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OCULAR DISEASE

CORNEAL DYSTROPHIES



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Introduction

- Corneal dystrophies are inherited disorders in which the cells have some inborn defects due to which pathological changes may occur with passage of time leading to development of corneal haze in otherwise normal eyes that are free from inflammation or vascularization.
- There is no associated systemic disease.
- Dystrophies occur bilaterally, manifesting occasionally at birth,



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CLASSIFICATION

- I. **Anterior dystrophies** (*superficial dystrophies*), primarily affecting **Epithelium and Bowman's layer.**
 1. Epithelial basement membrane dystrophy
 2. Reis-Buckler's dystrophy.
 3. Meesman's dystrophy.
 4. Recurrent corneal erosion syndrome.
 5. Stocker-Holt dystrophy.



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II. *Stromal dystrophies*

1. Granular (Groenouw's type I) dystrophy
2. Lattice dystrophy
3. Macular (Groenouw's type II) dystrophy
4. Crystalline (Schnyder's) dystrophy



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III. **Posterior dystrophies**, affecting primarily the corneal endothelium and Descemet's membrane.

1. Cornea guttata
2. Fuchs' epithelial-endothelial dystrophy (late hereditary endothelial dystrophy).
3. Posterior polymorphous dystrophy (of Schlichting).
4. Congenital hereditary endothelial dystrophy (CHED).



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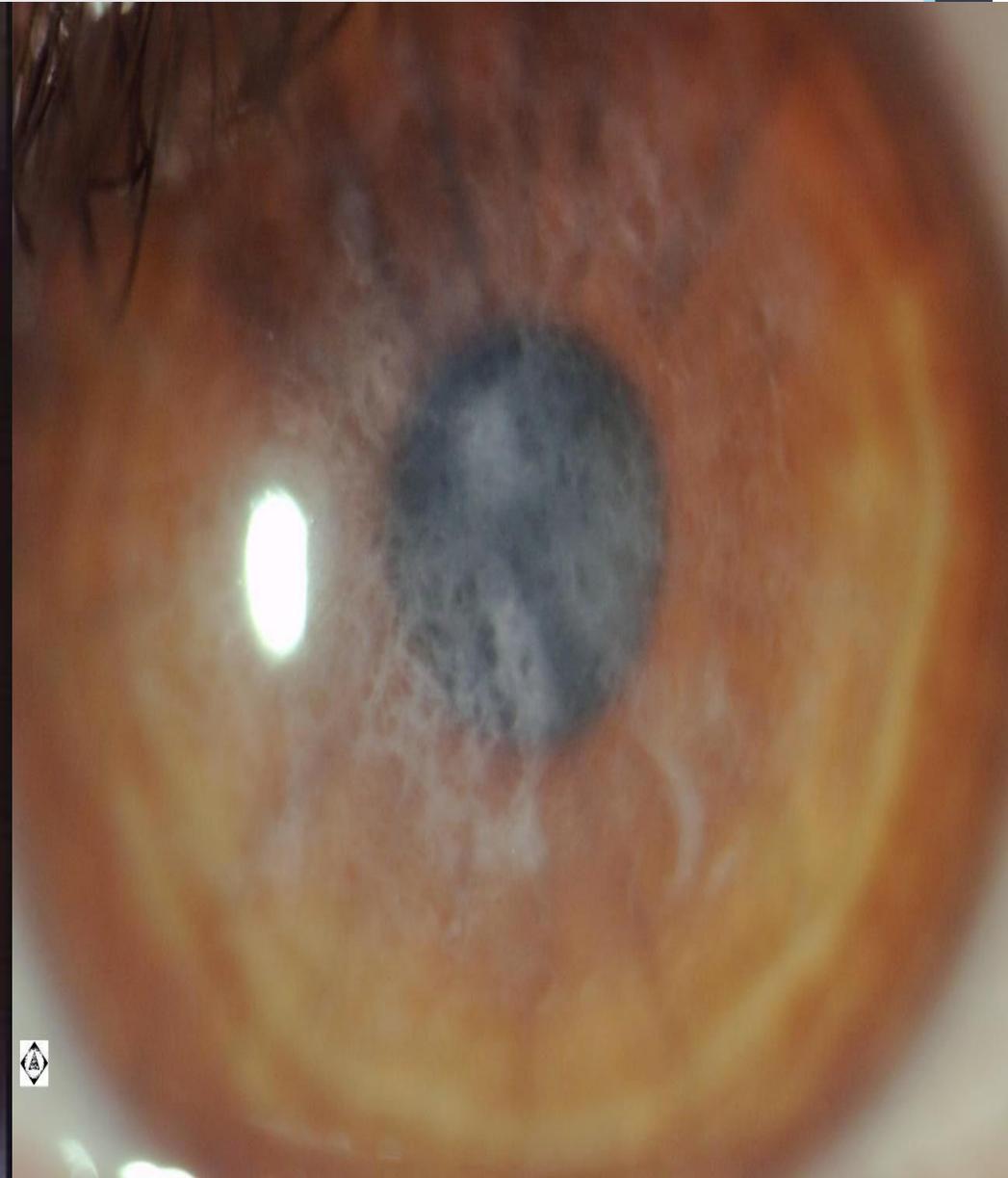
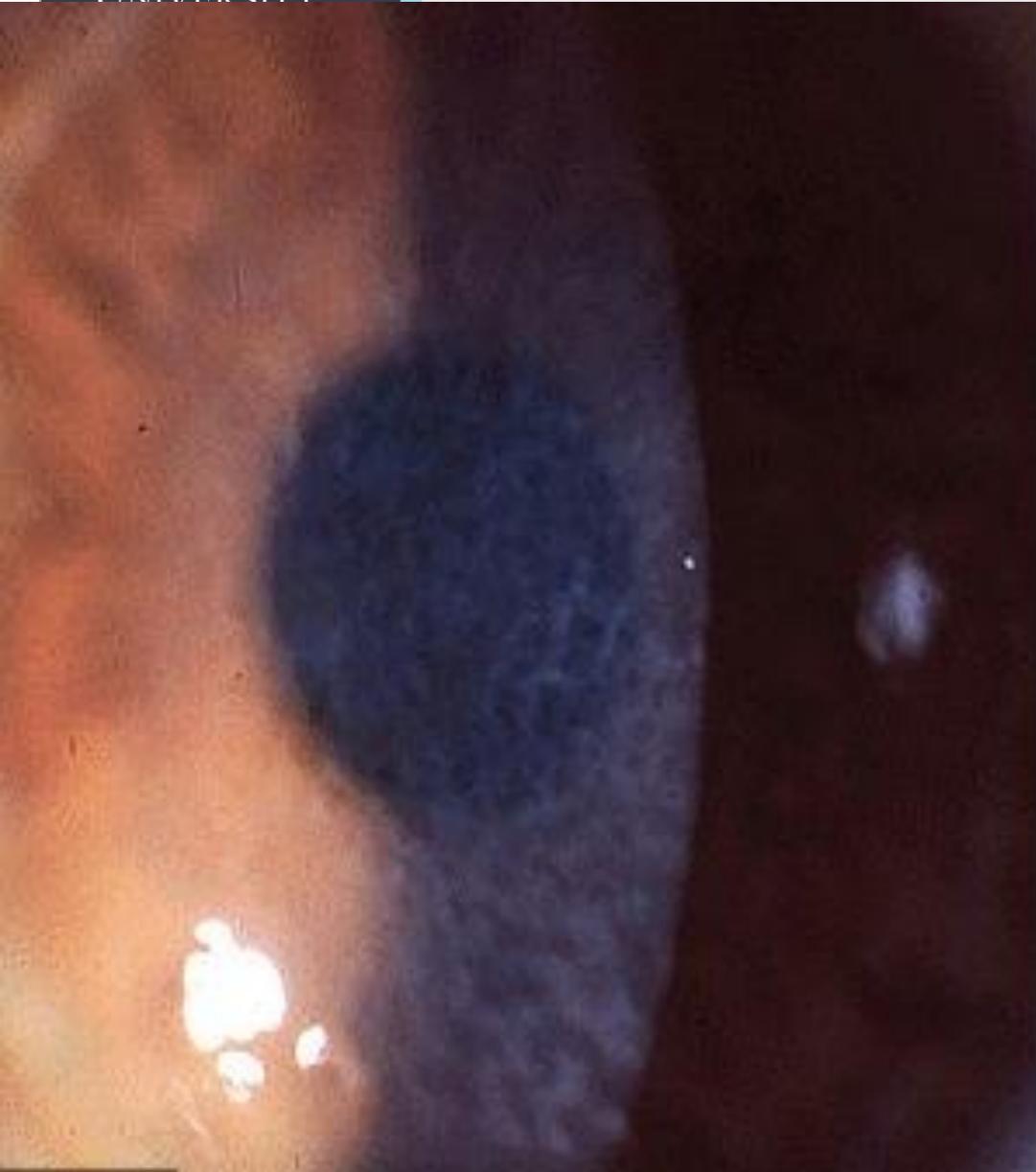
Reis-Buckler dystrophy

- Also known as ring-shaped dystrophy (due to the typical lesion) primarily involving the Bowman's layer is a progressive corneal dystrophy occurring in childhood.
- It has got autosomal dominant inheritance.
- Most patients get frequent attacks of recurrent corneal erosions that usually result in diffuse anterior scarring.



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Reis-Buckler dystrophy





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Treatment

- In early cases is same as that of recurrent corneal erosions, i.e. by patching.
- Patients ultimately need lamellar or penetrating keratoplasty.



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Recurrent corneal erosion syndrome

- It is often described as a type of dystrophy that typically follows trauma to cornea by finger nail or any other sharp edge.
- It has been shown that a lack of basement membrane and hemidesmosomes in the area of involvement, is the basic underlying cause.

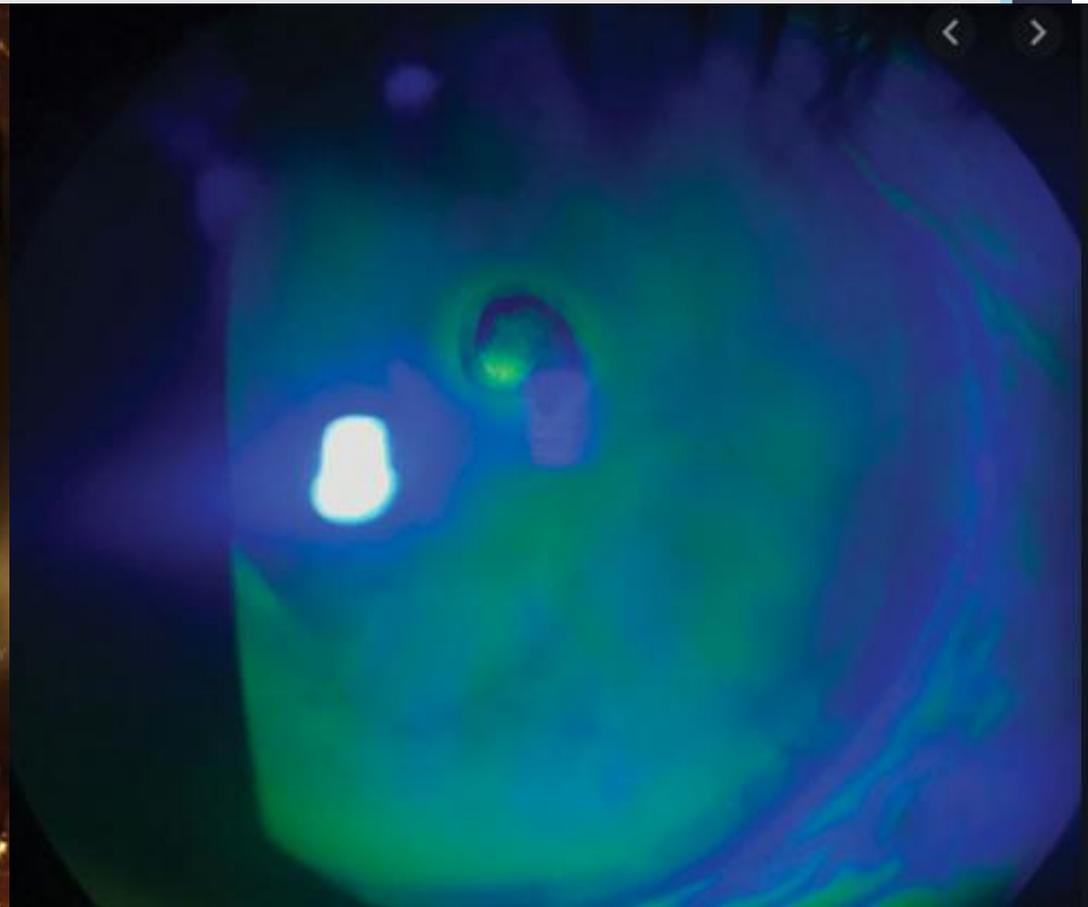


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- The condition is characterised by pain, photophobia, lacrimation and blurring of vision on awakening in the morning.





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Treatment

- It consists of patching with plain ointment for 1-2 days.
- Hypertonic saline drops or ointment decrease attack of erosions by reducing epithelial oedema.
- Severe cases may be treated by scraping the whole epithelium followed by pressure patching.



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Granular dystrophy

- Also known as 'Groenouw type I, is an autosomal dominant dystrophy characterised by milky-granular hyaline deposits in anterior stroma.
- Intervening stroma is clear.

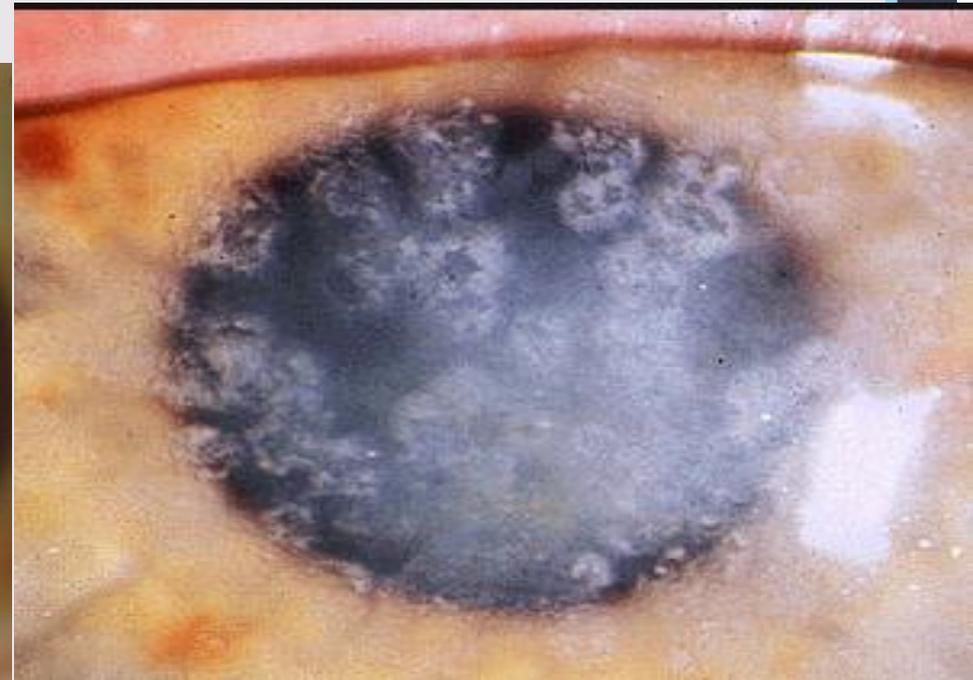
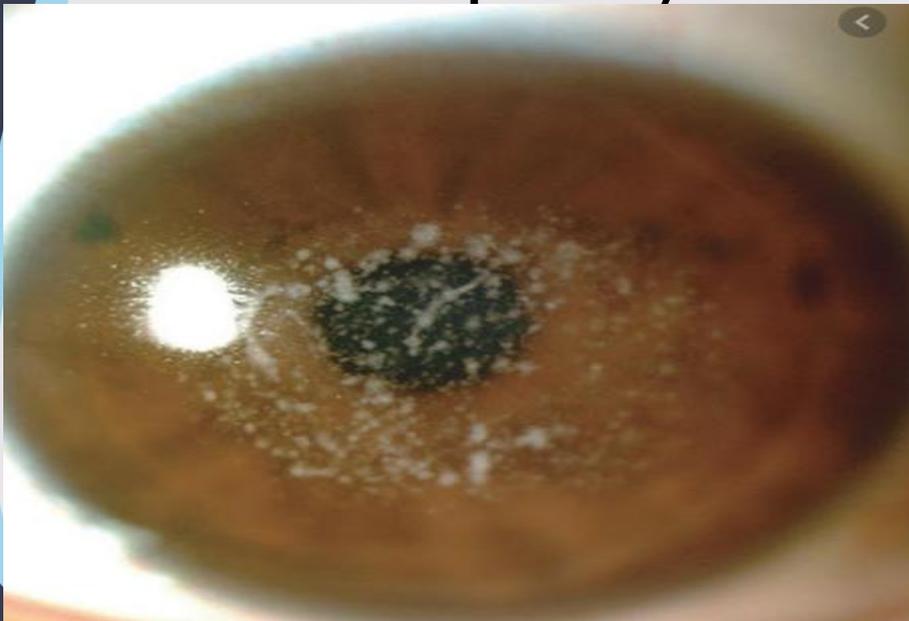


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- The condition developing in first decade of life is slowly progressive and usually asymptomatic.
- Occasionally visual acuity may be severely impaired, requiring keratoplasty.

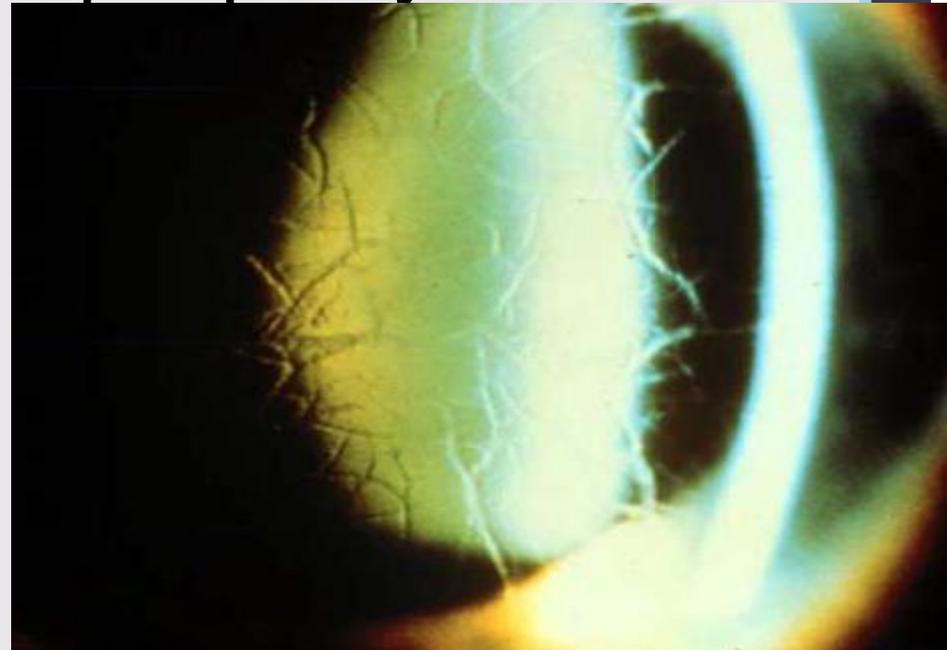
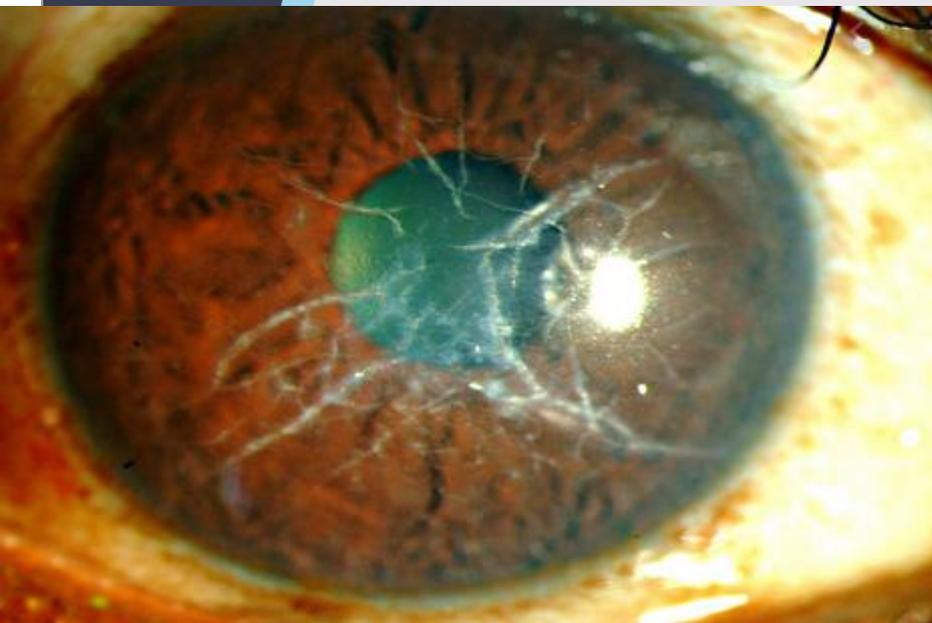




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Lattice dystrophy

- Also known as '*Biber-Haab-Dimmer dystrophy*'. It is an autosomal dominant disease characterised by branching spider-like amyloid deposits forming an irregular lattice work in the corneal stroma, sparing the periphery.





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- It appears at the age of 2 years, but the occurrence of recurrent erosions and progressive clouding of central cornea is apparent by the age of 20 years.
- Soon, visual acuity is impaired.
- Usually penetrating keratoplasty is required by the age of 30- 40 years.



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Macular dystrophy

- It is an autosomal recessive dystrophy characterised by appearance of dense grey opacity in the central cornea.
- The condition results due to accumulation of mucopolysaccharides owing to a local enzyme deficiency.



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- It occurs in childhood (5 to 10 years) and leads to marked defective vision in early life, which usually requires penetrating keratoplasty.



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Cornea Guttata of vogt

- This condition is characterised by drop-like excrescences involving the entire posterior surface of Descemet's membrane.
- These are similar to Hassal- Henle bodies which represent the age change and are mainly found in the peripheral part.



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- Cornea guttata may occur independently or as a part of early stage of *Fuch's dystrophy*.
- *The condition usually occurs in old age and is more common in females than males.*
- It rarely affects the vision and hence treatment is usually not required.



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Fuch's epithelial-endothelial dystrophy

- Fuchs dystrophy is frequently seen as a slowly progressive bilateral condition affecting females more than males, usually between fifth and seventh decade of life.
- Primary open angle glaucoma is its common association.





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Clinical features

- *Stage of cornea guttata:* It is characterised by the presence of Hassal-Henle type of excrescences in the central part of cornea.
- A gradual increase of central guttae with peripheral spread and confluence gives rise to the so called '*beaten-metal*' appearance.
- *This stage is asymptomatic.*

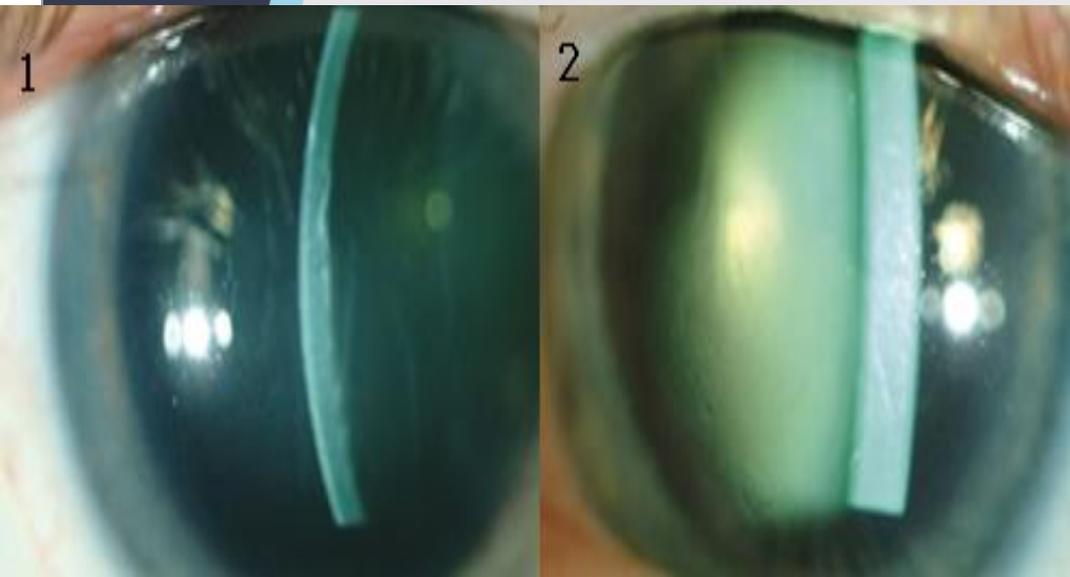




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2. **Oedematous stage or stage of endothelial decompensation** is characterised by the occurrence of early stromal oedema and epithelial dystrophy.
- Patients complains of blurring vision.



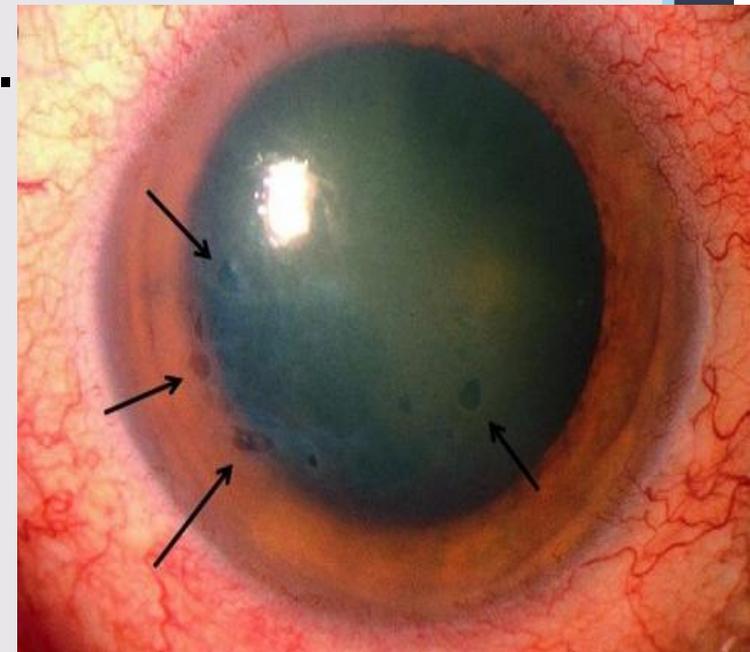


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3. Stage of bullous keratopathy. *This stage follows long-standing stromal oedema and is characterised by marked epithelial oedema with formation of bullae, which when rupture cause pain, discomfort and irritation with associated decreased visual acuity.*





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4. **Stage of scarring.** *In this stage epithelial bullae are replaced by scar tissue and cornea becomes opaque and vascularized.*
- The condition may sometimes be complicated by occurrence of secondary infection or glaucoma.





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Treatment

1. *In early oedematous stage use of 5 percent sodium chloride (hypertonic saline) may be of some use.*
2. *Bandage soft contact lenses provide some relief from disturbing symptoms in bullous keratopathy stage.*
3. *Penetrating keratoplasty is the treatment of choice when the visual acuity is reduced markedly.*



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Congenital hereditary endothelial dystrophy (CHED)

- This is a rare dystrophy associated with scanty or absent endothelial cells and thickened Descemet's membrane.
- The basic endothelial deficiency results in diffuse milky or ground glass opacification and marked thickening of corneal stroma.
- It may be inherited both dominantly and recessively.

