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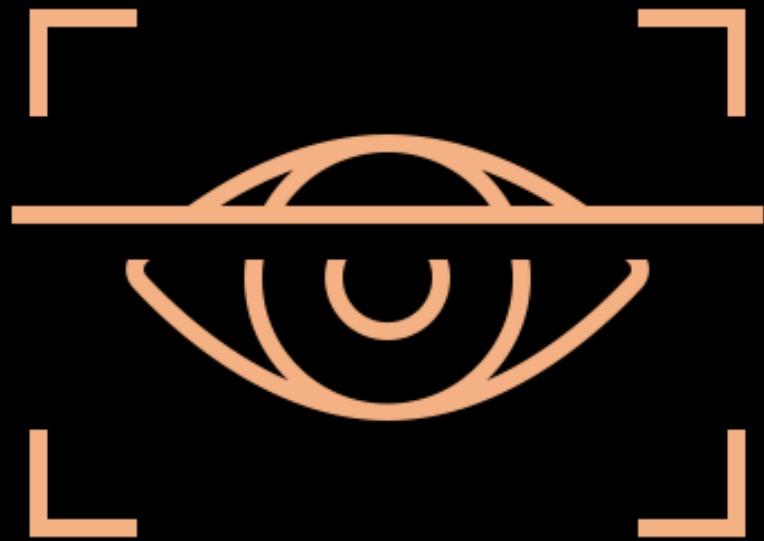
Fellowship in Glaucoma (Al-Shifa Trust, Pak)

Fellowship in Glaucoma (Univ. of Toronto, Canada)

Advance Fellowship in Glaucoma (BPOS, UK)

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# Corneal Ectasia Dystrophies & Degenerations

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# Learning objectives



Discuss the etiology, clinical features, investigation and management of keratoconus.



Give an overview of corneal dystrophies and degenerations.

# Corneal Ectasia

Abnormal shape of the cornea.

- Keratoconus
- Pellucid Marginal Degeneration
- Keratoglobus

# Keratoconus

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- Keratoconus is a progressive disorder.
- Cornea assumes a conical shape secondary to stromal thinning & protrusion.
- Onset around puberty
- Both eyes are affected.

# Keratoconus

- Presentation
  - Typically, during puberty
  - Unilateral impairment of vision due to progressive myopia & astigmatism.
  - 50% of normal fellow eyes progress to keratoconus within 16 years.

# Keratoconus

## Diagnosis

- Hallmark – central or paracentral stromal thinning, atypical protrusion & irregular astigmatism.
- Graded by keratometry according to the severity
  - Mild < 48 D
  - Moderate 48 – 54 D
  - Severe > 54 D

# Keratoconus

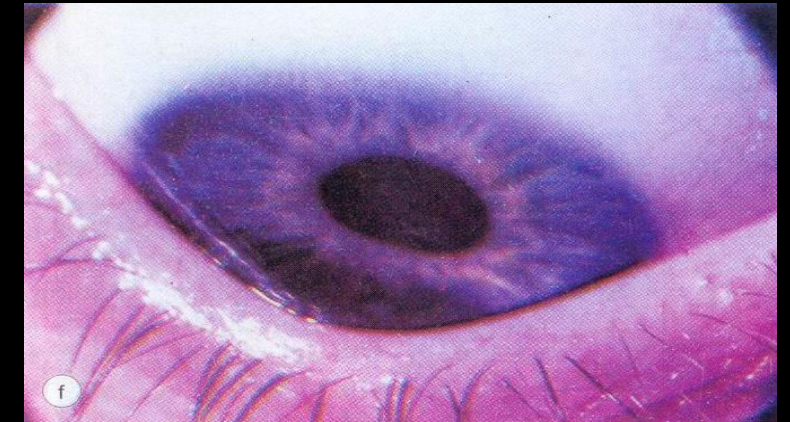
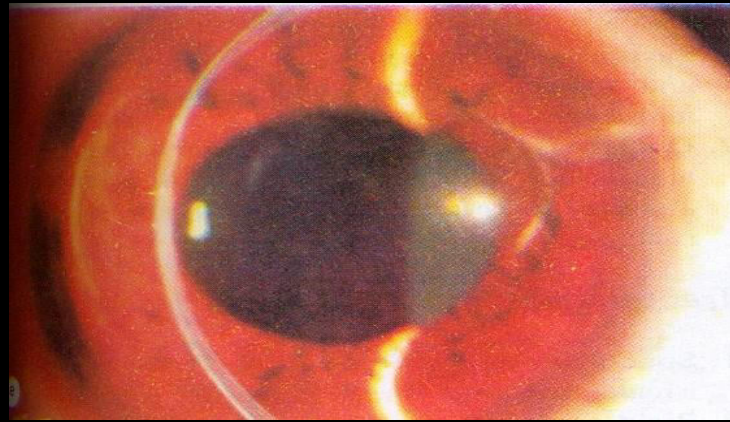
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- Signs
  - Direct ophthalmoscopy – oil droplet reflex
  - Retinoscopy – irregular scissor reflex
  - Slit lamp – fine vertical deep stromal striae
  - Epithelial iron deposits – surround the base of the cone (Fleischer ring)





# Diagnosis

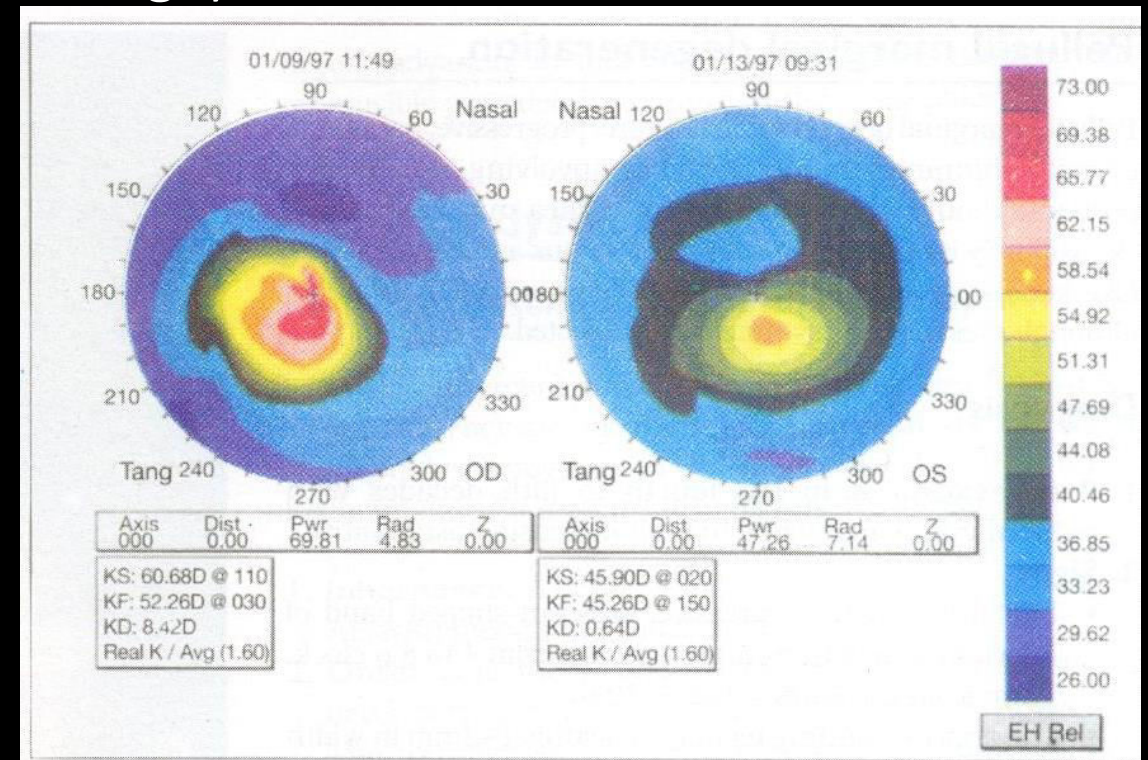


- Signs (Cont'd)

- Progressive corneal thinning
- Bulging of lower lid in down-gaze (Munson sign)

- Corneal Topography

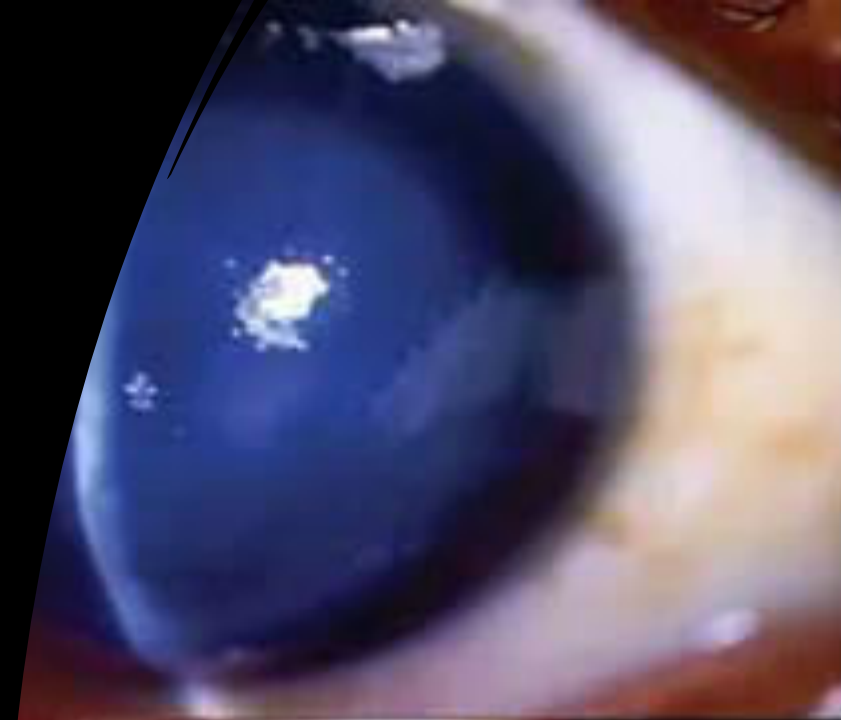
- Irregular astigmatism



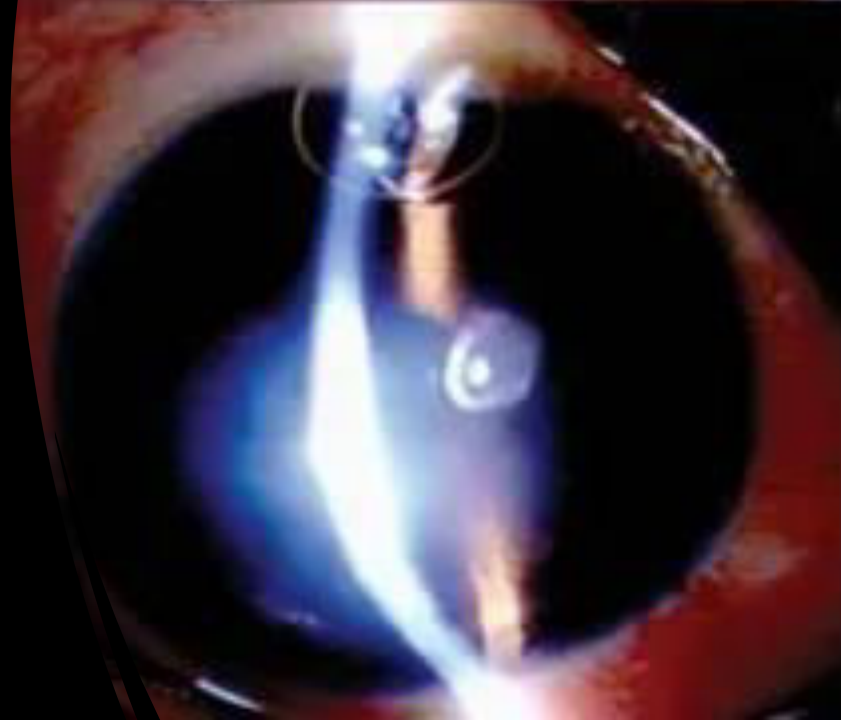
# Acute hydrops

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- Rupture in Descemet's membrane allows an influx of aqueous into the cornea.
- A sudden drop in visual acuity.



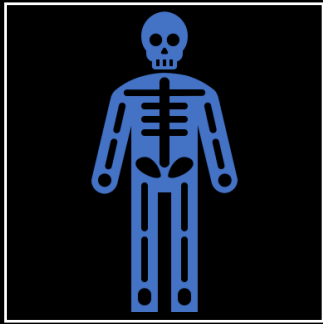
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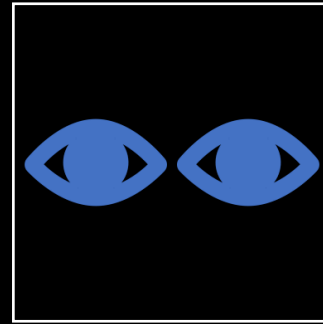


# Associations



## Systemic disorders

Down, Turner, Ehlers – Danlos syndrome, Marfan syndrome, Atopy.



## Ocular associations

Vernal keratoconjunctivitis, blue sclera, Aniridia, ectopia lentis, Retinitis pigmentosa

# Treatment



## Spectacles

In early cases

Regular & irregular astigmatism



## Rigid contact lenses

For higher degrees of astigmatism

Provide a regular refracting surface

Corneal cross-linking

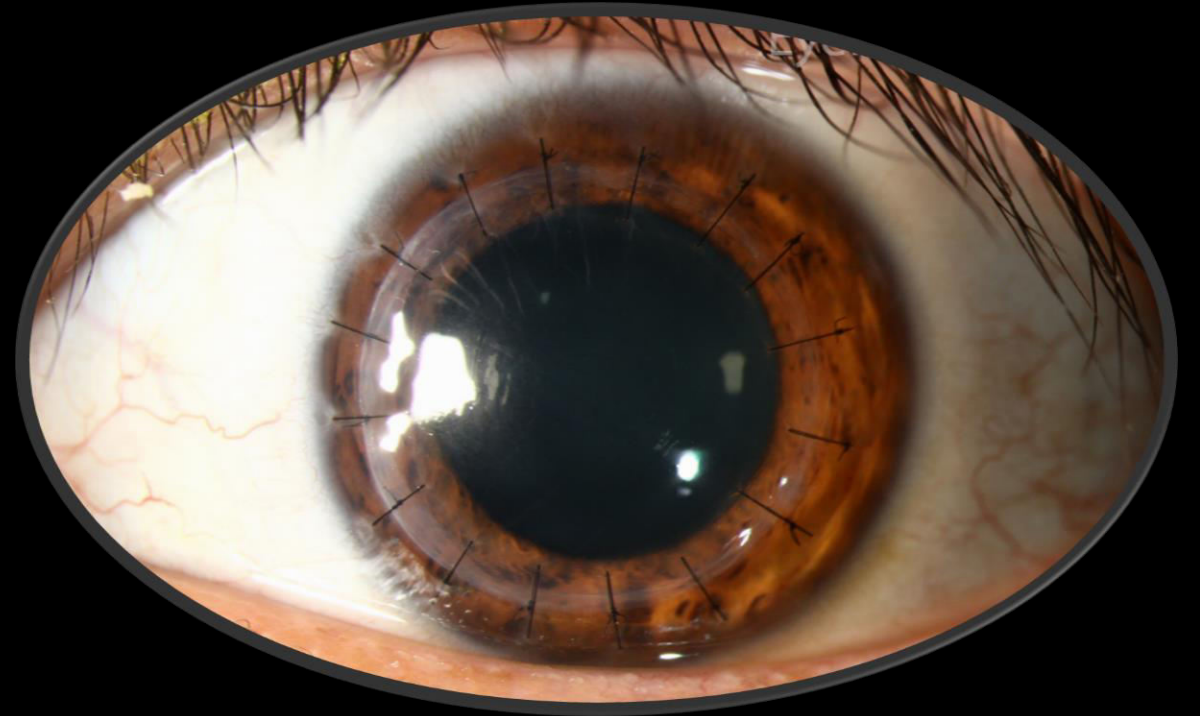
Intrastromal corneal rings



# Treatment

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- Keratoplasty
  - Penetrating or deep lamellar
  - Advanced progressive disease with corneal scarring
  - Clear grafts in 85% of cases



# Corneal Dystrophies

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A group of progressive disorders

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Bilateral, genetically determined

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Non-inflammatory opacifying disorders

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Age at the presentation – 1st & 4th decade.

# Classification

Based on biomicroscopic & histopathological features

Corneal dystrophies classified into

- Epithelial
- Bowman layer
- Stromal
- Endothelial

# Classification (Cont'd)

## Epithelial Dystrophies

- Epithelial basement membrane dystrophy.
- Meesman dystrophy
- Lisch dystrophy

## Bowman layer dystrophy

- Reis Buckler dystrophy
- Thiel Behnke dystrophy
- Central Schnyder dystrophy



# Classification (Cont'd)

- Stromal dystrophy
  - Lattice dystrophy 1
  - Lattice dystrophy 2
  - Lattice dystrophy 3 & 3A
  - Granular dystrophy type 1
  - Granular dystrophy type 2
  - Macular dystrophy
  - Gelatinous drop-like dystrophy
  - Central cloudy dystrophy of Francois

# Classification (Cont'd)

- Endothelial dystrophies
  - Fuchs endothelial dystrophy
  - Posterior polymorphous dystrophy
  - Congenital hereditary endothelial dystrophy

# Epithelial dystrophy

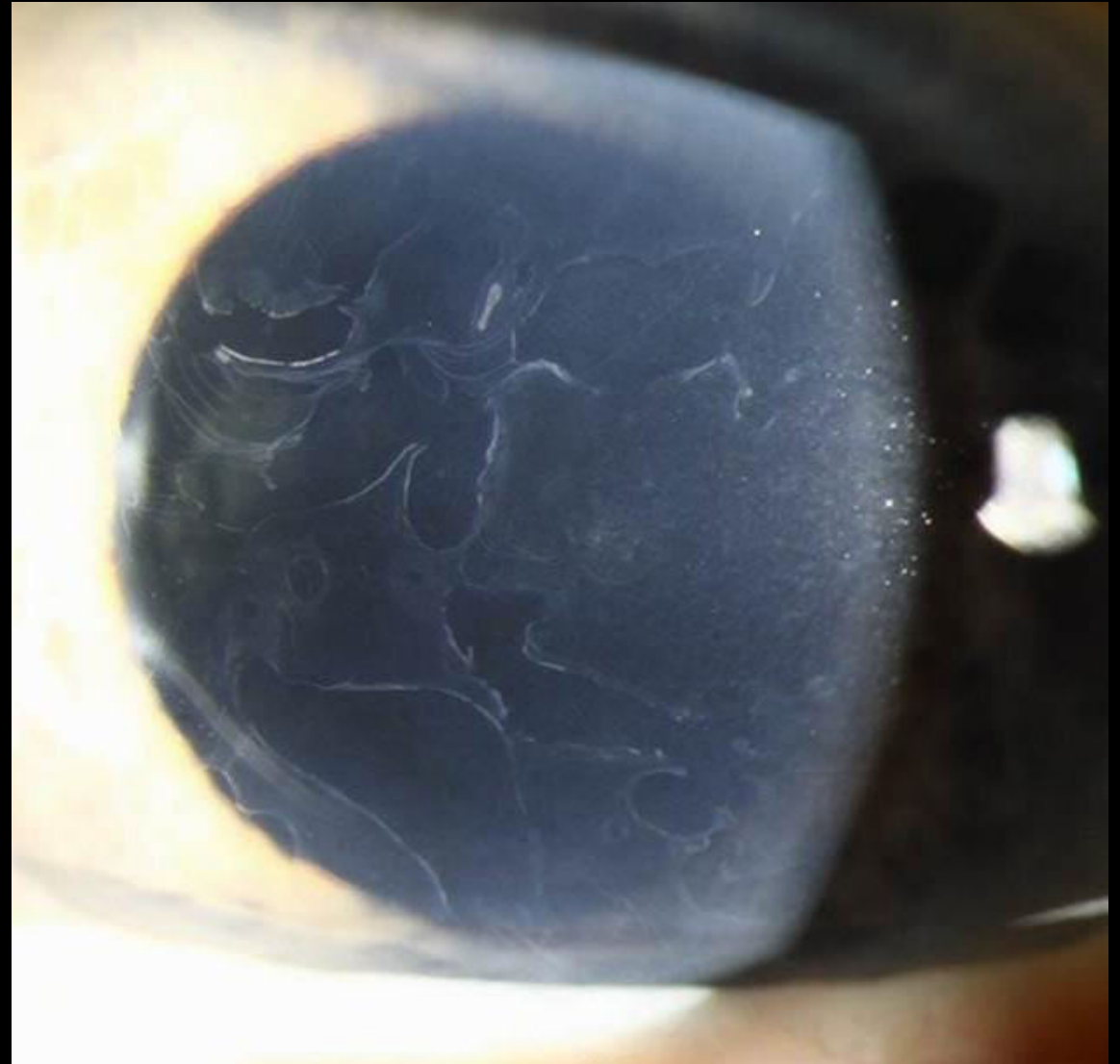
## Epithelial basement membrane Dystrophy

- Most common dystrophy
- Sporadic, rarely AD
- Onset 2nd decade
- 10% develop recurrent corneal erosion

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- Sign

- Dot-like opacities / epithelial micro-cysts
- A sub-epithelial map-like patterns
- Whorled fingerprint-like lines



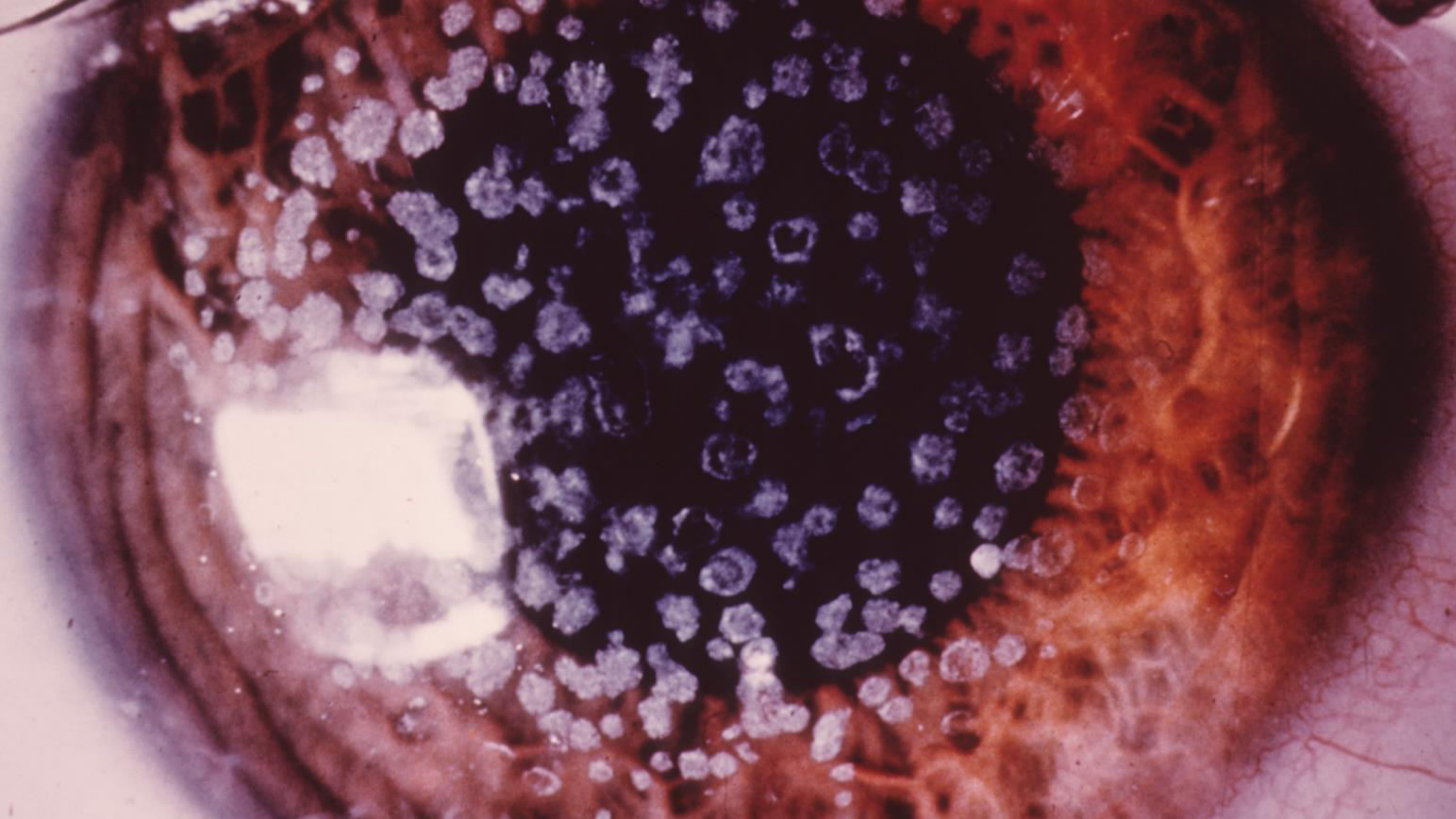
# Granular dystrophy type 1

Inheritance AD

Onset – 1st decade

Signs

- Small, sharply demarcated deposits resembling crumbs, and rings in the central anterior stroma.
- Central stroma between opacities is clear.
- Deposits spread outward, but not reaching limbus



# Macular Dystrophy

- Least common stromal dystrophy.
- Systemic inborn error of keratan sulfate metabolism
- Has corneal manifestations
- Inheritance AR
- Onset end of 1st decade with gradual impairment.







# Fuchs endothelial dystrophy

- A bilateral disease
- Accelerated corneal endothelial cell loss
- AD
- Slowly progressive disease in old age

# Stages

1

## Cornea guttata

- Irregular warts or excrescences of Descemet's membrane are secreted by abnormal endothelial cells

2

- Endothelial decompensation – central stromal edema

3

- Persistent epithelial edema – micro-cysts & bullae (bullous keratopathy)

# Treatment

- Topical
  - sodium chloride 5% drops or ointment.
- Bandage Contact lenses
  - Provide comfort by protecting exposed nerve endings.
- Penetrating Keratoplasty
  - High success rate.

# Corneal Degenerations

- Any of the several tissue changes
- Occur in previously normal tissues
- As a result of prior diseases.

- Classification

- Age-related degeneration
- Arcus Senilis
- Vogt limbal girdle
- Corneal Farinata
- Crocodile shagreen.

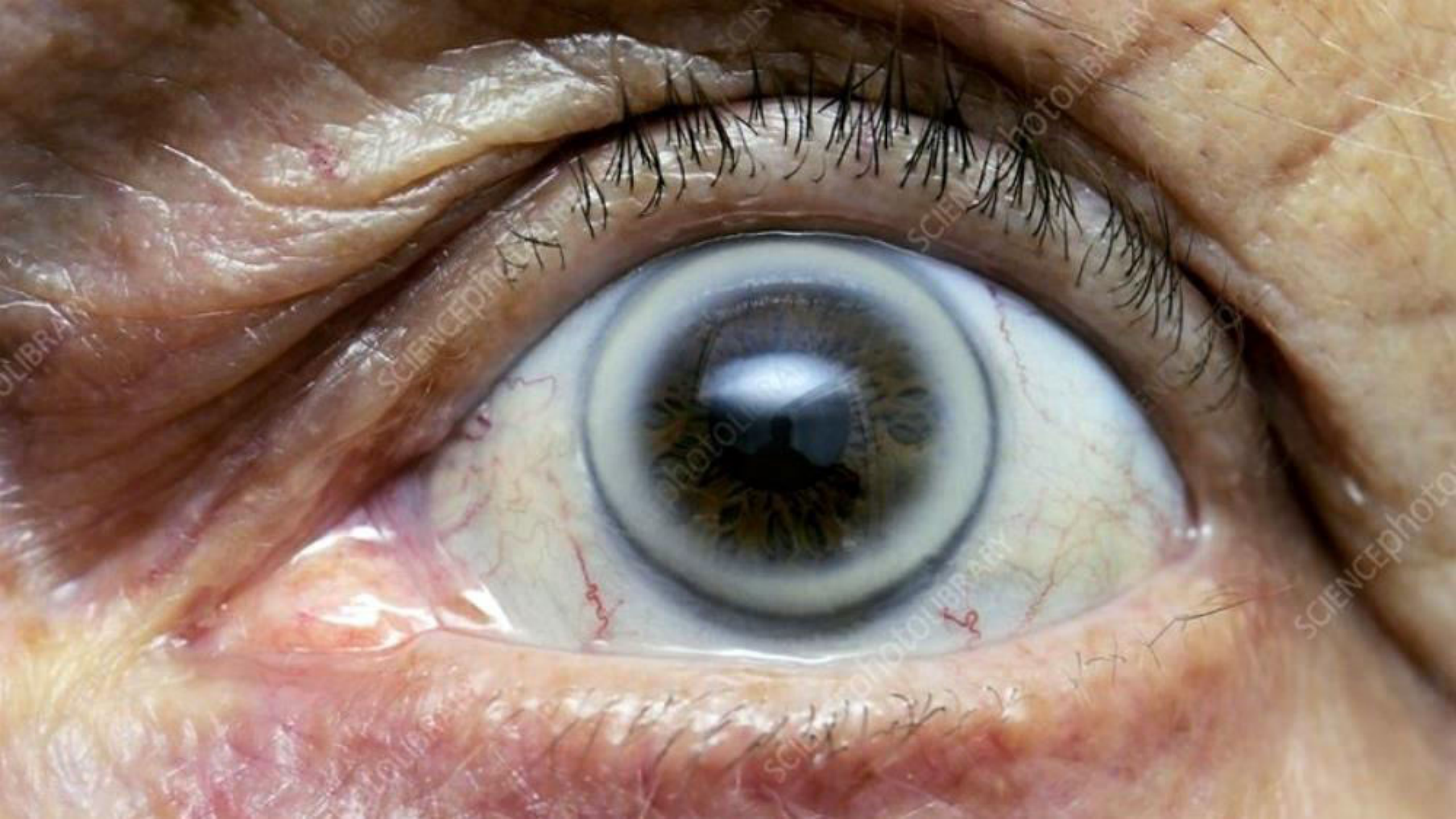
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## Lipid keratopathy

- ▶ Band keratopathy
- ▶ Spheroidal degeneration
- ▶ Salzmann nodular degeneration
- ▶ Terrien marginal degeneration

# Arcus-Senilis

- Most Common peripheral corneal opacity
- Frequently – without predisposing systemic conditions in the elderly.
- Occasionally – Familial & non-familial dyslipoproteinemias.
  - Hyperlipoproteinemia type II
  - Less common – type III, IV & V
- Unilateral arcus – rare entity carotid disease or ocular hypotony.





# Band Keratopathy (Cont'd)

- Ocular Causes
  - Chronic anterior uveitis
  - Phthisis bulbi
- Age-related
- Metabolic Causes
  - Metastatic calcification
  - Increased serum calcium & phosphorus, hyperuricemia & CRF.
- Hereditary
  - Familial cases

# Band Keratopathy (Cont'd)

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- Signs
  - Peripheral interpalpebral calcification with the clear cornea.
  - Gradual central spread to form a band-like chalky plaque.
  - Contains transparent small holes



- Treatment

- Indicated if vision is threatened.
- Chelation
  - Effective for mild cases
  - Ethylene diamine – tetra–acetic acid (EDTA).
- Diamond burr, Nd: YAG laser.

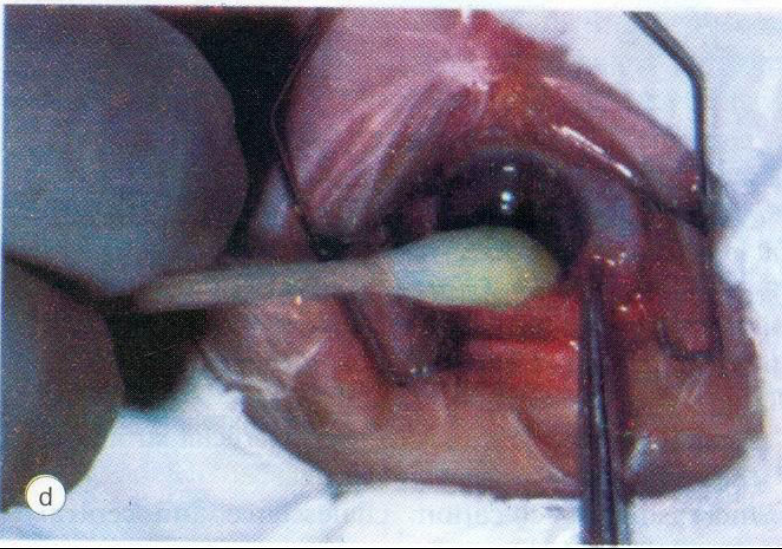
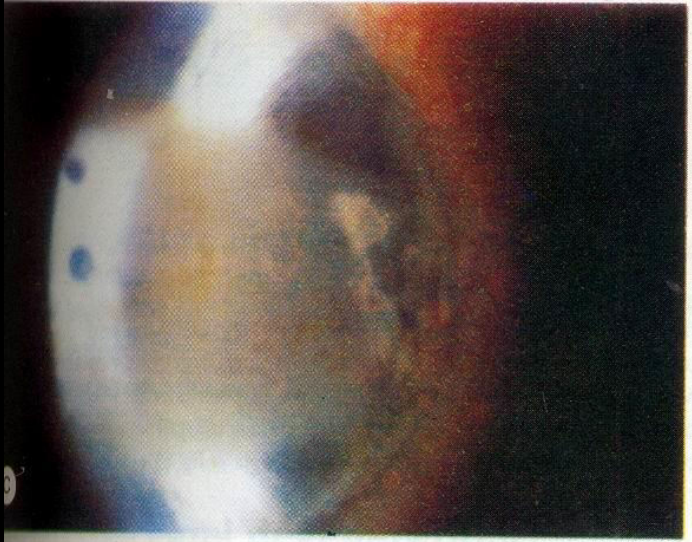
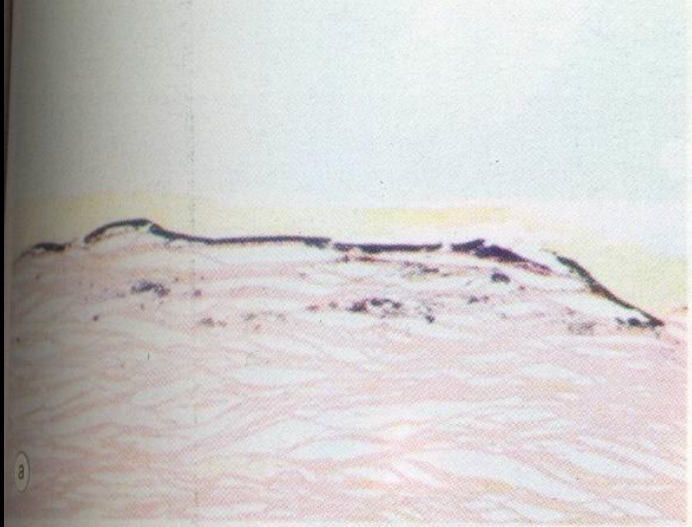


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# Learning objectives

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Thank you

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