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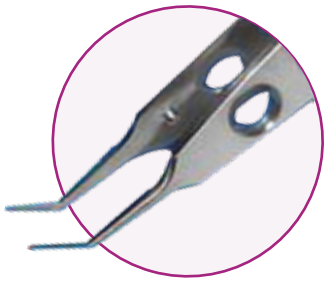
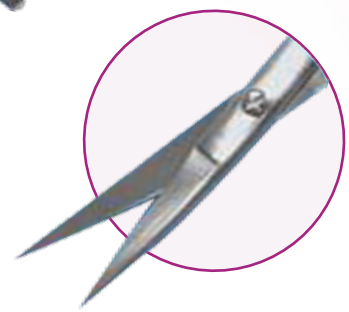
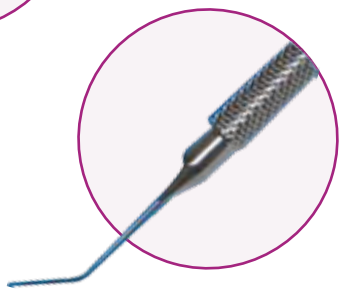
# Surgical Insights







## Keratoconus and its Management

### PART - I

ISSUE - 5

# KERATOCONUS AND ITS MANAGEMENT
# PART - I

Keratoconus, which was first described in detail in 1854, derives from the Greek words Kerato (cornea) and Konos (cone). Keratoconus is the most common primary ectasia. It is a bilateral and asymmetric corneal degeneration characterized by localized corneal thinning which leads to protrusion of the thinned cornea.[1]

Normal Cornea | Keratoconic cornea

**The reported incidence ranges from 1.3 to 25 per 100,000 per year across different populations, and a prevalence of 8.8–229 per 100,000.**[2] Prevalence of keratoconus varies widely depending upon the geographic location, diagnostic criteria used, and the cohort of patients selected. In Central India, the prevalence was found to be 2300 per 100,000 (0.0003%-2.3%) where the prevalence of keratoconus was defined as a corneal refractive power ≥48 D. Also studies have indicated that the prevalence is 4.4 and 7.5 times greater for Asian (Indian, Pakistani, and Bangladeshi) subjects compared with white Caucasians.[3]

## Aetiology and pathogenesis[1]

Despite the intensive research activity over the last few decades into the aetiology and pathogenesis of keratoconus, the cause(s) and possible mechanisms for its development remain poorly understood. Albeit, there have been several hypotheses proposed into the genetic and biochemical mechanisms

### a. Genetics

- Studies carried out before corneal topography techniques became commercially available reported that 6–8% of subjects with keratoconus had close relatives affected by the disease
- Studies assisted by corneal topography have shown that up to 50% of subjects with keratoconus have at least one close relative affected by the disease

**A recent study estimated that relatives of keratoconics have a 15–67 times higher risk of developing keratoconus than those who do not have relatives with keratoconus**

- Linkage studies carried out in families affected with keratoconus to identify the genetic regions (Loci) have reported genetic susceptibility to the disease. Several loci, have been associated to keratoconus disease in different studies:

- 20q-12
- 16q22.3-q23.1
- 15q22.33-24.2
- 3p14-q13
- 2p24
- 5q14.3-q21.1
- 9q
- 5q21.2

### b. Biochemical factors

**The excessive degradation of the corneal stroma commonly observed in keratoconus might be the result of proteolitic enzyme activity that can be initiated by an increased level of proteases and other catabolic enzymes, or decreased levels of proteinase inhibitors such as α2-macroglobulin and α1-antiprotease.**

- It has also been found that keratocytes in keratoconus have 4 times greater numbers of Interleukin-1 receptors compared to normal subjects
- As Interleukin-1 has been postulated to be a modulator of keratocytes proliferation, differentiation and death, it has been suggested that the loss of anterior stromal keratocytes might occur due to an excess of apoptotic cell death and stromal mass loss.
- Contact lens wear might be a precursor for ectasia development
- Interleukin- 6 cytokine is over expressed in early forms of keratoconus, which supports the development of chronic inflammatory events in the pathogenesis cascade of the disease

### c. Biomechanical factors

- The different distribution and lower number of stromal lamellae in keratoconic compared with normal corneas has been proposed as a precursor for corneal rigidity reduction and thinning, ultimately leading to keratoconus development
- Furthermore, oxidative damage has been described as a co-factor in keratoconus progression.
- The main factors related to increased oxidative damage are ultraviolet radiation, atopy and mechanical trauma; the latter could occur as a result of chronic eye rubbing and contact lens wear

**Keratoconus Risk Factors:**

- Eye rubbing
- Family history
- Use of contact lense for a prolonged period of time
- Genetics
- Certain diseases such as Down's syndrome, Ehlers - Danlos syndrome Leber's congenital amaurosis, or osteogenesis imperfect, etc.
- Refractive surgery procedures, etc.

**Protective steps to reduce oxidative damage and potentially prevent keratoconic development include:**

a. use of ultraviolet filters
b. improvement of ocular comfort with the use of non-steroidal anti-inflammatory medications
c. use of preservative-free artificial tears and allergy medications and
d. improved contact lens fit to minimize corneal microtrauma

### d. Related diseases

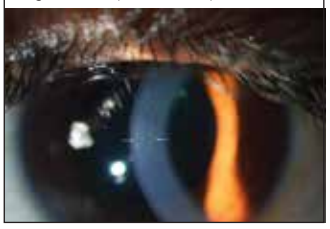
- Studies have reported that 0.5–15% of subjects with Down's syndrome suffer from keratoconus, leading to an association 10–300 times higher than that of the normal population
- This association has been suggested to occur as a result of eye rubbing owing to the increased rate of blepharitis seen in approximately 46% of Down's syndrome individuals
- It has also been found that 30–41% of subjects with Leber's congenital amaurosis, a rare genetic disorder, also suffer from keratoconus
- Other associations between keratoconus and connective tissue disorders, such as Ehlers-Danlos syndrome subtype VI, Osteogenesis Imperfecta and joint hypermobility have previously been reported.

## Clinical Features

**A typical patient with keratoconus presents in the teens or early twenties with complaints of blurring or distortion in vision and having to change glasses frequently due to changes in refractive error. Following are the various clinical features of keratoconus[2]:**

- Irregular myopic astigmatism (seen in retinoscopy)
- A scissoring reflex and an "oil-droplet" reflex (Charleux sign)
- Prominent corneal nerves on slit-lamp examination
- Corneal thinning, which is generally greatest at the apex of the cone
- Subepithelial and anterior stromal scars may be present, secondary to breaks in Bowman's membrane.
- Vogt's striae (fine parallel lines) in the posterior stroma
- Fleischer's ring, found around the base of the cone and best appreciated through a cobalt blue filter
- Gross clinical signs in advanced keratoconus include V-shaped distortion of the lower eyelid in down gaze due to an excessively protuberant conical cornea, known as Munson's sign, and a sharply focused light beam near the nasal limbus produced by lateral corneal illumination (Rizzuti's sign).

Slit-lamp photograph of eye showing apical scarring (yellow arrow) and Vogt's striae (white arrow)[2]



Slit-lamp photograph of eye showing Fleischer's ring [2]

- Acute hydrops caused by sudden breaks in Descemet's membrane (DM)
- A sudden decrease in vision accompanied by corneal clouding.
- Corneal edema with or without intrastromal clefts or blebs

## Diagnosis & Imaging

Keratometry using a manual keratometer may show a steep cornea, high astigmatism, and/or distorted mires in keratoconus. This can be used as a simple, inexpensive imaging device. Computerized videokeratography for diagnosis of keratoconus was first introduced in the 1980s.[2]

### Classically, keratoconus has been classified into:[1]

1. **Nipple**—The cone has a diameter ≤ 5mm, round morphology and is located in the central or paracentral cornea, more commonly in the infero-nasal corneal quadrant. Correction with contact lenses is normally relatively easy.[1]
2. **Oval**—The cone has a diameter > 5mm and a paracentral to peripheral location, more commonly in the infero-temporal corneal quadrant. Contact lens correction is more difficult.
3. **Keratoglobus**—The cone is located throughout 75% of the cornea. Contact lens correction is a difficult challenge, except in very limited cases.

The wide spread use of corneal topography has allowed the detection of new keratoconus patterns affecting the superior, nasal and central cornea. More recently, a new D-shape keratoconus pattern, which affects subjects who have undergone Lasik refractive surgery procedures, has been described in the literature.[1]

### Index-based systems for keratoconus detection

A variety of indices were developed for discriminating keratoconus from normal eyes as well as other conditions. Some of the popular ones included the keratoconus prediction index from the Klyce–Maeda group and the KISA index by the Rabinowitz group. These indices have been utilized to detect subtle anomalies and follow disease progression.[2]

**Index-based systems for keratoconus detection**

**A higher value than the point of cut value suggests the presence of keratoconus[1]**

| Author | Index | Point of Cut | Description |
|---|---|---|---|
| Rabinowitz/ Mc Donnel | K Value<br>I-S Value | 47.2<br>1.4 | Diagnosis is performed based on the central keratometry and the inferior–superior asymmetry in keratometric power |
| Maeda/Klyce | KPI | 0.23 | KPI is derived from eight quantitative videokeratography indexes.<br>KCI% is derived from KPI and other four indexes |
| Smolek/Klyce | KSI | 0.25 | Keratoconus detection and the level of severity is assessed using an artificial intelligent system |
| Schwiegerling/ Greivenkamp | Z3 | 0.00233 | Diagnosis is performed based in videokeratoscopic height data decomposed into orthogonal Zernike polynomials. |

| Index-based systems for keratoconus detection<br>A higher value than the point of cut value suggests the presence of keratoconus[1] | | | |
|---|---|---|---|
| **Author** | **Index** | **Point of Cut** | **Description** |
| Rabinowitz/ Rasheed | KISA% | 100% | Diagnosis is derived from K value, I-S value, AST and SRAX |
| Mc Mahon et al. | KSS | 0.5 | Diagnosis is performed based on slit-lamp findings, corneal topography, corneal power and higher order first corneal surface wavefront root mean square error |
| Mahmoud et al. | CLMI | >0.45 | Diagnosis based in detecting the presence or absence of keratoconic patterns and determining the location and magnitude of the curvature of the cone |

## Newer diagnostic devices aimed at early detection of subclinical keratoconus

1. **Orbscan** utilizes slit scanning technology to provide wide-field pachymetry and anterior and posterior elevation as well as keratometry maps. The Orbscan II, combines slit scanning with Placido-based topography analysis. This has been shown to be more sensitive than earlier devices for detection of keratoconus. Maximum posterior elevation compared with the best fit sphere (BFS), irregularity in the central 3 mm and 5 mm zones as well as pachymetry have been found to be useful in discriminating keratoconus suspects from normal subjects. Increase in apex elevation, displacement of the corneal apex, decrease in thinnest-point pachymetry, and an increase in the mean simulated keratometry minimum value have been documented on serial analysis in progressive keratoconus.[2]

2. **The Pentacam instrument,** which is based on the Scheimplug working principle, takes 12–50 images of the cornea at different angles using a rotating camera. This method evaluates disease severity and progression based on changes in corneal volume and anterior chamber angle, depth and volume. A much touted feature of the Pentacam is the Belin/Ambrosio-enhanced ectasia display, which excludes a 4 mm zone centered on the thinnest portion of the cornea from the reference shape calculation. The resulting "enhanced BFS" is supposed to approximate a normal cornea closely, making subtle elevations more pronounced and possibly aiding in detection of early or subclinical keratoconus.[2]

3. **The Ocular Response Analyzer** allows keratoconus diagnosis and classification by assessing corneal hysteresis and resistance.[1]

## Management

Keratoconus management varies depending on the disease severity. Traditionally, incipient cases are managed with spectacles, mild to moderate cases with contact lenses, and severe cases can be treated with keratoplasty. Other surgical treatment options include intra-corneal rings segments, corneal cross-linking, laser procedures (i.e., photorefractive keratectomy, phototherapeutic keratoctomy, lasik in situ keratomileusis) intra-ocular lens implants or a combination of these.[1]

### A. Optical correction

1. **Spectacles:** Spectacles are normally used in early cases of keratoconus only. As the disease progresses, irregular astigmatism develops and adequate visual acuity cannot be achieved with this type of visual correction.[1]
2. **Contact lenses:** Although contact lenses for keratoconus are manufactured with hydrogel, silicone hydrogel, gas permeable and hybrid (i.e., rigid centre and soft skirt) materials, gas permeable contact lenses remain the most commonly used contact lens type as high levels of irregular astigmatism cannot normally be corrected with other contact lens types (A popular design is the Rose K lens).[1,2] Moderate keratoconus may require the use of intralimbal rigid gas permeable lenses or miniscleral lenses. Very advanced cases with large, decentered cones, dry eye, or discomfort with conventional lenses may be corrected with the use of scleral lenses. Other options include piggyback lenses and hybrid lenses. The logistics and cost associated with use of these lenses often makes them an impractical choice.[2]

## Contact lens type for keratoconus[1]

| Lens Type | Proprietary name | Base curve (mm) | Diameter (mm) | Power (D) |
|---|---|---|---|---|
| Soft | Kerasoft | 8.00–9.00 | 14.00–15.00 | ±30.00 |
| | Soft K | 7.00–8.20 | 14.20 | +10.00 to –20.00 |
| Gas permeable | Rose K2 | 4.30–8.60 | 7.90–10.40 | ±30.00 |
| | IKone | 4.80–7.70 | 8.80–10.40 | ±30.00 |
| | Soper | 5.20–7.50 | 7.50–9.50 | ±30.00 |
| | McGuire | 5.60–7.35 | 8.60–9.60 | ±30.00 |
| | Dyna Intralimbal | 5.92–9.28 | 10.4–12.00 | ±25.00 |
| Mini-scleral | SoClear | 5.82–7.82 | 13.30–15.50 | +20.00 to –15.00 |
| | Digi Form | Any | 13.50–16.00 | Any |
| | Maxim | Any | 15.40–16.40 | Any |
| Scleral | Innovative | Any | 18.00–24.00 | Any |
| | GelFlex | Any | 18.00–24.00 | Any |
| | Tru-Scleral | 7.25–9.00 | 16.00–20.00 | Any |
| Hybrid | Clear-Cone | 100–600* | 14.50 | +5.00 to –15.00 |
| | SoftPerm | 6.50–8.10 | 14.30 | +6.00 to –16.00 |

*Clear Kone provides vault in microns instead of back surface curve in millimetres.

### To be continued……

Surgical management procedures of keratoconus for example, corneal cross-linking and its types, keratoplasty, photorefractive keratectomy, phakic intraocular lenses, refractive lens exchange and combination procedures, etc. would be highlighted in the next issue of Surgical Insights namely ‘Keratoconus and its management- Part 2’.

**Adapted from:**

1. Contact Lens & Anterior Eye 2010; 33: 157–166.
2. Clinical Ophthalmology 2013; 7: 2019–2030.
3. Indian J Ophthalmol 2013; 61: 382-3.
4. http://cdn.intechopen.com/pdfs-wm/31120.pdf Last accessed on 23rd February 2014

# NOTES

## Surgical Insights 1st Issue
## Immediate sequential bilateral cataract surgery (ISBCS)

**Summary :**

Cataract extraction with intraocular lens (IOL) implantation is the most common surgical procedure worldwide. The frequency of second-eye surgery accounting for approximately 30% to 45% of all cataract extractions has increased.

Today, majority of surgeons perform delayed sequential bilateral cataract surgery (DSBCS), scheduling surgery in the two eyes approximately 4 to 6 weeks apart. An emerging trend, however, is to perform both surgeries on the same day. Immediate sequential bilateral cataract surgery (ISBCS) is being performed with increasing frequency worldwide and a number of surgeons believe this may be the future of cataract surgery.

This issue of surgical insights gives a brief update on the ISBCS indications, contraindications, guidelines for performing ISBCS, benefits and a look at the controversies with the use of ISBCS.

## Surgical Insights 2nd Issue
## Femtosecond Laser Cataract Surgery

**Summary :**

Cataract surgery has evolved tremendously since it was first performed. In 2010, the US Food and Drug Administration (FDA) approved femtosecond laser system for cataract surgery. This technology has the potential to significantly impact cataract surgery as the laser offers an opportunity to better meet patients' expectations.

This issue of surgical insight is an update on femtosecond laser cataract surgery wherein it gives an overview of the uses of femtosecond laser in cataract surgery, its benefits, and limitations and also mentions Indian experience of femtosecond laser cataract surgery.

## Surgical Insights 3rd Issue
## Complications of Phacoemulsification & Manual Small-Incision Cataract Surgery (SICS)

For a number of years, phacoemulsification has been the method of choice for cataract extraction in developed countries. Phacoemulsification can offer a safe and elegant disassembly and aspiration of the lens and a rapid recovery for patients. However, it can also lead to serious complications when used to remove extremely dense cataracts commonly encountered in developing countries. Manual small-incision cataract surgery (SICS) has emerged as a cost-effective alternative to phacoemulsification in the developing world.

This issue of surgical insights gives a brief comparison on the complications occurring due to phacoemulsification as well as manual small-incision cataract surgery.

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*Ref:*

1. *Cipla Data on File*
2. *Invest Ophthalmol Vis Sci 2004;45: E-Abstract 4936*
3. *Katz et al. A fourth generation ocular fluoroquinolone. A suppl. To refractive care for ophthalmologists. Vol 7, no 3, march 2003*


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