



Parsons'

DISEASES OF THE EYE

22nd
EDITION

Editors

Ramanjit Sihota

Radhika Tandon

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Ramanjit Sihota MD, DNB, FRCS, FRCOphth

Professor of Ophthalmology

Dr Rajendra Prasad Centre for Ophthalmic Sciences

All India Institute of Medical Sciences, New Delhi

Radhika Tandon MD, DipNB, FRCOphth, FRCSEd (Gold Medalist)

Professor of Ophthalmology

Dr Rajendra Prasad Centre for Ophthalmic Sciences

All India Institute of Medical Sciences, New Delhi



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Sihota, Tandon

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Content Strategist: Renu Rawat

Senior Project Manager-Education Solutions: Shabina Nasim

Project Manager: Prasad Subramanian

Project Coordinator: Isha Bali, Shravan Kumar

Sr. Manager-Publishing Operations: Sunil Kumar

Production Manager: NC Pant

Sr. Production Executive: Ravinder Sharma

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Preface to the Twenty-Second Edition

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.

Ramanjit Sihota
Radhika Tandon

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Preface to the Nineteenth Edition

It was a privilege to update this classic textbook of ophthalmology, which has educated many generations of medical students and ophthalmologists. We have aimed to preserve the basic character of *Parsons'*: the coverage of essential topics remains concise and thorough in an easy-to-understand language for undergraduates as well as postgraduates at the start of their training in ophthalmology.

Over a decade has passed since the last edition was published. In view of the tremendous advances in the diagnosis and therapy of ophthalmological disorders during this period, obsolete portions have been deleted. The rest of the book has been almost completely rewritten to incorporate newer trends in classification, diagnosis and management. We have attempted to highlight the interlinkages between ocular and systemic diseases. Genetics is an integral part of medicine today and a detailed description of the presently known genetic associations and their possible utility in the management of ocular diseases was considered important

enough to warrant an independent chapter. In all, three new chapters have been added: Ocular Manifestations of Systemic Disorders, Genetics in Ophthalmology, and The Causes and Prevention of Blindness.

A major excitement of preparing the new edition was the introduction of colour. Numerous colour illustrations (both in line and halftone), tables and flowcharts have been added to highlight important aspects, facilitate understanding and make the text more interesting.

We hope that this revised edition, like its previous *avatars*, serves as a basic text to establish the foundations of knowledge of ophthalmology for undergraduates and a ready review or rapid revision guide for postgraduates, teachers and practising ophthalmologists.

**Ramanjit Sihota
Radhika Tandon**

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Consultant, Vitreoretina Services
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Bangalore

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Medical Director, Shroff Eye Centre
Past President, Vitreo Retinal Society of India
New Delhi

Dr. Jaya Devendra

Associate Professor
Department of Ophthalmology
Rohilkhand Medical College and Hospital
Bareilly

Dr. Kavita Bhatnagar

Professor, Department of Ophthalmology
Padmashree Dr. DY Patil Medical College
Pune

Dr. Kavitha Avadhani

Specialist Ophthalmologist
Uveitis and Ocular Immunology
New Medical Centre Specialty Hospital
Abu Dhabi

Dr. Lavanya G. Rao

Professor and Head
Department of Ophthalmology
Kasturba Medical College & Hospital
Manipal

Dr. Mathew Kurian

Medical Superintendent
Senior Consultant, Cataract and Refractive Lens Surgery
Narayana Nethralaya
Bangalore

Dr. Neelam Puthran

Professor and Head
Department of Ophthalmology
Bharati Vidyapeeth Deemed University Medical College
Pune

Dr. Nidhi Pandey

Associate Professor
Department of Ophthalmology
Pt. J.N.M. Medical College
Raipur

Dr. Parikshit Gogate

Consultant Pediatric Ophthalmologist
Dr. Gogate's Eye Clinic
Pune

Dr. Ronnie George

Senior Consultant, Glaucoma Services
Glaucoma Department and Research Director
Sankara Nethralaya, Chennai

Dr. SS Kubrey

Assistant Professor
Gandhi Medical College
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Radhika Tandon**

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Section I

Anatomy and Physiology

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Chapter 1

Embryology and Anatomy

Chapter Outline

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EMBRYOLOGY

The central nervous system is developed from the neural groove which invaginates to form the neural tube running longitudinally down the dorsal surface of the embryo. At either side from the lateral aspect of the anterior portion of this structure, which is the precursor of the forebrain, a thickening appears at an early stage (the *optic plate*) which then grows outwards as a diverticulum towards the surface to form the *primary optic vesicle* (Fig. 1.1A and B). From this pair of diverticula from the sides of the forebrain and the mesodermal and ectodermal structures in contact with it, the two eyes develop.

After it meets the surface ectoderm, the primary optic vesicle 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, the nerve fibres from which eventually grow backwards towards the brain. Its outer layer remains as a single layer of pigment epithelium; between the two lies a narrow space representing the original optic vesicle; and from its anterior border develops parts of the ciliary body and iris (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 forward 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; that 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; while 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).

Summary of ocular embryogenesis is given in Table 1.1.

In summary, the eye is essentially formed from both ectoderm and mesoderm. The ectoderm is of two types:

- (i) The neural ectoderm derived from the neural tube and
- (ii) The surface ectoderm on the side of the head (Table 1.2).

ANATOMY

The wall of the globe is composed of a dense, imperfectly elastic supporting tissue—the transparent cornea and the opaque sclera (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.

Inside the eye, posteriorly the sclera is lined by the uveal tract and retina and the globe are broadly divided into the anterior segment and posterior segment by the lens. The iris divides the *anterior segment* into an *anterior*

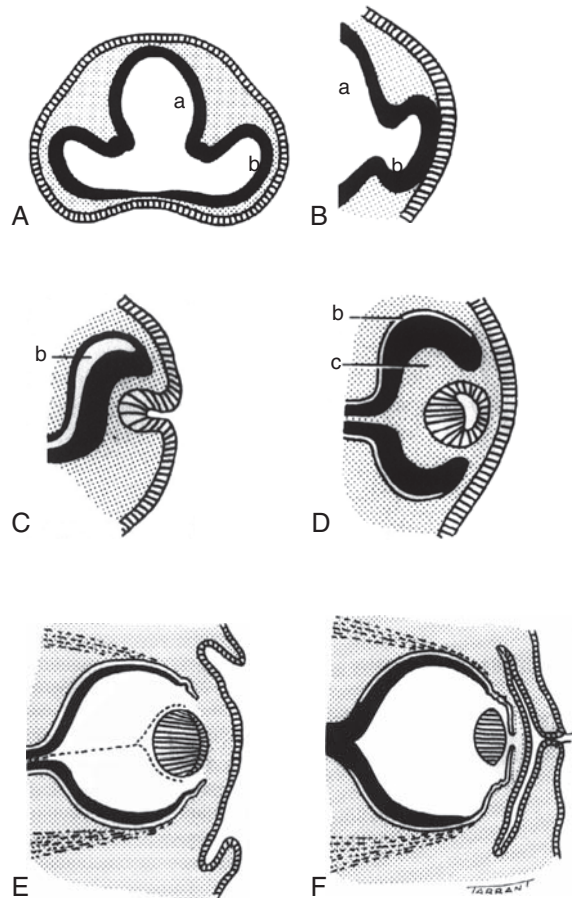


FIGURE 1.1 The development of the eye. In each case the solid black is the neural ectoderm, the hatched layer is the surface ectoderm and its derivatives, the dotted area is the mesoderm: a, cavity of the forebrain; b, cavity of the optic vesicle; c, cavity of the optic cup (or secondary optic vesicle) 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.

chamber bounded by the cornea anteriorly and **posterior chamber** bounded by the lens posteriorly. The cavity contains a clear watery fluid called aqueous humour. In the **posterior segment**, the **posterior chamber** lies behind the lens, has a cavity filled with a transparent gel-like substance called the vitreous humour and is bounded by the ciliary body and retina.

Cornea

The **cornea** is the transparent front part of the eye which resembles a ‘watchglass’ and consists of different layers and regions:

- Epithelium
- Bowman’s membrane

- Stroma (substantia propria)
- Dua’s layer (pre-Desemet’s layer)
- Desemet’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 \times . 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. 1.3). 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**

TABLE 1.1 Summary of Ocular Embryogenesis

Period After Conception	Major Milestone	
3 weeks	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	
	Migration of waves of neural crest	
	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	

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*	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

*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.

The structures are listed from anterior to posterior.

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.

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 out-pouchings 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 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. 1.4).

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 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

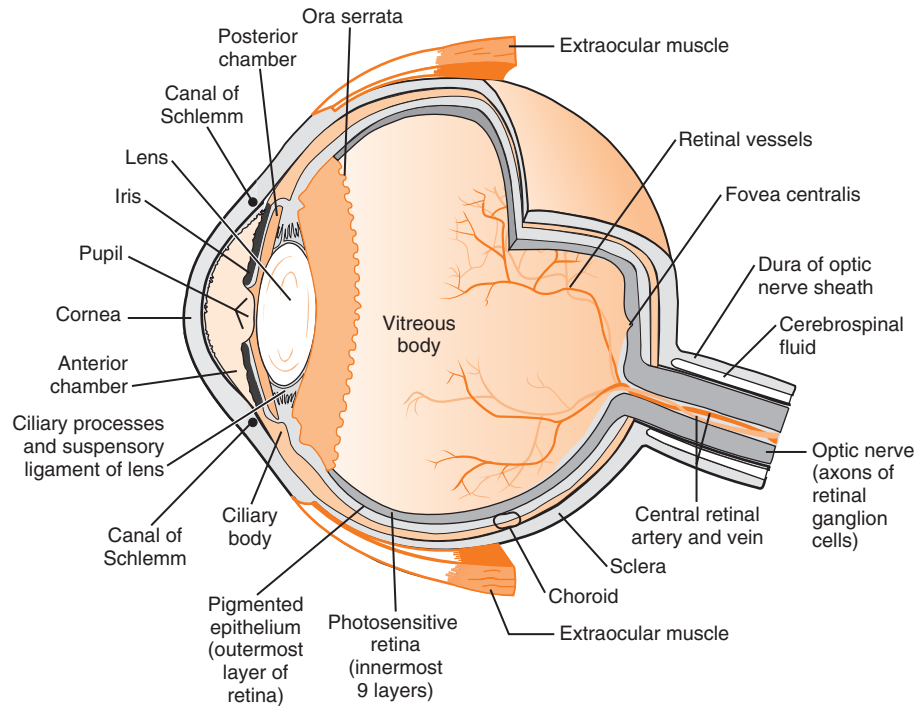


FIGURE 1.2 General anatomy of the eyeball, including its tunics and chambers. (From David Rolston, Craig Nielsen. *Chapter 2: Ophthalmology*. Rapid Review USMLE Step 3. St. Louis: Mosby; 2007. pp. 28–44)

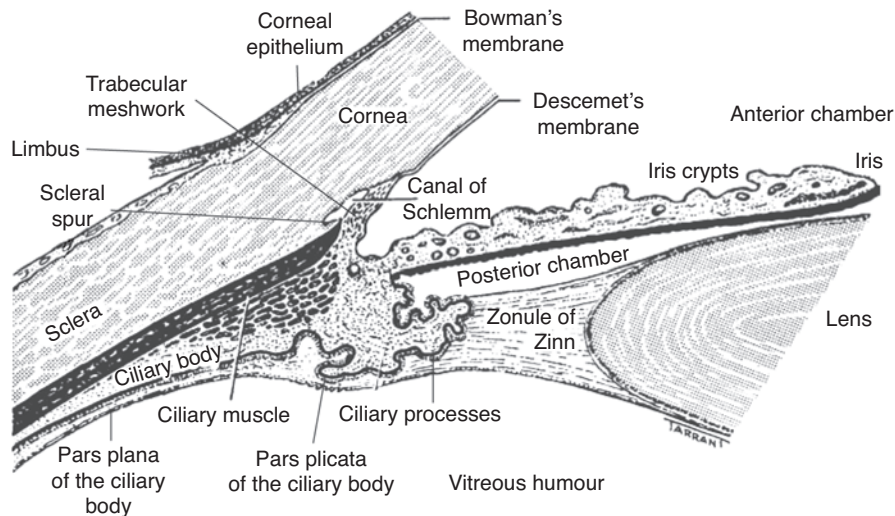


FIGURE 1.3 The region of the angle of the anterior chamber.

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 it 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