TABLE 7-1

Immune-Mediated Disorders Causing Corneal Irregularities

- Atopic keratoconjunctivitis
- Mooren's ulcer
- Nonulcerative keratitis
- Ocular mucous membrane pemphigoid
- Peripheral keratitis, marginal corneal infiltrates associated with blepharoconjunctivitis
- Peripheral ulcerative keratitis associated with systemic immune-mediated diseases, including rheumatoid arthritis, Wegener granulomatosis, systemic lupus erythematosus, and inflammatory bowel disease
- Scleritis
- Stevens-Johnson syndrome (erythema multiforme major)
- Ulcerative keratitis

Infectious Diseases

Infectious diseases can cause severe corneal scarring with resultant corneal flattening. Areas of extreme irregular astigmatism are not uncommon after severe infections. Particularly devastating to the cornea are infections with acanthamoeba keratitis (AK) (Figure 7-7A), bacterial keratitis, fungal keratitis (Figure 7-7B), microbial scleritis, and sclerokeratitis.

Acanthamoeba and fusarium keratitis have recently received attention due to an increased in incidence. Acanthamoeba keratitis is caused by a free-living protozoan ubiquitous in nature. In March 2007, data collected by the Centers for Disease Control (CDC) demonstrated an increase in culture-confirmed cases of AK with wide geographic distribution. The increase in cases began in 2004 and has continued to the present. The CDC initiated a multistate investigation to look for risk factors associated with this increase in AK cases. Preliminary results of that investigation indicated an association of AK in soft contact lens wearers who used Advanced Medical Optics (Santa Ana, CA) Complete MoisturePlus (AMOCMP) multipurpose cleaning solution, and prompted a recall of the solution.[49]

On March 8, 2006, the CDC began their investigation into an increased incidence in fusarium keratitis. The proportion of fungal keratitis attributable to *Fusarium* species also varies by region, from 25% to 62%. As of April 9, 2006, a total of 109 patients with suspected fusarium keratitis were under investigation in multiple states. Case finding was conducted through postings on the Epidemic Information Exchange (Epi-X) and ophthalmology listservs and through queries of clinical microbiology laboratories. Of 30 patients for whom complete data were available during the investigation, infection onset occurred during June 15, 2005, to March 18, 2006. One hundred percent of the infected contact lens wearers reported using a Bausch & Lomb (Rochester, New York) ReNu contact lens solution or a generic-brand solution manufactured by Bausch & Lomb, and the company announced a recall of ReNu multipurpose solutions with MoistureLoc.[50]

Immune-Mediated Disorders

Immune-mediated disorders are often linked with corneal pathology and the ability of the cornea to heal. Diseases range from the very mild to devastatingly severe and are listed in Table 7-1. Treatment includes topical steroids and systemic immunosuppressive agents, but clinical findings may progress despite aggressive treatment.

Corneal Dystrophies

Corneal dystrophies are typically divided into groups according to the anatomic level of involvement: epithelium, stroma, and endothelium. The 3 main epithelial dystrophies include epithelial basement membrane dystrophy (EBMD), Reiss-Buckler dystrophy, and Meesman's dystrophy.

Epithelial basement membrane dystrophy, or map-dot-fingerprint dystrophy, is the most common epithelial dystrophy. There is typically an autosomal-dominant inheritance. This can result in recurrent corneal erosions, dry eye symptoms, irregular astigmatism, and decreased vision, depending on the severity of the disease. An example of EBMD can be seen in Figure 7-8.

Reiss-Buckler dystrophy is a bilateral, centrally located epithelial irregularity. It typically shows a dominant inheritance, has severe effects on vision, making penetrating keratoplasty more likely than in cases with EBMD, and tends to recur in grafts. Meesman's dystrophy, is a rare, bilateral disorder with unknown etiology and typically dominant inheritance.[51] Cysts are seen microscopically,