Frederick A. Jakobiec

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Ocular Anatomy, Embryology, and Teratology

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Ocular Anatomy, Embryology, and Teratology

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Ocular Anatomy, Embryology, and Teratology is dedicated to my parents, Beatrice and Thaddeus, who are responsible for my physical and spiritual morphogenesis, and to my brother Teddy, blind from birth with retrolental fibroplasia, to whom I probably owe my interest in ophthalmology.

It is also dedicated to Victoria Ozanics, for the largest part of her career a research associate of the late George Smelser. Her quiet labors in the vineyard have yielded so much fruit for medical students, residents, and other investigators, and her contributions to this volume in her eighth decade represent the capstone of her career.

And lastly, this volume is dedicated to Ben S. Fine, M.D., whose early interest in electron microscopy has been so profitably applied to ocular anatomy and pathology, and whose personal modesty, intellectual vitality, and professional probity inform his work with its distinctive and lasting value.

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Preface

When Tom Duane approached me several years ago to organize the anatomy and embryology section of his new loose-leaf project, Biomedical Foundations of Ophthalmology (from which this hardcover volume is extracted), I had no idea that the undertaking would turn out to be so comprehensive, ambitious, and authoritative. Over 1,000 pages long and with over 2,000 illustrations and bibliographic citations, Ocular Anatomy, Embryology, and Teratology should be a valuable tool for medical students, residents, clinicians, and researchers and a unique pathfinder into the literature. I doubt that any other organ, including many that dwarf the eye in comparative size, has ever been the subject of a more extensive or detailed anatomical treatise. The devotees of the eye are enthralled by its ineffable beauty and complexity. In this volume they have paid untrammeled homage to it by documenting its anatomical relationships with all of the investigational techniques at their disposal. Early anatomists of the eve and its adnexa first labored strenuously to uncover gross anatomical relationships, and then later resorted to light-microscopic descriptions; such studies have been added to and increasingly superseded by data from transmission and scanning electron microscopy, from histochemistry and biochemistry, from autoradiography and heavy metal tracers, from freezefracture techniques, and, most recently, from hybridoma-derived monoclonal antibodies directed against specific cellular constituents.

Only a handful of individuals who are fully trained anatomists have completely dedicated themselves to the study of the structure and function of the eye, and several of these specialists have contributed to this volume. Ophthalmic pathologists are heavily represented in this enterprise, because they receive both normal and abnormal human ocular tissues in their laboratories. It should not be surprising that they are also willing to explore normal anatomic relationships to acquire a better understanding of the disease processes and "morbid anatomy" that they are expected to interpret. And, lastly, contributions have been made by an impressive number of clinical ophthalmologists who submerge themselves in anatomical studies in order to cope more intelligently with the clinical challenges of their work. From whatever direction it is approached, the eye transfixes its multifarious scholars.

No single investigator, however talented and monomaniac he may be, could ever produce a volume like this one, because he would lack the requisite material, time, investigational techniques, and knowledge to be able to encompass the entire set of topics. After having presided over this undertaking, it is with invigorated admiration that I regard the standard American textbooks on the structure of the eye that have preceded this one, namely, *Ocular Histology*, by Fine and Yanoff (Hagerstown, Harper & Row, 1979), and *Histology of the Human Eye*, by Hogan, Alvarado, and Weddell (Philadelphia, W.B. Saunders, 1971). Each of these other books is a precious and beautifully crafted study on ocular anatomy, to which the present volume should serve as a supplement and extension of our knowledge.

What this book offers is a fresh perspective on well-trod territory, reports

of newer developments, and expanded coverage of many new themes. Besides being textually longer than these other books, *Ocular Anatomy, Embryology, and Teratology* has been lavishly illustrated, with more numbered figures than are contained in the previous two textbooks put together. Since many of these figures have multiple parts, the total number of illustrations runs into the several thousands. The student of ocular anatomy is therefore presented with the luxury of learning the subject with more visual aids than have ever been available in one book before.

A few words are required to explain the design of the present volume and to highlight some of its distinctive features. When one dives into a highly detailed discussion of a special ocular tissue, certain absolute facts may be lost sight of; these "hard-core" anatomical relationships and measurements are therefore provided for the reader in one place in Chapter 1. Several chapters summarize and illustrate recent developments in our understanding of ocular, optic nerve, and adnexal embryology. For example, one of the most elegant contributions of experimental embryologists has been the elucidation of the role of the neural crest in providing much of the ocular and periocular supporting connective tissues. These chapters are therefore the first major update on ocular embryology since the classical work of Ida Mann. Throughout the textbook are scattered many chapters on functional anatomy that organize knowledge in new and exciting ways; these cover the anterior chamber angle, the vitreous, the wiring of the retina, axonal transport, and the extraocular muscles. The ocular adnexa have never been subjected to a more thorough treatment, and computed tomography has been included because it offers a new clinical dimension for appreciating adnexal anatomy. Finally, congenital anomalies have been canvassed because they represent a hieroglyphics of anatomical development gone awry and are best considered in conjunction with normal anatomy and embryology.

Ocular Anatomy, Embryology, and Teratology was many years in the making and a labor of love on the part of all who contributed. It should be the source of much pleasure and illumination for those who survey its contents.

Frederick A. Jakobiec, M.D.

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1

General Topographic Anatomy of the Eye

FREDERICK A. JAKOBIEC VICTORIA OZANICS

Many of the terms employed to describe the complicated and beautifully integrated organ by which we sense and react to light stem from the eye's overall shape as a globe. Thus, the anterior pole of the eye is the center of the corneal curvature; the posterior pole corresponds to the point of greatest curvature of the posterior segment. The optic nerve lies nasally 3 to 4 mm from the posterior pole and approximately 1 mm below the horizontal meridian. The meridians are imaginary lines connecting anterior and posterior poles.

The anatomical or geometric axis of the eye, which is also its largest sagittal diameter, coincides with the line between anterior and posterior poles. The optical axis is the straight line between the centers of curvatures of the refractive surfaces (specifically, the cornea and lens). Neither the anatomical nor optical axes are identical to the visual axis. The latter is on a straight line between the fovea centralis retinae and the nodal, or fixation, point.

One may speak of two equators. The "anatomical" equator is a line through all points around the circumference that are equidistant from the two poles. The "geometric" equator is perpendicular to the geometric axis and connects points around the largest circumference that are equidistant from the two poles. Since the shape and curvatures of the two halves of the globe are not equal, their distance from the poles would not be the same on the nasal and temporal hemispheres and the line connecting these points perpendicularly to the geometric axis would be slightly diagonal with respect to the anatomical axis.^{1,2}

The eye may be divided into three diameters:

- 1. The horizontal or anteroposterior, separating it into upper and lower halves.
- The sagittal, dividing it into nasal and temporal halves. Some asymmetry exists between these, with the temporal half being larger.
- 3. The vertical, dividing it into anterior and posterior hemispheres.

The sagittal diameter of the adult globe measures about 24 mm; its anteroposterior diameter about 23 mm. The weight of the globe has been given as 6.77 g³ or 7.5 g¹; the volume about 6.5 cc. The circumference is about 75 mm. The sagittal diameter is of importance because of its relation to refraction.

For histologic preparations one often designates meridional, tangential, or coronal (frontal) sections. The coronal plane passes at right angles to the sagittal plane, dividing the eye into anterior and posterior parts. It is identical with the vertical plane.

RELATION OF THE EYEBALL TO THE ORBIT

The globe occupies about one-fifth of the cavity, lying slightly nearer the upper and lateral sides but never in contact with the orbital bones. According to Whitnall,⁴ the average distances from the walls are

- 4.5 mm from the lateral and the superior walls
- 6.5 mm from the medial walls
- 6.2 mm from the inferior walls

The apex of the cornea is usually equidistant from the superior and inferior orbital margins. The lateral orbital margin is recessed so that about half of the eyeball is unprotected. Average distances between the equator of the globe from the orbital walls are

- 4.0 mm from the roof
- 4.5 mm from the lateral wall
- 6.8 mm from the floor
- 6.5 mm from the medial wall

The midpoints of the two pupils lie from 58 to 60 mm apart.⁴

FIBROUS COVERINGS

The cornea and the sclera form the outer fibrous envelope of the eye (Fig. 1). The anterior corneal surface is the major refracting ocular surface, accounting for 46 out of the total 50 diopters of refractive power of the eye. It is the most transparent tissue of the eye, with an index of refraction of 1.376.

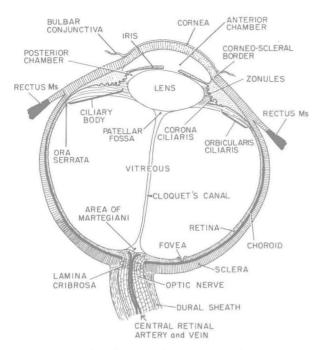


Fig. 1. Overall topographic anatomy of the eye.

The adult cornea approximates an ellipsoid in shape and extends over about one-fifth of the globe's outer aspect. The radius of curvature varies from the center to the periphery and is shortest, approximately 7.8 mm, near the center of the anterior surface. The posterior surface has a somewhat greater diameter, because the sclera slightly overlaps the anterior surface above and below. Average measurements for the external horizontal diameter are 11.75 mm; for the external vertical diameter, 10.6 mm. The anterior surface is formed by a stratified squamous epithelium and retains a precorneal tear film, whose air interface is the optical surface of the cornea. The stromal components are collagen fibrils in a regular array and fibroblasts (keratocytes) embedded in a mucopolysaccharide ground substance. The most anterior, noncellular layer of these fibrils, located beneath the epithelium, is referred to as Bowman's membrane.

The optical functioning of the cornea depends on the smoothness of its surface, regularity of curvature, and transparency. There are normally no blood vessels within this structure, but it is provided with numerous sensory nerve terminals.

The posterior mesenchymal epithelium or endothelium is a single layer of flat, hexagonal cells whose apical surface is bathed by the aqueous humor of the anterior chamber. Basally, these cells are separated from the corneal stroma back by a thick, specialized acellular collagenous sheath, Descemet's membrane. The endothelium regulates water transport from the aqueous through the stroma and epithelium, maintaining deturgescence and clarity.

Some dimensions of the corneal constituents are as follows:

Precorneal film, $7\mu m$ to $11\mu m$ Epithelium, about $50\mu m$ Bowman's membrane, $8\mu m$ to $14\mu m$ Stroma, 90% of the total thickness Descemet's membrane, $5\mu m$ to $10\mu m$ Endothelium, $5\mu m$

Total corneal thickness at the center, $510\mu m$ to $565\mu m$; at the periphery, $520\mu m$ to $720\mu m$

The remainder of the water-resistant skeleton of the eye, the sclera, covers about 80% of its total surface. Scleral thickness varies from 0.8 mm near the limbus, to 0.4 mm near the equator, and finally to 1 mm or more near the optic nerve. There are two large apertures in it: (1) the anterior foramen, closed by the cornea; and (2) the posterior scleral foramen, for the exit of the optic nerve.

The sclera is thinnest near the equator, but the width of the tissue is locally increased where the tendons of the rectus muscles insert into it (Fig. 1). It is composed of wide bundles of collagen fibrils that taper and split into intimately interwoven bands to produce a feltlike structure that gives strength and cohesiveness in all directions. The scleral collagen fibrils differ from those of the cornea in that they are larger, they vary more in diameter, they are randomly arranged, and they are coated with less abundant ground substance of different composition.

The sclera is rigid where support is essential (limbus and tendon insertions) and pliable in the posterior region around the optic foramen, where its fibrils are continuous with those of the dural sheath of the optic nerve. Scleral tissue has great tensile strength, extensibility, and flexibility, which enable it to equalize variations in the intraocular pressure.

There is no cellular investment on the outer surface. The episclera, or the superficial scleral stroma, is composed of looser, finer collagen fibers and is the locus of many vascular and venous plexuses. Anteriorly, it connects the sclera with the conjunctiva.

The fascia bulbi, or Tenon's capsule, is a thin, fibrous membrane enveloping the globe. It is connected to the episclera by delicate collagenous trabeculae. On its outer surface, Tenon's capsule is attached to the orbital muscle sheaths, to the connective tissue between the retrobulbar fat, to the

conjunctiva anteriorly, and to the suspensory ligament of Lockwood below.⁵

THE LIMBUS

All the tissues located between the termination of Bowman's and Descemet's membranes anteriorly, and the corneoscleral collagen lamellae, trabecular meshwork, the canal of Schlemm, and the tip of the scleral spur posteriorly, comprise the limbus.

The external scleral sulcus is the slight groove at the juncture of the flat scleral and more marked corneal curvatures at the limbus. It is also indicated externally by a circular area, about 1.5 mm wide and 1 mm deep—the "grey zone"—that marks the transition from clear cornea to opaque sclera.

The limbus defines a transition zone from corneal to conjunctival epithelium. Beneath the epithelium, the widened episcleral tissue of the scleral border merges with the juxtalimbal subepithelial conjunctival connective tissue and eventually condenses to become confluent with the most superficial corneal stroma.

The conjunctiva attaches the eyeball to the eyelids. It has a rich vascular supply and lymphatic drainage, the terminal loops of which are directed toward the cornea, but they do not encroach upon it.

The conjunctival epithelium contains goblet cells that are mucus glands; these glands provide the mucoid layer for the precorneal tear film.

THE ANGLE

The tissues of the angle (Fig. 2) are comprised of elements of the collagenous corneoscleral eye coats and the more cellular uveal eye coats.⁶

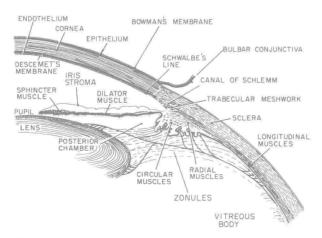


Fig. 2. Anatomical relationships of the anterior and posterior chambers.

The anterior scleral termination, facing the interior of the eye, is designated the scleral spur. It lies in the scleral groove at the limbus and is important because of its relation to the insertion of the ciliary muscle and the corneoscleral trabecular sheets.

The scleral spur is the forward extension of the posterior margin of the scleral septum or sulcus, which is a groove separating the interface of the corneal and scleral stroma.

The canal of Schlemm (Fig. 2), or sinus venosus, occupies the base of this groove. It lies on the scleral border of the trabecular meshwork, as a spongy, ringlike, completely avascular band about 1 mm wide, occupying the angle between the cornea, ciliary body, and iris. It is composed of 8 to 14 superimposed membranelike lamellae, consisting of a honeycomb of small beams or trabeculae whose openings are not in register. Anteriorly, their layers diminish to three or two and melt into the most posterior stromal lamellae of the cornea, or into Descemet's membrane. Their posterior attachment is to the tendinous fibers of the scleral spur and the connective tissue around it.

The trabecular meshwork and Schlemm's canal with its exit channels or aqueous veins constitute the "filtration angle." They are of importance in the drainage of aqueous humor from the anterior chamber through 20 to 30 excurrent channels, the aqueous veins (of Ascher), into the venus plexuses of the sclera and episclera.

DIMENSIONS OF STRUCTURAL COMPONENTS

The limboscleral junction measures about 1 mm in depth (horizontal meridian, 1.5 mm; vertical meridian, 2 mm); the width of the circular band of trabeculae is about 0.8 to 1 mm in meridional section; the width of Schlemm's canal is about 30μ m to 45μ m in meridional section.

The eye is turgid owing to the pressure within it, which is created by a pumplike secretory mechanism producing a constant influx of aqueous fluid from the ciliary processes. The aqueous drains out of the eye at a steady rate with respect to its production, thus maintaining a nearly constant intraocular pressure.⁷

THE UVEAL TRACT

The teleological function of retinal image perception, together with that of light perception, calls for complicated access aructures to form the image and to nourish, service, and protect the retina.

The choroidal coat or posterior portion of the