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Major review

LASIK and surface ablation in corneal dystrophies

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ABSTRACT

Corneal dystrophies are a rare group of hereditary disorders, that are bilateral, non-inflammatory, and progressive. Clinically, they can be classified based on the anatomic layer of the cornea affected. Refractive surgery and phototherapeutic keratectomy (PTK) can be performed with caution in patients with certain corneal dystrophies, but should be avoided in others. For epithelial basement membrane dystrophy, photorefractive keratectomy (PRK) is the procedure of choice for treatment of refractive error, and PTK may be performed for the treatment of recurrent erosions or irregular astigmatism. PRK and laser-assisted in situ keratomileusis (LASIK) have been associated with exacerbation of combined granular-lattice corneal dystrophy. LASIK and PRK appear to be safe in mild forms of posterior polymorphous corneal dystrophy, whereas LASIK should be avoided in Fuchs dystrophy. The safety of refractive surgery and PTK in the remainder of epithelial, Bowman layer, and stromal dystrophies has yet to be established.

1. Introduction

Corneal dystrophies are a group of inherited, bilateral, and symmetric disorders that are slowly progressive and generally have no relationship to environmental or systemic factors.[72] They are classified anatomically according to the layer of the cornea that is involved: epithelial, subepithelial, Bowman layer, stromal, Descemet membrane, and endothelial dystrophies.[72] Anterior corneal dystrophies include epithelial basement membrane, Meesmann, Lisch epithelial, Reis-Bücklers, and Thiel-Behnke dystrophy. Stromal dystrophies include granular, lattice, Avellino, macular, Schynder, fleck, and central cloudy dystrophy of Francois. The two most common posterior dystrophies are Fuchs and posterior polymorphous corneal dystrophy.

Corneal dystrophies are rare in the United States, with an estimated overall prevalence of 0.09%.[50] On the contrary, refractive error is quite common and may lead to patients with corneal dystrophies seeking refractive surgery. A number of corneal dystrophies have been reported to worsen after laser-assisted in situ keratomileusis (LASIK). We review use of and complications associated with LASIK in patients with corneal dystrophies. Photorefractive keratectomy (PRK) avoids some of these complications and may be a better option for

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correction of ametropia in eyes with corneal dystrophies, although PRK also has been associated with exacerbation of stromal dystrophies.

Phototherapeutic keratectomy (PTK), also used in the treatment of corneal dystrophies, utilizes a 193-nm excimer laser. PTK was approved by the U.S. Food and Drug Administration (FDA) in 1995 to treat anterior corneal dystrophies. FDA guidelines specify that no more than one-third of the corneal thickness be removed and that at least 250 μm of corneal stroma should remain after surgery.[66] A PTK procedure typically involves stromal ablation in the range of 40–100 μm.[18] Ideal candidates for PTK are dystrophies that involve only the anterior 10–20% of the cornea.[55,66] The main advantages of PTK are that it is minimally invasive, repeatable, allows for precise control of corneal ablation, provides a smooth base for corneal re-epithelialization, and may delay or prevent the need for invasive procedures such as lamellar or full-thickness penetrating keratoplasty. Disadvantages of PTK include the induction of a hyperopic shift, stromal haze, and postoperative discomfort.[66]

2. Epithelial and Bowman layer dystrophies

2.1. *Epithelial basement membrane dystrophy*

Epithelial basement membrane dystrophy (EBMD), also referred to as map-dot-fingerprint and Cogan microcystic dystrophy, is the most common corneal dystrophy, affecting 5–18% of the population.[15,54,58] The classic biomicroscopic findings result from a multilaminar basement membrane (resembling geographic maps and fingerprints) that blocks the normal migration of epithelial cells causing cysts to form that appear as dots.[15,39,54,58] Because of underdeveloped hemidesmosomes and anchoring fibrils, these patients are subjected to recurrent corneal erosions that typically occur from minor trauma, including friction from the eyelids during sleep, resulting in early morning pain.[54,65] They may also have decreased vision as a result of irregular astigmatism.

The poor attachments between the epithelial basement membrane and Bowman's layer make for an unstable corneal surface that is prone to sloughing during LASIK. Severe epithelial sloughing during LASIK may be the presenting sign for subclinical EBMD.[54] In a retrospective study, Dastgheib et al found that eyes with EBMD and epithelial sloughing during LASIK were predisposed to epithelial ingrowth, flap melting, and flap distortion.[15] Several other studies evaluating patients with EBMD who have undergone LASIK have also reported similar complications and recommend caution with this population.[15,54,58,67]

PRK is a safer alternative for the correction of refractive error in patients with EBMD as it may have a therapeutic effect due to the removal of the abnormal epithelium. Kymonis et al reported a single case of a myopic patient with EBMD who underwent bilateral PRK.[41] Although the patient had delayed epithelial healing and subepithelial haze formation in the first 3 months, one year postoperatively his uncorrected visual acuity was 20/20 in both eyes with no evidence of haze or recurrent erosions.

PTK is a treatment option for patients with EBMD unresponsive to conservative treatments such as lubrication and bandage contact lenses. With the removal of the abnormal epithelium and ultramicroscopic roughening of the underlying stroma and Bowman's membrane, a new and potentially more stable epithelium can regenerate.[76] In the largest series of patients with EBMD treated with PTK, Germundsson and colleagues followed 52 eyes post PTK for a mean follow-up of 43 months and found symptomatic recurrence occurred in 13% of eyes.[25] Of note, morphologic recurrence, defined as EBMD seen by slit-lamp or in vivo confocal microscopy, was present in 40% of eyes. Best spectacle-corrected visual acuity (BSCVA) remained unchanged or improved in 98% of patients.

Kremer et al followed 16 eyes for 26–42 months after a combined PRK/PTK treatment, all of which had complete alleviation of their symptoms without recurrence of erosions.[40] PTK may be used to treat symptomatic EBMD after LASIK.[20,24,61] Garcia-Gonzalez described two cases where PTK was performed to treat symptomatic EBMD one month after LASIK with good result.[24]

In a retrospective chart review comparing PTK versus diamond burr polishing of Bowman's membrane, Sridhar et al found that both groups obtained symptomatic relief[65]; patients treated with the diamond burr, however, had a lower incidence of postoperative haze and a lower recurrence rate.[65] Although effective, the diamond burr may not be the best option for patients who develop EBMD after LASIK as it could theoretically cause flap dislocation at a higher rate compared with PTK. Anterior stromal micropuncture is another less expensive alternative, although its use is limited to areas outside the visual axis, whereas PTK can safely be applied to the central cornea.

2.2. *Meesmann corneal dystrophy*

Meesmann corneal dystrophy (MECD) is a rare autosomal dominant (AD) dystrophy characterized by intraepithelial microcysts.[26] Although PRK and LASIK have not been used in MECD, Yeung and Hodge describe a 35-year old man treated with PTK.[74] There was recurrence 2 weeks later, with deterioration of vision due to haze and subepithelial fibrosis. The patient was retreated with manual keratectomy twice followed by manual keratectomy with mitomycin-C (MMC). There was minimal recurrence after 3 years' follow-up, and they conclude that topical MMC in conjunction with keratectomy may prevent the recurrence or mediate stabilization of MECD.

2.3. *Lisch epithelial corneal dystrophy*

Lisch epithelial corneal dystrophy (LECD) affects the epithelial basement membrane, causing densely crowded intraepithelial cysts, best appreciated with retroillumination.[73] Its appearance has been described as a "band shaped and whorl microcystic dystrophy."[9] Associated with the Xp22.3 gene locus, this disease is considered genetically unique from the other epithelial dystrophies.[39,73]

Despite various therapeutic measures including epithelial debridement and/or the use of contact lenses, there remains