MEMORIAL INSTITUTE FOR OPHTHALMIC RESEARCH

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Corneal Dystrophies; Easy to diagnose but hard to remember

Dr. Mohamed Abdelmoniem Mohamed Sedik
Ophthalmology resident, MIOR

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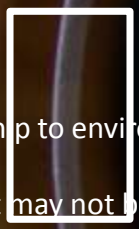
Corneal dystrophies are group of progressive, usually bilateral, mostly genetically determined, non-inflammatory opacifying disorders.



High magnification with Retro-illumination view

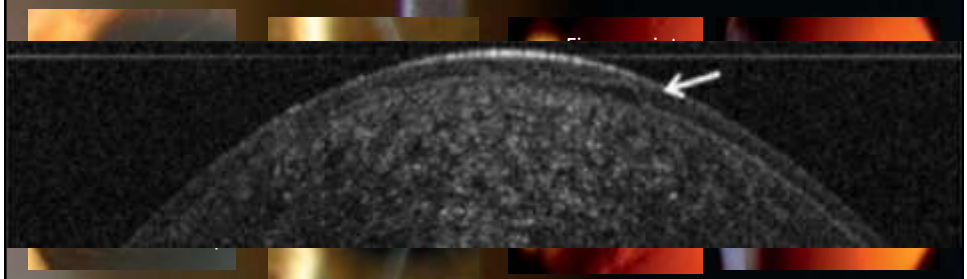
1. Bilateral.
2. Symmetrical.
3. Inherited condition.
4. Little or no relationship to environmental or systemic factors.
5. Begin in early life but may not become clinically apparent until later.
6. Slowly progressive.
7. Absence of inflammation.

CLASSIFICATION IN LAYERS

Epithelial dystrophies

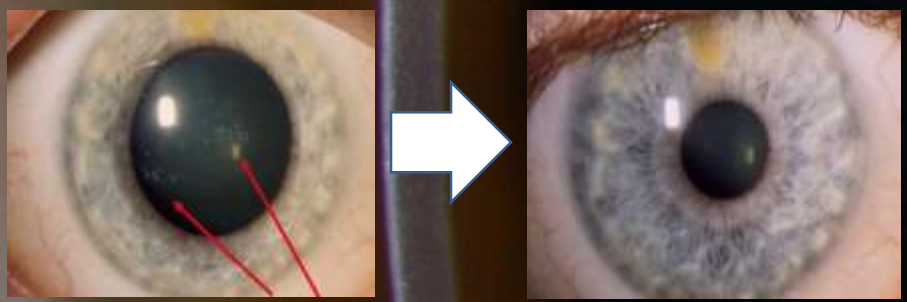
Cogan (epithelial basement membrane) dystrophy

- AD
- Absence of hemidesmosomes → Recurrent corneal erosions.
- Pain , lacrimation & blurred vision.



Treatment:

- Debridement.
- Lubricants.
- Bandage soft Contact Lens.
- Anterior stromal reinforcement or puncture.
- Phototherapeutic keratectomy (PTK).
- Superficial keratectomy – DALK.



Epithelial dystrophies

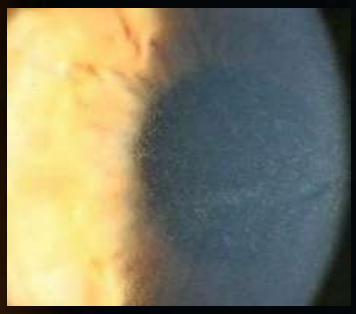
Meesmann epithelial dystrophy

•AD •Very rare, non-progressive

•Irregular thickening of the epithelial basement membrane & intraepithelial cysts maximal centrally & extend towards but do not reach the limbus.

•Asymptomatic or ocular irritation may begin in the first few months of life.

•There is a high risk of recurrence with debridment and keratectomy.



Epithelial dystrophies

Epithelial Recurrent Erosion Dystrophy

•AD

•Erosions seen during 4-6 years of age sometimes as early as 8 months of age & generally decline in frequency & intensity & cease by 50 years.

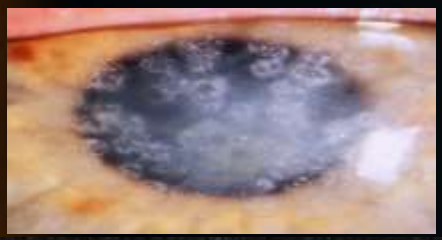
Subepithelial mucinous corneal dystrophy

•AD

•Bilateral subepithelial opacities most dense centrally,

•Onset is in first decade of life & progressive loss of vision occurs in adolescence.

• Painful episodes of recurrent corneal erosion which decrease during adolescence.

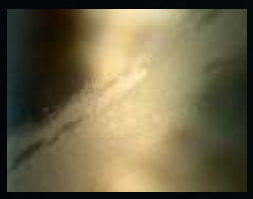


Epithelial dystrophies

Lisch-epithelial corneal dystrophy

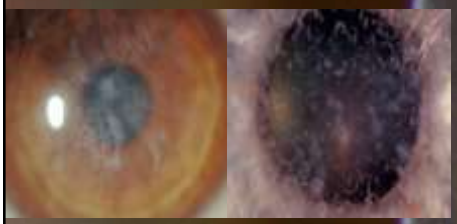
- AR
- X- Linked dominant
- Epithelial debridement or PTK
- Typically recurs

- Occurs in childhood with slow progression of localised gray opacities in different pattern: whorl like radial, band shaped ,flame.
- Asymptomatic, blurring of vision occurs if pupillary zone is involved.

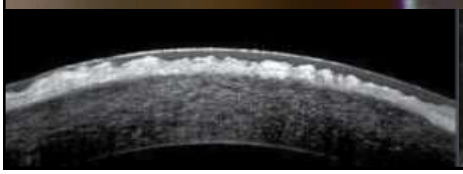


Reis–Bücklers corneal dystrophy

- AD
- Replacement of Bowmen and epithelial BM with fibrous tissue → recurrent corneal erosions.



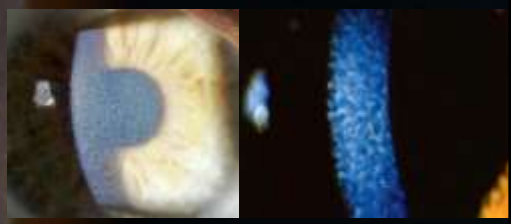
- Polygonal subepithelial opacities, most dense centrally.
- V.A affected due to scaring at the level of Bowman layer.



Bowman layer/anterior stromal dystrophies

Thiel–Behnke corneal dystrophy

- AD
- Recurrent corneal erosions



- Honey comb shaped
- OCT revealed saw-toothed deposits on top of the Bowman layer; the anterior stroma optically clear.

Lattice corneal dystrophy

Stromal dystrophies

•AD

•Recurrent erosions which precede typical stromal changes.

•Anterior stromal, glassy, fine lattice llnes, best seen on retroillumination.
•Deep & outward spread sparing the periphery.

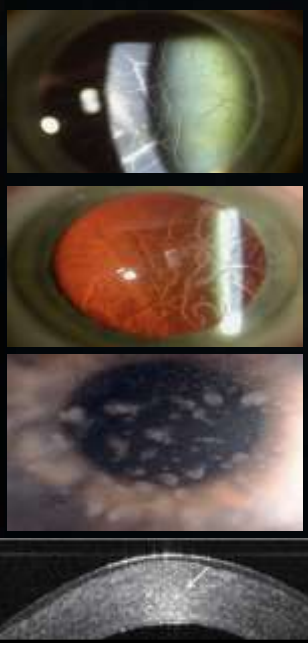
•Progressive cranial and peripheral neuropathy, mask-like facies and autonomic features in type 2.

•PTK
•PKP or DALK → Recurrence in graft may occur.

Macular corneal dystrophy •AR

•Anterior stromal haze involving central cornea.
•Greyish- white, dense, focal, poorly delineated spots in anterior stroma centrally & posterior stroma in the periphery.

•PTK
•PKP or DALK → Late recurrence in graft may occur.



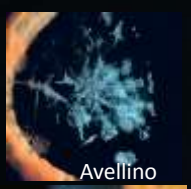
Stromal dystrophies

Granular corneal dystrophy •AD

•Onset is in 1st decade but vision is usually not affected in early stage of disease

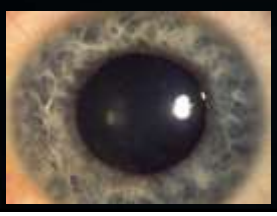
•Small, white, sharply demarcated deposits in central anterior stroma, overall pattern of deposition is radial or disc shaped or may be in the form of a christmas tree.

•PTK
•PKP or DALK is usually required by 5th decade and superficial recurrences may require repeated excimer laser PTK.



Avellino

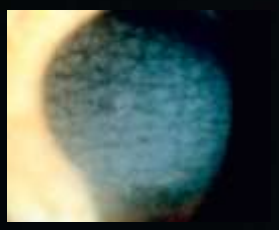
Granular corneal dystrophy before and 3 months after excimer laser PTK



Stromal dystrophies

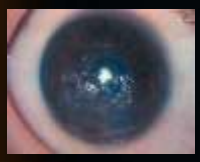
François central cloudy dystrophy

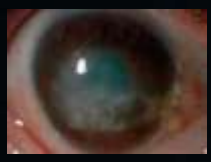
- AD
- Polygonal, cloudy grey opacities separated by relatively clear spaces.
- Posterior stroma most prominent centrally, creating a leather-like appearance.



Gelatinous drop-shaped corneal dystrophy

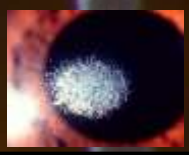
- AR
- Subepithelial nodules (Mulberry like) with anterior stromal involvement due to accumulation of amyloid.
- Severe photophobia, watering & visual impairement





Schnyder dystrophy

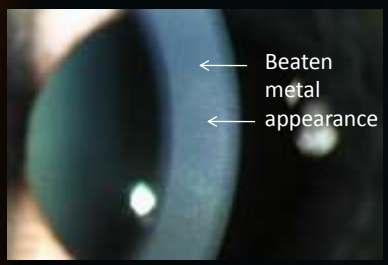
- AD
- Subepithelial crystalline opacity
- Diffuse corneal haze
- Excimer laser Keratectomy

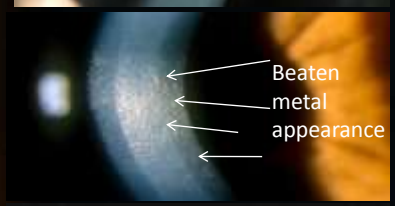


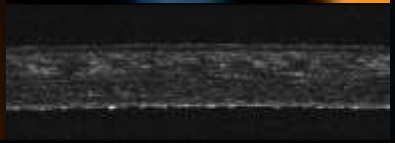
Descemet membrane and endothelial dystrophies

Fuchs endothelial corneal dystrophy

- AD or Sporadic
- Slowly progressive, bilateral, affecting females more than males usually 5th & 6th decade
- Open angle glaucoma is common association
- Cornea guttata, Tiny dark spots caused by disruption of regular endothelial mosaic.
- Endothelial decompensation gradually leads to central stromal oedema & blurred vision, worse in the morning & clearing later through the day.
- Epithelial oedema develops when stromal thickening has increased by about 30% → Persistent epithelial oedema → bullous Keratopathy → which causes pain & discomfort due to exposure of nerve endings.







Posterior polymorphous corneal dystrophy

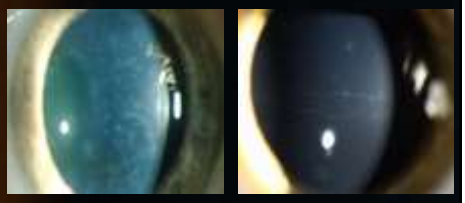
Descemet membrane and endothelial dystrophies

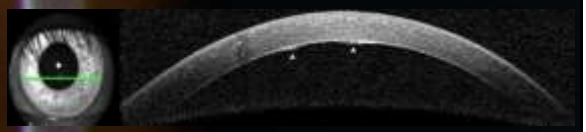
•AD

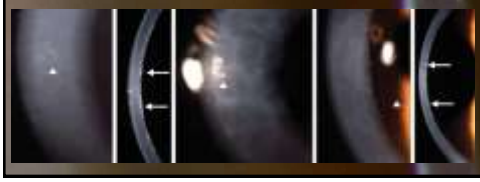
•Onset is at birth or soon thereafter, although it is most frequently identified by chance in later life.

•Subtle vesicular endothelial lesions that may become confluent band-like lesions or diffuse opacities which may be asymmetrical.

1. Iris abnormalities
2. Glaucoma
3. Alport syndrome







Take home massage to remember;

•Never forget to examine both eyes

•Family history, Past history and present history

•Retro-illumination and red reflex with dilated pupils

•Anterior segment OCT is very important to detect the pathology layer and treatment plan

•Moderate visual accuity is better to follow up rather than keratoplasty due to high recurrence

•Ask for systemic and ocular associations

•PTK with mytomycin is still a superior choice before keratoplasty


thank
you