Parsons' Diseases of the Eye

23rd Edition

Ramanjit Sihota
Radhika Tandon

ELSEVIER

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23RD EDITION

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Preface to the twenty-third edition

Ramanjit Sihota, Radhika Tandon

It is an honour and pleasure to present yet another edition of 'Parsons' Diseases of the Eye'. This classic textbook with its unique features provides a comprehensive compendium of information covering all the relevant aspects of ophthalmology for thorough knowledge of the subject. This 23rd edition has been updated keeping in view the changing disease spectrum, practice patterns and advancements in technology. We are happy that the MCI has provided an updated competency-based undergraduate curriculum which is to be rolled out in August 2019 and glad to provide the information that the contents of this book have been updated in compliance to fully cover all the aspects as prescribed. Additional information has been seamlessly integrated to provide a higher level of understanding of ophthalmology for those with a higher level of interest such as medical students with special curiosity about the subject, postgraduate students to revise their own understanding and teachers and clinicians who wish to refresh and update themselves. Core skills prescribed by the MCI are also adequately covered in the text and are supported by digital material. We hope you enjoy reading it and using the online resources to enrich yourselves as much as we did in preparing it for you.

Preface to the twenty-second edition

Ramanjit Sihota, Radhika Tandon

It is an honour and pleasure to present yet another edition of “Parsons’ Diseases of the Eye”. This classic textbook with its unique features provides a comprehensive compendium of information covering all the relevant aspects of ophthalmology for thorough knowledge of the subject. This 22nd Edition has been updated keeping in view the changing disease spectrum, practice patterns and advancements in technology. We hope you enjoy reading it and enrich your information spectrum as much as we did in preparing it for you.

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Ramanjit Sihota, Radhika Tandon

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We acknowledge and greatly appreciate the efforts and invaluable comments of our editors; we would especially like to thank Fariha for her diligent support and professional help throughout the revision process.

Last but not the least, we would like to make an endearing mention of our families who with their loyal forbearance allowed us to spend our spare time and devote our attention to this work, without which it would not have been possible to achieve this.

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SECTION I:

Anatomy and Physiology of the Eye and Visual Pathway

OUTLINE

1. Anatomy of the eye
2. Physiology of the eye
3. The physiology of vision
4. The neurology of vision

CHAPTER 1

Anatomy of the eye

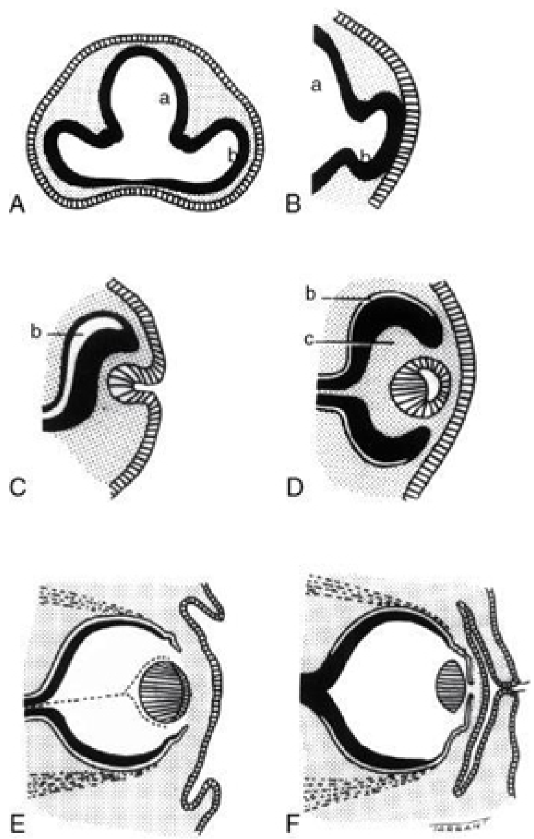
CHAPTER OUTLINE

Learning objectives

- To gain basic knowledge of the development of the eye.
- To develop essential understanding on how abnormalities at various stages of development can arrest or hamper normal formation of the ocular structures and visual pathways.
- To acquire adequate information about normal anatomy of the eye and related structures and develop a strong foundation for a better understanding of common ocular problems and their consequences.

Development of the eye

The formation, early growth and development of the eyes and visual system in utero, is fascinating and of critical importance in understanding the aetiopathogenesis of congenital disorders, providing an insight in to genetic evolution and a better understanding of developmental disorders. The eyes and visual system develop as a part of the central nervous system with contributions from the adjacent mesodermal and ectodermal tissues (Table 1.1).



• FIG. 1.1 The development of the eye. In each case, the solid black is the neural ectoderm, the hatched layer is the surface ectoderm and its derivatives, the dotted area is the mesoderm: a, cavity of the forebrain; b, cavity of the optic vesicle; c, cavity of the optic cup (or secondary optic vesicle) formed by invagination. **(A)** Transverse section through the anterior part of the forebrain and optic vesicles of a 4-mm human embryo. **(B)** The primary optic vesicle. **(C)** The formation of the optic cup by invagination at the embryonic fissure and invagination of the surface epithelium. **(D)** The optic cup and lens vesicle. **(E)** The formation of the ciliary region and iris, the anterior chamber, the hyaloid artery and the lid folds. The lens is formed from the posterior cells of the lens vesicle. **(F)** The complete eye.

TABLE 1.1

Summary of Ocular Embryogenesis

Period after Conception	Major Milestone	
3rd week	Optic groove appears	
4th week	Optic pit develops into optic vesicle	
	Lens plate forms	
	Embryonic fissure develops	Fig. 1.1A–D
1st month	Lens pit and then lens vesicle forms	
	Hyaloid vessels develop	
1½ months	Closure of embryonic fissure	
	Differentiation of retinal pigment epithelium	
	Proliferation of neural retinal cells	
	Appearance of eyelid folds and nasolacrimal duct	
7th week	Formation of embryonic nucleus of the lens	
	Sclera begins to form	
	First wave: formation of corneal and trabecular endothelium	
	Second wave: formation of corneal stroma	
	Third wave: formation of iris stroma	Fig. 1.1E
3rd month	Differentiation of precursors of rods and cones	
	Anterior chamber appears	
	Fetal nucleus starts to develop	
	Sclera condenses	
	Eyelid folds lengthen and fuse	
4th month	Formation of retinal vasculature begins	
	Hyaloid vessels begin to regress	
	Formation of physiological optic disc cup and lamina cribrosa	
	Canal of Schlemm appears	
	Bowman's membrane develops	
	Formation of major arterial circle and sphincter muscle of iris	
5th month	Photoreceptors differentiate	
	Eyelid separation begins	
6th month	Differentiation of dilator pupillae muscle	
	Nasolacrimal system becomes patent	
	Cones differentiate	Fig. 1.1F
7th month	Rods differentiate	
	Myelination of optic nerve begins	
	Posterior movement of anterior chamber angle	
	Retinal vessels start reaching nasal periphery	
8th month	Completion of anterior chamber angle formation, hyaloid vessels disappear	
9th month	Retinal vessels reach temporal periphery, pupillary membrane disappears	
After birth	Macular region of the retina develops further	

The anterior portion of the neural tube is the precursor of the forebrain, and its lateral aspect develops a thickening (the optic plate) which then grows outwards as a diverticulum towards the surface to form the primary optic vesicle (Fig. 1.1A and B).

***In the developing human embryo, the two eyes develop from the neural ectoderm derived from the neural tube as a pair of diverticula (primary optic vesicle) arising from the sides of the forebrain with contributions from the adjacent mesoderm and surface ectoderm in contact with it (Table 1.2*).**

TABLE 1.2

Primordial Tissue and Its Derivatives

Precursor	Derivatives
Neural ectoderm	Smooth muscle of the iris Optic vesicle and cup Iris epithelium Ciliary epithelium Part of the vitreous Retina Retinal pigment epithelium Fibres of the optic nerve
Surface ectoderm	Conjunctival epithelium Corneal epithelium Lacrimal glands Tarsal glands Lens
Mesoderm	Extraocular muscles Corneal stroma Sclera Iris Vascular endothelium of eye and orbit Choroid Part of the vitreous
Neural crest[a]	Corneal stroma, keratocytes and endothelium Sclera Trabecular meshwork endothelium Iris stroma Ciliary muscles Choroidal stroma Part of the vitreous Uveal and conjunctival melanocytes Meningeal sheaths of the optic nerve Ciliary ganglion Schwann cells of the nerve sheaths Orbital bones Orbital connective tissue Connective tissue sheath and muscular layer of the ocular and orbital blood vessels

[a]During the folding of the neural tube, a ridge of cells comprising the neural crest develops from the tips of the converging edges and migrates to the dorsolateral aspect of the tube. Neural crest cells from this region subsequently migrate and give rise to various structures within the eye and the orbit.

Note: The structures are listed from anterior to posterior.

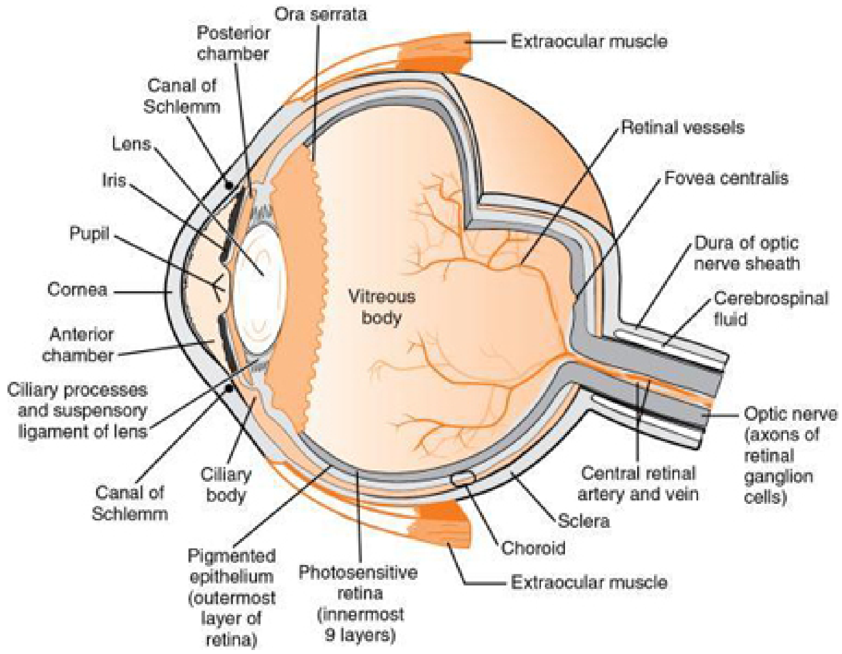
The *primary optic vesicle* grows outwards, meets the surface ectoderm, and then invaginates from below (the *optic cup*), the line of invagination remaining open for some time as the *embryonic fissure* (Fig. 1.1C). The inner layer of the cup forms the main structure of the retina, from which the nerve fibres eventually grow backwards towards the brain. The outer layer of the cup remains as a single layer of pigment epithelium. A narrow space representing the original optic vesicle remains as a potential space between the retina and the retinal pigment epithelium, and this space may get manifest in diseases such as retinal detachment or subretinal haemorrhage. The anterior border of the *optic vesicle and optic cup* form parts of the ciliary body and iris, both having corresponding inner and outer layers which are contiguous with the retina or retinal pigment epithelium, respectively (Fig. 1.1E). At the point where the neural ectoderm meets the surface ectoderm, the latter thickens to form the *lens plate*, invaginates to form the *lens vesicle* (Fig. 1.1C) and then separates to form the lens (Fig. 1.1D). The hyaloid artery enters the optic cup through the embryonic fissure and grows forwards to meet the lens, bringing temporary nourishment to the developing structures before it eventually atrophies and disappears; as it does so, its place is taken by a clear jelly (the vitreous) largely secreted by the surrounding neural ectoderm.

While these ectodermal events are taking place, the mesoderm surrounding the optic cup differentiates to form the coats of the eye and the orbital structures; the mesoderm between the lens and the surface ectoderm becomes hollowed to form the anterior chamber, lined by mesodermal condensations which form the anterior layers of the iris, the angle of the anterior chamber and the main structures of the cornea; whereas the surface ectoderm remains as the corneal and conjunctival epithelium. In the surrounding region, folds grow over in front of the cornea, unite and separate again to form the lids (Fig. 1.1E and F).

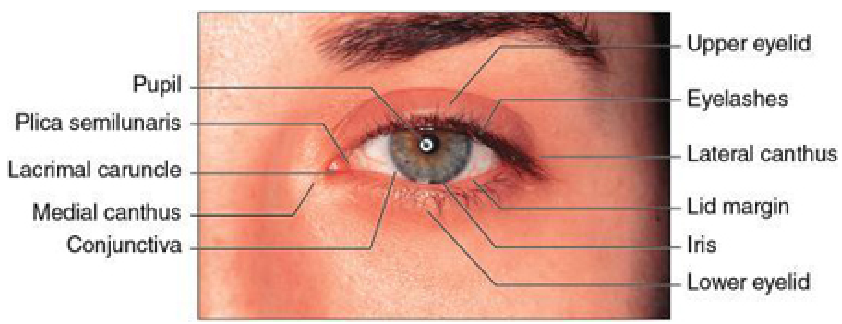
Anatomy

Knowledge of the anatomy of the eye is important for a proper understanding of ocular health and disease conditions affecting it.

The eye, also known as the eyeball, is a globe. The wall is composed of an outermost coat which is a dense, imperfectly elastic supporting tissue, the transparent cornea forming the anterior one-quarter and the opaque sclera forming the remainder three-quarters of the eye (Fig. 1.2). The anterior part of the sclera is covered by a mucous membrane—the conjunctiva—which is reflected from its surface onto the lids. The external appearance of the eye as seen clinically is illustrated in Fig. 1.3.



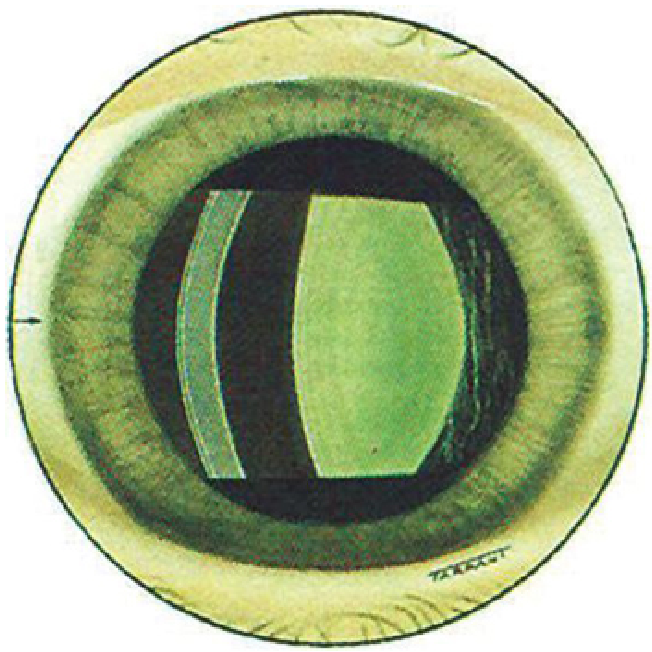
• **FIG. 1.2** General anatomy of the eyeball, including its tunics and chambers. Source: (From Rolston D, Nielsen C. Chapter 2: Ophthalmology. In: Rapid review USMLE step 3. St. Louis: Mosby; 2007. p. 28–44.)



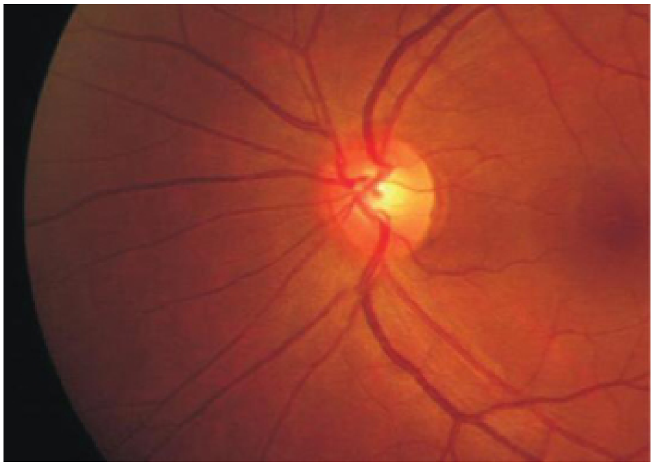
• **FIG. 1.3** The eyelids and anterior aspect of the eyeball. Source: (Adapted from Standring S. Chapter 39: The orbit and accessory visual apparatus. In: Standring S, Borley NR, Collins P, editors. Gray's anatomy: The anatomical basis of clinical practice. 40th ed. Edinburgh: Churchill Livingstone; 2009. p. 655–74.)

Inner to the sclera lies the middle layer of the wall consisting of the uveal tract comprising the choroid posteriorly and the ciliary body and iris anteriorly. The retina form the innermost layer of the posterior two-thirds to three-quarters of the eye lining the posterior part of the eye up to its anterior 360° circumferential boundary called the ora serrata.

Inside the eye, the globe is broadly divided into the anterior segment and posterior segment by the lens. The iris divides the anterior segment into an anterior chamber bounded by the cornea anteriorly and posterior chamber bounded by the lens posteriorly (Fig. 1.4). The posterior segment, i.e. the part of the eye that lies behind the lens, is bounded internally by the ciliary body anteriorly and retina posteriorly. Hence, posteriorly the sclera is lined by the uveal tract and inner to that lies the retinal pigment epithelium and retina. The optic nerve head is visible as the optic disc and represents the retinal nerve fibres leaving the eye as a tract nasal to the posterior pole of the eye (Fig. 1.5). The cavity of the eye contains a clear watery fluid called aqueous humour in the anterior segment, and posterior to the lens, the cavity is filled with a transparent gel-like substance called the vitreous humour.



• **FIG. 1.4** Optical section of the normal eye, as seen with the slit lamp. The light (arrowed) comes from the left and, in the beam of the slit lamp, the sections of the cornea and lens are clearly seen.



• **FIG. 1.5** The normal fundus. The disc or optic nerve head is approximately 1.5 mm in diameter and the centre of the fovea or macula is located about 2 disc diameters temporal to it.

Cornea

The ***cornea*** is the transparent front part of the eye which resembles a 'watch glass' and consists of different layers and regions:

- Epithelium
- Bowman's membrane
- Stroma (substantia propria)
- Dua's layer (pre-Descemet's layer)
- Descemet's membrane
- Endothelium

Transparency of cornea

Transparency of the cornea is related to the regularity of the stromal components. The stromal collagen fibrils are of regular diameter, arranged as a lattice with an interfibrillar spacing of less than a wavelength of light so that tangential rows of fibres act as a diffraction grating resulting in destructive interference of scattered rays. The primary mechanism controlling stromal hydration is a function of the corneal endothelium which actively pumps out the electrolytes and water flows out passively. The endothelium is examined by a specular microscope at a magnification of ×500. Endothelial cells become less in number with age, and the residual individual cells may enlarge to compensate.

Blood supply and innervation

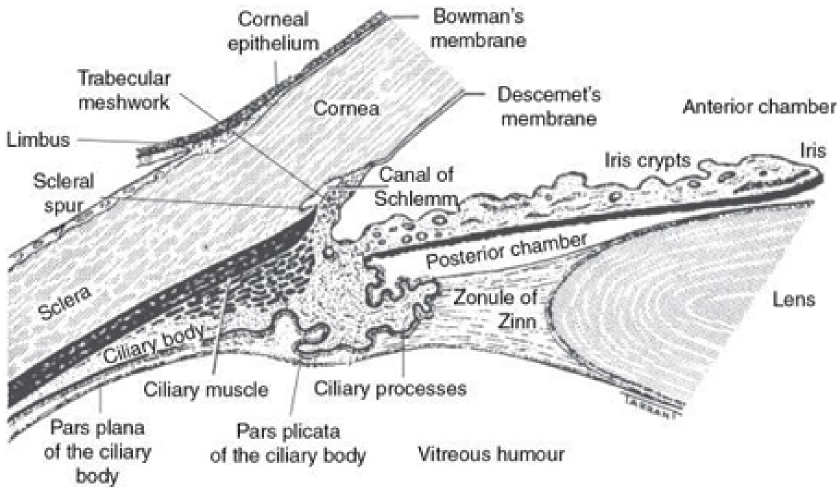
The cornea is avascular with no blood vessels with the exception of minute arcades, extending about 1 mm into the cornea at the limbus. It is dependent for its nourishment upon diffusion of tissue fluid from the vessels at its periphery and the aqueous humour. The cornea is very richly supplied with unmyelinated nerve fibres derived from the trigeminal nerve.

Sclera

The ***sclera*** is the 'white' supporting wall of the eyeball and is continuous with the clear cornea. It is a dense white tissue, thickest in the area around the optic nerve. The outer surface of the sclera is covered by the conjunctiva, beneath which is a layer of loose connective tissue called ***episclera*** and the innermost layer of the sclera consists of elastic fibres called the ***lamina fusca***. Lining the inner aspect of the sclera are two structures—the highly vascular ***uveal tract*** concerned chiefly with the nutrition of the eye, and within this a nervous layer, the true visual nerve endings concerned with the reception and transformation of light stimuli, called the ***retina***.

Anterior chamber

The ***anterior chamber*** is a space filled with fluid, the ***aqueous humour***; it is bounded in front by the cornea, behind by the iris and the part of the anterior surface of the lens which is exposed in the pupil. Its peripheral recess is known as the ***angle of the anterior chamber***, bounded posteriorly by the root of the iris and the ciliary body and anteriorly by the corneosclera (Fig. 1.6). In the inner layers of the sclera at this part, there is a circular venous sinus, sometimes broken up into more than one lumen, called the ***canal of Schlemm***, which is of great importance for the drainage of the aqueous humour. At the periphery of the angle between the canal of Schlemm and the recess of the anterior chamber, there lies a loosely constructed meshwork of tissues, the ***trabecular meshwork***. This has a triangular shape, the apex arising from the termination of Descemet's membrane and the subjacent fibres of the corneal stroma and its base merging into the tissues of the ciliary body and the root of the iris. It is made up of circumferentially disposed flattened bands, each perforated by numerous oval stomata through which tortuous passages exist between the anterior chamber and Schlemm's canal. The extracellular spaces contain both a coarse framework (collagen and elastic components) and a fine framework (mucopolysaccharides) of extracellular materials, which form the probable site of greatest resistance to the flow of aqueous.



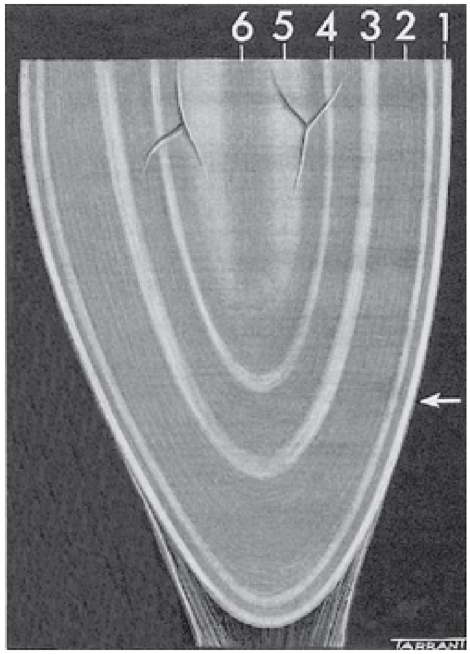
• **FIG. 1.6** The region of the angle of the anterior chamber.

The endothelial cells of Schlemm's canal are connected to each other by junctions which are not 'tight' but this intercellular pathway accounts for only 1% of the aqueous drainage. The major outflow pathway appears to be a series of transendothelial pores, which are usually found in outpouchings of the endothelium called 'giant vacuoles'.

The anterior chamber is about 2.5 mm deep in the centre in a normal adult; it is shallower in very young children and in old people.

Lens

The ***lens*** is a biconvex mass of peculiarly differentiated epithelium. It has three main parts: the lens capsule, the lens epithelium and the lens fibres. The outer capsule lined by the epithelium and the lens fibres and is developed from an invagination of the surface ectoderm of the fetus, so that what was originally the surface of the epithelium comes to lie in the centre of the lens, the peripheral cells corresponding to the basal cells of the epidermis. Just as the epidermis grows by the proliferation of the basal cells, the old superficial cells being cast off, so the lens grows by the proliferation of the peripheral cells. The old cells, however, cannot be cast off, but undergo changes (sclerosis) analogous to that of the stratum granulosum of the epidermis, and become massed together in the centre or nucleus; moreover, the newly formed cells elongate into fibres. The lens fibres have a complicated architectural form, being arranged in zones in which the fibres growing from opposite directions meet in sutures. Without going into details, it is important to bear in mind that the central nucleus of the lens consists of the oldest cells and the periphery or cortex the youngest (Fig. 1.7).



• FIG. 1.7 The structure of the lens in an adult 40 years of age, as shown in the optical beam of the slit lamp: 1, anterior capsule; 2, cortex; 3, adult nucleus; 4, infantile nucleus; 5, fetal nucleus; 6, embryonic nucleus (see Fig. 15.9).

The fibres of the lens are split into regions depending on the age of origin. The central denser zone is the nucleus surrounded by the cortex. The oldest and innermost is the central ***embryonic nucleus*** (formed during 6–12 weeks of embryonic life), in which the lens fibres meet around Y-shaped sutures. Outside this embryonic nucleus, successive nuclear zones are laid down as development proceeds, called, depending on the period of formation, the ***fetal nucleus*** (3–8 months of fetal life), the ***infantile nucleus*** (last month of intrauterine life till puberty), the ***adult nucleus*** (corresponding to the lens in early adult life) and finally and most peripherally, the ***cortex*** consisting of the youngest fibres. In this part of the lens, also the fibres meet along the sutures with a general stellate arrangement. The mass of epithelium which constitutes the lens is surrounded by a hyaline membrane, the ***lens capsule***, which is thicker over the anterior than over the posterior surface and is thinnest at the posterior pole; the thickest basement membrane in the body, the lens capsule, is a cuticular deposit secreted by the epithelial cells, having on the outside a thin membrane, the ***zonular lamella***.

The lens in fetal life is almost spherical; it gradually becomes flattened so as to assume a biconvex shape. It is held in place by the ***suspensory ligament*** or **zonule of Zinn**. This is not a complete membrane, but consists of bundles of strands which pass from the surface of the ciliary body to the capsule where they join with the zonular lamella. The strands pass in various directions so that the bundles often cross one another. Thus, the most posterior arise from the pars plana of the ciliary body almost as far back as the ora serrata; these lie in contact with the ciliary body for a considerable distance and then curve towards the equator of the lens to be inserted into the capsule slightly anterior to the equator. A second group of bundles springs from the summits and sides of the ciliary processes, i.e. far forwards, and passes backwards to be inserted into the lens capsule slightly posterior to the equator. A third group passes from the summits of the processes almost directly inwards to be inserted at the equator.

Uveal tract

The uveal tract consists of three parts, of which the two posterior, the ***choroid*** and ***ciliary body***, line the sclera while the anterior forms a free circular diaphragm, the ***iris***. The plane of the iris is approximately coronal; the aperture of the diaphragm is the ***pupil***. Situated behind the iris and in contact with the pupillary margin is the crystalline lens.

Iris

The ***iris*** is thinnest at its attachment to the ciliary body, so that if torn it tends to give way in this region (Fig. 1.6). It is composed of a stroma containing branched connective tissue cells, usually pigmented but largely unpigmented in blue irides, with a rich supply of blood vessels which run in a general radial direction. The tissue spaces communicate directly with the anterior chamber through crypts found mainly near the ciliary border; this allows the easy transfer of fluid between the iris and the anterior chamber. The stroma is covered on its posterior surface by two layers of pigmented epithelium, which developmentally are derived from the retina and are continuous with each other at the pupillary margin. The anterior layer consists of flattened cells and the posterior of cuboidal cells. From the epithelial cells of the former, two unstriped muscles are developed which control the movements of the pupil, the ***sphincter pupillae***, a circular bundle running round the pupillary margin, and the ***dilator pupillae***, arranged radially near the root of the iris.

The anterior surface of the iris is covered with a single layer of endothelium, except at some minute depressions or crypts which are found mainly at the ciliary border; it usually atrophies in adult life.

The iris is richly supplied by sensory nerve fibres derived from the trigeminal nerve. The sphincter pupillae is supplied by parasympathetic autonomous secretomotor nerve fibres derived from the oculomotor nerve, while the motor fibres of the dilator muscle are derived from the cervical sympathetic chain.

Ciliary body

The ***ciliary body*** in anteroposterior section is shaped roughly like an isosceles triangle, with the base forwards. The iris is attached about the middle of the base, so that a small portion of the ciliary body enters into the posterior boundary of the anterior chamber at the angle (Fig. 1.6). The chief mass of the ciliary body is composed of unstriped muscle fibres, the ***ciliary muscle***. This consists of three parts with a common origin in the ciliary tendon, a structure which runs circumferentially round the globe blending with the 'spur' of the sclera and related to the trabecular mesh work. The greater part of the muscle is composed of meridional fibres running anteroposteriorly on the inner aspect of the sclera to find a diffuse insertion into the suprachoroid. Most of the remaining fibres run obliquely in interdigitating V-shaped bundles so as to give the impression of running in a circle around the ciliary body, concentrically with the base of the iris. The third portion of the muscle is composed of a few tenuous iridic fibres arising most internally from the common origin and finding insertion in the root of the iris just anterior to the pigmentary epithelium in close relation to the dilator muscle.

The inner surface of the ciliary body is divided into two regions—the anterior part is corrugated with a number of folds running in an anteroposterior direction while the posterior part is smooth. The anterior part is therefore called the ***pars plicata***; the posterior is called the ***pars plana***. About 70 plications are visible around the circumference macroscopically, but if microscopic sections are examined, many smaller folds, the ***ciliary processes***, will be seen between them. These contain no part of the ciliary muscle, but consist essentially of tufts of blood vessels, not unlike the glomeruli of the kidney. They are covered upon the inner surface by two layers of epithelium, which belong properly to the retina, and are continuous with similar layers in the iris; the outer layer, corresponding to the anterior in the iris, consists of flattened cells, the inner of cuboidal cells, but only the outer layer in the ciliary body is pigmented.

The ciliary body extends backward as far as the ***ora serrata***, at which point the retina proper begins abruptly; the transition from the ciliary body to the choroid, on the other hand, is gradual, although this line is conveniently accepted as the limit of the two structures. The ora serrata thus circles the globe, but is slightly more anterior on the nasal than on the temporal side.

The ciliary body is richly supplied with sensory nerve fibres derived from the trigeminal nerve. The ciliary muscle is supplied with motor fibres from the oculomotor and sympathetic nerves.

Choroid

The ***choroid*** is an extremely vascular membrane in contact everywhere with the sclera, although not firmly adherent to it, so that there is a potential space between the two structures—the ***epichoroidal*** or ***suprachoroidal space***. On the inner side, the choroid is covered by a thin elastic membrane, the ***lamina vitrea*** or ***membrane of Bruch***.

The blood vessels of the choroid increase in size from within outwards, so that immediately beneath the membrane of Bruch there is a capillary plexus of fenestrated vessels, the ***choriocapillaris***. Upon this is the layer of medium-sized vessels, while most externally are the large vessels, the whole being held together by a stroma consisting of branched pigmented connective tissue cells. The choroid is supplied with sensory nerve fibres from the trigeminal as well as autonomic nerves, presumably of vasomotor function.

Posterior chamber and vitreous humour

It will be noticed that there is somewhat a triangular space between the back of the iris and the anterior surface of the lens, having its apex at the point where the pupillary margin comes in contact with the lens; it is bounded on the outer side by the ciliary body. This is the ***posterior chamber*** and contains aqueous humour.

Behind the lens is the large vitreous chamber, containing the ***vitreous humour***. This is a jelly-like material, chemically of the nature of an inert gel containing a few cells and wandering leucocytes. As in other gels, the concentration of the micellae on the surface gives rise to the appearance of a boundary membrane in sections—the so-called ***hyaloid membrane***.

The ***vitreous body*** is attached anteriorly to the posterior lens surface by the ligament of Weigert. In the region of the ora, the vitreous cortex is firmly attached to the retina and pars plana and this attachment is referred to as the vitreous base.

Posteriorly, the vitreous body is attached to the margin of the optic disc and to the macula forming a ring around each structure and also to the larger blood vessels. The primary vitreous is concentrated into the centre of the globe by the secondary vitreous and forms the canal of Cloquet which contains material less optically dense than the secondary vitreous.

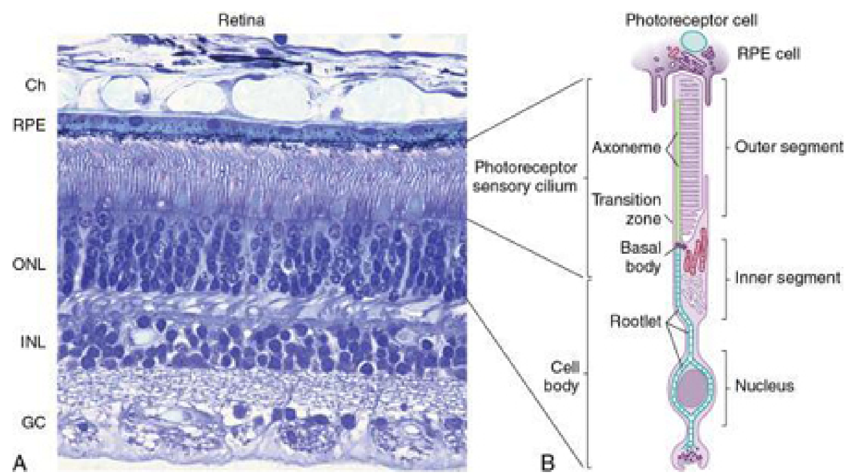
The body of the vitreous has a loose fibrous framework of collagenous fibres, whereas its cortex is made up of collagen-like fibres and protein.

Retina

The retina corresponds in extent to the choroid, which it lines, although the same embryological structure is continued forwards as a double layer of epithelium as far as the pupillary margin. If the two layers of epithelium are traced backwards, the anterior layer in the iris is found to be continuous with the outer layer in the ciliary body, and this again is continued into the pigment epithelium of the retina as a single layer of hexagonal cells lying immediately adjacent to the membrane of Bruch. Similarly, the posterior layer in the iris, although pigmented, passes into the inner unpigmented layer of the ciliary body, and this suddenly changes at the ora serrata into the highly complex visual retina.

The retina develops from the neural ectoderm by invagination of the optic vesicle—the outer layer forming the retinal pigment epithelium, the inner layer the transparent neurosensory retina and the cavity of the embryologic optic vesicle remains as a potential space between them. The two layers are largely lightly apposed to each other in the healthy eye except at the edges of the optic disc and the ora serrata where the two are firmly attached and inseparable.

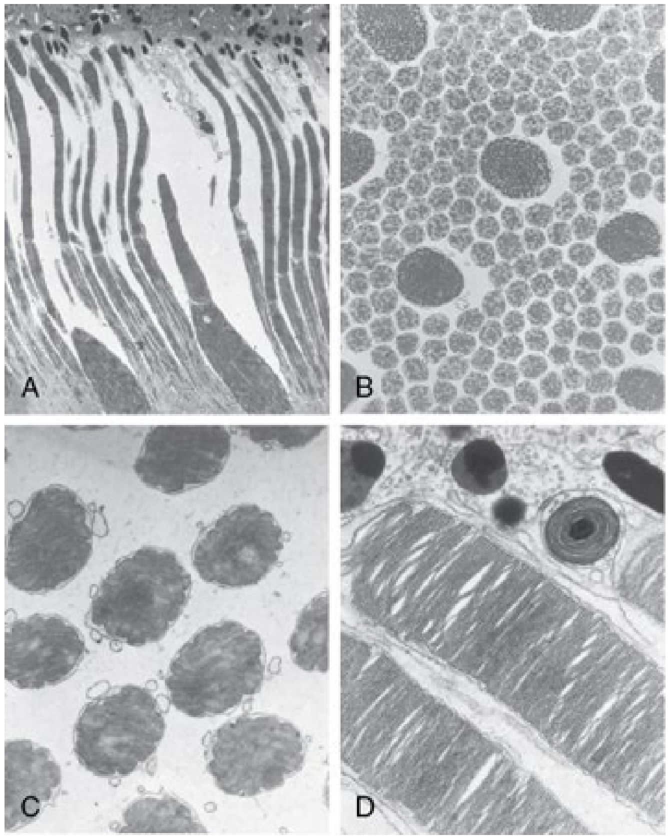
Histologically, the retina consists of 10 layers (Fig. 1.8A and B) formed by three strata of cells and their synapses, namely outermost visual cells or photoreceptors, the bipolar cells in the middle and the ganglion cells innermost.



• FIG. 1.8 Retina and photoreceptor cell structure. **(A)** Cross-section of human retina, showing retinal layers. **(B)** Drawing of rod photoreceptor cell, showing different portions of the cell. The photoreceptor sensory cilium is indicated. Ch, choroid; GC, ganglion cell layer; INL, inner nuclear layer; ONL, outer nuclear layer; RPE, retinal pigment epithelium. Source: (From Levin LA, Albert DM. Chapter 74: Retinitis pigmentosa and related disorders. In: Ocular disease: mechanisms and management. Edinburgh: Saunders; 2010. p. 579–89.)

Retinal pigment epithelium: The pigment epithelium consists of a single layer of hexagonal cells lying between the retinal photoreceptor outer segments and Bruch's membrane of the choroid. They assist the metabolism of the retina by transporting selected substances to the receptor cells. Products of metabolism are freely exchanged between the receptor cells and the pigment epithelium. The most striking inclusions in the pigment epithelium are the melanin granules responsible for its colour. Most of the light which passes through the retina and is not absorbed by the photopigments in the photoreceptor outer segments is absorbed by these granules. The cells also contain important organelles called phagosomes which are known to be discarded rod discs that have been engulfed by the pigment epithelium. The phagocytic capacity of the pigment epithelium is demonstrated in the response of the retina to injury as by laser irradiation, when the number of phagosomes in the underlying epithelial cells increases significantly.

Photoreceptors (rods and cones): Most externally, in contact with the pigment epithelium is neural epithelium, the rods and cones, which are the end-organs of vision (Fig. 1.9). The microanatomy of the rods and cones reveals the transductive region (outer segment), a region for the maintenance of cellular homoeostasis (inner segment), a nuclear region (outer nuclear layers) and a transmissive region (the outer plexiform or synaptic layer). When the outer segments of the rods are sectioned parallel to their long axes, they are seen by the electron microscope to consist of a boundary or cell membrane, which encloses a stack of membrane systems. The discs in the rods are continuously renewed throughout life. New discs are formed in the region of the inner segment and are progressively displaced towards the pigment epithelium. Rod discs have a limited life and are eventually lost to the pigment epithelium. At the junction of the inner and outer segments, the cell body of both rods and cones constricts. The electron microscope reveals a connecting cilium which is always eccentric and provides the only link between the inner and outer segments.



• FIG. 1.9 Anatomical features of rods and cones revealed by electron microscopy. **(A)** Cross-section of the human retina (×2440) demonstrating rod and cone outer segments adjacent to the pigment epithelium. **(B)** Tangential section through the inner segments of the human photoreceptor layer (×3750). The large inner segments belong to cones, and the smaller inner segments are those of rods; note the large number of mitochondria in the inner segments. **(C)** Tangential section of human retina at the outer segment level, showing rod discs contained within the cell membrane (×14,110). **(D)** Rod outer segment showing discs contained within the cell. A phagosome within a pigment epithelial cell is on the upper right (rhesus monkey ×23,000). Source: (From Ryan S, Schachat A, Wilkinson C, Hinton D, Wilkinson C, editors. Retina. 4th ed. Edinburgh: Elsevier; 2005.)

Histologically distinguishable 10 layers of retina (outermost to innermost) are listed as follows:

1. Retinal pigment epithelium
2. Outer segment of rods and cones
3. External limiting membrane
4. Outer nuclear layer: the outer nuclear layer (the nuclei of the rods and cones)
5. The outer plexiform layer consisting of synapses
6. The inner nuclear layer (the nuclei of the bipolar cells)

7. The inner plexiform layer (again synaptic)
8. The ganglion cell layer
9. The nerve fibre layer composed of the axons of ganglion cells running centrally into the optic nerve
10. The internal limiting membrane

These special nervous constituents are bound together by neuroglia, the better developed vertical cells being called the fibres of Müller, which in addition to acting as a supportive framework, have a nutritive function. The structure is completed by two limiting membranes, the outer perforated by the rods and cones, and the inner separating the retina from the vitreous.

To excite the rods and cones, incident light has to traverse the tissues of the retina but this arrangement allows these visual elements to approximate the opaque pigmented layer to form a functional unit, and their source of nourishment is the choriocapillaris.

At the posterior pole of the eye, which is situated about 3 mm to the temporal side of the optic disc, a specially differentiated spot is found in the retina, the *fovea centralis*, a depression or pit, where only cones are present in the neuroepithelial layer and the other layers are almost completely absent. The fovea is the most sensitive part of the retina, and is surrounded by a small area, the *macula lutea*, or yellow spot which, although not so sensitive, is more so than other parts of the retina. It is here that the nuclear layers become gradually thinned out, while parts of the plexiform layers are especially in evidence. The ganglion cells too, instead of consisting of a single row of cells, are heaped up into several layers. There are no blood vessels in the retina at the macula, so that its nourishment is entirely dependent upon the choroid (see Table 1.3).

TABLE 1.3

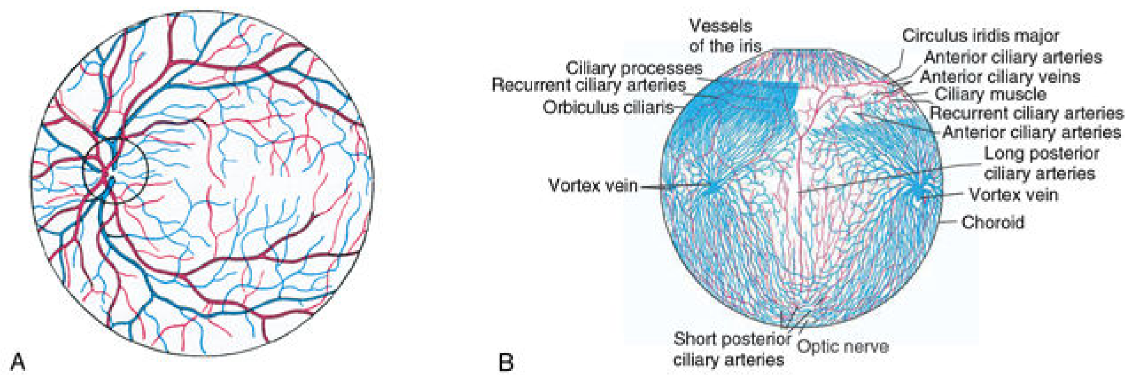
Special Features of Fovea Centralis and Macula Lutea

Fovea Centralis	Macula Lutea
• Most sensitive for vision	• Nuclear layers get gradually thinned out
• Contains only cones	• There are several layers of ganglion cells
	• There are no blood vessels in this region
	• Entirely dependent on choroid for nourishment

At the optic disc, the fibres of the nerve fibre layer pass into the *optic nerve* (*see* Chapter 14, Diseases of the Uveal Tract), the other layers of the retina stopping short abruptly at the edge of the aperture in the scleral canal. This is spanned by a transverse network of connective tissue fibres containing much elastic tissue, the *lamina cribrosa*, through the meshes of which the optic nerve fibres pass. The optic nerve fibres which are the axons of the ganglion cells of the retina are afferent or centripetal fibres and they become surrounded by medullary sheaths as soon as they exit the eye.

The blood supply of the eye

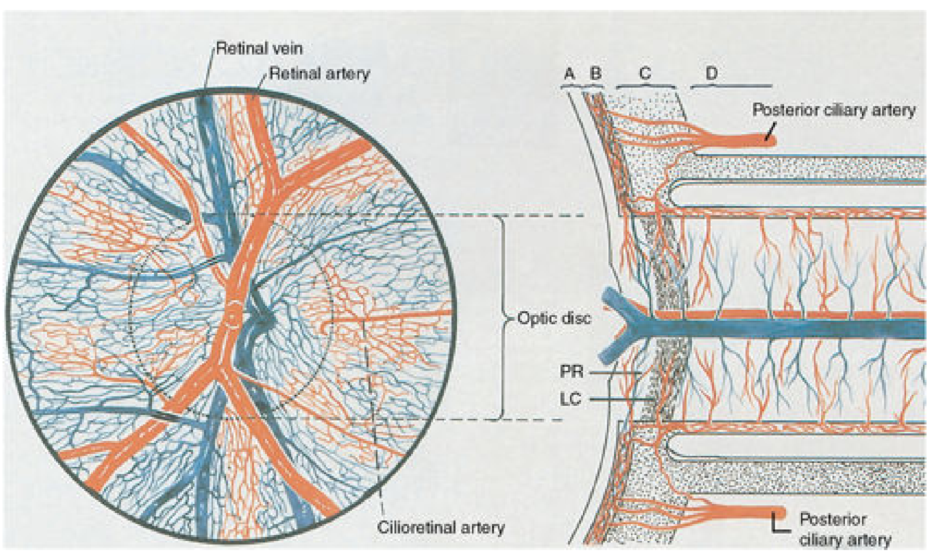
The **arteries of the eye** are all derived from the ophthalmic artery (Fig. 1.10A and B), which is a branch of the internal carotid artery. The ophthalmic artery has few anastomoses, so that on the arterial side the ocular circulation is an offshoot of the intracranial circulation. As far as the venous outflow is concerned, most of the blood passes to the cavernous sinus by way of the ophthalmic veins. It is to be noted that these veins anastomose freely in the orbit, the superior ophthalmic vein communicating with the angular vein at the root of the nose and the inferior ophthalmic vein with the pterygoid plexus. This has important clinical implications.



• **FIG. 1.10** **(A)** The retinal circulation. **(B)** The choroidal circulation.

The retina is supplied by the central retinal artery, which enters the optic nerve on its lower surface, 15–20 mm behind the globe. The central artery divides on, or slightly posterior to, the surface of the disc into the main retinal trunks, which will be considered in detail later (Fig. 1.10A). The retinal arteries are end arteries and have no anastomoses at the ora serrata. The only place where the retinal system anastomoses with any other is in the neighbourhood of the lamina cribrosa. The veins of the retina do not accurately follow the course of the arteries, but they behave similarly at the disc, uniting on, or slightly posterior to, its surface to form the central vein of the retina, which follows the course of the corresponding artery.

The blood supply of the optic nerve head in the region of the ***lamina cribrosa*** is served by fine branches from the arterial circle of Zinn but mainly from the branches of the posterior ciliary arteries (Fig. 1.11). The central retinal artery makes no contribution to this region. The ***prelaminar region*** is supplied by centripetal branches from the peripapillary choroidal vessels with some contribution from the vessels in the lamina cribrosa region. The central artery of the retina does not contribute to this region either. The surface layer of the ***optic disc*** contains the main retinal vessels and a large number of capillaries in addition to some small vessels. The capillaries on the surface of the disc are derived from branches of the retinal arterioles. In this part of the disc, vessels of choroidal origin derived from the adjacent prelaminar part of the disc may be seen usually in the temporal sector of the disc, and one of them may enlarge to form a cilioretinal artery. The capillaries on the surface of the disc are continuous with the capillaries of the peripapillary retina. These capillaries are mainly venous and drain into the central retinal vein. In the retrolaminar part of the optic nerve, blood is supplied by the intraneural centrifugal branches of the central artery of the retina with centripetal contributions from the pial branches of the choroidal arteries, circle of Zinn, central artery of the retina and the ophthalmic artery.



• FIG. 1.11 Blood supply of the optic nerve. Region marked: A, represents the surface of the disc and peripapillary nerve fibre layer; B, portion anterior to the lamina cribrosa; C, portion related to the lamina cribrosa; D, portion behind the lamina cribrosa; LC, lamina cribrosa; PR, prelaminar. Source: (Reproduced with kind permission from Hayreh SS. Arch Ophthalmology 1977;95:1560.)

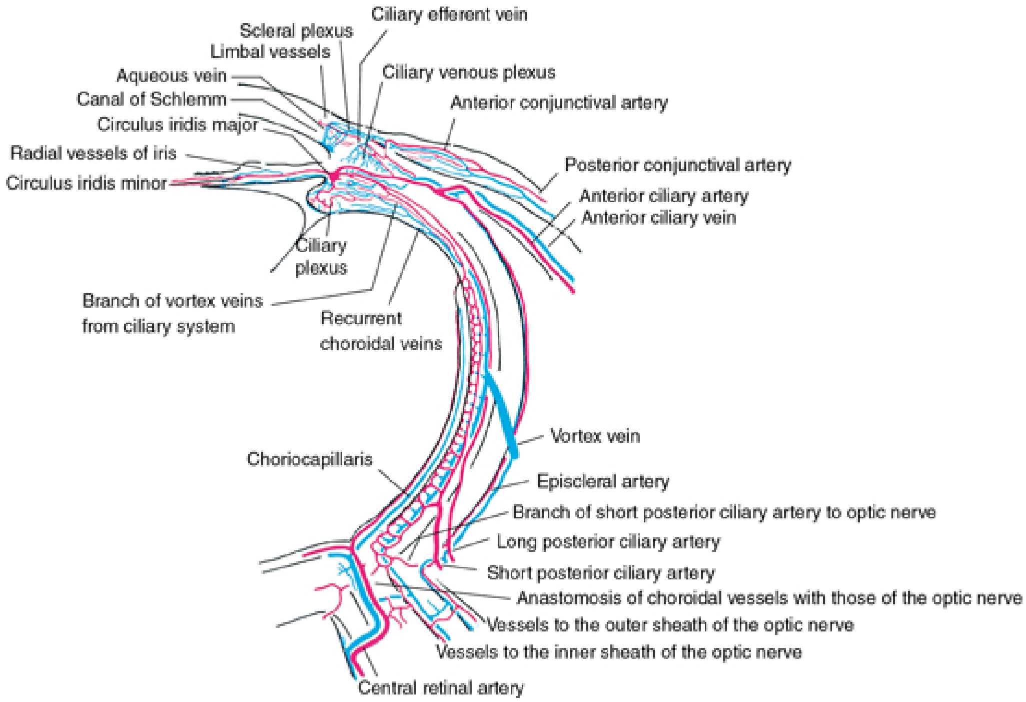
Venous drainage of the optic disc is mainly carried out by the central retinal vein. The prelaminar region also drains into the choroidal veins. There is no venous channel corresponding to the circle of Zinn. The central retinal vein communicates with the choroidal circulation in the prelaminar region.

The uveal tract is supplied by the ciliary arteries, which are divided into three groups—the short posterior, the long posterior and the anterior (Figs 1.10B and 1.12). The short posterior ciliary arteries, about 20 in number, pierce the sclera in a ring around the optic nerve, running perpendicularly through the sclera, to which fine branches are given off. The long posterior ciliary arteries, two in number, pierce the sclera slightly farther away from the nerve in the horizontal meridian, one on the nasal, the other on the temporal side. They traverse the sclera very obliquely, running in it for a distance of 4 mm. Both these groups are derived from the ophthalmic artery, while the anterior ciliary arteries are derived from the muscular branches of the ophthalmic artery to the four recti. They pierce the sclera 5 or 6 mm behind the limbus, or corneoscleral junction, giving off twigs to the conjunctiva, the sclera and the anterior part of the uveal tract.

The ciliary veins also form three groups—the short posterior ciliary, the vortex veins and the anterior ciliary. The short posterior ciliary veins are relatively unimportant; they do not receive any blood from the choroid, but only from the sclera. The vortex veins or venae vorticosae are the most important, consisting usually of four large trunks which open into the ophthalmic veins. They enter the sclera slightly behind the equator of the globe, two above and two below, and pass very obliquely through this tissue. The anterior ciliary veins are smaller than the corresponding arteries, since they receive blood from only the outer part of the ciliary muscle.

Of these ciliary vessels, the short posterior ciliary arteries supply the whole of the choroid, being reinforced anteriorly by anastomoses with recurrent branches from the ciliary body. The ciliary body and iris are supplied by the long posterior and anterior ciliary arteries. The blood from the whole of the uveal tract, with the exception of the outer part of the ciliary muscle, normally leaves the eye by the vortex veins only.

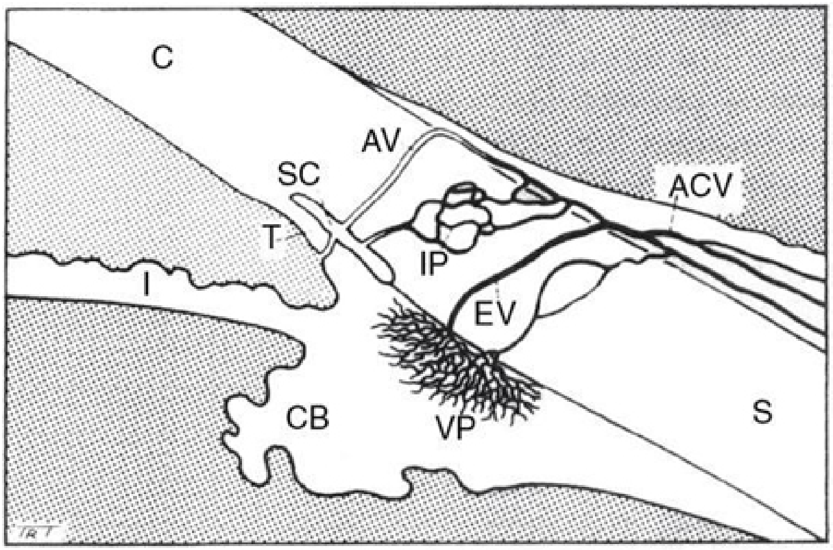
The two long posterior ciliary arteries pass forward between the choroid and the sclera, without dividing, as far as the posterior part of the ciliary body. Here each divides into two branches (Fig. 1.12); they run forward in the ciliary muscle, and at its anterior part bend round in a circular direction, anastomosing with each other and thus forming the ***circulus arteriosus iridis major***. This is situated in the ciliary body at the base of the iris; from it, the ciliary processes and iris are supplied. Other branches from the major arterial circle run radially through the iris, dividing dendritically and ending in loops at the pupillary margin. A circular anastomosis takes place a little outside the pupillary margin, the ***circulus arteriosus iridis minor***.



• FIG. 1.12 The ciliary circulation.

The tributaries of the vortex veins, which receive the whole of the blood from the choroid and iris, are arranged radially, the radii being bent, so as to give a whorled appearance—hence their name. The veins of the iris are collected into radial bundles which pass backwards through the ciliary body, receiving tributaries from the ciliary processes. Thus reinforced, they form an immense number of veins running backwards parallel to each other through the smooth part of the ciliary body. After reaching the choroid, they converge to form the large anterior tributaries of the vortex veins.

The veins from the outer part of the ciliary body, on the other hand, pass forward and unite with others to form a plexus (the ***ciliary venous plexus***), which drains into the anterior ciliary veins and the episcleral veins. These vessels communicate directly with the canal of Schlemm, which is intimately connected with the anterior chamber by means of numerous tortuous channels through the loose tissue of the trabecular meshwork. From this canal, the efferent channels form a complex system (Fig. 1.13); some of them drain into efferent ciliary veins in the sclera while others traverse the sclera and only join the venous system in the subconjunctival tissues (***aqueous veins***).



• FIG. 1.13 The exit channels of the aqueous humour in man: C, cornea; S, sclera; I, iris; and CB, ciliary body. The primitive drainage channels of lower animals are seen in VP, the ciliary venous plexus, draining by EV, the ciliary efferent veins into ACV, and the anterior ciliary veins. Superimposed on this is the drainage system peculiar to primates, represented by T, the trabeculae; SC, the canal of Schlemm; IP, the intrascleral plexus; and AV, an aqueous vein emptying into the anterior ciliary veins.

The marginal loops of the cornea and the conjunctival vessels are branches of the anterior ciliary vessels (Fig. 1.12).

Clinical anatomy of the eye

On clinical examination, the parts of the external surface of the eye appear as shown in Fig. 1.3. The appearance of the anterior segment from the cornea to the lens is as shown in Fig. 1.4 and the posterior segment behind the lens as shown in Fig. 1.5.

Summary

The human eye and its adnexal structures develop from the neuroectoderm of the neural groove and the adjoining surface ectoderm, mesoderm and cells of neural crest origin. Though the development takes place by a predetermined sequence of events, local interactions and trophic influences affect the chain of interrelated processes which take place both simultaneously and sequentially. Teratogenic influences like intrauterine infections, noxious stimuli, maternal intake of drugs or alcohol and exposure to radiation can affect the normal course of development leading to abnormalities and congenital deformities. The milestones in embryological development are not absolute and are more representative of a time

period than an actual finite time. Any disruption in that period will have an effect on the structures forming at that particular phase of development. As expected, abnormal developmental influences have a more severe impact if they occur early when the system is more immature and more prone to major developmental defects.

The eye is a complex anatomical structure consisting of delicate tissues. It is a sense organ which is designed to capture and focus light to form a retinal image which is translated into electrical signals and transmitted to the central nervous system via the optic nerve. The eye is protected from the environment by the eyelids, lashes and the orbital wall. The extraocular muscles function in a synchronized fashion to stabilize the globes, enable binocular vision and the full functional field of vision by allowing the full range of ocular movements. The blood supply of the eye and orbit is derived from the ophthalmic artery.

There are certain facts which are noteworthy as clinically relevant. The major part of ocular development occurs from week 3 and week 10. The neural tube ectoderm is the precursor of the retina, optic nerve, epithelium of iris and ciliary body, and smooth muscles of the iris. The surface ectoderm gives rise to the lens, the epithelium of cornea and conjunctiva, eyelids, and lacrimal system. The other ocular structures form from mesenchyme. The PAX 6 gene plays a significant part in ocular development..

Suggested reading

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