



CORNEAL DYSTROPHIES



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EPITHELIAL CORNEAL DYSTROPHY

| Type | Cogan | Meesman | Lisch |
|-------------|--|---|------------------------------------|
| Inheritance | Sporadic, AD | AD | AD, X-linked |
| Symptom | Recurrent corneal erosion | Ocular irritation | Ocular irritation |
| Sign | Dot-like, map-like, epithelial microcyst, finger print. | Myriad intraepithelial cyst, thin cornea, reduced sensation | Whorled configuration, micro cyst. |
| Histology | Thick BM with deposition of fibrillary protein between BM and Bowman's layer, Absent hemidesmosome | Thick BM. | Thick BM. |
| T/t | Lubrication | Lubrication | Lubrication |

BOWMAN'S AND ANTERIOR STROMAL DYSTROPHY

| Type | CDB1 | CDB2 | Schnyder |
|-------------|--|---|--|
| Inheritance | AD | AD | AD |
| Symptoms | Recurrent corneal erosion. | Recurrent corneal erosion. | ↓ VA, Glare. |
| Signs | Fine, round, polygonal subepithelial opacities, visual impairment due to scarring of BL. | Subepithelial opacities (honeycomb) configuration involving central cornea. | Central crystalline opacity. |
| Histology | B.L & B.M replaced by fibrous tissue. | B.L replaced by curly fibers. | Phospholipid & cholesterol deposition. |
| Treatment | Lubricant, E.L keratectomy | Not necessary | E.L keratectomy |

STROMAL CORNEAL DYSTROPHY

| Type | Lattice I | Lattice II | Lattice III |
|-------------|--|--|------------------------------------|
| Inheritance | AD | AD | AD |
| Symptoms | Recurrent corneal erosion | Recurrent corneal erosion | Photophobia, ↓VA, Watering. |
| Signs | Glassy dots, fine lattice, outward spread, stromal haze, ↓corneal sensation | Radial periphery located short, fine lattice lines, ↓corneal sensation. | Thick rope-like band lattice. |
| Histology | Amyloid deposition | Amyloid deposition | Amyloid deposition |
| Treatment | Keratoplasty if affect the vision. | Keratoplasty if affect the vision. | Keratoplasty if affect the vision. |



TABLE 1

Corneal Dystrophies Categorized by Corneal Layer with Inheritance and IC3D Category (Weiss et al, 2008)

| DYSTROPHY | Inheritance | IC3D category |
|--|------------------|---------------------|
| EPITHELIAL AND SUBEPITHELIAL | | |
| Epithelial basement membrane dystrophy (EBMD) | Sporadic | 1 (when applicable) |
| Epithelial recurrent erosion dystrophy (ERED) | AD | 4, 3 |
| Subepithelial mucinous corneal dystrophy (SMCD) | AD | 4 |
| Meesmann's corneal dystrophy | AD | 1 |
| Lisch epithelial corneal dystrophy | XD | 2 |
| Gelatinous drop-like corneal dystrophy | AR | 1 |
| BOWMAN'S LAYER DYSTROPHIES | | |
| Reis-Buckler's corneal dystrophy | AD | 1 |
| Thiel-Benke corneal dystrophy | AD | 2 |
| Grayson-Wilbrandt corneal dystrophy | AD | 4 |
| STROMAL DYSTROPHIES | | |
| Lattice type 1 corneal dystrophy | AD | 1 |
| Granular corneal dystrophy | AD | 1 |
| Macular corneal dystrophy | AR | 1 |
| Schnyder corneal dystrophy | AD | 1 |
| Congenital stromal corneal dystrophy | AD | 1 |
| Fleck corneal dystrophy | AD | 1 |
| Posterior amorphous corneal dystrophy | AD | 3 |
| Central cloudy dystrophy of Francois | Unknown | 4 |
| Pre-Descemet's corneal dystrophy | Unknown | 4 |
| DESCEMET'S MEMBRANE AND ENDOTHELIAL DYSTROPHIES | | |
| Fuchs' endothelial corneal dystrophy, late onset | Unknown, some AD | 2 |
| Fuchs' endothelial corneal dystrophy, early onset | AD | 1 |
| Posterior polymorphous corneal dystrophy 1 | AD | 2 |
| Posterior polymorphous corneal dystrophy 2 | AD | 1 |
| Posterior polymorphous corneal dystrophy 3 | AD | 1 |
| Congenital hereditary endothelial dystrophy 1 | AD | 2 |
| X-linked endothelial corneal dystrophy | XS | 2 |