

MANAGEMENT OF KERATOCONUS WITH CONTACT LENS

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CERTIFICATE

This is to certify that this dissertation entitled “MANAGEMENT OF KERATOCONUS WITH CONTACT LENS” is a bonafide work done by Dr.ARUNA.P.J under the guidance and supervision in the Department of Cornea, Aravind Eye Hospital and Post Graduate Institute of Ophthalmology, Madurai during the period of her Post Graduate training in Ophthalmology from May 2011 to April 2014.

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PROFORMA

ABBREVIATIONS

MASTER CHART

INTRODUCTION

Keratoconus , a condition of obscure etiology has been known from as early as 17th century and was first described by Nottingham. Most of associations of keratoconus are incidental probably, though multitude of predisposing causes exist as well. Recent advances in videokeratoscopy gives us opportunity to evaluate interesting details in the disease. The advent of specialised contact lenses and revolutions in surgeries like keratoplasty has made a break through in the management of keratoconus .

Keratoconus is a degenerative disorder of the eye in which structural changes within the cornea cause it to thin and change to a more conical shape than its normal gradual curve. The word keratoconus is derived from greek word kerato-cornea, konos-cone ¹. Keratoconus is a non-inflammatory asymmetrically progressive disease of cornea characterised by progressive corneal thinning and protrusion. The protrusion usually affects the central and paracentral cornea (inferonasal). The disease is usually bilateral¹, although unilateral cases have also been reported.

Epimediology:

Keratoconus is a common corneal disorder that is found to have a prevalence ranging from 50 to 230 per 100,000⁶. The incidence is reported to be 2 per 1,00,000 every year. Keratoconus affects males and females

equally with an onset during early adulthood. The disease affects all races. It is typically diagnosed during adolescence and becomes most severe between the ages of 20 and 40.

Etiopathogenesis

Etiology:

Etiology is multifactorial and mostly unknown. The most likely factors are:

- 1) Minor traumas caused externally
- 2) Poorly fitted contact lenses
- 3) Ocular allergies such as vernal keratoconjunctivitis
- 4) Eye rubbing due to atopy^{3,4,6}

All the above causes lead to release of cytokines from corneal epithelium that stimulates apoptosis of keratocytes i.e. the programmed death of cells that occurs following injury. Researchers also suggest that the apoptosis of keratocytes is accelerated in keratoconus. Studies demonstrate higher levels of gelatinolytic and collagenolytic activities in corneas with keratoconus. Though keratoconus is a disease of non-inflammation, inflammatory molecules like interleukins and TNF (Tumour Necrosis Factor) are found to be elevated. These interleukins and TNF mediate production and activation of proteases. Proteases breakdown the collagen cross-linkages in the corneal stroma of cornea. There is a simultaneous

reduced expression of protease inhibitors. The ratio of keratan sulfate to dermatan sulfate is altered in the stromal matrix when compared to normal corneas. Patients with keratoconus have also shown enzymatic changes in the epithelium with increased expression of lysosomal and proteolytic enzyme.

Pathology:

Keratoconus involves every layer of cornea.

Epithelium and Bowman's layer:

Enlarged, elongated epithelial cells.

Degeneration of basal epithelial cells



Disruption of the basement membrane



Growth of epithelium posterior to the Bowman's layer and collagen anterior to the epithelium



Z-shaped interruptions or breaks in the Bowman's layer



Scarring of the Bowman's layer and the anterior stroma



Stroma:

Thinning of stroma due to reduction of number of collagen fibres.

However, the size of collagen fibres are normal.

Histopathology shows fibrillation, fragmentation of collagen and increase in activity of fibroblasts.

Endothelium:

Pleomorphism of endothelial cells

Polymegathism of endothelial cells

With increasing duration and severity, base undergoes more pathological changes than the apex of the cone.

FAMILIAL KERATOCONUS:

Majority keratoconus patients presenting to practitioners have sporadic form of disease. However, the involvement of genetic factors and familial keratoconus has also been reported. Autosomal dominant pattern of inheritance with reduced penetration is displayed in familial keratoconus.

Higher concordance rate has been reported in monozygotic twins⁸. 6-8% of keratoconus cases reveal positivity of family history⁹ and first-degree relatives have 15-67-times of more prevalence when compared with general population. In addition, first-degree relatives who are unaffected have a increased incidence of abnormal topography than general population. Genetic heterogeneity are of two types.

1) Allelic heterogeneity - same locus with different mutations

2) Locus heterogeneity - different loci producing same phenotype.

The basis of genetics in keratoconus is being studied through mutation analysis and linkage mapping to reveal its molecular basis. Mapping studies identified many loci for autosomal-dominant pattern of inherited keratoconus. They are: 16q22.3-q23.1, 2p24, 20p11-q11^{12,13}, 15q22.32-24, 3p14-q13 and 5q14.3-q21.1¹⁸. Numerous potential loci are reported, supporting genetic heterogeneity.

CLINICAL PRESENTATION:

SYMPTOMS:

Patients with keratoconus usually present in their adolescence with:

Blurred/ distorted vision

Ghost images/ monocular diplopia

Photophobia

Glare at night

Haloed around lights

Frequent change of glasses

SIGNS:

Munson's sign- When a keratoconic patient is asked to look down, angulation of the lower lid occurs due to corneal protrusion.

Rizzuti's sign - When a light is shone temporally, conical reflection of the nasal cornea is seen.

Moderate: beam central to limbus

Advanced: beam displaced peripherally.

Vogt's striae- Deep vertical lines parallel to the axis of the cone at the level of stroma and Descemet's membrane which disappear temporarily on digital pressure.



Fleischer's ring- Yellowbrown pigment ring seen in the deep epithelial layer. The significance of the ring is that it delineates the base of the cone. It may be partial or complete. The ring is better seen with cobalt blue light. The colour of the ring depends on the amount of ferritin deposited.

Prominent corneal nerves.

Scissoring of retinoscopic reflex- Appreciated better when pupil is dilated. It is produced due to presence of two conjugate foci in the axis of pupil and high irregular astigmatism. The central part of cone is hypermetropic compared to myopic periphery. During retinoscopy, at the point of neutralization the central zone produces a reflex that moves with the streak due to hypermetropia and peripheral zone produces a reflex that moves against the streak due to myopia which produces scissoring reflex.

Direct ophthalmoscopy from a distance - Shows an oil droplet reflex "Charleaux sign" that is seen with dilated fundus examination. Annular dark shadow separates bright reflex of central and peripheral areas. This occurs because of total internal reflection of light.

Placido disc – Distortion of the mires

Keratometry- shows irregular astigmatism where the horizontal and vertical meridian are not 90degrees apart and superimposition of mires do not occur.

Acute Hydrops :

In a keratoconic eye, rupture of the descemet's membrane and the endothelium that is overlying leads to imbibition of aqueous into the corneal stroma through the ruptured DM which causes stromal swelling (stromal edema) with overlying epithelial edema (bullae). Patient presents with redness, severe pain and a drop in visual acuity. The condition resolves in a few weeks to few months with resultant scarring.

Associations of keratoconus:

Systemic associations:

Down's syndrome

Ehlers-Danlos syndrome

Osteogenesis Imperfecta

Marfan's syndrome

Crouzon's syndrome

Laurence Moon Bardet Biedl syndrome

Nail patella syndrome

Neurofibromatosis

Pseudoxanthoma elasticum

Turner's syndrome

Xeroderma pigmentosa

Ocular associations

Corneal disorders:

Atopic keratoconjunctivitis

Axenfeld's anomaly

Corneal amyloidosis

Essential iris atrophy

Fuch's corneal dystrophy

Microcornea

Lattice dystrophy

Non corneal disorders:

Retinitis Pigmentosa

Vernal conjunctivitis

Leber's Congenital Amaurosis

Gyrate atrophy

Aniridia

Congenital Cataract

Ectopia lentis

Lenticonus

Macular coloboma

Retinal dysplasia

Floppy eyelid syndrome

Modified Krumeich classification of keratoconus :

Stage	Characteristics
I	Eccentric corneal steepening Induced myopia and/or astigmatism <5D Corneal radii $\leq 48D$ Vogt's striae, no scars
II	Induced myopia and/or astigmatism >5D, < 8 D Corneal radii $\leq 53D$ No central scars Corneal thickness $\geq 400 \mu m$
III	Induced myopia and/or astigmatism >8D, <10D Corneal radii >53D No central scars Corneal thickness 200—400 μm
IV	Refraction not measurable Corneal radii >55D

Central scars, perforation

Corneal thickness <200 μm

Imaging in keratoconus¹⁰⁵ :

Numerous imaging modalities are now available to identify subtle abnormalities in curvature of cornea, corneal thickness and tissue architecture. In the initial days, imaging of keratoconus was done with photographic placido disc studies, keratometry, photokeratoscopy and computer-assisted videokeratoscopy. Corneal topography used widely nowadays has evolved through placido disc based devices to slit scanning and Scheimpflug imaging devices and serves as the most important diagnostic tool in imaging for keratoconus. Though placido based devices are highly sensitive in diagnosing curvature changes on the anterior surface of cornea, they might miss early posterior corneal ectatic changes. Newer devices like Scheimpflug imaging and (OCT)optical coherence tomography of anterior segment are useful adjuncts in imaging early indicators of keratectasia.

Computer-assisted videokeratoscopy/placido disc based corneal topography:

The common topographers used nowadays are based on principles of placido disk. Though many such devices are currently available elevation

based topography is mostly used. These instruments primarily consist of either a placido disk-type nose cone or large placido disc consisting of different number of dark and light rings and sometimes colors. Central camera captures a placido disk image that is reflected from thin tear film on the cornea into system based on computer. This computer based system analyzes the data. A scan of good quality gives accurate measurements of corneal curvature and the pre-requisites for a good scan are :

- 1)Stable tear film
- 2)Good patient fixation
- 3)Adequate exposure of cornea
- 4)Superior and inferior quadrants of cornea not obscured by lids.

Topographic scales

The following colours are true for most of the “standard” scales. But different topographers use different steps of colors, hence comparing two different topographers is difficult.

Red and orange colours on the map are considered warmer colours and they indicate steep cornea with high keratometric dioptric power.

Violet and blue colors are considered cooler colours and they indicate flat cornea with low dioptric power, Green and yellow indicate colors found in the normal cornea. Topography of same cornea may look different

with change in steps of the color. Smaller steps help to increase the sensitivity to identify early keratoconus, however it can diagnose a normal cornea falsely as keratoconus, whereas larger steps of colours may miss out the early changes. Hence, the topography must not be evaluated only based on colors .

Absolute or standardized scale:

Absolute scale map has a fixed color-coding system for that particular instrument. Same colours always represent same dioptric steps, minimal and maximal diopters. These maps taken over a period of time are good for direct comparison to analyze the progression of keratoconus and to detect gross pathologies. However, the colour steps are in large increments (some 0.5 D and others 1.5 D) and the disadvantage is that subtle changes of curvature are not identified and subtle local changes like early keratoconus can be missed.

Normalized or relative scale:

These maps have different color scales that are assigned to each map. Here computer identifies the minimal and maximal dioptric values in the map and distributes the range of colors automatically. According to the range in a given cornea, computer contracts or expands its range of colours. Advantage of normalized map is that it shows more topographic details

because dioptric range assigned to every color is smaller when compared to the absolute map. Disadvantage is that colors of the different maps from even the same cornea cannot be compared directly because they have different steps. A normal cornea may have different color and if interpreted only based on the colors it may look abnormal.

Curvature/power map:

Axial curvature map or sagittal curvature map:

Most commonly used map. This map measures the curvature at a certain point on the surface of cornea in axial direction relative to center. It helps in evaluating overall corneal shape. The advantage of the map is that pattern diagnosis of the map can be done and the map can be distinguished into normal or abnormal. Typical topographic patterns of diseases is used to identify them easily. For example: keratoconus has asymmetric bow tie pattern and skewing of radial axis and pellucid marginal degeneration has “butterfly” or “crab-claw” pattern. Disadvantages of the map are inaccurate measurement of peripheral curvature and smaller or local irregularity can be missed.

Tangential curvature map or instantaneous map or meridional curvature map:

This map measures the curvature on the surface of cornea at a certain point in a meridional direction relative to other points on the particular ring. Tangential curvature maps are very sensitive in detecting local change of corneal curvature, hence useful in detecting early changes that could have been missed by axial map. Tangential curvature maps are more accurate than the axial map in periphery of cornea. The disadvantage is that it is subjected to variations because it detects localized changes and so for the same disease similar topography may not be obtained making pattern diagnosis difficult.

Elevation map :

Elevation is not measured by placido based topographers directly. However, certain assumptions lead to the construction of elevation maps. Elevation of a point on the surface of the cornea displays the height of the point (in micron) on the corneal surface relative to a reference surface. This reference surface in most of the instruments is a sphere. Some systems also allow various other shapes like ellipsoid, toric ellipsoid, torus as reference surface. The best mathematical approximation of actual surface of cornea is called best-fit sphere and is calculated using instrument software for each elevation map separately. Also in the same individual, size or radius of

curvature of the best-fit sphere might differ from test to test. The same surface may appear different when mapped against different surfaces of reference. Hence, direct comparison of two elevation maps that have slight difference in best-fit spheres as reference values is difficult and comparison can only be intuitive. Some elevation based topographers have an option of changing and matching the radius of curvature of the best-fit sphere of two different maps. It is also important to check for the scan quality using their raw data or “Quality Score”. Also direct comparison of two maps requires x-y alignment of the two maps, and a few elevation based topographers are capable of doing this.

Statistical indices:

Common indices are:

1) Simulated keratometry (SimK): Equivalent to keratometry and is calculated at steepest axes and at 90° to the steepest axes from the average power at the central 3 mm zone. Difference is calculated as cylinder (Cyl).

Flattest axes (MinK) can also be measured.

2) Surface asymmetry index (SAI): Difference in corneal power on the same ring between points that are 180° apart, that quantifies progression of keratoconus.

3)Surface regularity index (SRI): Points that are in the central 4.5 mm are compared with the surrounding points. Presence of high values suggest high irregularity in the corneal surface.

4)Inferior-superior value (I-SV): Calculated from the difference of power between five superior points and five inferior points 3 mm from center at intervals of 30 degrees.

5)Other indices specific for each instrument do exist. Example: corneal uniformity index (CUI), Point spread function(PSF), Predicted corneal acuity (PCA). Patients with normal corneal indices may have poor vision due to disturbances in any other part of the eye.

Diagnostic evaluation¹:

Corneal topography is a valuable tool to diagnose subclinical keratoconus and to track the progression of keratoconus. Rabinowitz has suggested the following quantitative videokeratographic indices for screening keratoconus¹⁰⁰.They include:

- 1)Central corneal power >47.2 Dipoters
- 2)Inferior-superior dioptric asymmetry > 1.2
- 3)Sim-K astigmatism >1.5 Diopters
- 4)Skewing of radial axis $>21^\circ$.

Maeda and Klyce used eight indices and developed a topographic classifier for diagnosing keratoconus¹⁰¹. In this classification, keratoconus prediction index (KPI) > 0.23 is indicative of keratoconus. KCI% is derived using a binary decision-making tree with input from KPI and four other indices. KCI% value > 0 is indicative of keratoconus.

Evolution in detection of keratoconus has resulted in indices refinement continuously such as KISA% as described by Rabinowitz and Rasheed¹⁰². The product of the following four indices gives KISA% index. The following equation is used:

$$\text{KISA\%} = (K) \times (I - S) \times (\text{AST}) \times (\text{SRAX}) \times 1/3$$

K value - Expression of steepening of central cornea

I-S value - Expression of inferior-superior dioptric asymmetry

AST - corneal astigmatism index that quantifies regular corneal astigmatism (Sim K1-Sim K2)

SRAX - skewed radial axis index, Expression of irregular astigmatism that occurs in keratoconus.

KISA value $\geq 100\%$, patient is likely to have clinically detectable keratoconus.

KISA value of 60 to 100% - indicates keratoconus suspects (less than 0.5% overlaps with normal population). This is a useful index for screening of

candidates undergoing refractive surgery. Patients suspected to have the disease should be followed over time for monitoring of signs that indicates further keratoconus development. Such patients are better deferred from any refractive surgery.

The Orbscan (Bausch and Lomb) provides data on anterior elevation, posterior elevation, best-fit sphere and corneal pachymetry and can be used as screening tool to rule out patients suspected to have keratoconus. Pentacam is an imaging device that is recently introduced which provides accurate measurement of corneal power, elevation and pachymetry (Oculus). Pentacam uses a rotating Scheimpflug camera. This Scheimpflug system determines net corneal power, elevation maps, AC (anterior chamber) depth and corneal wavefront. It is an excellent method for detecting forme fruste keratoconus and suspects keratoconus which is done by calculating corneal thickness spatial profile and corneal volume distribution.

Topographic pseudokeratoconus:

Contact lens wear is the common culprit (both soft hard and) for this condition. Contact lens wear induces inferior steepening pattern that is difficult to distinguish from keratoconus. However such patterns disappear when contact lens wear is stopped. Topographic pseudokeratoconus can result from technical errors during the procedure such as compression of

inferior eyeball while retracting the eye lids, misalignment of eye due to inferior or superior rotation of eye ball, incomplete mires digitization, dry spots formation that simulates inferior steepening. Conditions like pellucid marginal degeneration, Terrien's marginal degeneration, keratoglobus, corneal scar , previous ocular surgery can also cause inferior steepening.

Corneal OCT:

Corneal thickness (pachymetry) measurement has important applications related to diagnosis and surgery in keratoconus and other corneal ectasias. The Ocular Coherence Tomography is a noncontact modality for imaging which provides cross-sectional analysis of corneal thickness with high resolution. Though OCT scanners of retina can measure central corneal thickness, due to slow scanning speed and motion artefacts pachymetric mapping is not possible. Corneal OCT provides a precise pachymetric map, unlike ultrasound pachymeters which provides only spot pachymetry. The five OCT parameters in pachymetry which shows high sensitivity and specificity in diagnosis of established keratoconus are:

- 1) Minimum-median – A cut off value of 62.6 μ
- 2) The I-S: Difference of average thickness of inferior (I) octant and superior(S) octant (A cut off value of 31.3 μ).

3)The IT-SN:Difference of average thickness of IT octant and SN octant (A cut off value of 48.2 μ).

4)Minimum thickness (A cutoff value of 491.6 microns).

5)Vertical location of the minimum. Locations superior to vertex of cornea - positive values, locations inferior to vertex of cornea -negative values (A cut off value of 716 μ).

With Fourier domain OCT, epithelial thickness profile maps have been recently used to detect subtle changes in epithelium, a sign of early keratoconus. Thinning of epithelium over the corneal apex in early ectasias may mask the topographic changes on anterior surface of cornea. In the past, ultrasound of high frequency was used to demonstrate precise epithelial thickness profiles which were useful in diagnosing keratoconus at an early stage. However Fourier domain OCT provides a simpler and noninvasive method to perform corneal epithelial analysis.

The OCT is also useful in studying optical characteristics of cornea after surgical interventions like collagen cross-linking. After cross-linking:

1)First few weeks - faint hyperreflectivity in anterior stroma.

2)Around 1 month - distinct demarcation between cross-linked and the non-cross-linked areas of cornea is noted.

3)By 3 months - demarcation line fades and is sometimes replaced with faint irregular hyperreflective lines in deeper stroma.

In previous cases of hydrops, OCT is extremely useful to identify Descemet's membrane irregularity and helps in deciding about deep anterior lamellar keratoplasty. Hand held OCT with high resolution is a useful tool to identify the extent of residual cornea in cases that underwent lamellar keratoplasty. OCT also shows the corneal stroma that is posterior to intracorneal ring segments. OCT in cases of post keratoplasty helps us to study the architecture of wound and wound apposition posteriorly and suture removal can be decided.

Management OF KERATOCONUS:

Treatment of keratoconus varies considerably depending on stage of disease patient presents with. There are various modalities available today and they can be broadly divided into non-surgical & surgical.

NON-SURGICAL MODALITIES:

Spectacles:

Early stages of keratoconus is managed by use of spectacles. As the disease progresses, irregular astigmatism sets in due to changes in corneal shape. This results in poor quality (suboptimal) of vision with spectacles. Due to increasing levels of irregular astigmatism and higher-order

aberrations, patient with keratoconus will no longer be able to obtain good visual acuity and contact lenses will be required.

Contact lenses:

Contact lenses are the main stay of nonsurgical management in moderate & advanced keratoconus. The primary goal of fitting different types of contact lenses is to provide optimum visual acuity with good comfort without affecting the health of cornea. Barr *et al.*, in his study has shown that modifying the contact lens fitting can reduce corneal scarring. Selection of suitable contact lens is on the basis of manifest refraction and degree of keratoconus.

Contact lens fit- Parameters:

Diameter of the contact lens

Base curve and

Power.

Contact lenses that are used in the management of keratoconus are broadly classified into-

Soft contact lens:

Indicated in early keratoconus where soft lens with toric design adequately corrects myopia and the regular astigmatism. Soft lenses are known for their comfort. Glasses can be prescribed over these soft contact

lenses. Selection of soft lens – thicker lens and low water content neutralizes irregular astigmatism. Larger diameter soft lenses are used for severe apical displacement and globus cone²⁶.The various soft toric lenses available are Soft K (Advanced Vision Technologies),) Ocu-Flex Toric (Ocu Ease), HydroKone (Medlens Innovations SpecialEyes 59/54, Solus Soft K (Strategic Lens Innovations). Soft contact lenses are also used in patients intolerant to rigid lenses and discomfort with rigid lenses.

Rigid Gas Permeable lens:

Corneal RGP lenses are the lens of choice for visual improvement in keratoconus.Rigid lenses usually rest on the corneal apex, tear fluid fills the gap between the regular smooth inner surface of lens and the irregular outer corneal surface, creating the effect of smoother cornea, thus producing improved vision. To prevent hypoxia related changes on the cornea lens material with high oxygen transmissibility should be selected. Dynamic and static fit to be assessed after insertion of lens on the eye for 30 minutes (adaptation period).



Dynamic fit -lens fit is acceptable:

- 1) when the lens is centered on the cornea adequately during post-blink movements
- 2) good stability in different gazes
- 3) patient is comfortable during all eye movements
- 4) The movement should be not more than 1mm with every blink
- 5) The lens should not cross the limbus.

Static fit : Assessed after instilling fluorescein in the eye and examining with cobalt blue filter with or without the Wratten filter. Yellow gelatine paper can be used if Wratten filter is not available to assess the fitting.

Advantages of RGP Lenses :

- 1) Produces a smooth corneal surface
- 2) Provides good tear exchange

Disadvantages of RGP lenses:

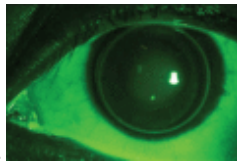
- 1) Stability (more likely to decenter)
- 2) Difficulty in adaptation.

RGP CONTACT LENS FITTING PHILOSOPHIES:

Apical clearance:

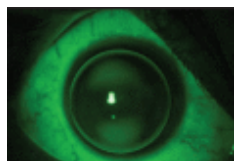
There is no bearing or touch in the corneal apex. i.e., vault is present (apical area is clear) and the lens bearing is directed towards the periphery

(corneal periphery bears the lens). Usually practised in smaller diameter steeper lenses, centered over the cone (usually decentered in keratoconus). This sometimes results in lens edge in the visual axis or optic zone bifurcating the pupil and flare or fluctuating vision. Advantages are: reduced risk of scarring, erosions and whorl keratopathy. However tightening at the periphery may result in sealing of tear exchange.²⁸



Apical bearing:

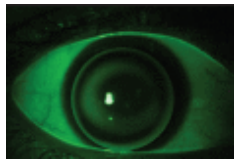
The optic zone of contact lens bears or touches on the apex of the cone. This can cause heavy bearing on the cornea because of the flatter fitting of the lens on the cornea, which might result in corneal scarring and intolerance to contact lens over long term use. However, visual quality is better with apical bearing.



Apical bearing i.e. bearing in the centre seen as dark area because of lack of fluorescein in this area

Three point touch:

This is also called divided support. Preferred type of lens fitting by many optometrists. The lens bearing is shared between the apex and the midperipheral cornea. Thus helps in minimizing the risk of apical scarring and also facilitates tear exchange. Advantages are good vision improvement, better patient comfort and can be worn for a prolonged time.



Three point touch-Diffuse fluorescein in the centre and midperiphery on both sides

An ideal fit will show a central feather touch and a peripheral edge lift of 0.5-0.7 mm. As the cone advances, there can be air bubbles under the peripheral edge and lens edge may stand off with pooling.

Rose K lens:

The Rose K is a unique keratoconus lens design that has complex computer-generated peripheral curves based on data collected by Dr. Paul Rose of Hamilton, New Zealand. The system 26 lens set) incorporates a triple peripheral curve system ^{29,30}

These lenses are multicurve lenses and has a small optical zone. This small optic zone snugly fits the cone, for the so called 'feather touch' to the

cornea. When the cone is a central nipple cone, a Rose K lens can be recommended.

Selection of trial lenses:

KERATOCONUS	AVERAGE K VALUE	TRIAL LENS K
Mild to moderate	Flatter than 6.0 mm	0.2mm steeper than average K
Severe	5.1-6.0mm	Same as average K
Advanced	Steeper than 5.0mm	0.3mm flatter than average K

Selecting the initial trial lens base curve based on the 5-mm average K reduces the number of trial lenses and shortens chair time in fitting Rose K lenses in eyes with keratoconus³¹. When the optimal fit is achieved, final power is calculated by performing a spherical over refraction over the trial lens. Rose K2 XL semi-scleral contact lens also improves visual acuity and comfort and is shown to avoid the need for piggy back contact lens in managing irregular cornea³².

Intralimbal lenses:

These are lenses with larger diameter ranging between 10.5-12 mm. Useful in moderate keratoconus and low sagging cones. The larger diameter

helps in reducing movements of lens, improving lens centration. Three point touch method of fitting is preferred. As these lenses are of larger diameter, patients find it cumbersome to use it for the first time. examples of Intralimbal lenses are KBA 10.2 mm (Precision Technology Services), DynaZ Intralimbal

11.2 mm (Lens Dynamics), Rose K2 IC 11.2 mm (Blanchard CL), GBL 11.2 mm (ABB-Concise), I Kone 10.4 mm (Medlens and Valley Contax) and XL-T 11.0 to 12.0 mm (Medlens Innovations).

The I Kone lens has bisurface aspheric design which are used when rigid gas permeable lenses either do not center or causes bearing on center of cornea. The posterior surface of I Kone lens has four conic zones: central portion vaults the cornea. The first and second zones manage corneal ectasia by distributing pressure over larger area. The third and fourth zones in peripheral curve system provide an alignment fit over non-ectatic cornea. central portion vaults cornea. The asphericity of the anterior surface of the lens aims to reduce spherical aberrations. I Kone lenses are available in diameters of 9.6 mm, 8.8 and 10.4 mm. However the standard diameter is 9.6mm.

Fitting of intralimbal lenses:

Light feather touch or central corneal vault with moderate peripheral clearance and midperipheral bearing. Though these lenses do not move much like the traditional rigid lenses, they provide adequate tear exchange.

Piggy Back Contact Lens:

This contact lens system consists of two lenses in one eye i.e. rigid lens fitted on top of a soft contact lens. Aim is to obtain the same visual acuity as with a single lens. These lenses are used in difficult situations like cases of RGP lens intolerance, pellucid marginal degeneration, proud nebulae in keratoconus, apical dimpling, post graft, recurrent epithelial erosion, post LASIK ectasia, unstable RGP on the eye, lens popping out, staining at 3 and 9'0 clock with RGP fit.

Fitting of piggy back contact lens:

First a diagnostic soft contact lens, preferably silicone hydrogel lens is fitted – usually a lens with minus power. After obtaining an optimal fit with soft contact lens, corneal topography or keratometry is performed over the surface of the soft lens to select the base curve of the rigid gas permeable lens. RGP lens base curve is selected on flat K value with 9 -9.5 mm diameter. Both dynamic and static fit are to be assessed for RGP. Movements of soft lens and rigid lens are independent of each other. Once

optimal fits for both the soft lens and RGP lens are obtained, over refraction is performed and final subjective acceptance is added to rigid lens power. It is wise to choose both lenses of high Dk material to reduce the risk of corneal hypoxia. Final fit of RGP lens can be modified by modifying power of soft contact lens. Plus power soft lens to flatten the RGP and minus power soft lens to steepen RGP²⁶. Custom piggy back lenses which are lathe cut lenses has a groove that is present in soft contact lens. RGP lens 1mm smaller than the groove diameter of soft lens is selected and placed in the groove of soft contact lens. This allows the RGP lens to center well on the surface. Also presence of RGP with a less thick lens edge in the groove provides better comfort because of minimal interaction with the lids. The reduced thickness reduces the recurrences of complications related to hypoxia. Disadvantages are:

- 1)lost RGP

- 2)Difficulty in handling and maintaining two lenses

- 3)Giant papillary conjunctivitis

- 4)Increased risk of hypoxia due to presence of two lenses

on corneal surface³³.

Hybrid lenses:

Hybrid lenses have a soft skirt in periphery and rigid lens in centre. Recently introduced hybrid lenses are the SynergEyes lens (SynergEyes, Inc, Carlsbad, CA) with multiple designs. These SynergEyes lens have a central rigid gas permeable portion made of Paragon HDS100 material (also called Paflucocon D) which is bonded nonionic hydrophilic skirt material containing 27% water (PolyHEMA hem-iberfilcon A). The diameter of rigid central portion is 8.4mm and the total diameter of lens is 14.5mm. The curvature of posterior portion of RGP is described as a 'prolate ellipsoid'.

Fitting of hybrid lenses:

Hybrid lenses are fitted with no or minimal apical touch in the centre of cornea. The vault should be between 100-600 microns. Soft skirt should exhibit blink induced movements of 0.25mm. Nowadays lens is fitted towards flatter side³⁴ to avoid lens tightening over a period of time. Rigid lens and soft skirt can be changed individually for different lenses. These lenses can be fitted on all types of cones and cones of any severity. SynergEyes lens allows more amount of oxygen to reach the cornea during wear than the Soft lens in the region of central cornea, 2.0 mm and 4.5 mm temporal to the centre of cornea³⁵. Indications for hybrid lenses are:

- 1) Inability to obtain an optimal RGP fitting
- 2) RGP intolerance
- 3) Poor centering of RGP lens
- 4) Reduced RGP lens wearing time

Complications of hybrid lens:

- 1) Giant Papillary Conjunctivitis
- 2) Soft skirt tear
- 3) Infection
- 4) Discomfort
- 5) Corneal vascularisation
- 6) Central corneal clouding³⁶

Scleral lenses:

The availability of high Dk material³⁷ and multiple newer designs has led to the development of sclera lenses. True scleral lenses do not touch cornea and limbus but rest on sclera thus leaving a clear area between the contact lens and cornea. Scleral lenses can either be fenestrated or non fenestrated. Indications for scleral lenses are:

- 1) Suboptimal fit with RGP
- 2) Intolerance to RGP lens
- 3) corneal staining at 3 and 9'o clock

4) vascularisation due to PBCL use

5) advanced keratoconus

6) corneal scarring⁴⁰

7) can also be used in the presence of vernal kerato conjunctivitis

8) Prior to penetrating keratoplasty.

Contraindications are:

1) Acute hydrops. In cases of hydrops scleral lens can be fitted once the hydrops heals⁴¹.

2) In presence of oedema of the cornea.

3) Trabeculectomy and other post filtration surgery.

Fitting of scleral lenses:

Here fitting of scleral lens of non fenestrated type (PROSE) is described. (PROSE- Prosthetic Replacement of Ocular Surface Ecosystem). These lenses are fluid filled before insertion in the eye. PROSE is a lathe cut lens that is manufactured with use of software to customize lens design for haptic bearing or scleral portion of lens and vaulting for clearance of cornea^{39,42}.

With Slit lamp biomicroscopy two parameters are to be assessed: Haptic and the vault. An acceptable sclera lens fit is characterized by adequate clearance of cornea (no touch) without air bubble in fluid and

without impingement i.e. no edge lift or no conjunctival vessel compression. After attaining an acceptable fit, lens is left in eye for the patient to experience comfort and improved vision after trial. Trial can either be continued on same day or patient can be called back later for extended trial of continuous lens wear of four to six hours. This is an important requirement as lens gets settled on ocular surface.

Corneal clearance should be assessed by measuring the distance between cornea and back surface of lens in the central corneal area and compared with peripheral corneal thickness or to known thickness of lens. Acceptable corneal clearance is of 400-600 microns. The vault shall be kept in excess in patients with keratoconus considering progression of the disease over follow-up. Haptic portion lens is assessed by the presence of suction during removal of the lens from eye and from fluorescein staining of conjunctiva after the removal of lens. If there is no suction felt during lens removal and no staining of conjunctiva and no collection of debris in vault after 4 hours of continuous lens wear, lens is ordered with adjustment of power after doing subjective refraction. Flatter haptic is needed when there is presence of suction during lens removal which indicates a tight lens on the surface of eye. Flatter haptic settles more on ocular surface, hence vault is to be increased.

Scleral lenses are custom made based on fluorescein staining of conjunctiva. A pattern of staining in 360 degree requires a flatter haptic and vault increased by 0.1 to 0.2. Staining in one or two quadrants indicates toric sclera and haptic should be designed with toricity³⁹. Patients must be re-examined for final assessment of scleral lens fit after 3 to 4 weeks of dispensing the lens. Normal saline is preferred to fill these scleral lenses before inserting in the eye. Cornea clearance is between 150-600 μ without air bubble in fluid filled lenses in keratoconic eyes. Corneal clearance can be between 40-80 microns in scleral lenses of fenestrated type.

Removal of sclera lenses of fenestrated type is aided by applying plunger at centre and non- fenestrated lenses are removed by applying plunger at junction of the vault and the haptic . Advantages of scleral lenses are:

- 1) Improved comfort with lens
- 2) Stable visual acuity
- 3) Delays or obviates need for keratoplasty.

Disadvantages are:

- 1) Care of scleral lenses
- 2) Removal of lens with plungers
- 3) Requires frequent change of saline bottles.

Contact lens following keratoplasty:

Penetrating keratoplasty or full-thickness transplantation of cornea has been common surgical intervention for irregular astigmatism due to keratoconus. The main cause of decreased visual acuity following keratoplasty is high astigmatism. Even after removal of sutures, residual astigmatism causes visual problems. Various contact lenses have been tried following keratoplasty to improve lens and optical stability. They are: rigid gas permeable lens, reverse geometry hydrogel lens, sclera lens and silicone hydrogel contact lens⁴⁴. In cases with post residual astigmatism less than 4D, a hard gas permeable contact lens with larger diameter is recommended (diameter of donor cornea should be less than the diameter of the contact lens). If astigmatism is less than 1D, refractive status can be successfully corrected with soft contact lenses. Intralimbal lenses are also found effective in increasing visual acuity following penetrating keratoplasty. Special design contact lenses are available to improve vision after penetrating keratoplasty. To date many contact lenses are available for correction of post keratoplasty astigmatism and they can be tried before conducting other surgical methods.

Complications of contact lens wear:

Contact lens compromise the eye by inhibiting tear film cleansing action, compromising barrier function of epithelium and by introducing more micro organisms.

Adverse effects related to corneal hypoxia:

- 1) Corneal stria – Due to buckling of the Descemets membrane and endothelium.
- 2) Microcysts
- 3) Stromal edema
- 4) Neovascularization at the limbus
- 5) Myopic creep- Refractive change towards increase in myopia of 0.5D, which is reversible after shifting to high Dk (diffusion coefficient) lens.

Inflammatory adverse effects:

- 1) Contact lens induced peripheral ulcer
- 2) Superior arcuate epithelial lesions
- 3) Contact lens associated red eye
- 4) Giant papillary conjunctivitis

Mechanical adverse effects:

- 1) Deposits on the lens – protein, lipids, inorganic salts, filamentous deposits.

- 2) Inadequate blinking
- 3) Tight lens syndrome.

Corneal Collagen Cross linking:

Corneal cross-linking (CXL) is an enthusiastically used method for treatment of corneal ectasia and keratoconus. CXL is proved to strengthen corneal structure thereby inhibiting progression of keratoconus⁴⁶. CXL in keratoconus is achieved by generating chemical bonds within stroma of cornea through photopolymerization (localised) that strengthens the cornea while exposure to surrounding eye structures is minimised⁴⁷. Studies have also shown that this procedure has effect on corneal biomechanical properties that results in increase in rigidity of cornea of approximately 70%⁴⁸ which is due to creation of new chemical bonds within the corneal stroma.

Procedure of CXL:

- 1) Preparation of corneal epithelium with weck cell sponge that removes phospholipid layer from corneal surface
- 2) Corneal soakage of photosensitiser – Hypoosmolar 0.5% solution of riboflavin. Cornea and the anterior chamber are seen with slit-lamp to ensure if riboflavin has soaked into cornea.

3)After confirming riboflavin soak, exposure to ultraviolet-A (UV-A) light for 30 minutes is commenced.



Corneal epithelium of approximately 50 μm thickness forms a barrier to penetration of both the riboflavin and UV-A. Hence removal of the epithelium enhances penetration and absorption of riboflavin into the stroma of cornea and anterior chamber that is required for UV-A light to illuminate the cornea efficiently and then to excite the riboflavin. Treatments that are performed with removal of epithelium are called “epi-OFF” treatments and those performed with intact epithelium are called “epi-ON” treatments. New formulations aim at “epi-ON” treatments. During illumination of UV light, riboflavin functions as shield during irradiation to cornea that protects the deeper ocular structures like endothelium, lens, retina from UV-A irradiances which are high⁴⁹. Combination of riboflavin and ultraviolet A light creates 80–95% absorption into stroma of cornea during cross-linking⁴⁸. This depends on concentration and the thickness of cornea.

Throughout the procedure, the irradiation dose used is constant- 5.4 J/cm^2 . Photochemical processes occurring in the stroma of cornea depends on radiant exposure of the ultraviolet-A light. To ensure no damage to corneal endothelium by UV-A light, CXL should effectively occur in the superficial $200\text{--}250 \text{ }\mu\text{m}$ of the stroma of cornea[49]. Damage to endothelium, lens and retina can be avoided if the following criteria are fulfilled:

- 1) Wavelength of $360\text{--}370 \text{ nm}$
- 2) Ultraviolet light intensity of 3 mW/cm^2
- 3) Ultraviolet light energy of 5.4 J/cm^2 .
- 4) Corneal thickness of atleast $400 \text{ }\mu\text{m}$.

Riboflavin also prevents corneal dehydration during the procedure⁵⁰. Modifications of original Dresden protocol includes use of hypo-osmolar 0.1% riboflavin to make thinner corneas swell artificially to reduce the risk of cytotoxicity of ultraviolet light to endothelium, transepithelial crosslinking with use of different compounds that are designed to improve penetration of riboflavin or using iontophoresis, measures to reduce the duration of procedure by increasing the radiance of ultraviolet light⁵¹. However safety and efficacy of the above modifications is yet to be proved.

Adverse effects:

- 1) Bacterial, acanthamoebal, fungal and sterile keratitis ^{58,59}
- 2) Significant loss of endothelium after the procedure in thin corneas ⁵¹.
- 3) Persistent corneal edema ^{65,66}
- 4) Decompensation of cornea ^{65,66}
- 5) Damage to structures of anterior segment ^{65,66,67}

Epi-ON CXL (also called trans –epithelial CXL) is reported to produce less pain for patient and reduces the risk of postoperative infection due to the presence of intact epithelium. The riboflavin concentration in the corneal stroma was found to be 40 times lesser in transepithelial CXL when compared with standard CXL. This reduces the long term success of cross-linking effect on the shape of the cornea⁶⁸. Hence epi -OFF CXL is preferred.

Intrastromal Corneal Ring Segments:

Keratoconus patients intolerant to contact lens and those with clear central corneas benefit from insertion of intracorneal ring segments. ICRS were initially developed for correction of myopia and is now approved for the reduction of myopia and keratoconus associated irregular astigmatism. It is postulated that ICRS placement induces displacement of the local anterior surface of the cornea, causes peripheral corneal steepening and flattening of

central portion of anterior corneal surface by adding extramaterial at midperiphery of the cornea⁶⁹ which provides a good biomechanical support for thin cornea⁷⁰. Intrastromal corneal ring segments are manufactured using polymethyl methacrylate material. These segments contain 150 degrees crescent-shaped arc length. The inner diameter of segment is 6.8 mm and outer diameter of the segment is 8.1 mm when placed in stroma of cornea. The thickness of Intacs varies between 0.25 to 0.45 mm, with 0.05 mm increments. Ring segments are inserted into corneal stroma through femtosecond laser-assisted and mechanically created corneal tunnels.

In mechanical method of insertion of segment, a radial incision of length 1.8 mm is approximately made using calibrated diamond knife at approximately 70% of corneal thickness which is determined by pachymetry (ultrasonic). Corneal pockets are created using pocketing hooks on either side of incision. Then two semicircular dissectors - clockwise and counterclockwise⁷¹ are placed one after the other into the lamellar pocket and steadily advanced into stroma by rotational movement. In recently introduced femtosecond laser-assisted procedure, disposable glass lens of laser system is applanated to the cornea first to fixate eye and this helps to maintain precise distance from laser head to focal point. Then continuous circular corneal stromal tunnel is created at 80% of corneal depth

approximately within 15s. Further corneal manipulation should be avoided⁷². This procedure, performed on outpatient basis which is done under local anaesthesia, is safe and reversible, also restores functional vision in most patients with keratoconus. As no eye tissue is removed, the procedure is potentially exchangeable^{73,74}. Early insertion of Intacs (Addition Technology Inc., Sunnyvale, USA)) also limits the development of scarring of cornea associated with use of contact lens. Colin *et al.* reported improvement of visual acuity and astigmatism which were stable for time and also quite a good amount of reduction in keratometry readings at follow-up of 1 and 2 years compared with the baseline. They also found that creation of more regularly shaped cornea after insertion of intacs facilitates easier fitting of contact lens in patients with keratoconus. Newer modifications to these Intacs segments are reported. A lamellar pocket with adjustable intracorneal ring is one of the reported modification. In this technique, a 9 mm diameter closed pocket of about 300 µm depth is created within the stroma of cornea and a single full-ring implant that is flexible is inserted into this closed corneal pocket through narrow incision tunnel. After insertion of the implant and evaluation of clinical data, the position of implant could be adjusted inside corneal pocket. This helps to achieve optimal treatment result. This technique provides the surgeon the

sophistication of choosing the diameter of the implant, thickness of the implant and position of the implant position⁷⁷.

Intacs SK for Severe Keratoconus - a newer modification of ICRS has a small 6mm optical zone that can correct high grades of keratectasia. It also has an elliptical cross-section that minimises the glare. Rodriguez *et al.* reported significant reduction of keratectasia, improved uncorrected visual acuity (UCVA), keratometry and spherical equivalent after placing Intacs SK in eyes with severe post LASIK ectasia⁷⁸.

The Ferrara ring is another modification of intracorneal ring and is made out of polymethyl methacrylate (Perspex CQ segments of acrylic). These segments vary in their thickness-0.15mm, 0.20mm, 0.30mm and 0.35 mm. Nevertheless, cross-section of the segment is triangular and base of the segment for all thickness is 0.60mm wide. These segments also have 160° of arc and provides 5 mm optic zone. Torquetti *et al.* followed up the keratoconus patients for 5-years after placement of Ferrara ring and reported improvement in uncorrected visual acuity (UCVA) and best corrected visual acuity (BCVA) and significant post-operative flattening of cornea, that remained stable⁷⁹.

Ferrara reported recently results of insertion of a 210° arc new Ferrara ring. These Perspex CQ acrylic segments has a 2.5mm inner radius

of curvature and thickness of 150 to 300 μm . This new model of Ferrara ring is assumed to have the following advantages over conventional ring:

- (1) Minimal induction of astigmatism
- (2) Flattening of cornea
- (3) Single segment implantation.

These rings also showed significantly improved UCVA, BCVA and flattening of central cornea as indicated by decreased keratometry values⁸⁰.

The primary purpose of implantation of ring segments in patients with keratoconus is to reshape cornea and to improve topographic irregularities and hence the visual acuity. Though the results are encouraging in keratoconus after implantation of intracorneal ring segments, residual myopia and astigmatism has to be corrected with use of either spectacles or contact lenses.

Complications:

- 1) Epithelial defects
- 2) Anterior perforations
- 3) Posterior perforations during creation of channel
- 4) Incision extension toward visual axis
- 5) Uneven or shallow implant placement
- 6) Decentration of implant

7) Infectious keratitis

8) Superficialization of segment

9) Exposure of segment

10) Stromal thinning

11) Breakdown of epithelium

12) Corneal melting ⁸²

13) Lamellar deposits in the channel that primarily consists of intrastromal accumulations of lipid (as a response to injury to cornea). Incidence and density of intrastromal deposits was directly related to thickness of segment and duration since segment implantation. However, intrastromal deposits do not alter optical performance of Intacs and also do not interfere with physiological and anatomical changes induced by the insertion of segments in the cornea.

Epikeratophakia:

Epikeratophakia is nothing but suturing a small donor corneal lenticule or epikeratophakia lens to the anterior surface of cornea. Typical epikeratophakia lens consists of central optical zone and a thin wing at the periphery that is sutured to keep lens in place. Most of the epikeratophakia lenses were made using a specially designed cryolathe (Citycrown Sales, 14 Kempston Close, Gatehouse Way, Aylesbury, Buckinghamshire) that

facilitates a smooth transition automatically between radius of cut used for central optical zone and radius of cut used for peripheral wing. Alternatively, lenses are also made with single radius of cut and the central optical zone in these lenses is extended to edge of lens. The procedure of epikeratophakia consists of removal of corneal epithelium initially followed by dissection of peripheral pocket that is used to suture the wing of lens in place. The technique employed nowadays evolved over this series. A scalpel blade is used for removal of epithelium and the use of absolute alcohol should be deferred since use of the same was reported to interfere with re-epithelialisation subsequently. Corneal epithelium is removed thoroughly to avoid unnecessary cellular proliferation subsequently at interface between the host cornea and epikeratophakia lens. Re-epithelialisation over epikeratophakia lens occurs from corneal periphery and hence the epithelium peripheral to the lens is to be left intact. Failure to re-epithelialise promptly may lead to melting of epikeratophakia lens and infective keratitis. The next step is creating peripheral pocket for inserting the lens wing. Partial thickness trephination of 7 or 7.5 mm diameter to a depth of 180 microns is made using a Hessburg-Baron suction trephine. A 21 gauge needle that is bent 2 mm from the end to 90degrees is used for dissection of pocket parallel to surface of cornea from the base of partial thickness trephination

extending to periphery. The final step of the surgery is to fix the lens in place. 10-0 monofilament sutures are used for fixation. Eight sutures are usually sufficient though 16 sutures were used initially. These lenses can also be fixed with fibrin glue. In keratoconus sutures are tied very tight while assistant presses firmly to reduce corneal ectasia. Here the sutures are tied with the help of an operative placido disc and sutures are removed or added as dictated by symmetry of corneal reflex in an attempt to reduce suture induced astigmatism. These sutures are left in place unless they become loose or are inducing astigmatism. Patients were refracted at regular intervals, starting as soon as the epikeratophakia lens clears sufficiently. Reepithelialisation when successful completes by seven days postoperatively.

Toric implantable collamer lens:

The toric implantable collamer lens (Eg: Visian - Staar Surgical AG, Switzerland) is a single-piece plate haptic monoblock lens made out of collamer. Collamer is a highly hydrophilic and extremely biocompatible flexible copolymer of collagen. Refractive index of collamer is 1.452. The material is permeable to oxygen and other nutrients). The lens is designed for implantation in posterior chamber of the eye behind the iris. A layer of aqueous humor separates it from the natural lens with haptics resting on

ciliary sulcus. Toric phakic intraocular lenses⁹⁰ are implanted in keratoconus patients unfit for procedures on the cornea, having poor quality of vision with spectacles, intolerant to contact lens. This can also be combined with other procedures like placement of intacs^{91,92,93}. Other indications are for correction of myopia (-3 to -23.0 dioptres) and astigmatism (upto 6 dioptres). Pre-requisites for implantation:

- 1)ACD - Anterior Chamber Depth of 2.8 mm
- 2)Best Corrected Visual acuity at distance - 20/50 minimum
- 3)No prior ocular surgery
- 4)Normal intraocular pressure
- 5)Open angle of anterior chamber (Shaffer grade 3 and grade 4)
- 6)Normal peripheral retina or done with laser photocoagulation if necessary
- 7)Stable keratoconus - stable refraction for 2 years without any surgical intervention and in eyes that have undergone collagen cross linking refraction should be stable for 3 consecutive months.
- 8)Keratoconus with clear cornea in the centre
- 9) K value \leq 52.00 D
- 10)Centralized cone
- 11)A decentered cone that is centralized with use of intracorneal ring segments as an adjunct^{94,95}.

Pre- operative evaluation:

- 1) Gonioscopy to look for configuration of peripheral iris
- 2) Dilated fundus examination
- 3) Both manifest and cycloplegic refraction
- 4) Corneal curvature using Orbscan and Pentacam
- 5) Intraocular pressure
- 6) Measurement of anterior chamber depth using anterior segment ocular coherence tomography
- 7) White-to-white diameter measured using Orbscan and Digital Calliper
- 8) Size of pupil
- 9) Specular microscopy for corneal endothelial count
- 10) Measurement of diameter of horizontal sulcus using ultrasound biomicroscope.

The power of the lens, size of the lens which is based on horizontal white-to-white diameter and axis of placement of the phakic intraocular lens are calculated with the help of software that is provided by manufacturer. The postoperative target should be close to emmetropia as much as possible.

Procedure of lens implantation:

Prior to surgery, 0-180 degree horizontal axis is marked with a reference marker with the patient in sitting position in primary gaze to avoid

cyclotorsion. Then intraoperatively, the required rotation from the horizontal position is marked using an axis marker. The implantation of lens is performed through clear corneal tunnel of size 3mm on the temporal side. This is to be done under hydroxypropyl methylcellulose. After injecting lens into anterior chamber, haptics of the collamer lens are tucked under iris. The lens is then aligned to desired axis with the help of Vukich manipulator. Hydroxypropyl methylcellulose is finally washed at the end of surgery. Two laser iridotomies are pre-operatively recommended. Alternatively, surgical iridectomy can be performed using vitrectomy cutter with minimum cut rate - 10 cuts per minute and high vacuum (250 mmHg). This is done after injecting preservative-free pilocarpine 0.13 mg/mL intra-camerally which causes constriction of pupil.

Contraindications:

- 1) Corneal degeneration and/or dystrophy
- 2) Low count of endothelium
- 3) Lenticular opacity
- 4) Pseudoexfoliation
- 5) Pigment dispersion
- 6) Glaucoma
- 7) Intraocular inflammation

8)Macular pathology

9)Neovascularization.

Deep Anterior Lamellar Keratoplasty:

Keratoplasty is resorted when all the above measures fail. The common indications for keratoplasty in keratoconus are poorer visual acuity with use of contact lenses, intolerance to contact lens, poor contact lens fit and corneal hydrops that does not resolve. Lamellar Keratoplasty is technique of corneal graft that consists of partial-thickness donor cornea transplantation, without endothelium, Descemets membrane and deep stroma into recipient stromal bed that is healthy after dissecting pathological anterior stroma. In Deep Anterior Lamellar Keratoplasty pathological stroma of cornea is removed down to Descemets membrane that is followed by of endothelium devoid donor cornea transplantation over host bed.

Surgical techniques:

Various methods are described and used based on different pathology of cornea and personal experience of surgeon. They are:

- 1)Direct open dissection
- 2)Dissection with intrastromal air injection
- 3)Dissection with hydrodelamination
- 4) Closed dissection

- 5) Dissection with viscoelastics
- 6) Dissection with “big bubble” technique
- 7) Microkeratome-assisted lamellar Keratoplasty
- 8) Femtosecond laser-assisted lamellar transplantation

Direct open dissection:

In this method, 60–80% of thickness of cornea is trephined using a guarded trephine and a paracentesis is made for injection of fluid and air into anterior chamber and for aqueous removal when required. Stretching and lifting of superficial stromal corneal layers and sweeping of blade in the flat plane over stroma of cornea facilitates dissection. This dissection may be done in several layers but as the dissection progresses to deeper stroma, there is risk of injuring Descemet's membrane as deep stromal fibres are visualized with difficulty. No air or fluid is injected into host cornea to facilitate removal of tissue upto Descemet's membrane.

Dissection with intrastromal air injection:

An air filled tuberculin syringe is connected to 26-gauge needle and is then inserted obliquely into mid-peripheral corneal stroma. As a result of injection of air the corneal stroma becomes opaque. The host cornea is now trephined to a depth to leave behind a safety margin. Dissection of corneal layers is continued, until Descemet's membrane is touched. Endothelium of

donor cornea is removed and the remaining full thickness button is sutured to the recipient cornea.

Dissection with hydrodelamination:

In this procedure, three quarters of thickness of cornea is trephined, then followed by an appropriately deep lamellar keratectomy. Collagen fibers in the corneal stroma are cut across and down and a depression is produced. A 27-gauge needle is used to inject saline at the bottom of the depression. The injected fluid penetrates between stromal collagen fibers, which becomes white. A fine spatula is inserted rectilinearly into this hydrodelaminated stroma and is moved repeatedly like a fan in different directions. The remaining fibers in the stromal bed over Descemets membrane is removed with corneal scissors and a fine forceps. Finally, Descemets membrane in the donor cornea is removed and full-thickness cryolathed donor cornea is sutured to the recipient bed. Recently, a sclerolimbic approach was reported by Senoo et al. in 2005 to detach Descemets membrane from corneal stroma by using hydrodelamination before trephination. Flaps are created as in trabeculectomy and these flaps are used to approach the Descemets membrane. A hydrodelamination pocket is made by inserting a spatula from the flap into the space between Descemets and stroma of the cornea. Now the Descemets membrane is

completely separated from the deep stroma by hydrodelamination. Viscoelastic material is filled in the space between the two and trephination is done.

Closed dissection:

In this procedure, “air to endothelium” interface is created by exchanging aqueous with air that helps in visualization of posterior surface of cornea. A 5mm scleral incision is made using custom made dissection blade and a deep stromal pocket is created across cornea (air to endothelium interface is used as reference plane for depth of dissection). The anterior corneal lamella is excised after filling the pocket with viscoelastic. Descemet's membrane in the donor cornea is stripped and the full-thickness button is sutured to the host cornea. Intraoperative complications like Descemet's membrane rupture and the double anterior chamber formation can be reduced by using a blunt spatula instead of blade for closed dissection and by leaving a minimal thickness of deep stroma over the host bed intentionally.

Dissection with viscoelastics:

Two techniques are described: 1) An “air to endothelium” interface is created by exchanging aqueous with air that helps in visualization of

posterior surface of cornea. A 30G needle is inserted anterior to Descemet's membrane and viscoelastic substance is injected. This dissects the Descemet's membrane from posterior stroma. Anterior corneal lamella is excised after trephination. Descemet's membrane in the donor cornea is stripped off and the full-thickness button is sutured to the recipient cornea.

In second technique, after a deep trephination (comparatively), a pocket is created in the deep fibers of the stroma. With a 25G blunt cannula, viscoelastic is injected through this pocket. Under the injection force, viscoelastic material enters the plane between deep stroma of cornea and Descemet's membrane and opens up the potential space.

Dissection with “big bubble” technique:

This popular technique was demonstrated by Anwar and Teichmann. Corneal stroma is trephined upto 60-80% depth before injection of air. An air-filled syringe is attached to 30 gauge needle that is bent at 60° (bevel facing down) 5 mm from tip., Then this needle is carefully inserted into stroma of cornea in the paracentral area under direct visual control. As a result injection of air, a whitish semiopaque disc that is circular is achieved. This indicates big bubble formation. The big bubble detaches the central part of Descemet's membrane from the deep stromal layers. Anterior stroma of the host cornea is excised by partial keratectomy. At the anterior wall of this

big air bubble, small incision is made and after inserting a fine blunt spatula into the bubble, stroma over spatula is excised. Descemet's membrane in the donor cornea is stripped off and the full-thickness button is sutured to the recipient cornea.

Microkeratome-assisted lamellar Keratoplasty:

The use of advanced microkeratome instrumentation in LASIK and newer microkeratome modifications has allowed us to perform semi automated microkeratome-assisted LK. This has a good precision, consistency and smooth lamellar dissection unlike manual lamellar dissection and hence the visual potential. This procedure can be done for conditions that affect the clarity of superficial 250 microns of cornea such as early cases of granular and lattice dystrophy, moderate Keratoconus and post-PRK scarring of cornea. In cases of deeper stromal opacification, excimer laser is applied to recipient bed when there is enough residual corneal tissue for structural integrity. For Deep Anterior Lamellar Keratoplasty surgery, semi automated lamellar anterior corneal dissection of host cornea is performed using a microkeratome. Then donor lamellar graft is cut using the same microkeratome with aid of AC(Anterior Chamber) maintainer to match the stromal bed of host and is sutured in place.

Femtosecond laser-assisted lamellar transplantation:

Femtosecond laser is evolving as a new alternative to microkeratomes. Femtosecond laser is a laser of infrared type that theoretically ablates tissue with lower interference. This property allows for deeper cuts in cloudy and edematous corneas. It is a new option in lamellar corneal transplantation surgery. The two currently available forms of Femtosecond are: the Intralase and the Femtec. The difference between two systems is in the appplanation of cornea during application of laser. The Femtec uses patented interface(concave) that requires less suction and also minimally distorts corneal surface than flat Intralase. Femtosecond laser has greater precision in anterior LK as it produces highly reproducible cut dimensions at graft–host junction and also has vertical (mechanical microkeratome - horizontal) side cut orientation. Since the corneal dimensions of donor and recipient can be customized by the surgeon, there is improved resulting fit that leads to less surgically induced irregular astigmatism and the interface haze. This feature results in better and also faster wound healing. In the presence of post hydrops scar, anterior segment OCT(ocular coherence tomography) is used to assess the depth of scar preoperatively. This is done to fashion the donor lenticule thickness. Both the donor and the recipient corneal lenticules are created with the aid of an artificial Anterior Chamber. This procedure

can be made sutureless if the donor lenticule and the recipient stromal beds are cut at proper orientation and depth (i.e. top hat). It is also reported that Intralase laser keratome provides accurate depth and diameter of corneal discs with smooth bed even at increased depth settings with raster pattern. If needed, stromal bed can be further be smoothed by excimer phototherapeutic keratectomy.

Donor cornea:

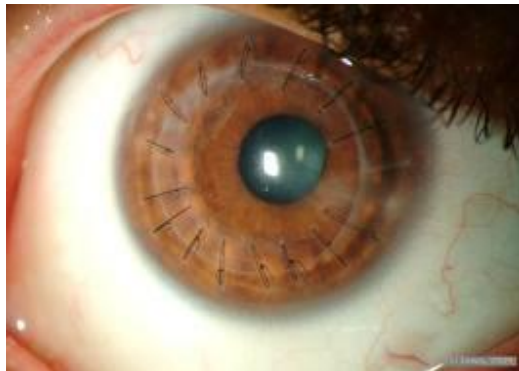
Both preserved and fresh corneal grafts are used in Deep Anterior Lamellar Keratoplasty. Most of the surgeons strip descemets membrane and endothelium because descemets may cause scarring at the interface and wrinkling at the graft host junction and endothelium may present antigens that can lead to immunological rejection of the graft. The diameter of donor corneal button should be of either same size or larger than recipient bed. The graft is sutured to recipient with interrupted sutures, continuous suture or combined interrupted-continuous 10-0 nylon sutures. Selective removal of suture is possible for reduction of surgically induced astigmatism if interrupted sutures have been used. This suture removal can be done even at an earlier period of 3 weeks after surgery. If the sutures are not excessively tight or loose and does not attract blood vessels, the sutures can be left in place for 1 year.

Advantages of DALK:

DALK protects the intraocular structures of the eye during surgery.

Hence there is reduced chance of:

- 1) Postoperative glaucoma
- 2) Formation of cataract
- 3) Retinal detachment
- 4) Cystoid macular edema
- 5) Expulsive choroidal hemorrhage
- 6) Ingrowth of epithelium
- 7) Endothelial rejection
- 8) Low rate of endothelial loss following surgery.



Disadvantages of DALK:

- 1) Requires technical expertise
- 2) Long surgery time

3) Descemet's membrane perforation during dissection. Smaller perforations can be sealed by injecting air or gas mixture of air and C3F8. Tissue glue can also be used. However, larger perforations require conversion to penetrating keratoplasty.

4) Stromal graft rejection.

Penetrating keratoplasty:

A procedure of corneal transplantation where a full thickness recipient corneal tissue is replaced by full thickness donor corneal tissue.

Intraoperative complications:

- 1) Scleral perforation
- 2) Improper trephination
- 3) Retained Descemet's membrane
- 4) Damage to endothelium
- 5) Intraocular hemorrhage
- 6) Vitreous loss

Postoperative complications:

- 1) Wound leak
- 2) Persistent epithelial defect
- 3) Suture related infiltrates and vascularisation
- 4) Formation of anterior synechiae

5) Postkeratoplasty astigmatism

6) Graft infection

7) Rejection of graft

8) Post keratoplasty glaucoma.

REVIEW OF LITERATURE

Xie PY et al, in their study on The evaluation of Visual quality in keratoconus eyes corrected by rigid gas-permeable contact lens analysed 30 eyes of mild keratoconus, 13 eyes of moderate keratoconus and 11 eyes of severe keratoconus and reported that average myopia was 6.92 ± 3.66 Dioptres and average astigmatism was 5.75 ± 1.85 Dioptres. UCVA in 96.3% of keratoconus eyes was ≤ 0.3 . RGP contact lens corrected visual acuity was increased significantly compared with spectacles corrected VA. The visual acuity in 59.3% of eyes corrected by spectacles were ≥ 0.6 , while 100% by RGPCL. The VA in 20.4% of eyes corrected by spectacles were ≥ 1.0 , while 66.7% by RGPCL. Root-mean-square (RMS) of wavefront aberration decreased from (3.63 ± 2.08) microm (without RGP) to (1.39 ± 0.82) microm (with RGP) ($P < 0.01$). RMS of Zernike function in each order was decreased as well, total high order from (2.35 ± 1.57) microm to (1.10 ± 0.62) microm, second order from (2.64 ± 1.74) microm to (0.81 ± 0.66) microm and third order from (1.78 ± 1.28) microm to (0.68 ± 0.39) microm With RGPCL fitting.

Erdurmus M et al, in their study on Contact lens related quality of life in patients with keratoconus analysed 71 patients whose mean age was 42.6 ± 13.1 years. The study included only one eye of each patient, of

which 40 eyes were fitted with rigid gas-permeable lenses, 20 eyes with hybrid lenses and 11 eyes with soft toric lenses. The mean CLIQ (The contact lens impact on quality of life) person measure was found to be 45.5 +/- 8.2 in rigid gas permeable lens group, 45.4 +/- 7.5 in hybrid group and in soft toric group 48.4 +/- 10.5. The study concluded that there is no significant difference among the three groups in self-reported results from the CLIQ questionnaire (P = 0.8).

Gothwal VK et al, in their study on Assessment of the impact of keratoconus on vision-related quality of life analysed 160 keratoconus patients and patients were classified to have mild keratoconus (average Sim K < 45 diopters), moderate keratoconus (average Sim K 45-52 D) and severe keratoconus (average Sim K > 52 D) based on keratometry. The vision-related quality of life was measured using Impact of Vision Impairment Questionnaire. They reported that the mean age affected by keratoconus was 23.3 years with 63% of males being affected, 37% had moderate keratoconus and 63% had severe keratoconus. They also found that there was no significant difference in visual function (mean change -0.16, P = 0.55) and emotional well being scores (mean change -0.32, P = 0.23) between patients who had moderate and severe keratoconus .

Joon Seo Hwang et al, in their study on Effects of Multicurve RGP Contact Lens Use on Topographic Changes in Keratoconus reported that in multicurve RGP lens wearing group corrected logarithm of the minimum angle of resolution visual acuity improved significantly from -0.016 ± 0.065 to -0.032 ± 0.10 at follow-up ($p=0.05$). In advanced keratoconus ,in lens-wearing group, the Sim Kmax significantly decreased from 57.68 ± 4.26 to 55.51 ± 4.28 D, Sim Kmin from 50.50 ± 2.32 D to 49.62 ± 3.26 D, apical power from 62.79 ± 5.11 D to 60.31 ± 5.41 D, astigmatic index from 7.20 ± 0.55 D to 5.90 ± 0.51 D and anterior elevation diopter from 67.36 ± 16.30 μm to 60.61 ± 16.09 μm (paired t -test, $p < 0.05$).

Yamazaki ES et al, in their study on Keratoconus special soft contact lens fitting found that of 80 eyes with keratoconus, the mean age of presentation was 29 years, 51.5% were men and 48.5% women. According to the groups: 15.0% had incipient keratoconus, 53.7% had moderate keratoconus, 26.3% had advanced keratoconus and 5.0% had severe keratoconus. 91.5% of the eyes of patients using contact lenses (91.25%) achieved visual acuity better than 20/40. 88 eyes 58% were fitted with lens with spherical power (mean -5.45 diopters) , 41% with spherocylinder power (-0.5 to -5.00 cylindrical diopters). The most frequent base curve was 7.6 in 61% of the eyes. The main reasons for these soft

contact lens fitting were due to intolerance and inability to achieve good fitting pattern with other lenses.

AIM AND OBJECTIVES

Aim:

To assess the outcomes of use of different types of contact lens used for the management of keratoconus

Primary objective:

To assess the visual improvement with good fitting contact lens.

Secondary objectives:

Contact lens associated problems like intolerance, infection and acute red eye

Progression or inability to fit contact lens

Comfort and quality of life with contact lens

Type of study:

Prospective, non randomized study of patients attending Aravind Eye Hospital, Madurai.

Duration of study:

Jan 2011 – June 2012 and a follow up 12 ± 6 months.

Inclusion criteria:

Newly diagnosed keratoconus patients eligible for contact lens wear in one or both eyes.

Advanced cases needing surgery in other eye are also eligible.

Exclusion criteria:

Post C3R

Post penetrating keratoplasty

Post refractive surgery

Post pseudophakia

Pellucid marginal degeneration

Patients with active vernal keratoconjunctivitis

Patients with corneal opacities other than hydrops

Patients with advanced keratoconus requiring surgery

MATERIALS AND METHODS:

An informed consent was obtained from the patients explaining the nature of study. All the patients in the study underwent retinoscopy, Cycloplegic refraction (when necessary), torch light examination, slit lamp examination, direct ophthalmoscopy and documentation of signs. Contact lens trial was given and suitable contact lens prescribed according to patient's satisfaction. Quality of life scores were assessed at the time of entry into study and patients were asked to come for follow up and repeat assessment at the end of 1 year.

Types of contact lenses prescribed:

Rigid gas permeable lens

Piggy back lens

Hybrid lens

Rose k lens. Rose k lenses were used as the last resort because of high costs.

Quality of life was assessed with a self – administered National Eye Institute –Visual Function Questionnaire .The questionnaire had 25 questions and they were subdivided into general health, mental health, pain, role playing, social activities, dependency, distant visual acuity, near visual acuity and

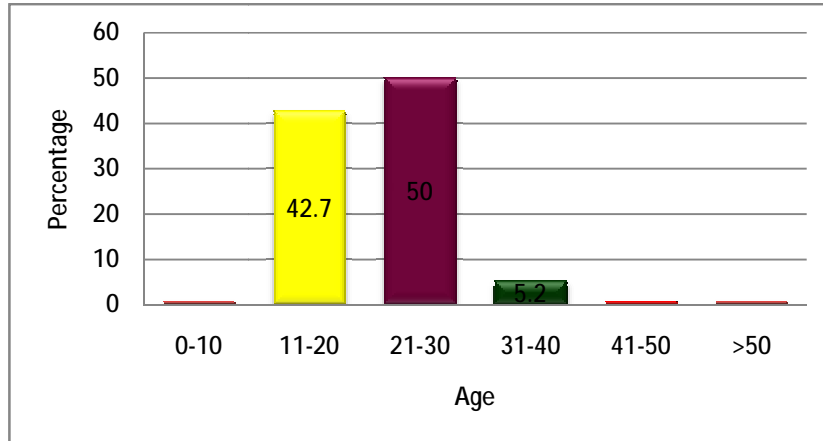
driving. Higher scores represent poorer visual functioning (Rasch analysis)⁹⁹.

OBSERVATIONS AND RESULTS

A prospective study on management and Quality of Life of keratoconus patients with contact lens conducted at cornea clinic and contact lens clinic at Aravind Eye Hospital, Madurai over a period of 18 months from January 2011 to June 2012. A total of 221 eyes of 136 patients were analysed in our study.

Age distribution:

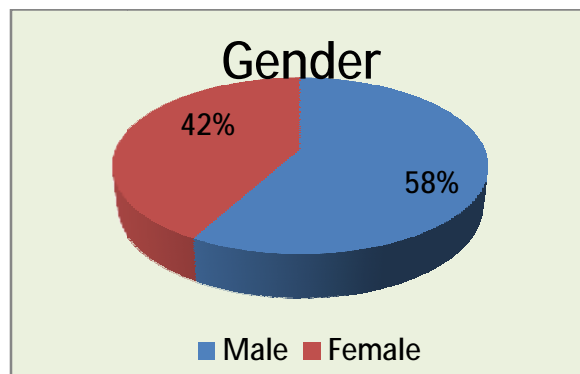
Age	Number of patients	Percentage
0-10	1	0.7%
11-20	58	42.7%
21-40	68	50.0%
31-40	7	5.2%
41-50	1	0.7%
>50	1	0.7%
Total	136	100%



The mean(SD) age of presentation was 22.16(5.83) years. The youngest age of presentation was 10 years and the oldest was 53 years of age.

Gender distribution:

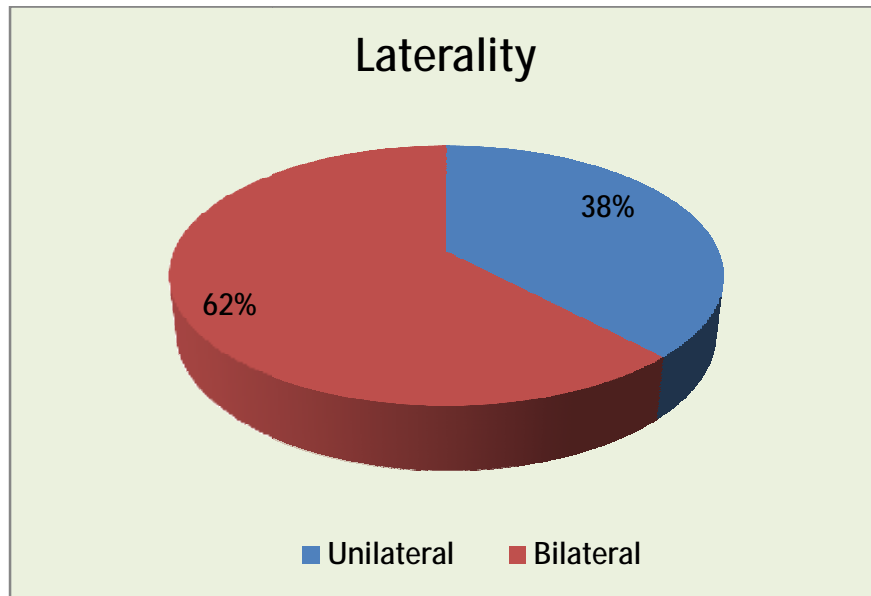
Gender	Number of patients	Percentage
Male	79	58
Female	57	42
Total	136	100



Of the 136 patients, 79(58%)were males and 57(42%)were females.

Laterality:

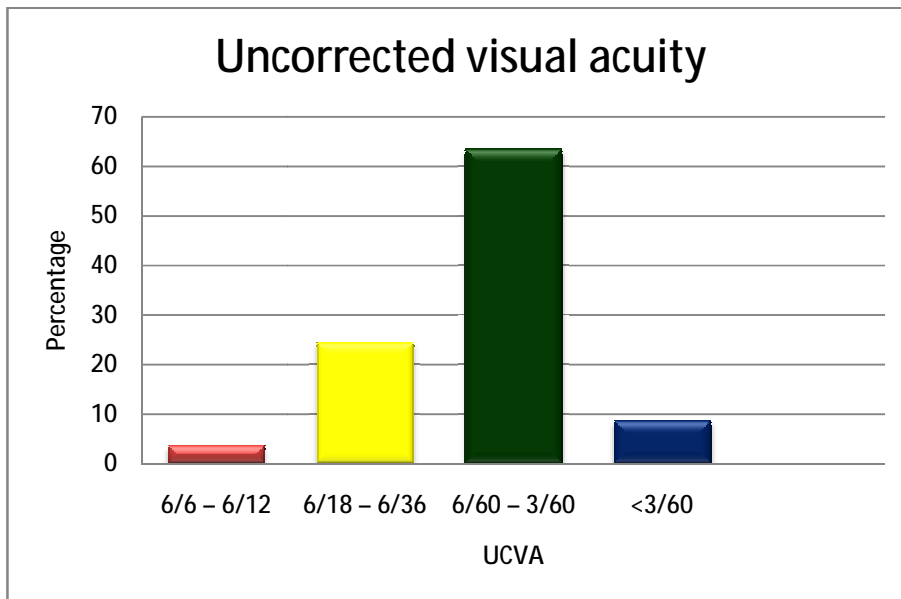
Laterality	Number of patients	Percentage
Unilateral	51	38
Bilateral	85	62
Total	136	100



Keratoconus was found to be unilateral in 51(38%) patients and bilateral in 85(62%)patients based on keratometry and/or orbscan.

UNCORRECTED VISUAL ACUITY:

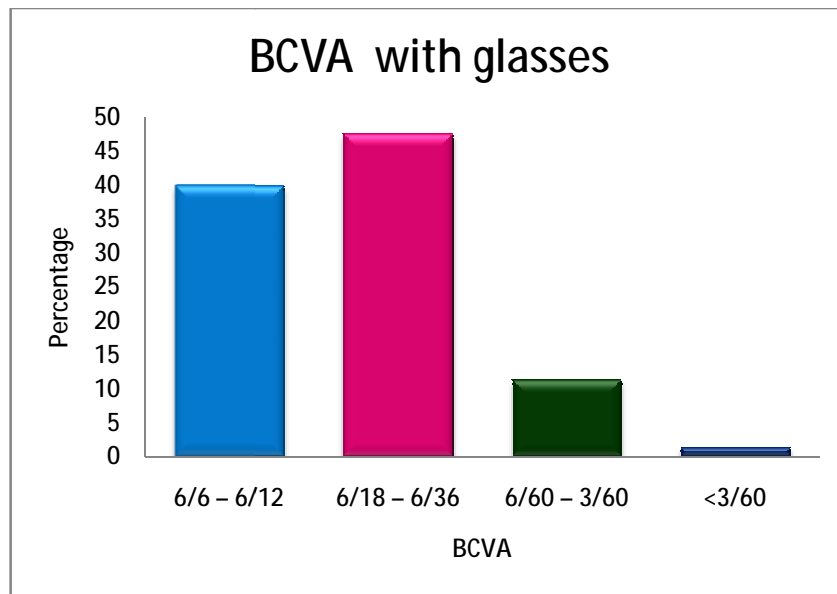
UCVA	Number of eyes	Percentage
6/6 – 6/12	8	3.6
6/18 – 6/36	54	24.4
6/60 – 3/60	140	63.4
<3/60	19	8.6
Total	221	100.0



About 8(3.6%) patients had UCVA of 6/6-6/12, 54(24.4%) patients had UCVA of 6/18-6/36, 140(63.4%) of patients had UCVA of 6/60-3/60, 19(8.6%) patients had visual acuity of less than 3/60 before contact lens fitting.

BEST CORRECTED VISUAL ACUITY WITH GLASSES:

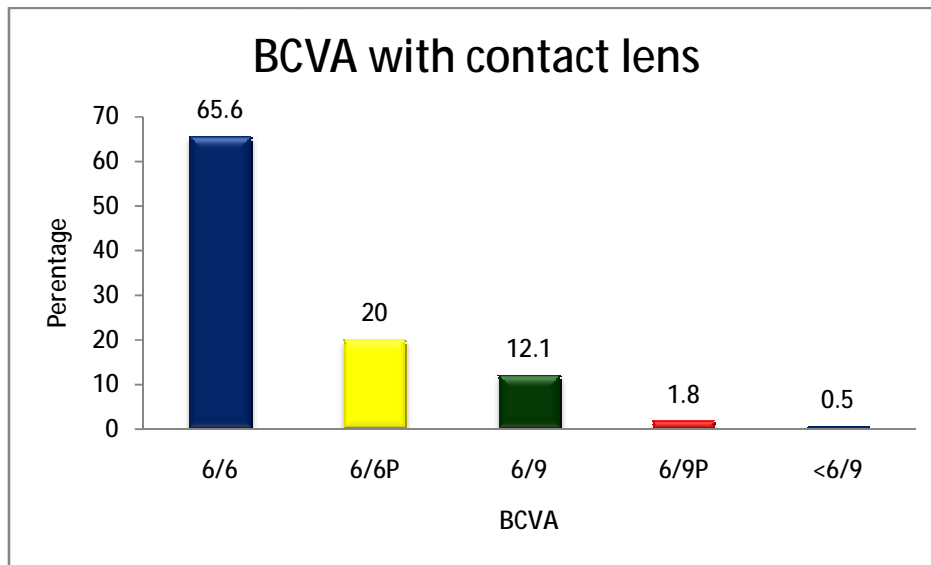
VA with glasses	Number of eyes	Percentage
6/6 – 6/12	88	39.8
6/18 – 6/36	105	47.5
6/60 – 3/60	25	11.3
<3/60	3	1.4
Total	221	100.0



About 88(39.8%) patients had an acuity of 6/6-6/12, 105(47.5%) patients had an acuity of 6/18-6/36, 25(11.3%) patients had an acuity of 6/60-3/60, 3(1.4%) patients had an acuity of less than 3/60 with glasses.

BEST CORRECTED VISUAL ACUITY WITH CONTACT LENS:

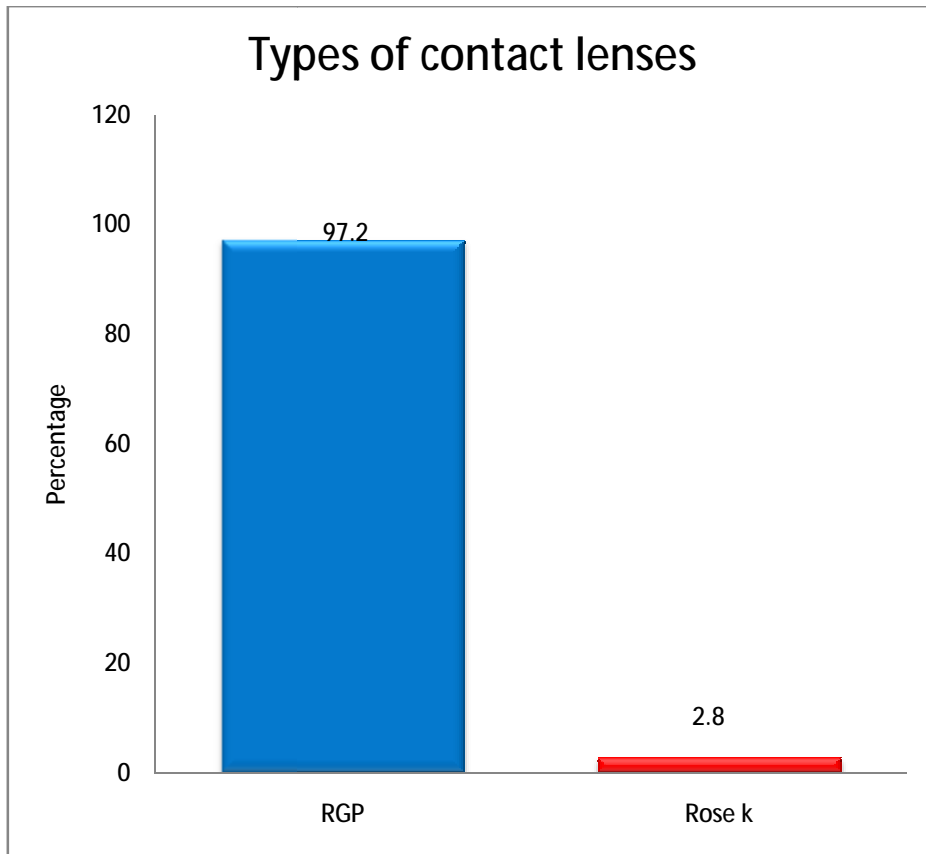
VA with Contact lens	Number of eyes	Percentage
6/6	143	65.0%
6/6P	46	20.5%
6/9	26	11.8%
6/9P	5	2.2%
<6/9	1	0.5%
Total	221	100.0%



Of the 221 eyes studied at the initial visit, 143(65%) eyes improved to 6/6, 46(20.5%) eyes improved to 6/6 partial, 26(11.8%) eyes improved to 6/9, 5(2.2%)eyes improved to 6/9 partial and only 1(0.5%) eye had vision less than 6/9 with contact lens.

TYPES OF CONTACT LENSES:

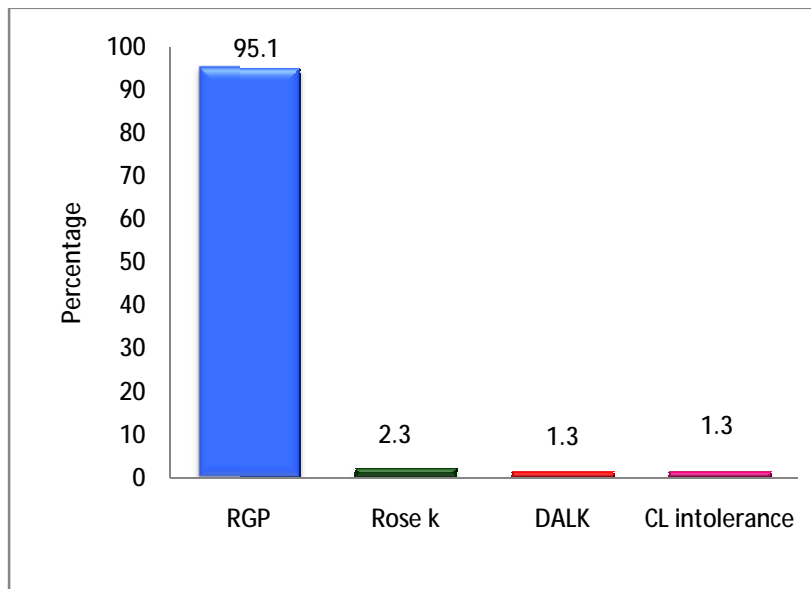
Lens	Number of eyes	Percentage
RGP	215	97.2
Rose K	6	2.8
Total	221	100



At the initial visit, 215(97.2%) eyes were fitted with RGP lens and 6(2.8%)eyes were fitted with Rose K lens.

AT THE END OF 1 YEAR:

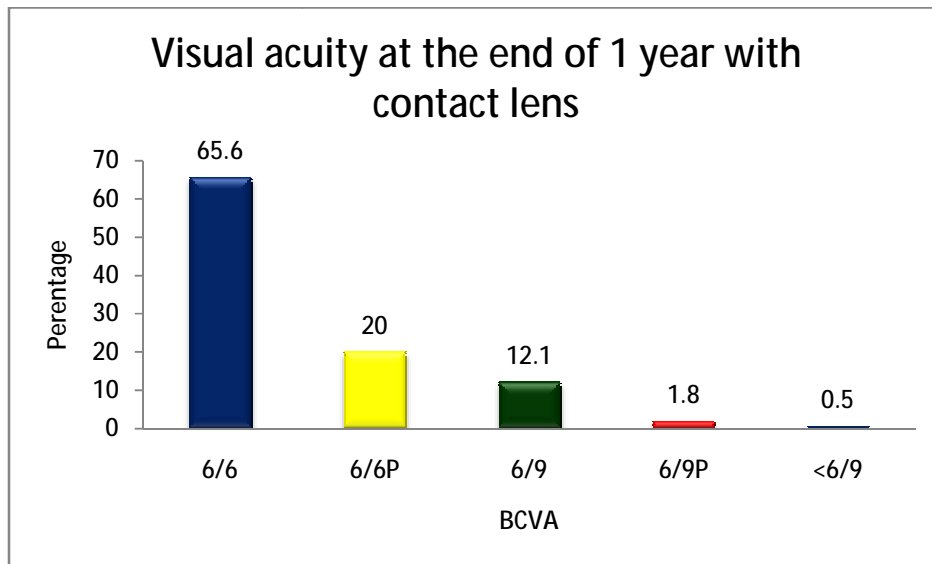
At the end of 1 yr	No. of eyes	Percentage
RGP	210	95.1
Rose K	5	2.3
DALK	3	1.3
CL intolerance	3	1.3
Total	221	100



At the end of 1 year, 210(95%)eyes continued to use RGP lens and 5(2.3%)eyes continued to use Rose K lens, 3(1.3%)eyes underwent Deep Anterior Lamellar Keratoplasty and 3(1.3%)eyes discontinued contact lens due to intolerance to the same.

**VISUAL ACUITY WITH CONTACT LENS AT THE
END OF 1 YEAR:**

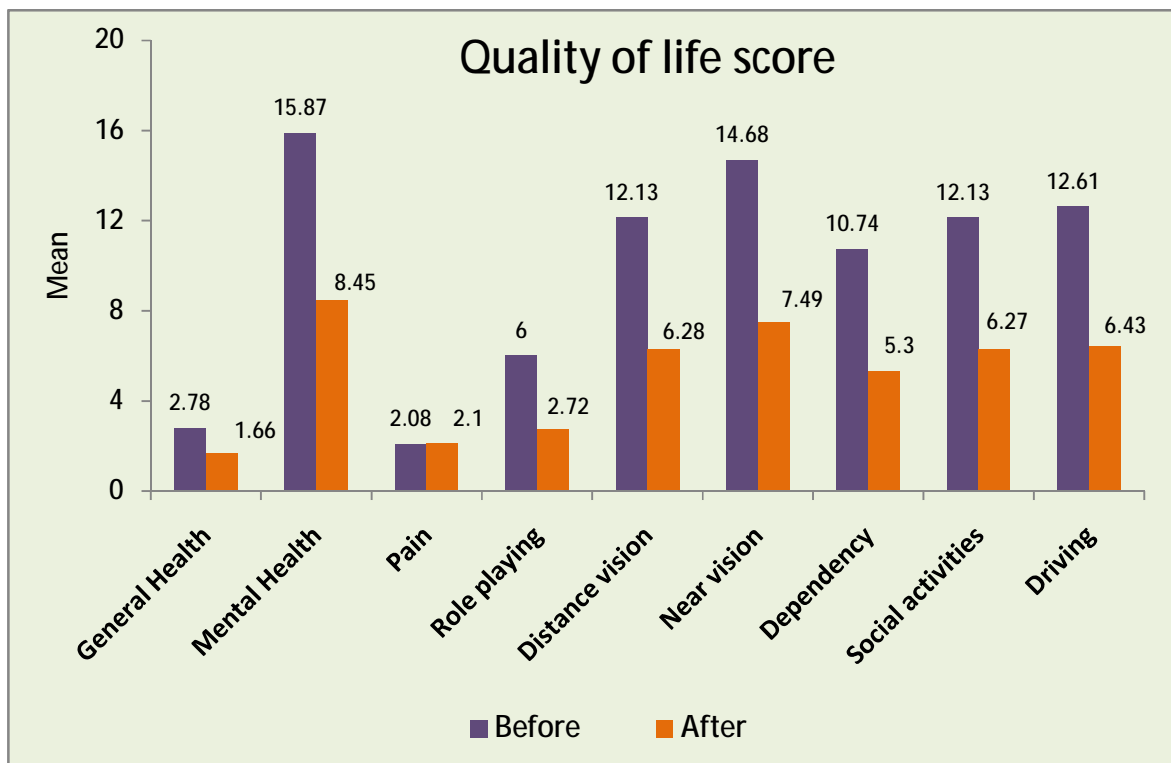
VA with contact lens	Number of eyes	Percentage
6/6	141	65.6%
6/6p	43	20.0%
6/9	26	12.1%
6/9p	4	1.8%
<6/9	1	0.5%
Total	215	100%



At the end of 1 year, 141(65.6%) eyes improved to 6/6, 43(20.0%) eyes improved to 6/6 partial, 26(12.1%) eyes improved to 6/9, 5(1.8%) eyes had visual acuity of 6/9 partial and 1(0.5%) eye had vision less than 6/9.

QUALITY OF LIFE SCORES:

The scores show a higher value before contact lens use and lower values after contact lens use for 1 year (Higher scores indicate poorer visual functioning as shown in Rasch analysis)(99). This decrease in scores shows their in decrease in difficulty in day to day activities in distant vision, near vision, role playing, dependency, social functions, general health and mental health except for pain.



Questionnaire	Quality of life score					P-value
	n	Before		After		
		Mean(SD)	Min – Max	Mean(SD)	Min - Max	
General Health	134	2.78(0.58)	2 – 5	1.66(0.55)	1 – 4	<0.001
Mental Health	134	15.87(2.60)	10 – 22	8.45(2.13)	3 – 14	<0.001
Pain	134	2.08(0.46)	2 – 6	2.10(0.49)	2 – 6	0.64
Role playing	134	6.00(1.28)	4 – 9	2.72(0.92)	2 – 6	<0.001
Distance vision	134	12.13(1.75)	8 – 16	6.28(1.44)	3 – 11	<0.001
Near vision	134	14.68(2.13)	10 – 22	7.49(1.59)	4 – 12	<0.001
Dependency	134	10.74(1.78)	4 – 15	5.30(1.39)	3 – 8	<0.001
Social activities	134	12.13(1.56)	7 – 16	6.27(1.27)	3 – 9	<0.001
Driving	122	12.61(1.78)	9 – 16	6.43(1.57)	4 – 11	<0.001
Total	134	87.91(7.86)	59 – 109	46.13(6.98)	25 – 64	<0.001

Paired t test

DISCUSSION

Keratoconus is a progressive non-inflammatory disorder of cornea that usually manifests during adolescence. Kennedy et al, in their clinical and epidemiologic study of keratoconus reported keratoconus prevalence rate of 54.5 per 1,00,000 in the general population. Prevalence of keratoconus as reported by Rabinowitz is between 50 and 230 per 1,00,000 in the population⁶. The incidence and prevalence could not be commented in our paper because the study was conducted only among the patients attending our hospital. The disease has an insidious onset and a variable severity and patients may not seek ophthalmic care for a long time. As the disease is asymmetric, one eye may be affected more than the other. Some patients are able to perform their routine activities with the vision in the better eye. This possibly explains the reason for loss of follow up of some patients at 1 year who were initially eligible for the study. Rabinowitz YS, in his study on Keratoconus has reported that keratoconus usually presents at puberty and remains progressive till the third and sometimes fourth decades of life. Sainai et al, in their study of keratoconus among Asian patients reported that about 50% of patients presented before 20 years of age¹⁰³ whereas Wagner et al, in their Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study reported only 4% presented before the age of 20

years¹⁰⁴. Tufts et al in a study of 2723 keratoconus patients reported mean age at diagnosis of 22.4 years for males and 23.3 years for females. Crews *et al*, in a retrospective study on keratoconus reported the mean age of patients with disease presenting to hospital as 28 years. Our study showed that the mean age of presentation was 22.16 ± 5.83 years (range 10 to 53 years). The reported gender predilection may vary for keratoconus. Thomas CI et al in his study on The cornea and Nuel JP et al in his study on keratoconus reported that females slightly outnumbered males. Sainai et al, study on keratoconus among Asian eyes in India, reported female preponderance of the disease. Rabinowitz in a review of keratoconus in the year 1998 reported that there is no sex predilection in keratoconus. Kennedy et al in his study on clinical and epidemiologic study of keratoconus, a 48-years clinical trial of keratoconus found annual incidence rate to be more among males. However, this difference was not statistically significant. A study conducted by N Lim at the Western eye Hospital reported a increased incidence of disease in the males. Our study showed a male preponderance, male:female ratio of 8:6. Keratoconus is essentially a bilateral disease of cornea, although presentation may be grossly asymmetric. It may take a few years after the initial diagnosis of keratoconus in one eye for the condition to become apparent in his/her fellow eye. The term “forme fruste keratoconus” is used

for less affected fellow eyes that display no clinical findings but certain changes in corneal topography. A descriptive study on keratoconus conducted by N Lim at Jordan eye hospital reported a bilateral incidence of 92%. The reported incidence of unilaterality of keratoconus varies from 14.4%–41% .Our study showed that 68% of patients had bilateral disease and 32% had unilateral disease. Keratoconus, a degenerative disorder of cornea leading to thinning and ectasia of cornea, causes characteristic irregular light reflexes on distant direct ophthalmoscopy and streak retinoscopy and irregular mires on keratometry. The patient develops a irregular myopic astigmatism that is gradually progressive, due to which patients complain of blurred vision and reduced visual acuity. Xie PY et al, in a study of visual acuity in keratoconus reported average myopia and average astigmatism as (6.92 +/- 3.66) Dioptres and (5.75 +/- 1.85)Dioptres respectively. They reported the uncorrected visual acuity in 96.3% of keratoconic eyes to be very low.The visual acuity in 59.3% of eyes corrected by spectacles were ≥ 0.6 . The VA in 20.4% of eyes corrected by spectacles were ≥ 1.0 . Cesneková T et al in a study on visual functions and quality of life in keratoconus reported the uncorrected visual acuity was 0.20 (+/- 0.18). Our study showed that about 96% of patients had an uncorrected visual acuity of less than 6/18, which would be very disabling and only 3.6%

of patients had an acuity of 6/12 and more. When glasses were prescribed, 39.8% of patients improved to 6/12 and more, 47.5% had visual acuity of 6/18- 6/36, 11.3% had visual acuity of 6/60-3/60, 1.4% had visual acuity of less than 3/60. High corneal irregular astigmatism induced by corneal distortion is the most frequent cause of increased aberration and reduced ocular optical quality in keratoconus. Xie PY et al reported that proper Rigid Gas Permeable Contact Lens fitting provided an effective method of reshaping of ocular surface combined with tear film and RGP contact lens could reduce corneal aberrations and corneal astigmatism leading to improved optical quality and finally increased visual acuity and contrast sensitivity. Cesneková T et al in their study on visual functions and quality of life in keratoconus concluded that the use of RGP correction can improve visual acuity significantly, however did not lead to significant improvement in corneal aberrations and contrast sensitivity. Jinabhai A et al in their study on visual performance and optical quality with soft lenses in keratoconus patients concluded that the RGP lenses provided superior visual performance and caused more reduction of 3rd-order aberrations when compared to toric SCL. He also found out that the visual performance with the toric SCL was comparable to that measured with spectacles. However, toric Soft Contact Lenses were successful in reducing only

uncorrected higher-order aberrations significantly with the exception of spherical aberration. Betts AM et al, in their study on Visual performance and comfort with the Rose K lens for keratoconus reported that there were no statistically significant changes in high- or low-contrast visual acuity with the Rose K lenses. They also added that there was statistically significant improvement in self-reported assessment of vision and comfort in the eyes with more advanced keratoconus after using Rose K lenses. Our study showed that 97% of patients improved to 6/9 and more with contact lens wear. In this study, Rigid Gas Permeable lenses provided optimal fit in 97.2% of contact lens users and Rose k lenses provided optimal fit in 2.8% of patients. Rose K lenses were tried when patients did not report improvement in visual acuity with the other lenses used in the study and their use is often limited by high costs. Keratoconus is one of the common indication for Penetrating Keratoplasty. Dana *et al* in their study confirmed the following three main reasons for Penetrating Keratoplasty the primary cause being unsatisfactory visual acuity of under 20/40 (43%), followed by intolerance to contact lens (32%) and frequent lens displacement (13%). Weed *et al* in their study on keratoconus over years reported that the main reason was contact lens discomfort (67.5%), followed by falling out of contact lenses (19%) and poor visual acuity with contact lens (13.5%).

Betts AM et al in their study on Rose K lens for keratoconus reported that 72% of patients preferred Rose K lenses over habitual lenses and 87% reported that they were willing to continue on Rose K lenses. In our study, over a follow up of 1 year 97.4% continued to use contact lenses, 1.3% underwent Deep anterior lamellar keratoplasty, and the rest 1.3% discontinued contact lenses due to intolerance to the same. Since keratoconus disease affects adults during their active years, the disease may have impact on their financial and social health, hence quality of life.

Kymes SM et al in their study on Quality of life in keratoconus found that binocular visual acuity worse than 6/12 was associated with lower quality of life scores on all scales except General Health and Ocular Pain and a steep keratometric reading was associated with lower scores on the Mental Health, Role Difficulty, Driving, Dependency, and Ocular Pain scales. Quality of life scores revealed betterment of life in day to day activities after contact lens use. Jones-Jordan LA et al in their study on Asymmetry in keratoconus and vision-related quality of life concluded that increase in ocular asymmetry and decrease in Visual acuity and steepening of cornea in the better eye were associated with decreasing vision-related quality of life, although the magnitude of the changes were not meaningful clinically. However, eye with better had greater effect on the vision-related quality of

life. Quality of life scores done using NEI-VFQ 25 in our study showed that patient had a better quality of life with regards to general health, mental health, dependency, role playing, driving, social activities, distant vision, near vision($p < 0.001$) except for ocular pain.

CONCLUSION

In this study we concluded that that keratoconus commonly presents during adolescence, mean age being 22.16(SD 5.83) years. Male preponderance of the disease was found, 58% of males were affected and 42% of females were affected. Bilateral presentation of the disease was found to be more common (based on keratometry and/or orbscan). 62% of patients had bilateral involvement and 38% of patients had unilateral involvement. Uncorrected visual acuity was less than 6/36 in 28% of eyes of patients, 63.4% of eyes of patients had UCVA between 6/60 and 3/60, 8.6% of eyes of patients had UCVA less than 3/60. When glasses were prescribed, 39.8% of eyes of patients improved to 6/12 and more, 47.5% of eyes had visual acuity of 6/18-6/36, 11.3% of eyes of patients had visual acuity of 6/60-3/60 and 1.4% of eyes of patients had visual acuity less than 3/60. On using contact lens, 97.3% of eyes of patients improved to 6/9 and more. 97.2% of eyes of patients were successfully fitted with Rigid Gas Permeable lens and 2.8% of eyes of patients were fitted with Rose k lens. At the end of 1 year, 97.7% of eyes of patients improved to 6/9 and more with contact lens and 97.4% of eyes of patients continued using contact lens, 1.3% of eyes of patients underwent Deep anterior lamellar keratoplasty due to inability to fit

contact lens, 1.3% of eyes of patients discontinued contact lens due to intolerance.

No case of infection or contact lens associated acute red eye due to contact lens wear was reported in our study. The quality of life scores showed marked improvement after contact lens wear except for pain and hence betterment of life. Contact lenses continues to play a predominant role in the management of keratoconus successfully and a regular follow up is necessary to monitor the progression of disease.

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PROFORMA

1.Name

2.MRNumber

3.Date

4.Age

5.Gender

1.Male

2.Femele

6.Laterlity

1.RE

2.LE

3.BE

7.Torch Light

Munson

1..RE

2.LE

3.BE

Rizutti

1.RE

2.LE

3.BE

8.Slit lamp examination:

Position of the cone

1.Central

2.Eccentric

Prominent corneal nerves

0.No

1.Yes

Vogt striae

0.No

1.Yes

Stromal thinning 0.No 1.Yes

Fleischers ring 0.No 1.Yes

Hydrops 0.No 1.Yes

10.Cycloplegic refraction:	Sphere	Cylinder	
Axis			
RE	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
LE	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

11.Uncorrected visual acuity
RE..... LE.....

12.Best corrected visual acuity with glasses
RE..... LE.....

13. Best corrected visual acuity with contact lens
RE..... LE.....

14. Type of contact lens suggested

- 0.No contact lens
- 1.RGP lens
- 2.Piggy back lens
- 3.Hybrid lens

4. Rose K lens

15. Contact lens

Base curve.....

Power.....

Diameter.....

17. Quality of life scores

General health.....

Mental health.....

Pain.....

Distant vision.....

Near vision.....

Role playing.....

Social activities.....

Dependency.....

Driving.....

FOLLOW UP AT 1 YEAR

1. Date.....

2. Uncorrected visual acuity

RE.....

LE.....

3. Best corrected visual acuity with contact lens

RE.....

LE.....

4. Outcomes

1. Able to continue contact lens
2. Contact lens intolerance
3. Infection due to contact lens
4. Frequent fall of contact lens
5. DALK
6. PKP

5. Quality of life scores

- General health.....
- Mental health.....
- Pain.....
- Distant vision.....
- Near vision.....
- Role playing.....
- Social activities.....
- Dependency.....
- Driving.....

Quality of life scores were assessed using National Eye Institute Visual Function Questionnaire 25.

- General health.....Question 1
- Mental health.....Question 2,3,21,22,25
- Pain.....Question 4,19
- Distant vision.....Question 8,9,10

Near vision.....Question 5,6,7,12

Role playing..... Question 17,18

Social activities..... Question 11,13,14

Dependency..... Question 20,23,24

Driving..... Question 15,16

National Eye Institute Visual Function Questionnaire 25.

1. In general, would you say your overall health is:

Excellent 1

Very Good..... 2

Good..... 3

Fair..... 4

Poor..... 5

2. At the present time, would you say your eyesight using both eyes (with glasses or contact lenses, if you wear them) is excellent, good, fair, poor, or very poor or are you completely blind?

Excellent 1

Good..... 2

Fair..... 3

Poor..... 4

Very Poor 5

Completely Blind..... 6

- 3 - *version 2000*

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3. How much of the time do you worry about your eyesight?

None of the time..... 1

A little of the time..... 2

Some of the time..... 3

Most of the time 4

All of the time? 5

4. How much pain or discomfort have you had in and around your eyes (for example, burning, itching, or aching)? Would you say it is:

None 1

Mild..... 2

Moderate 3

Severe, or..... 4

Very severe?..... 5

5. How much difficulty do you have reading ordinary print in newspapers?

Would you say you have:

No difficulty at all..... 1

A little difficulty..... 2

Moderate difficulty.....	3
Extreme difficulty.....	4
Stopped doing this because of your eyesight	5
Stopped doing this for other reasons or not interested in doing this	6

- 4 - *version 2000*

© R 1996

6. How much difficulty do you have doing work or hobbies that require you to see well up close, such as cooking, sewing, fixing things around the house, or using hand tools? Would you say:

No difficulty at all.....	1
A little difficulty.....	2
Moderate difficulty.....	3
Extreme difficulty.....	4
Stopped doing this because of your eyesight	5
Stopped doing this for other reasons or not interested in doing this	6

7. Because of your eyesight, how much difficulty do you have finding something on a crowded shelf?

No difficulty at all.....	1
---------------------------	---

- A little difficulty..... 2
- Moderate difficulty..... 3
- Extreme difficulty..... 4
- Stopped doing this because of your eyesight 5
- Stopped doing this for other reasons or not
interested in doing this 6

8. How much difficulty do you have reading street signs or the names of stores?

- No difficulty at all..... 1
- A little difficulty..... 2
- Moderate difficulty..... 3
- Extreme difficulty..... 4
- Stopped doing this because of your eyesight 5
- Stopped doing this for other reasons or not
interested in doing this 6

- 5 - version 2000

© R 1996

9. Because of your eyesight, how much difficulty do you have going down steps, stairs, or curbs in dim light or at night?

- No difficulty at all..... 1

- A little difficulty..... 2
- Moderate difficulty..... 3
- Extreme difficulty..... 4
- Stopped doing this because of your eyesight 5
- Stopped doing this for other reasons or not
interested in doing this 6

10. Because of your eyesight, how much difficulty do you have noticing objects off to the side while you are walking along?

- No difficulty at all..... 1
- A little difficulty..... 2
- Moderate difficulty..... 3
- Extreme difficulty..... 4
- Stopped doing this because of your eyesight 5
- Stopped doing this for other reasons or not
interested in doing this 6

11. Because of your eyesight, how much difficulty do you have seeing how people react to things you say?

- No difficulty at all..... 1
- A little difficulty..... 2
- Moderate difficulty..... 3

Extreme difficulty.....	4
Stopped doing this because of your eyesight	5
Stopped doing this for other reasons or not interested in doing this	6

- 6 - *version 2000*

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12. Because of your eyesight, how much difficulty do you have picking out and matching your own clothes?

No difficulty at all.....	1
A little difficulty.....	2
Moderate difficulty.....	3
Extreme difficulty.....	4
Stopped doing this because of your eyesight	5
Stopped doing this for other reasons or not interested in doing this	6

13. Because of your eyesight, how much difficulty do you have visiting with people in their homes, at parties, or in restaurants ?

No difficulty at all.....	1
A little difficulty.....	2
Moderate difficulty.....	3

Extreme difficulty..... 4
Stopped doing this because of your eyesight 5
Stopped doing this for other reasons or not
interested in doing this 6

14. Because of your eyesight, how much difficulty do you have going out to see movies, plays, or sports events?

No difficulty at all..... 1
A little difficulty..... 2
Moderate difficulty..... 3
Extreme difficulty..... 4
Stopped doing this because of your eyesight 5
Stopped doing this for other reasons or not
interested in doing this 6

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15. Now, I'd like to ask about driving a car. Are you currently driving, at least once in a while?

Yes 1 *Skip To Q 15c*

No..... 2

15a. IF NO, ASK: Have you never driven a car or have you given up driving?

Never drove 1 *Skip To Part 3, Q 17*

Gave up..... 2

15b. IF GAVE UP DRIVING: Was that mainly because of your eyesight, mainly for some other reason, or because of both your eyesight and other reasons?

Mainly eyesight 1 *Skip To Part 3, Q 17*

Mainly other reasons 2 *Skip To Part 3, Q 17*

Both eyesight and other reasons ... 3 *Skip To Part 3, Q 17*

15c. IF CURRENTLY DRIVING: How much difficulty do you have driving during the daytime in familiar places? Would you say you have:

No difficulty at all 1

A little difficulty 2

Moderate difficulty 3

Extreme difficulty 4

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16. How much difficulty do you have driving at night? Would you say you have:

No difficulty at all..... 1

A little difficulty..... 2

Moderate difficulty..... 3

Extreme difficulty..... 4

Have you stopped doing this because
of your eyesight 5

Have you stopped doing this for other
reasons or are you not interested in
doing this 6

16a. How much difficulty do you have driving in difficult conditions, such
as in bad weather, during rush hour, on the freeway, or in city traffic? Would
you say you have:

No difficulty at all..... 1

A little difficulty..... 2

Moderate difficulty..... 3

Extreme difficulty..... 4

Have you stopped doing this because
of your eyesight 5

Have you stopped doing this for other
reasons or are you not interested in

doing this 6

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17. Do you accomplish less than you would like because of your vision?

None of the time.....1

A little of the time.....2

Some of the time.....3

Most of the time.....4

All of the time.....5

18. Are you limited in how long you can work or do other activities because of your vision?

None of the time.....1

A little of the time.....2

Some of the time.....3

Most of the time.....4

All of the time.....5

19. How much does pain or discomfort in or around your eyes, for example, burning, itching, or aching, keep you from doing what you'd like to be doing? Would you say:

None of the time.....1

- A little of the time.....2
- Some of the time.....3
- Most of the time.....4
- All of the time.....5

- 10 - *version 2000*

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20. I stay home most of the time because of my eyesight

- Definitely False.....1
- Mostly False2
- Not Sure3
- Mostly True4
- Definitely True5

21. I feel frustrated a lot of the time because of my eyesight.

- Definitely False.....1
- Mostly False2
- Not Sure3
- Mostly True4
- Definitely True5

22. I have much less control over what I do, because of my eyesight.

- Definitely False.....1
- Mostly False2
- Not Sure3
- Mostly True4
- Definitely True5

23. Because of my eyesight, I have to rely too much on what other people tell me.

- Definitely False.....1
- Mostly False2
- Not Sure3
- Mostly True4
- Definitely True5

24. I need a lot of help from others because of my eyesight.

- Definitely False.....1
- Mostly False2
- Not Sure3
- Mostly True4
- Definitely True5

25. I worry about doing things that will embarrass myself or others, because of my eyesight.

Definitely False.....1

Mostly False2

Not Sure3

Mostly True4

Definitely True5

ABBREVIATIONS

UCVA	-	Uncorrected Visual Acuity
BCVA	-	Best Corrected Visual Acuity
CL	-	Contact Lens
PG	-	Present Glass
RGP	-	Rigid Gas Permeable Lens
SCL	-	Soft Contact Lens
PBCL	-	Piggy Back Contact Lens
QOL	-	Quality Of Life
DALK	-	Deep Anterior Lamellar Keratoplasty
PKP	-	Penetrating Keratoplasty

The Tamil Nadu Dr. M.G.R. Medical College - July 2016-2017

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INTRODUCTION

Keratoconus, a condition of obscure etiology has been known from as early as 17th century and was first described by Nottingham. Most of associations of keratoconus are incidental probably, though multitude of predisposing causes exist as well. Recent advances in videokeratography gives us opportunity to evaluate interesting details in the disease. The advent of specialised contact lenses and revolutions in surgeries like keratoplasty has made a break through in the management of keratoconus.

Keratoconus is a degenerative disorder of the eye in which structural changes within the cornea cause it to thin and change to a more conical shape than its normal gradual curve. The word keratoconus is derived from

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123	3469688	16	F	5/60	6/24	6.60	-2.0	9.30	6/9	6/6	4/60	6/18P	6.90	-2.25	9.30	6/6P	6/6	1	1	1	Irregular mires	irregular mires	70@70	60.4@160	3	2	16	8	2	2	4	2	12	6	14	5	10	4	13	7	13	6
124	3471160	15	M	2/60	2/60	7.20	-6.0	11.0	6/9	6/6	2/60	6/36	7.50	-4.0	11.0	6/9	6/6	1	1	1	54.7@92	52.9@2	47@15	41@105	3	2	15	9	2	2	6	2	12	5	12	5	12	6	11	6	14	8
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126	3461264	26	M	2/60	6/18	7.05	-2.0	9.30	6/6P	6/6	6/6	NIL GLASS						1	0	1	66.9@42	55.7@132	54.4@88	48.3@178	3	2	14	7	2	2	5	2	16	7	14	7	11	5	16	7	16	9
127	3473698	16	M	6/6	NIL GLASS					6/6	2/60	NIG	7.0	-4.0	11.0	6/6P	6/6	0	1	1	45.1@127	44.6@37	>70	>70	2	1	14	8	2	2	7	2	10	5	15	8	15	7	14	7	12	6
128	3463653	26	F	6/18	6/6	7.60	-1.0	9.10	6/6	6/6	6/36P	6/9	7.60	-2.50	9.20	6/6	6/6P	1	1	1	60.01@33	54.17@123	46.55@57	42.84@147	2	2	15	8	4	2	6	2	14	7	18	10	9	5	13	8	14	9
129	3478844	41	F	6/6	NIL GLASS					6/6	6/60	6/12	7.50	-2.0	9.10	6/6	6/6	0	1	1	44.25@30	43.75@120	45.50@75	42.50@165	2	1	17	7	2	2	7	2	13	9	14	7	14	7	15	6	11	5
130	3478243	20	F	6/60	6/9	7.35	-2.50	9.10	6/6	6/6	6/36P	6/9	7.40	-2.0	9.10	6/6	6/6	1	1	1	49.35@52	44.55@142	54.50@35	50.3@125	3	2	16	7	2	2	6	2	10	6	14	8	12	6	12	7	11	5
131	3484367	15	F	6/24	6/18	6.90	-5.0	9.20	6/6	6/6	6/9	6/6P	0	-0.75	0	6/6	6/6	1	1	1	50.4@55	44.50@145	55.75@55	53.75@145	2	1	18	9	2	2	8	3	12	5	14	7	13	7	15	8	13	6
132	3482976	18	M	6/18	6/9P	7.60	-0.50	9.20	6/6	6/6	6/6	NIL GLASS						1	0	1	47.9@112	44.9@22	45.3@132	45.0@42	3	2	17	6	2	2	7	4	13	4	16	9	13	7	11	6	10	5
133	3253406	31	F	6/36	6/18	7.40	-1.0	9.20	6/6	6/6	6/36	6/18	7.35	-3.0	9.20	6/6	6/6	1	1	1	56.25@120	52.50@30	60.50@60	55.50@150	2	1	13	7	2	2	5	3	14	8	17	9	12	7	10	6	12	7
134	3481530	24	M	5/60	6/18	7.55	-4.0	9.0	6/6	6/6	6/24	6/6	7.85	-1.0	9.20	6/6	6/6	1	1	1	53.1@127	47.7@37	43.4@30	41.6@120	2	2	15	9	2	2	6	2	15	5	15	7	10	5	12	6	12	6
135	3311317	12	M	6/6	NIL GLASS					6/6	2/60	6/60	6.50	-4.50	9.30	6/24	6/6	0	1	1	60.05@24	55.05@114	59.7@75	53.4@165	2	1	14	6	2	2	6	3	12	5	13	7	7	3	11	5	14	6
136	3311352	21	M	6/60	6/18	7.10	-3.0	9.30	6/6	6/6	2/60	6/36P	7.0	-4.75	9.30	6/6P	6/6	1	1	1	54.2@106	47.1@16	67.3@87	57.2@177	3	2	12	6	2	2	5	2	12	7	16	10	10	5	9	4	13	7