasound biomicroscopy

27.8.11 Measuring Intraocular Pressure (IOP)

Elevated IOP can result from a variety of conditions, including hyphema, glaucoma, retrobulbar haemorrhage, tumours, thyroid eye disease or carotid-cavernous fistula. Decreased IOP can result from open-globe injury, uveitis, cyclodialysis (separation of the ciliary body from the sclera), or retinal detachment. Determining the IOP is therefore an important part of the diagnosis and helps triage patients requiring urgent treatment. A number of techniques are available for measuring intraocular pressure (IOP), but require specialist training and expertise. Portable tonometry can be used in patients with suspected glaucoma and following trauma. Handheld devices are now available but can be unreliable if not used correctly. They require a cooperative patient and administration of local anaesthetic. If a penetrating injury or laceration is suspected, the eye are should be protected and pressure measurements should only be performed by an ophthalmologist (Fig. 27.37).

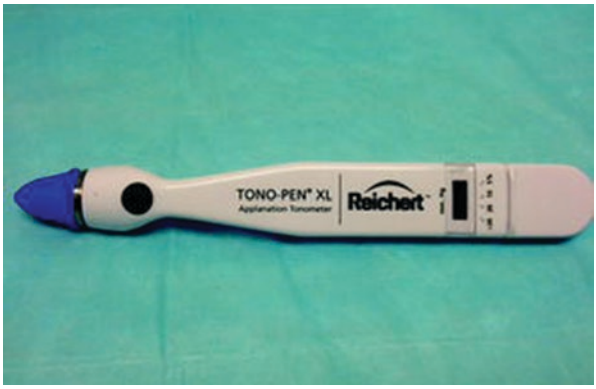


Fig. 27.37 Handheld applanation tonometer with 1 mm tip

27.9 Imaging of the Eye

27.9.1 Plain Xrays

Although plain orbital X-rays may reveal fractures and retained foreign bodies, CT scan is today the investigation of choice if the history suggests the presence of either. In some congenital ophthalmic conditions the bony orbit may appear abnormally small. Alternatively the orbit can appear to be paradoxically large if there is an associated mass or cyst within it. Associated facial dysplasia, clefts and other malformations may be seen.

Dacryocystography may be performed in patients with suspected nasolacrimal duct obstruction, usually presenting with epiphora after imaging has shown no obvious obstructing mass lesions. This technique can be used to determine the precise level of obstruction. It can also demonstrate mucosal lesions and concretions or a fungus balls within the ductal system. These studies are useful in the assessment of the chronic tearing eye.

27.9.2 CT/MRI Scanning

CT scan is commonly requested in the assessment of complex trauma and foreign body localisation. This may be supplemented with ultrasound, magnetic resonance imaging and fluorescein angiography. Lens dislocation, ocular hypotonia from a penetrating injury, intraocular gas, and haemorrhage may all be visible on both CT and MRI) (Figs. 27.38, 27.39, and 27.40).

However as a first line investigation, CT remains the single most useful imaging study. 3-mm slices are usually sufficient, unless a small foreign body is being sought or 3D model fabrication is required. Images should be viewed in both the axial and

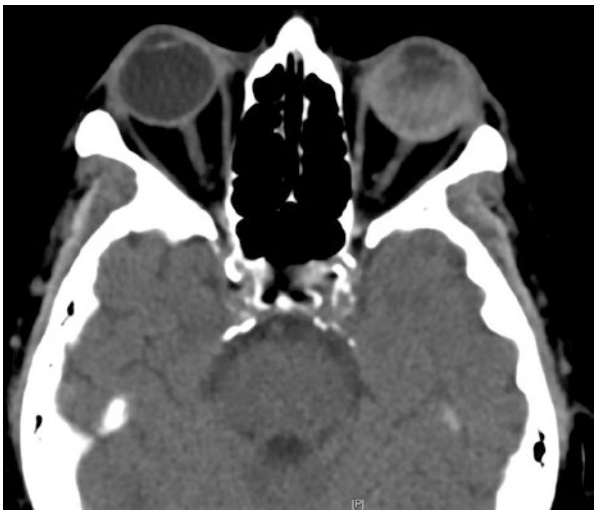


Fig. 27.38 Intraocular blood

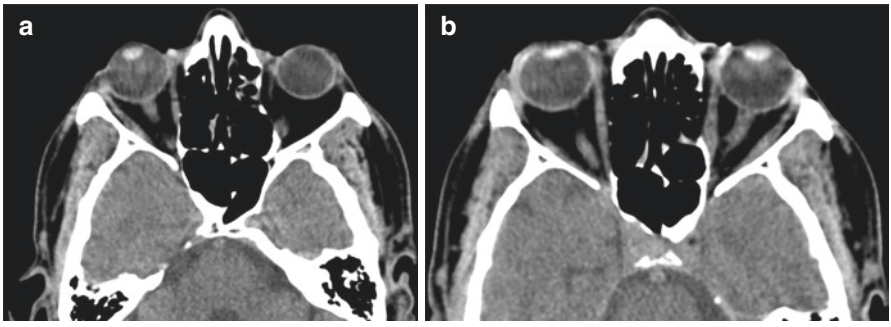


Fig. 27.39 CT orbits/globes

coronal planes and sagittal plane if available. CT is also preferred for inflammatory disease and in detecting subtle optic sheath lesions, because of its clearer depiction of fluid collections, the nature of any surrounding inflammatory reaction, and bone detail. However today, contrast-enhanced and plain CT offer limited advantages over MRI for most mass lesions and therefore MRI may be preferred. MRI is particularly useful in the assessment of the globe, optic nerve and in the assessment of orbital swellings/masses. It is often the preferred choice for imaging the cavernous sinus and orbital apex. Most of the individual cranial nerves related to the cavernous sinus are visible on high resolution studies. Suspected disorders of the optic pathways should also be studied with MRI. This will assess the entire visual pathway, including the optic nerve, chiasm, and tracts as well as the lateral geniculate body,

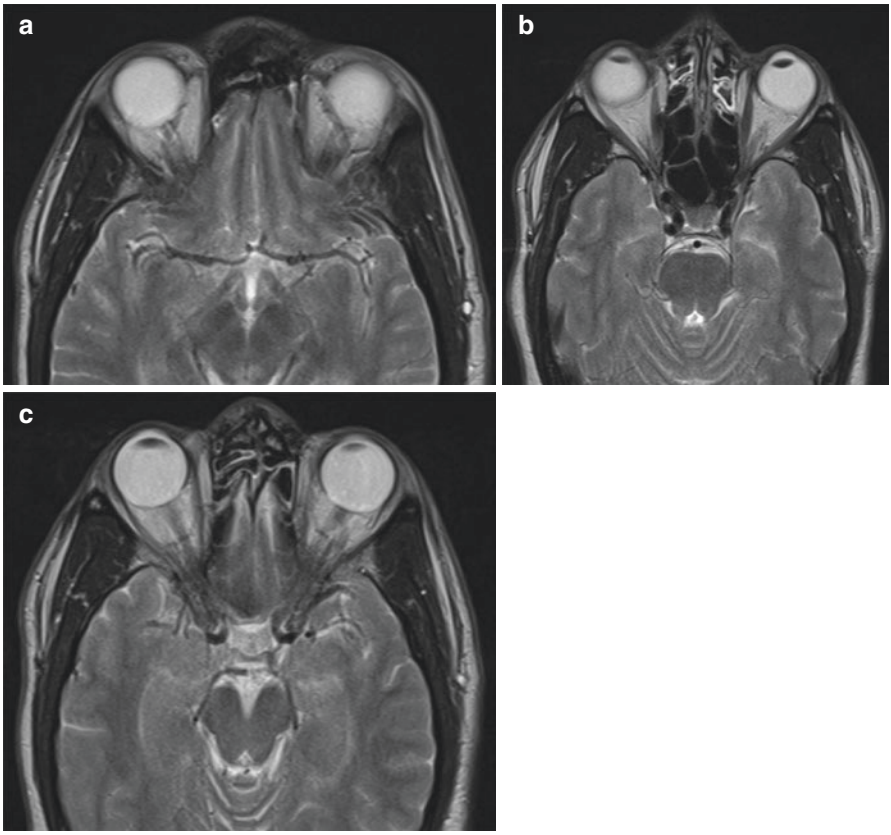


Fig. 27.40 (**a**–**c**) MRI is very useful in evaluating globe/orbital pathology

optic radiations, visual cortex, cavernous sinus, and brain stem. MRI is contraindicated if a metallic foreign body is suspected. It is also more difficult to obtain quickly, and does not show bone as well. Imaging of the eye and orbit is also susceptible to motion artefacts. In some centres MRI has become the modality of choice for much of the imaging of the orbit and visual pathways, in a non-traumatic context. However it lacks sensitivity to calcification, bone detail and can experience difficulties with motion degradation compared to CT. MRI scans in paediatric cases may require a general anaesthetic. Scanning before and after a Valsalva manoeuvre and with the orbit in different dependent positions, or with venous compression may be useful in rare cases of suspected orbital varices. MRA may be used to define pathologies related to the ophthalmic artery, the cavernous portion of the carotid artery and its cerebral branches.

27.9.3 Ultrasound (US)

The superficial location and cystic structure of the eye make ultrasound (US) an ideal modality for imaging, but requires specialist equipment. Since the late 1980s, colour flow Doppler techniques have further increased the utility of ultrasound as a primary diagnostic tool. However currently, ultrasound is mostly used by ophthalmologists. Few diagnostic radiologists perform these, particularly acutely.

Nevertheless, ocular B-mode ultrasonography can often detect intra ocular foreign bodies (IOFB), retinal detachment and determine globe integrity. Lens dislocation, ocular hypotonia from penetrating injury, intraocular gas and haemorrhage may all be visible on US. Penetrating objects can result in residual foreign bodies remaining embedded in the sclera, anterior segment or the vitreous body. In general, these will show up as echogenic focus with some degree of reflectivity of the sound.

Intraocular gas can also mimic foreign body, therefore it is very important to know the history.

US is the quickest and simplest method of imaging the eye, but requires specialist training. B-mode US may be useful when ophthalmoscopy is not possible (due to intra ocular opacifications). It is also particularly useful in the detection of retinal, vitreous, and choroidal detachments, tumours (e.g. choroidal melanoma, metastases, haemangioma) and other pathologies in the posterior segment of the eye. Scanning in real time, provides better evaluation of the vitreous, retina and choroid. Calcification is the hallmark of retinoblastoma. These may be seen within the tumour, as opposed to ocular wall calcification, which can occur in other conditions that mimic retinoblastoma (causing leukocoria) (Fig. 27.41).

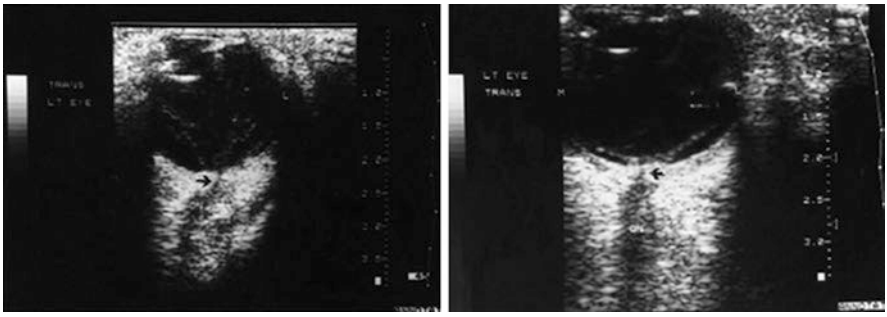


Fig. 27.41 B-scans demonstrating hypolucencies posterior to optic nerve head (arrows). From Sawhney et al. [54]. Reproduced with permission from Nature Publishing Group

27.10 Tools to Study Ocular Blood Flow

There are a number of tools that can provide information regarding ocular blood flow (OBF) in and around the eye. These include:

1. Colour Doppler imaging (CDI)—a non-invasive ultrasound-based technique that uses the Doppler effect to measure blood velocities. This can provide information on the ophthalmic artery, short posterior ciliary arteries and the central retinal artery.
2. Laser Doppler flowometry (LDF)—uses a fundus camera and the Doppler effect to measure retinal capillary blood flow.
3. Doppler optical coherence tomography (OCT)—this uses the Doppler frequency shift to determine the velocity of the blood inside the major retinal vessels and the cross-sectional diameter of these vessels throughout the cardiac cycle.
4. The retinal vessel analyser (RVA)—uses a fundus camera and analysis software. It allows for real-time measurements of retinal vessel diameter.
5. Retinal oximetry is a non-invasive method for assessing the haemoglobin oxygen saturation in the retinal vessels. It is used to study the metabolic needs of the retina and its ability to react to stimuli.
6. Dynamic contour tonometry (DCT) is a device for non-invasive continuous measurement of IOP.
7. Autofluorescence is a technique which is based upon fluorescent emission of ocular tissues. It is used to assess retinal diseases and vascular abnormalities. Fluorescein angiography is the process whereby following injection of sodium fluorescein into the systemic circulation, fluorescence of the retina (when illuminated) is recorded. This enables diabetic retinopathy, vascular occlusions, oedema of the optic disc, and tumours to be detected.