OPHTHALMOLOGY

PRINCIPLES AND CONCEPTS

FRANK W. NEWELL

FIFTH EDITION

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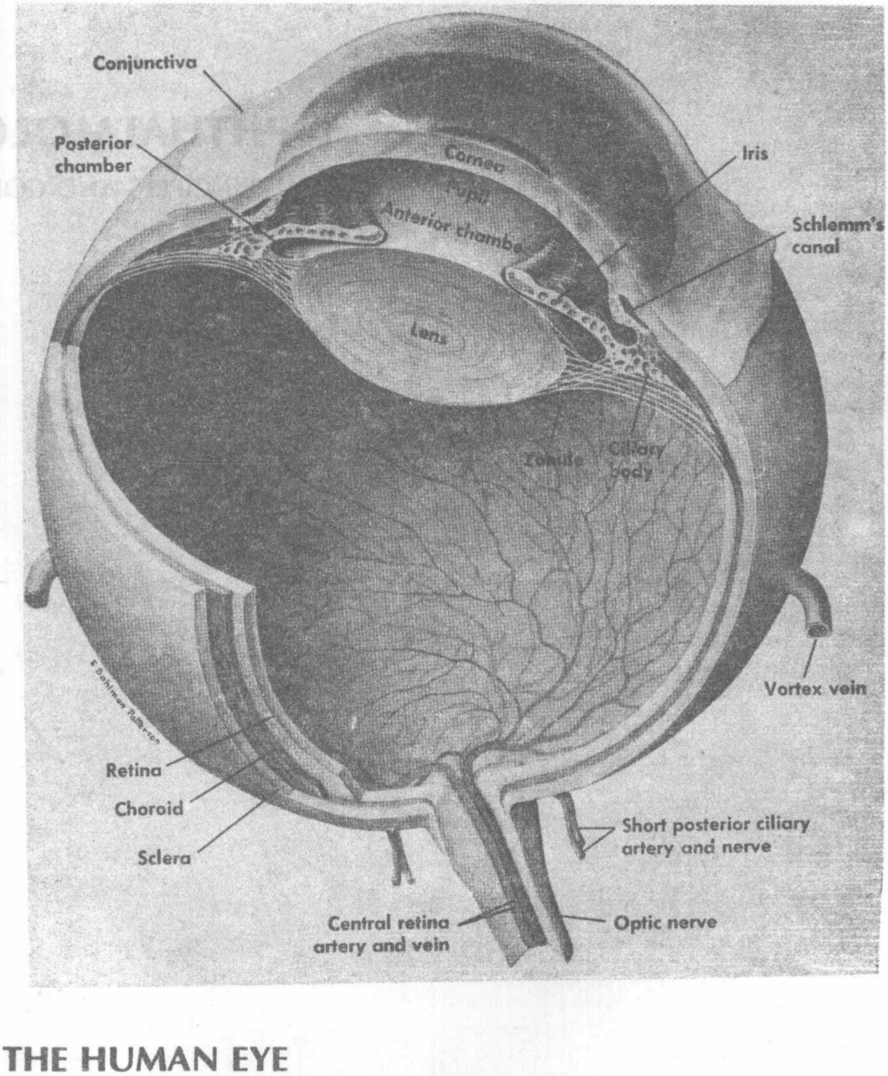

THE HUMAN EYE

PREFACE

It is gratifying to prepare this fifth edition of *Ophthalmology: Principles and Concepts* 16 years after the first edition appeared. They have been exciting years in ophthalmology, possibly the most important since the invention of the ophthalmoscope by Helmholtz in 1851 and the description of iridectomy for glaucoma by von Graefe in 1857. Photocoagulation has revolutionized the management of diabetic retinopathy and the treatment not only of ocular neovascularization but also of open-angle and angle-closure glaucoma. Closed systems of ophthalmic surgery permit early ambulation, removal of nonmagnetic intraocular foreign bodies, vitrectomy, retinal surgery, lens removal, and surgical procedures only dreamed of a decade ago. The enzymatic abnormalities in a host of disorders have been identified. Intraocular lenses minimize many of the optical disadvantages of aphakia. New drugs have emerged in the treatment of glaucoma. It has been an exciting time that in many respects has made many topics in earlier editions of this text obsolete.

I have extensively revised this edition to reflect as accurately as possible modern ophthalmology. During 1980 and 1981, the residents at the University of Chicago, Drs. Steven L. Berk, William S. Blakemore, Jr., Martin Ehrenberg, Victor M. Elner, Joseph P. Kiernan, Albert S. Leveille, Leon G. Partamian, and Larry S. Stone reviewed paragraph by paragraph the material in the fourth edition. Their corrections, suggestions, and criticism were used as a basis of revision. Dr. Leveille then reviewed the revisions and made further suggestions. This has resulted in a modern textbook that reflects the present and anticipates the future.

Ms. Kathy Hirsh, in the audiovisual department of the University of Chicago Medical Center, prepared many new illustrations. Mr. Charles Wellek, of the audiovisual department, and Mr. Ernest Heath, of the audiovisual section of the Department of Ophthalmology at the University of Chiçago, prepared many photographs. Dr. Victor Elner and Mrs. Ursula Williams provided some of the photomicrographs. Mrs. Karin Cassel, my capable assistant for 18 years, was, as always, exceptional in the care with which she prepared material. It should be evident, therefore, that I have had unusually expert assistance in preparing this fifth edition.

Frank W. Newell

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PART ONE

BASIC MECHANISMS

1

ANATOMY AND EMBRYOLOGY

ANATOMY

Dissection of a fresh animal eye readily reveals the interrelationship of the intraocular tissues and the organization of the eye as a multichambered, nearly spherical structure. The surface anatomy is easily studied in a living subject by direct inspection with a small penlight for illumination and a +20 diopter lens for magnification.

THE EYE

The eye (frontispiece) rests in the front half of the cavity of the orbit upon a fascial hammock surrounded by fat and connective tissue. Only its anterior aspect is exposed, and it is protected by the bony orbital rim. Attached to the eye are four recti and two oblique muscles. These are innervated by the oculomotor (N III), trochlear (N IV, superior oblique muscle), and abducent (N VI, lateral rectus muscle) cranial nerves, which enter the orbit through the superior orbital fissure in the posterior orbit. The ophthalmic branch of the trigeminal nerve (N V) that transmits sensory fibers from the upper face and the eye also enters the cranial cavity through the superior orbital fissure. The optic nerve leaves the orbit through the optic foramen, which also transmits the ophthalmic artery and the sympathetic innervation of the eye. The exposed anterior one third of the eye consists of a central transparent portion, the cornea, and a surrounding opaque portion, the sclera. The sclera is covered with the bulbar conjunctiva, which is continuous with the palpebral conjunctiva lining the inner surface of the protective tissue curtains, the eyelids. The lacrimal gland is located in the upper outer portion of the bony orbit.

The anterior pole of the eye is the center of curvature of the cornea. The posterior pole marks the center of the posterior curvature of the globe, and it is located slightly temporal to the optic nerve. The geometric axis is a line connecting these two poles. The equator encircles the eye midway between the two poles (Fig. 1-1).

The anteroposterior diameter of the normal eye, measured by ultrasonic methods, is about 22 to 27 mm. The circumference is between 69 and 85 mm. In the average eye (24 mm in diameter), the equator is on the surface of the sclera 16 mm posterior to the corneoscleral limbus. The posterior pole is 32 mm behind the corneoscleral limbus. The anterior termination of the sensory retina is 5.75 to 6.5 mm posterior to the termination of the Descemet membrane of the cornea (Fig. 1-2).

The globe has three main layers, each of which is further divided (see Table 1-1). The outer supporting coat consists of the transparent cornea, the opaque sclera, and their junction, the corneoscleral sulcus or limbus. The middle vascular layer, or the uvea, consists of the choroid, the ciliary body, and the iris, which contains a central opening, the pupil. The inner layer consists of the retina, which is composed of

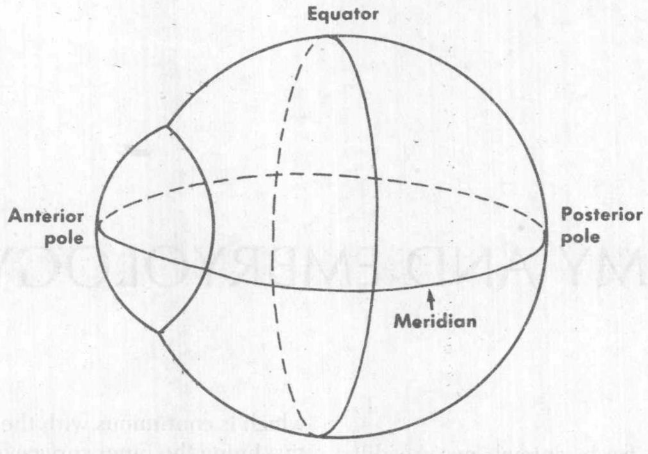

Fig. 1-1. The principal coordinates of the eye. The visual line that connects an object in space with the fovea centralis does not exactly correspond to the geometric axis.

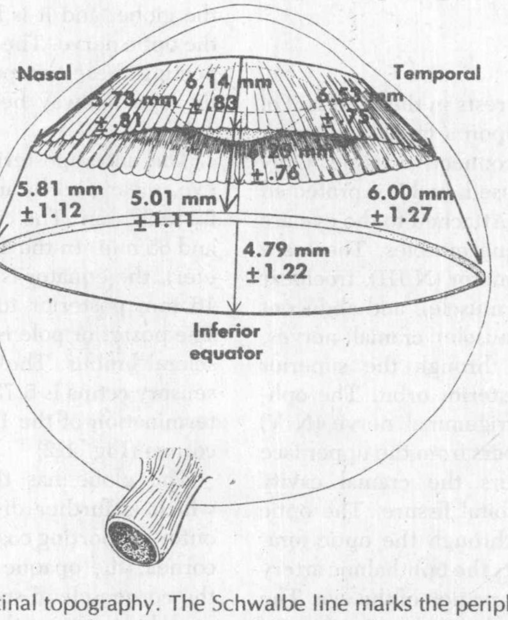

Fig. 1-2. Peripheral retinal topography. The Schwalbe line marks the periphery of the Descemet membrane of the cornea. The relationship of the Schwalbe line to the ora serrata and the relationship of the ora serrata to the ocular equator are shown in the four principal meridians. The average measurements and the standard deviation are given in millimeters. (Redrawn from Straatsma, B.R., Landers, M.B., and Kreiger, A.E.: Arch. Ophthalmol. **80:**30, 1968. Copyright 1968, American Medical Association.)

two parts, a sensory portion and a layer of pigment epithelium.

The lens is a transparent structure located immediately behind the iris and supported in position by a series of fine fibers, the zonule. These are attached to the ciliary body and the capsule of the lens.

The eye encloses three chambers: (1) the vitreous cavity, (2) the posterior chamber, and (3) the anterior chamber. The *vitreous cavity*, by far the largest, is located behind the lens and zonule and is adjacent to the retina throughout. The *posterior chamber* is minute in size and is bounded by the lens and zonule behind and the iris in front. The *anterior chamber* is located between the iris and the posterior surface of the cornea and communicates with the posterior chamber through the pupil. Aqueous humor is secreted by the ciliary processes into the posterior chamber and passes through the pupil into the anterior chamber. The trabecular meshwork opens into the canal of Schlemm, an endothelium-lined channel that encircles the anterior chamber.

Outer coat

The outer coat of the eye consists of relatively tough fibrous tissues shaped as segments of two spheres: the sclera, with a radius of curvature of about 13 mm, and the cornea, with a radius of curvature of about 7.5 mm. The white, opaque sclera constitutes the posterior five sixths of the globe, and the transparent cornea provides the anterior one sixth of the globe. The junction of the cornea and the sclera, the corneoscleral limbus, contains the trabecular meshwork and the

A

B

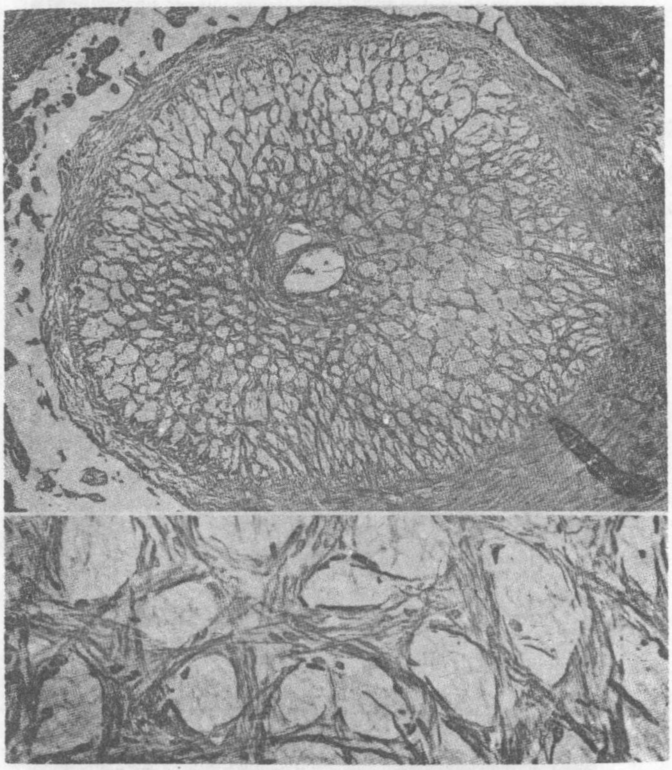

Fig. 1-3. A, Flat section of human lamina cribrosa (posterior scleral foramen). (Wilder stain; ×45.) **B,** The fibers are continuous with the scleral fibers but are lined with microglia. (Wilder stain; ×400.) (Courtesy Ramesh C. Tripathi.)

aqueous humor drainage system, the canal of Schlemm, which is an important functional and anatomic area.

THE SCLERA. The sclera is a dense, fibrous, collagenous structure that comprises the posterior five sixths of the eye. Anteriorly, it forms the "white" of the eye and is covered with richly vascular episclera, the fascia bulbi (Tenon capsule), and the conjunctiva. The fine blood vessels of the episclera are visible anteriorly through the transparent conjunctiva. Posteriorly, the sclera is connected by loose, fine collagen fibers to the dense fascia bulbi (Tenon capsule).

The sclera has two large openings, the anterior and posterior scleral foramina, and numerous smaller openings through which nerves and blood vessels pass. The sclera is perforated 3 mm medial to the posterior pole by the posterior scleral foramen, the canal through which the optic nerve passes from the eye. The canal is cone-shaped and measures 1.5 to 2.0 mm in diameter on the inner surface of the sclera and 3.0 to 3.5 mm on the outer surface. The scleral foramen is bridged by a sievelike structure, the lamina cribrosa (Fig. 1-3), the most posterior portion of which is formed by scleral fibers. The anterior portion, derived from the choroid and Bruch membrane, is rich in elastic tissue.

The anterior scleral foramen is a transitional area between the cornea and sclera. On its inner surface is the scleral spur to which the longitudinal portion of the ciliary muscle (N III) is attached. Slightly anterior to this is the canal of Schlemm. The sclera is thickest (1.0 mm) in the region surrounding the optic nerve, where the

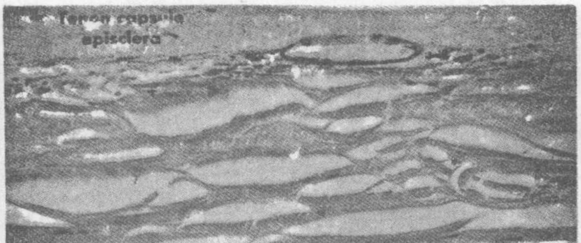

Fig.1-4. Transverse section of sclera. (Masson trichrome stain; ×160.).

meningeal coverings of the nerve blend into the sclera. It is thinnest (0.3 mm) immediately posterior to the insertions of the recti muscles. About 4 mm posterior to the equator in the region between the recti muscles are the openings for the vortex veins that are the collecting channels for choroidal veins. In the area surrounding the optic nerve, the sclera is perforated by the long and short ciliary nerves. About 4 mm posterior to the corneoscleral limbus and just anterior to the recti muscles insertions, the anterior ciliary arteries pierce the sclera at a site sometimes marked with a dot of uveal pigment. Occasionally, a loop of a long ciliary nerve extends through the sclera, returns to the ciliary body, and with uveal melanin appears as a small pigmented dot 2 to 4 mm from the corneoscleral limbus.

Structure. The sclera (Fig. 1-4) has three parts: (1) the episclera, (2) the scleral stroma, and (3) the lamina fusca.

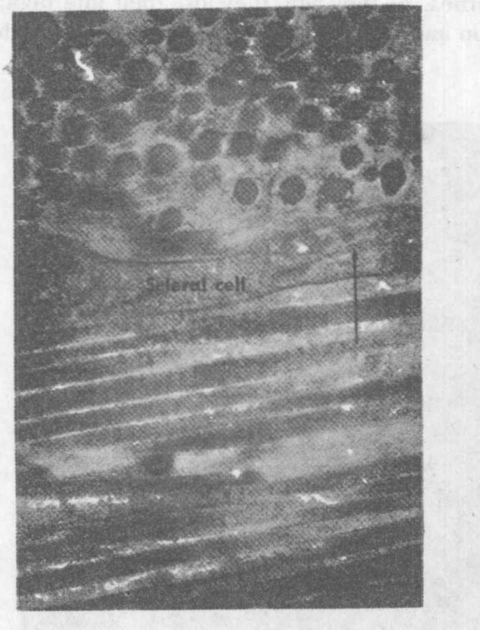

Fig. 1-5. Electron micrograph of the sclera in cross section showing collagen bundles of the sclera. The collagen fibrils in lamellae are of irregular diameter and much more irregularly arranged than those of the cornea. Part of a scleral cell is seen in the interlamellar space (*arrow*). (×40,000.) (Courtesy Ramesh C. Tripathi.)

The *episclera* is the outermost layer. It is a moderately dense, vascularized connective tissue that merges with the scleral stroma and sends connective tissue bundles into the fascia bulbi (Tenon capsule). The episclera becomes progressively thinner toward the back of the eye. Both the fascia bulbi and the episclera are attenuated behind the equator; this accounts for the relative avascularity of the posterior sclera.

The dense *scleral stroma* consists mainly of bundles of typical collagen fibers (Fig. 1-5) that vary in diameter from 10μ to 16μ and in length from 30μ to 140μ. Fibers are oriented parallel to the corneoscleral limbus to form an interlacing basket weave in that region. In the region of the insertion of the extraocular muscles, they become more meridional, apparently in response to mechanical stresses. The sclera is white because of the variable diameter and irregular arrangement of the collagen fibers of the stroma. When the water content of the sclera (usually between 65% and 70%) is reduced to less than 40% or increased to more than 80%, the sclera becomes transparent.

The *lamina fusca* is the innermost layer of the sclera; it is located adjacent to the choroid, which provides many melanocytes that give it a brown color. Fine collagen fibers blend with the choroid and form delicate connections between the choroid and the sclera.

Blood supply. The scleral stroma derives its nutrition from the episcleral and choroidal vascular network. Anterior to the insertion of the recti muscles, the anterior ciliary arteries form a dense episcleral plexus. These vessels become congested in "ciliary injection." Small branches of the posterior ciliary arteries supply the scleral stroma posterior to the recti muscles.

Nerve supply. The posterior sclera is innervated by branches of the short ciliary nerves that enter the sclera close to the optic nerve. The long ciliary nerves provide sensory innervation anteriorly. Because of the generous innervation, inflammations of the sclera are extremely painful.

THE CORNEA. The cornea is the transparent tissue at the anterior one sixth of the eye. Its anterior peripheral portion is covered with conjunctiva, whereas its posterior margin terminates at the trabecular meshwork. Anteriorly, it measures about 10.6 mm vertically and about 11.7 mm horizontally; posteriorly. it is circular with a diameter of 11.7 mm. The central portion is 0.52 mm thick with almost parallel anterior and posterior surfaces. It thickens to about 1.0 mm at the periphery. Its growth is complete in humans at about 6 years of age. The radius of curvature of the anterior surface is 7.8 mm, and the radius of curvature of the concave posterior surface is 6.2 to 6.8 mm. The cornea separates air with an index of refraction of 1.00 and aqueous humor with an index of refraction of 1.34. It is the main refracting structure of the eye. Variations in the radius of curvature in different corneal meridians cause astigmatism.

Structure. The cornea (Fig. 1-6) has three layers: (1) the epithelium and its basement membrane, (2) the substantia propria (stroma) and its anterior condensation, Bowman zone, and (3) the endothelium and its basement membrane (Descemet membrane), which separates the endothelium from the stroma.

The *epithelium* is 50μ to 90μ thick and covers the stroma anteriorly. It is continuous with the epithelium of the conjunctiva. The epithelium is stratified, five to six cell layers thick. It has an outermost layer two to three cell layers thick, a midzone layer formed by two or three layers of polyhedral cells (wing cells), and a single layer of tall, columnar germinal cells that rest upon a basement membrane. Epithelial cells form in the basal layer, become progressively flatter, and are shed from the superficial layer 7 days later. The superficial squamous cells (Fig. 1-7) have many microvilli. These cells are flat, have horizontal nuclei, and are joined to each other with zonulae occludentes that prevent aqueous solutions (such as tears) from penetrating the cornea. As they age, they lose their interdigitation and disintegrate or are swept away by the

A

B

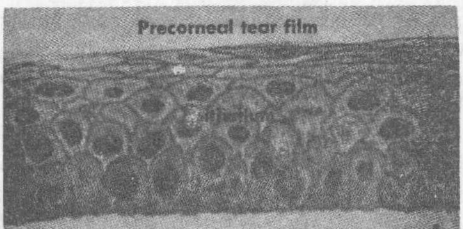

Fig. 1-6. Cross section of the axial area of the cornea. The substantia propria constitutes 90% of the thickness. **A,** The basement membrane of the epithelium is firmly adherent to the Bowman zone, the anterior condensation of the substantia propria. **B,** The lamellae of the posterior substantia propria are much more regularly arranged than those of the anterior substantia propria. (×500.) (Courtesy Ramesh C. Tripathis.)

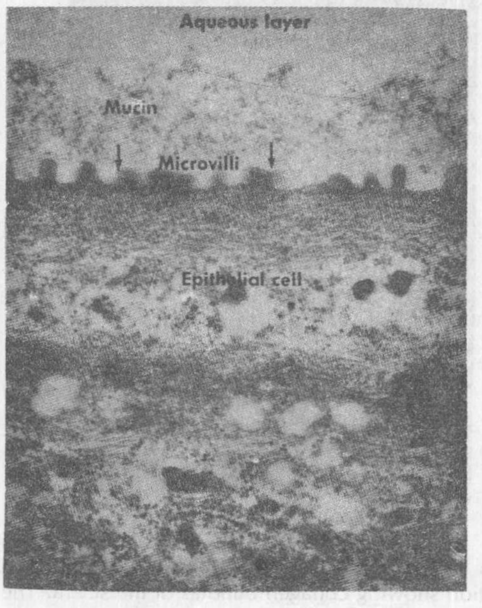

Fig. 1-7. The anterior surface of the corneal epithelium shows a degenerating epithelial cell with its microvilli covered with mucin. (×25,000.) (Courtesy Ramesh C. Tripathi.)

eyelids in blinking. Cells in the midzone are polyhedral with a convex anterior surface, parallel with the surface of the cornea, and a concave posterior surface. Those immediately adjacent to the columnar epithelium have round nuclei that become successively flatter as the cells approach the surface. The cells are joined together by desmosomes and maculae occludentes. The basal cells are tall and columnar in shape. They have a flattened base that rests on the basement membrane and is attached to it by hemidesmosomes. The interdigitating adjacent cell borders are joined by desmosomes. Cells often show mitosis. The basement membrane is PAS-positive and is firmly attached to the underlying anterior condensation of the substantia propria (Bowman zone) by irregular filaments. After injury, this attachment may take up to 6 weeks to reestablish itself, and it generally constitutes a barrier, separating superficial processes in the cornea from the underlying substantia propria.

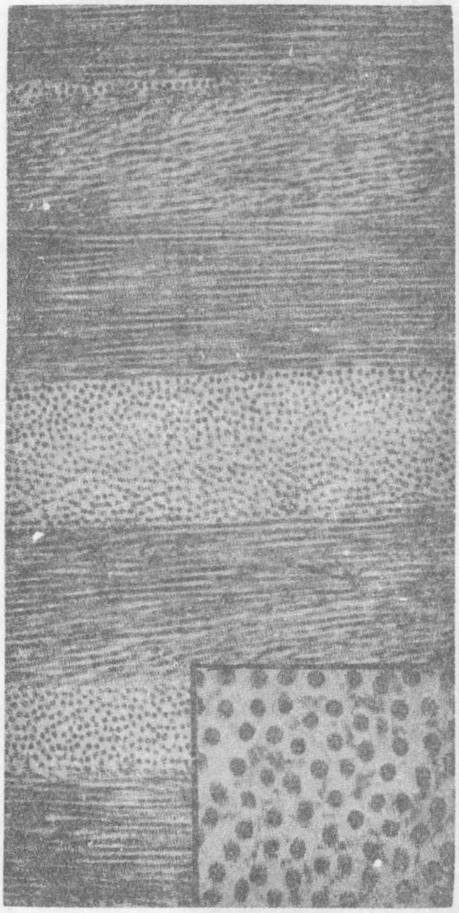

Fig. 1-8. Cross section of the substantia propria of the cornea. The collagen fibers are in bundles of approximately 200 lamellae that are arranged tangentially and at right angles to each other. Collagen fibrils are embedded in a mucopolysaccharide matrix and are separated by a distance of about 200 nm (one-half the wavelength of blue light, so that the cornea is transparent). The collagen fibrils (*inset,* ×90,000) generally are of uniform size; interspersed among them is a microfibrillar structure that may be a precursor of collagen. (×32,500.) (Courtesy Ramesh C. Tripathi.)

The *substantia propria* (Fig. 1-8), or stroma, constitutes 90% of the corneal thickness. Its anterior portion, the Bowman zone (Bowman membrane to the light microscopist), is made up of randomly oriented collagen fibers that form an acellular region resistant to deformation, trauma, and the passage of foreign bodies or infecting organisms. Once destroyed, its typical architecture is not restored, causing scarring or irregularity in corneal thickness that results in irregular astigmatism. The stroma is composed of lamellae of collagen fibers of uniform diameter that extend the entire width of the cornea. In the posterior cornea, the lamellae are of almost equal thickness; they become more irregular in the anterior portion. All fibers within a lamella are parallel but at a right angle to fibers in the adjacent lamellae. They are enmeshed in a ground substance consisting predominantly of mucoproteins and glycoproteins. Scattered throughout are fixed, long, and flat cells known as keratocytes or corneal corpuscles that function as fibroblasts do in other tissues. There are a few wandering cells (leukocytes and macrophages).

The posterior surface of the stroma is lined with a PAS-positive glassy membrane, the Descemet membrane. This is formed of atypical collagen fibers together with amorphous material and fine fibrils arranged in a hexagonal pattern. The Descemet membrane is the basement membrane of endothelial cells.

The *endothelium* of the cornea is a single layer of endothelial cells (Fig. 1-9) with centrally located large oval nuclei. These cells are rich in intracellular organelles. The posterior border is