

2.2 Cornea: Keratopathies

Plan

Anatomy
General terms
Keratopathy

Red in module Red eye

Black in module decrease vision

Inflammatory (Keratitis)

Infective (Bacterial, fungal, viral & amoeba ulcers)

Non infective (Immune & hypersensitivity)

Corneal ulcer treatment

Corneal ulcer complications & their management

Ectasias: (Keratoconus; Etiology, Features, Investigations, Treatment- keratoplasty)

Dystrophies

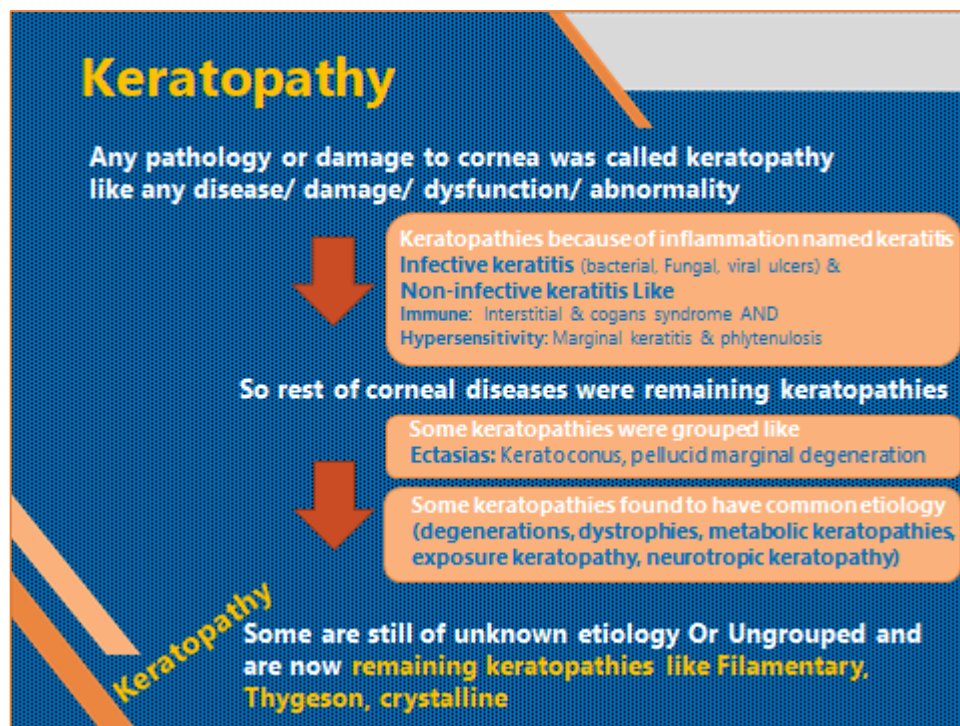
Degenerations

Metabolic keratopathies

Exposure keratopathies

Neurotropic keratopathies

Ungrouped keratopathies



Keratopathy is any pathology, disease, dysfunction or abnormality of cornea

Keratopathy

- **Infective Keratitis:** Keratopathy due to infective inflammation
 - Bacterial keratitis, Fungal keratitis, Viral keratitis, Protozoa keratitis
- **Non-Infective Keratitis:** Keratopathy because of non infective inflammation
 - Immune: Interstitial, Cogan & Hypersensitivity: Marginal Phlyctenolosis
- Some Keratopathies are grouped like Ectasias (Keratoconus)
- Keratopathies because of systemic disease or aging or trauma are called **Degenerations**
- Hereditary Keratopathies are called **Dystrophies**
- Some keratopathies of common etiology metabolic, exposure, neurotropic
- Keratopathies which have unknown cause and not grouped are called **Unclassified keratopathies** like **Filamentary, Thygeson, Crystalline**

General terms:

Keratopathy: Any disease, damage or pathology of cornea was initially called keratopathy.

Keratitis: Keratopathy because of inflammation is called keratitis. Keratitis may be infective (bacterial, fungal, viral or amoeba) or non-infective (Immunogenic or hypersensitivity).

Corneal ulcer: Usual clinical name for infective keratitis like bacterial ulcer, fungal ulcer and viral ulcer.

Ectasias: Keratopathies causing protrusion of cornea

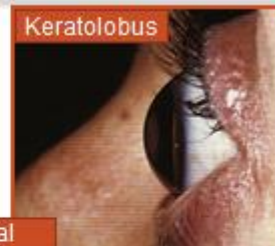
Classification & Examples

- Keratitis: Inflammatory Infective & Inflammatory non-Infective
- **Ectasias**
- Dystrophies: lesions inherited; not related to any disease
- Degenerations: non hereditary; changes under circumstances such as systemic disease, ageing, trauma
- Metabolic
- Exposure
- Neurotropic
- Unclassified Keratopathies
- Keratoconus
- Pellucid marginal degeneration
- Keratoglobus

Details in chapter
VHB 7.5

Corneal Ectasia

- Inflammation
 - Ectatic cicatrix
(Ulcer makes cornea thin and cornea bulges with normal IOP)
 - Anterior staphyloma
(Bulging cornea with increased IOP)
- Congenital
 - Keratoglobus (symmetrical bulging)
 - Pellucid marginal deg (inferior bulging)
 - Keratoconus (conical bulging)



Keratoconus

Introduction

Keratoconus presents in early teens or twenties.

It is progressive thinning of central or paracentral cornea.

Usually, one eye is involved followed by other eye.

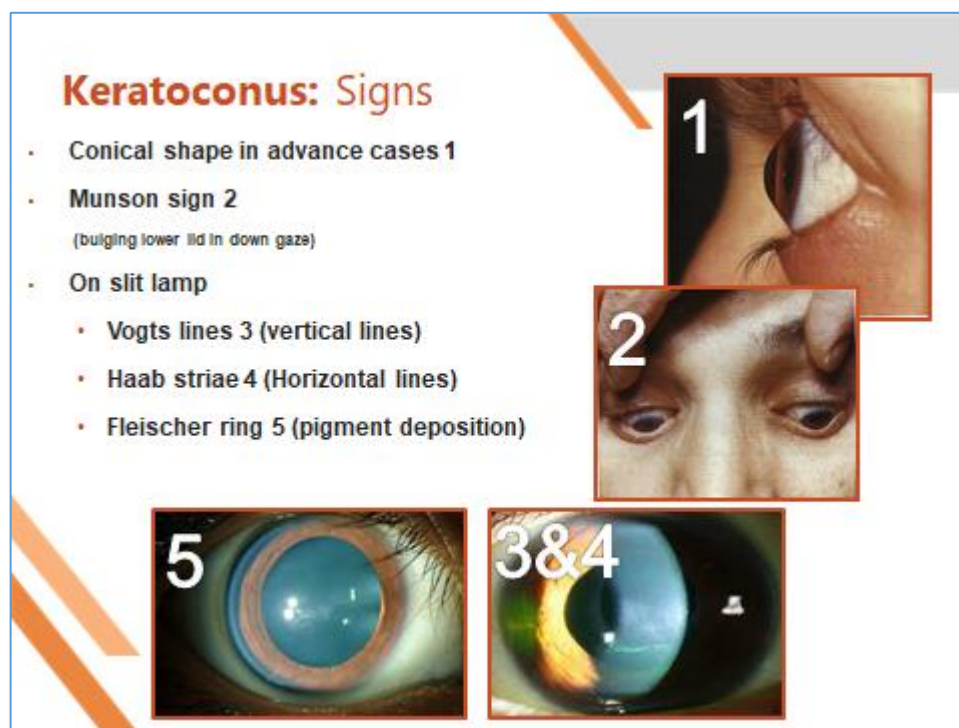
Keratoconus occurs mostly without systemic involvement but It is also associated with systemic diseases like Downs's syndrome, Marfan's syndrome and Ehlers-Danlos syndrome and also associated with ocular diseases like vernal keratoconjunctivitis, blue sclera, retinitis pigmentosa and aniridia.

Etiology

Exact etiology is not known but defective collagen causes weakness of cornea which results in bulging forward of cornea which is called keratoconus.

Symptoms & signs

1. Less than normal visual activity
2. Frequently changing glasses prescription
3. Irregular astigmatism



Investigations

Direct ophthalmoscope and retinoscopy are clinical tests.

Best investigation is corneal topography.


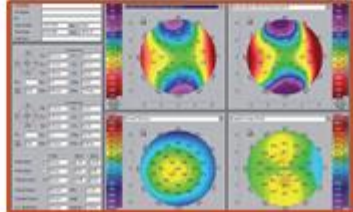


Corneal topography has four colored maps.

Parameters to look for on topography are

- Red colors on map show abnormality
- Increased anterior elevation
- Increased posterior elevation
- More than 470 micron thin cornea
- K-max more than 47 diopter

Keratoconus: Investigations

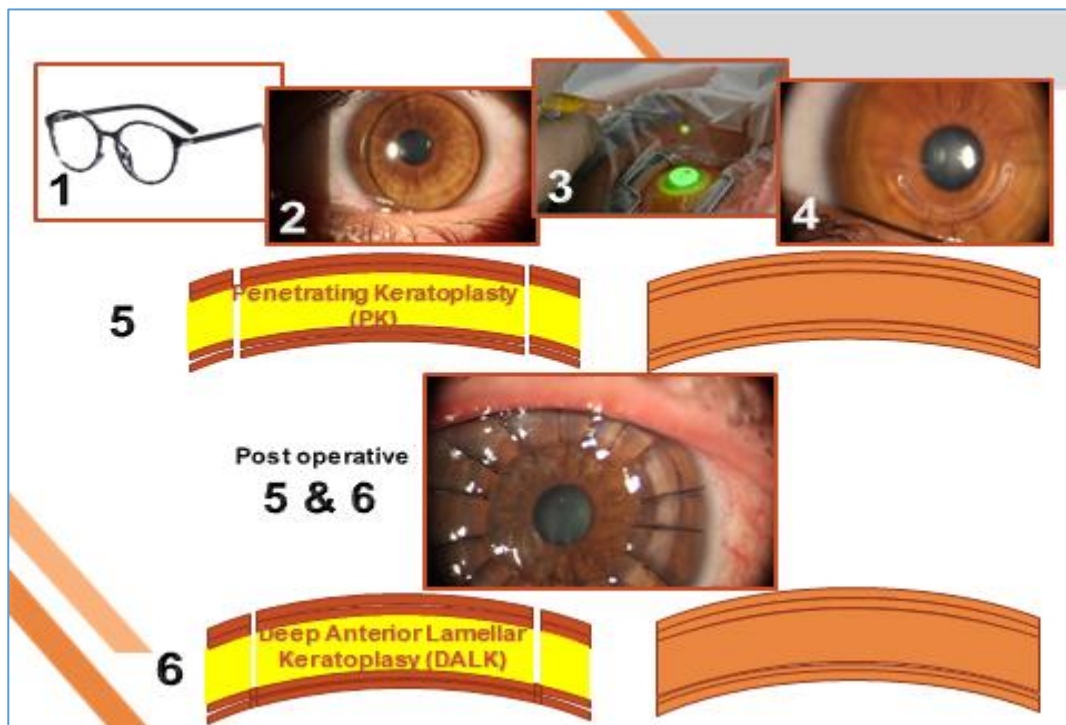
- Direct Ophthalmoscope
 - Tear drop sign
- Retinoscopy
 - Irregular astigmatism
 - Scissors reflex
- Keratometry
 - >47 diopter
- Topography
 - Corneal thickness
 - Corneal bulge



Treatment

Treatment of keratoconus may be one of following or combination of following.

- 1. Glasses:** Glasses are simple answer to keratoconus in early cases. Astigmatism is usually unstable and keeps changing.
- 2. Contact lenses:** When glasses do not work properly because of irregular astigmatism then contact lenses may be tried. Contact lenses are soft and hard. Initially soft contact lenses are tried which easy and comfortable to wear. Hard contact lenses are next option.
- 3. Collagen cross linking (CXL):** Keratoconus is progressive disease because of defective collagen which weakens cornea. To strengthen cornea CXL procedure is done. It involves putting riboflavin eye drops on cornea. Then ultraviolet light is focused on cornea which causes cross linking of collagen fibers and hence strengthens cornea.
- 4. Intra stromal ring segment:** These are half circle rings which are surgically placed in corneal stroma to improve vision by making cornea regular.
- 5. Penetrating keratoplasty (PK):** PK is replacing all five layers of cornea. It is further explained in keratoplasty section.
- 6. Deep anterior lamellar keratoplasty (DALK):** Pathology is cornea stroma and ALK replaces top three layers with donor tissue. It is latest surgery and carries less risk of rejection.



Keratoplasty

Definition

Keratoplasty is replacing abnormal host cornea with healthy donor cornea. There are different indications for replacing cornea.

Indications

1. Optical Keratoplasty:

Optical Keratoplasty is only to improve vision like in keratoconus, corneal dystrophies, endothelial decompensation.

2. Therapeutic Keratoplasty:

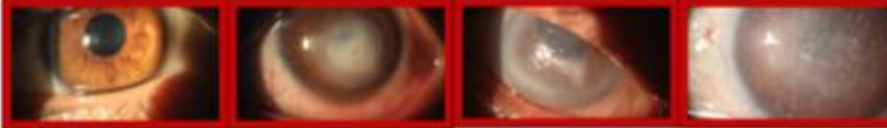
This is for therapeutic purpose like in non-responding corneal ulcer.

3. Tectonic Keratoplasty:

This is regain integrity of eye ball in corneal perforations or threatening perforations like in descematocele and stromal thinning.

4. Cosmetic Keratoplasty:

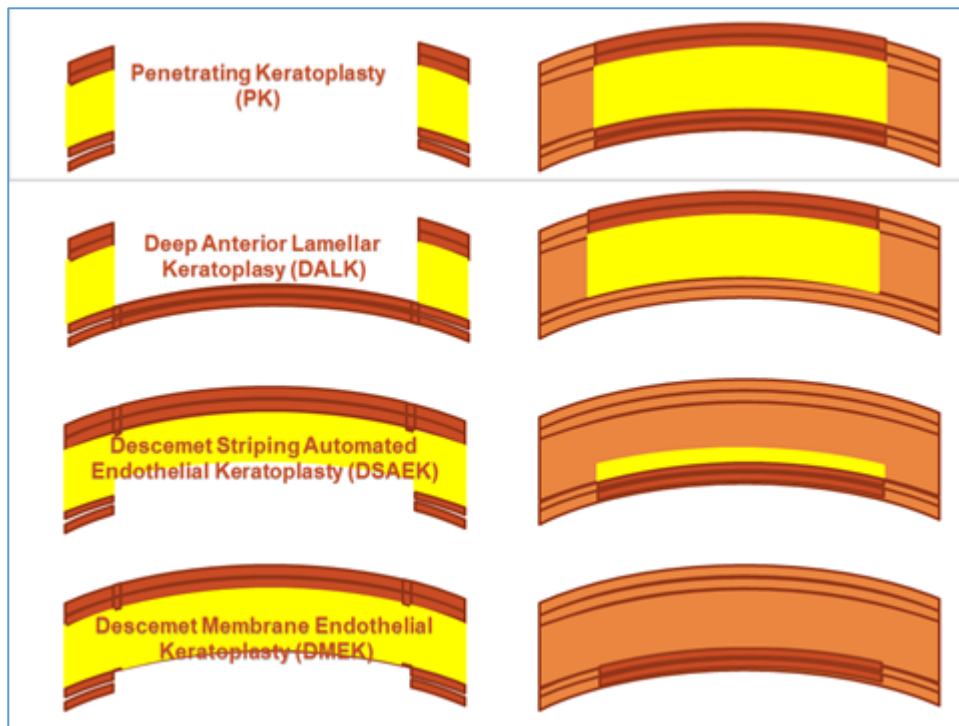
In cosmetic Keratoplasty white scarred cornea is replaced with transparent cornea only to improve cosmetic look. It is neither for improving vision nor for therapeutic or tectonic purpose.



Indications

- 1. Optical:** purely to improve vision
-Keratoconus/scarring/Bullous keratopathy/ Dystrophies
- 2. Therapeutic:** To treat disease
-Corneal ulcer
- 3. Tectonic:** To restore structure
-Stromal thinning /
-Descemetocele
- 4. Cosmetic:** for cosmetic
-White cornea in blind eye

Types of keratoplasty



Postoperative complications

Post-op Complications

Early complications

- persistent epithelial defect
- Irritation by protruding sutures
- Papillary hypertrophy
- Wound leak / flat anterior chamber
- Iris prolapse
- Increased IOP & infection
- Early graft failure: recovers

Late complications

- Astigmatism
- Late wound separation
- Recurrence of initial disease process in graft
- Retro-corneal membrane formation
- Glaucoma
- Cystoid macular edema
- Late graft failure: needs treatment

Dystrophies: Inherited keratopathies

This is a group of corneal dystrophies which are inherited and associated with disease.

Dystrophies are classified in to **anterior, stromal and posterior dystrophies**.

Anterior dystrophies involve epithelium and bowman membrane.

Stromal dystrophies involve corneal stroma.

Posterior dystrophies involve descemet membrane and endothelium

Treatment depends on vision and symptoms.

In early disease nothing needs to be done but in advanced cases Keratoplasty is done to replace diseased tissue with healthy tissue.

Classification & Examples

- Keratitis: Inflammatory Infective & Inflammatory non-Infective
- Ectasias
- **Dystrophies:** lesions inherited not related to any disease
- Degenerations: non hereditary; changes under circumstances such as systemic disease, ageing, trauma
- Metabolic
- Exposure
- Neurotropic
- Unclassified Keratopathies
- **Anterior dystrophies (Epi+Bowman)**
 - Cogan's microcystic
 - Recurrent corneal erosion
 - Reis-Buckler (Bowman membrane)
- **Stromal dystrophies**
 - Granular
 - Macular
 - Lattice
 - crystalline
- **Posterior dystrophies (Endo+Desemet)**
 - Fuchs' endothelial (Cornea guttata)
 - Posterior polymorphous
 - Congenital Hereditary Endothelial Dystrophy (CHED)

Degenerations: Keratopathies under circumstances

This is group of keratopathies which is Keratopathy because of local or systemic circumstances or aging or trauma.

Depending on location degeneration are classified in to central and peripheral degeneration.

Treatment depends on location and severity.

Peripheral degeneration do not obstruct vision so do not need treatment.

Central degenerations may need keratoplasty.

Classification & Examples

- Keratitis: Inflammatory Infective & Inflammatory non-Infective
 - Ectasias
 - Dystrophies: lesions inherited: not related to any disease
 - **Degenerations:** non hereditary; changes under circumstances such as systemic disease, ageing, trauma
 - Metabolic
 - Exposure
 - Neurotropic
 - Unclassified Keratopathies
- **Central Degenerations**
 - Cornea farinata
 - Band degeneration / keratopathy
 - Lipid degeneration / keratopathy
 - Spheroidal degeneration
 - Salzmann nodular degeneration
 - Hyaline degeneration
 - **Peripheral Degenerations**
 - Arous senilis
 - Limbal girdle of voght
 - Crocodile shagreen
 - Terriens marginal degeneration
 - Moorens ulcer??

Metabolic Keratopathy because of metabolic diseases

This group of keratopathies is because of metabolic diseases. Metabolic diseases cause deposit of metabolic materials in cornea.

Treatment is treating cause. No specific eye treatment usually needed.

Classification & Examples

- Keratitis: Inflammatory Infective & Inflammatory non-Infective
- Ectasias
- Dystrophies: lesions inherited; not related to any disease
- Degenerations: non hereditary; changes under circumstances such as systemic disease, ageing, trauma
- **Metabolic**
- Exposure
- Neurotropic
- Unclassified Keratopathies

- Cystinosis
- Muchopolysaccharidosis
- Wilson disease

Exposure Keratopathy because of exposure

This is group of keratopathies because of exposure of cornea.

It occurs when lids cannot close in diseases like facial palsy or when cornea is protruded out in proptosis.

Treatment is lubrication of cornea and closing lids.

Classification & Examples

- Keratitis: Inflammatory Infective & Inflammatory non-Infective
 - Ectasias
 - Dystrophies: lesions inherited: not related to any disease
 - Degenerations: non hereditary; changes under circumstances such as systemic disease, ageing, trauma
 - Metabolic
 - **Exposure**
 - Neurotropic
 - Unclassified Keratopathies
- Exposure because of incomplete lid closure in
- Paralytic: facial palsy
 - Reduced tone: parkinsonism
 - Mechanical: lid scarring
 - Proptosis

Neurotropic keratopathies because of sensory loss

This group of keratopathies is because of loss of corneal sensations. This happens in lesions or diseases which cause damage to sensory trigeminal nerve.

Treatment is usually treating cause and lubrication.

Classification & Examples

- Keratitis: Inflammatory Infective & Inflammatory non-Infective
 - Ectasias
 - Dystrophies: lesions inherited; not related to any disease
 - Degenerations: non hereditary; changes under circumstances such as systemic disease, ageing, trauma
 - Metabolic
 - Exposure
 - **Neurotropic**
 - Unclassified Keratopathies
- Because of loss of trigeminal innervation in
- Tumours
 - Peripheral neuropathy like diabetes
 - Viral keratitis

Ungrouped keratopathies

This group of keratopathies is those keratopathies which can not be grouped on basis of inheritance (dystrophies), circumstances (degenerations), metabolic disease (metabolic diseases), exposure of cornea (exposure Keratopathy), loss of sensation (neurotropic). These remaining ungrouped keratopathies are filamentary Keratopathy, thygeson Keratopathy and crystalline Keratopathy.

Classification & Examples

- Keratitis: Inflammatory Infective & Inflammatory non-Infective
 - Ectasias
 - Dystrophies: lesions inherited; not related to any disease
 - Degenerations: non hereditary; changes under circumstances such as systemic disease, ageing, trauma
 - Metabolic
 - Exposure
 - Neurotropic
- Filamentary keratopathy
 - Thygeson keratopathy
 - Crystalline keratopathy

Unclassified Keratopathies