Keratoconus (KC)

Definition:

KC is a unilateral or bilateral progressive non-inflammatory stromal thinning with apical protrusion and visual deterioration.

Aetiology:

- Hereditary: autosomal dominant transmission (10% offspring with KC).
- Unknown.

Risk Factors:

- Positive family history.
- Teens and twenties.
- Puberty in pediatrics.
- Pregnancy and lactation.
- Chronic eye rubbing.
- Vernal keratoconjunctivitis *(VKC)*.
- Dry eye disease *(DED)*.
- Thyroid eye disease *(TED)*.
- Down and Marfan syndromes.

Classification (Amsler-Krumeich Classification):

Stage	K reading (Diopter)	Corneal thickness (µm)	Myopia and Astigmatism (Diopter)
Stage I	< 48	> 450	> 5
Stage II	< 53	> 400	5 – 8
Stage III	53 - 55	300-400	8 – 10
Stage IV	> 55	< 300	> 10

Diagnosis:

Symptoms: painless diminution of vision.

Signs:

- Direct ophthalmoscopy → oil droplet reflex.
- Retinoscopy → scissoring reflex.
- Vogt striae → fine vertical stromal stress lines disappear with globe pressure.
- Fleischer ring → epithelial iron deposits around base of the cone.
- Munson sign → bulging of the lower lid in downgaze.
- Acute hydrops: sudden rupture in the Descemet membrane → influx of aqueous into the cornea → corneal edema → pain, photophobia and decreased vision.
- Corneal opacity: resulting from old recurrent hydrops episodes.

Investigations (Corneal Topography: Pentacam and Orbscan):

- ***KC diagnosis:*** the 3 diagnostic points meet within KC cone:
  - ✓ Steepest point of the corneal curvature.
  - ✓ Thinnest point of the corneal thickness.
  - ✓ Highest point on the posterior corneal surface.
- ***KC progression diagnosis:*** with the follow-up CT criteria:
  - ✓ ↑↑ Keratometry (corneal curve and power).
  - ✓ ↓↓ Corneal thickness.
  - ✓ ↑↑ Height of corneal elevations.

Treatment:

A) Therapeutic:

Corneal Collagen Cross-Linking *(CXL)* is the only proven true therapy that halts KC progression. CXL means corneal irradiation with UVA after corneal saturation with riboflavin solution.

- **Types of CXL:**
  - ✓ Standard CXL *(S-CXL)*: best but slow with low UVA energy for 30 minutes irradiation time.
  - ✓ Accelerated CXL *(A-CXL)*: less effective but rapid with high UVA energy up to 3 minutes irradiation time.
- **Techniques:**
  - ✓ Epithelium- off *(Epi-off CXL)*: difficult, painful but effective.
  - ✓ Epithelium- on *(Epi-on CXL)*: easy, painless but inefficient.

B) Refractive:

Adding a refractive tool to correct the refractive status and improve vision.

- **Refractive tools:**

- *Non-Surgical(Cheap with Best Vision Ever):*
  - ✓ Spectacles: in low errors.
  - ✓ RGP Contact Lens: all errors.
  - ✓ Scleral Contact Lens: all errors comfortable.
- *Surgical(Costy with unexpected outcomes):*
  - ✓ Wavefront-guided Photorefractive Keratectomy *(WFG PRK)*.

- ✓ Intracorneal Ring Segments *(ICRS)*: femtosecond laser-assisted implementation.
- ✓ Toric Implantable Collamenar Lens *(T-ICL)*.

C) Combination Procedures:

To combine CXL with a refractive surgical procedure *(CXL-Pus)*.

Types of CXL-Plus:

- ✓ CXL + WFG PRK: in low errors and to correct anterior corneal surface irregularities.
- ✓ CXL + ICRS: in moderate to high errors.
- ✓ CXL + T-ICL: in high errors.

D) Keratoplasty (KP):

Corneal Replacement Therapy in advanced KC.

Types of KP:

- ✓ Deep Lamellar Keratoplasty *(DALK)*: replacement of corneal stoma preserving endothelium to avoid rejection with excellent outcomes.
- ✓ Penetrating Keratoplasty *(PKP)*: replacement of the entire cornea if DALK is not suitable.

Prognosis:

Good prognosis especially in early treated cases.

Further Reading:

Types of ICRS:

- *Segmented rings (better astigmatic correction):*
  - ✓ Kera.
  - ✓ Ferara.
  - ✓ Intacts.
- *Continous rings (better myopic correction):*
  - ✓ Myoring.
- *Corneal Allogenic Intrastromal Ring Segments (CAIRS): Most recent but still experimental.*

TTT of acute hydrops:

- ✓ Cycloplegia.
- ✓ Hypertonic (5%) saline eye drops and eye gel.
- ✓ Soft bandage contact lens and patching.