epithelial erosions. Scarring in the vicinity of Bowman layer may generate an irregular corneal surface, further compromising vision.

The identification of specific mutations responsible for various stromal dystrophies is generating a new molecular classification of these disorders that has been correlated with the conventional phenotypic classifications. One such dystrophy, inherited as an autosomal dominant, is caused by mutations in the *TGFB1* gene which encodes an extracellular matrix protein called keratoepithelin.Some mutations cause improper folding of this protein which in turn causes depositions in the cornea.

KEY CONCEPTS

- The cornea—not the lens—is the major refractive surface of the eye. **Keratoconus** is an example of a condition that distorts the contour of the cornea and alters this refractive surface, producing an irregular form of astigmatism.
- The normal cornea is avascular, a feature that contributes to transparency and the low incidence of graft rejection after corneal transplantation.
- Inflammations of the cornea may be accompanied by a non-infectious exudative process in the anterior chamber that may organize to distort anterior segment anatomy and contribute to secondary glaucoma and to cataract.
- **Corneal dystrophies** are generally inherited and **degenerations** are typically not inherited. Fuchs dystrophy and pseudophakic bullous keratopathy both produce visual loss through the final common pathway of corneal edema and both of these conditions are leading indications for corneal transplantation in the United States.

Anterior Segment

Functional Anatomy

The anterior chamber is bounded anteriorly by the cornea, laterally by the trabecular meshwork, and posteriorly by the iris (Fig. 29-11). Aqueous humor, formed by the pars plicata of the ciliary body, enters the posterior chamber, bathes the lens, and circulates through the pupil to gain access to the anterior chamber. The posterior chamber lies behind the iris and in front of the lens.

The lens is a closed epithelial system; the basement membrane of the lens epithelium (known as the lens capsule) totally envelops the lens. Thus, the lens epithelium does not exfoliate like the epidermis or a mucosal epithelium. Instead, the lens epithelium and its derivative fibers accumulate within the confines of the lens capsule, thus "infoliating." With aging, therefore, the size of the lens increases. Neoplasms of the lens have not been described.

Cataract

The term *cataract* describes lenticular opacities that may be congenital or acquired. Systemic diseases (e.g., galactosemia, diabetes mellitus, Wilson disease, and atopic dermatitis), drugs (especially corticosteroids), radiation, trauma, and many intraocular disorders are associated with cataract. Age-related cataract typically results from opacification of the lens nucleus *(nuclear sclerosis)*. The accumulation of urochrome pigment may render the lens nucleus brown, thus distorting the individual's perception of blue color (the predominance of yellow hues in Rembrandt's paintings later in life might have been a consequence of nuclear sclerotic cataracts). Other physical changes in the lens may generate opacities. For example, the lens cortex may liquefy. Migration of the lens epithelium posterior to the lens equator may result in *posterior subcapsular cataract* secondary to enlargement of abnormally positioned lens epithelium. The technique that is most commonly used to remove opacified lenses extracts the lens contents, leaving the lens capsule intact (extracapsular cataract extraction). A prosthetic intraocular lens is typically inserted into the eye. Residual lens epithelial cells may migrate over the lens capsule, contributing to opacification of the capsule and reduction in vision after surgery.

Occasionally, the lens cortex may liquefy nearly entirely, a condition known as hypermature or *morgagnian cataract*. High-molecular-weight proteins from liquefied lens cortex may leak through the lens capsule *(phacolysis)*. This phacolytic protein—either free or contained within macrophages—may clog the trabecular meshwork and contribute to elevation in intraocular pressure and optic nerve damage; phacolytic glaucoma is an example of secondary open-angle glaucoma.

The Anterior Segment and Glaucoma

The term *glaucoma* refers to a collection of diseases characterized by distinctive changes in the visual field and in the cup of the optic nerve. Most of the glaucomas are associated with elevated intraocular pressure, although some individuals with normal intraocular pressure may develop characteristic optic nerve and visual field changes (*normal* or *low-tension glaucoma*). The relationship between intraocular pressure and optic nerve damage is discussed later under "Optic Nerve."

To understand the pathophysiology of glaucoma it is useful to consider the formation and drainage of aqueous humor. As Figure 29-11 illustrates, aqueous humor is produced in the ciliary body and passes from the posterior chamber through the pupil into the anterior chamber. Although there are multiple pathways for the egress of fluid from the anterior chamber, most of the aqueous humor drains through the trabecular meshwork, situated in the angle formed by the intersection between the corneal periphery and the anterior surface of the iris. With this background, glaucoma can be classified into two major categories.

- In *open-angle glaucoma* the aqueous humor has complete physical access to the trabecular meshwork, and the elevation in intraocular pressure results from an increased resistance to aqueous outflow in the open angle.
- In *angle-closure glaucoma* the peripheral zone of the iris adheres to the trabecular meshwork and physically impedes the egress of aqueous humor from the eye.