Recent Advancements in Corneal Ectatic Conditions and their Management: A Mini Review

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Abstract

**Background:** Recent advancements in the corneal diagnostics have helped us a lot in effective management of ectatic conditions of the cornea. This article gives a brief overview about the developments in management and various ways of vision rehabilitation for the condition of corneal ectasia.

**Methods:** After reviewing several studies in the field of ophthalmology and optometry, we tried to understand the various techniques and management plans for improving vision in the ectatic corneal conditions effectively and to update them in this article.

**Conclusion:** During recent years, there are various combination strategies which can be combined to give a productive outcome to these patients who are suffering with ectasia disorders. Procedures like C3R or DALK in conjunction with contact lens can help us achieve better visual outcomes. In severe conditions, it is judicious to check the visual outcome with a soleral lens before planning for a corneal transplant which can avoid the waiting time for the donor cornea and other complications of the surgery beforehand.

**Keywords:** Ectasia; Penetrating keratoplasty; Corneal collagen cross linking; Riboflavin; Keratoconus; Keratoglobus; Pellucid marginal degeneration; Post LASIK ectasia; corneal rehabilitation; Kerasoft IC

Introduction

In the recent years, technology has developed in many folds so is the diagnostic, rehabilitation and surgical aspects of eye care system. Cornea being avascular, transparent structure covering the one third anterior surface of the eye is one of the important vision contributing structure not only for accounting the major refracting power, but also by acting as a primary interface for the light that is entering our eyes.

This article mainly discusses about the recent advancement in the management of corneal ectatic conditions and vision rehabilitation with the help of contact lenses and some surgical interventions. In normal conditions, Cornea being a strong layer of the eyeball has the strength and ability to withstand the pressure of intraocular contents excreting on it, however, there are few ectatic conditions which will distort the shape of the cornea leading to unclear image to fall on the retina making a patient to complain about his unstable and unclear vision even after wearing a standard spectacle correction.

Types of ectasia

Conditions of ectasia are primary and secondary in origin. Primary conditions like keratoconus, keratoglobus, pellucid marginal degeneration and terrien’s marginal degeneration, which have shown a few characteristics like progressive thinning (stroma), bulging and irregular astigmatisms [1]. Secondary reasons are acquired in nature due to corneal diseases or reshaping procedures like refractive surgery procedures are noted.

Etiology and stages of ectasia

In the initial stages, these ectatic disorders are undiagnosed and are usually managed by a common spectacle correction. Decreased quality of vision achieved by the patient spectacles, frequent change in the spectacle prescriptions and intermittent contrast or glare problems are a few common complaints seen in the initial stages of ectasia [2].

With the increasing severity of ectasia gradually patient will have a clinically significant decrease in best corrected, pin hole and uncorrected visual acuities. To investigate the cause of the vision loss, the diagnostic tests like keratometry, Placido’s disc, pachymetry, slit scanning topography, scheimpflug tomography, optical coherence tomography, corneal biomechanics will help us reveal the underlying cause and nature of the ectatic disorder. Based on the parameters like simulated Keratometry reading, Anterior Steeper curvature, Inferior-superior value and other parameter analysis will reveal the category and type of corneal disorder. Predisposing factors like connective tissue disorders, vernal kerato-conjunctivitis, chronic marginal blepharitis and other observations like frequent rubbing of the eyes are to be noted to rule out the cause of corneal ectasia [3].

Keratoconus being the most common condition is usually bilateral in nature with an age of onset in Puberty, which usually leaves a scar in the later stages. Pellucid marginal degeneration and Terrien’s marginal degeneration are rare with the onset of age around 20 to 40 years and are usually bilateral in nature with a thinning of the inferior and superior corneal areas respectively [1]. Keratoglobus is a rare condition which shows bilateral thinning and globular protrusion in the cornea with increased fragility due to the extreme thinning with unclear genetic origin. Based on the origin, keratoglobus has primarily been a congenital disorder in nature with no genetic or inherited patterns defined. Recent studies reveal an acquired type of keratoglobus too exists [3-6].

After ultrastructure observations of corneal ectatic tissue, a significant decrease in the collagen lamellae in the ectatic region and accumulation of lamellae in the surrounded non-ectatic regions were noted, indicating a change in the normal anatomical structure of the cornea [7,8].

**Keywords:** keratoconus; keratoglobus; corneal ectasia; corneal biomechanics; Placido’s disc; pachymetry; slit scanning topography; scheimpflug tomography; optical coherence tomography, corneal biomechanics; keratoconus; keratoglobus; corneal ectasia; corneal biomechanics; Placido’s disc; pachymetry; slit scanning topography; scheimpflug tomography; optical coherence tomography.
Advancements in management of ectasia

With newer imaging and topography techniques, more accurate and early diagnosis is possible leading to a significant decrease in the chances of post refractive ectasia. Studies show a reduced rate of complications post lasik and, ectasia is a very rare vision deteriorating complication when not diagnosed or detected in time [9].

During histopathology and ultrastructure examination of corneal ectatic zone reveal few common signs like epithelial hypoplasia with rare hyperplastic foci, breaks in Bowman’s membrane, reduction in number of stromal lamellae and thin residual stromal bed and thin collagen fibrils are a few correlated signs in explaining the pathophysiology of ectatic disorders [10].

Use of femto second laser for a most predictive flap removal and in place LASIK are more common strategies which surgeons are adopting for preserving the residual stromal bed which help them in reducing post LASIK ectatic complications and predictable visual outcomes. However, in rare cases, post LASIK corneal ectatic complications can occur. These ectatic complications are observed as early as a weekday after surgery can be seen. But, majority of these ectasia signs are identified as surgical complications within the first and second year after surgery [11].

The pathophysiology of the post lasik corneal ectasia is due to incomplete or splits seen in the interweaving of stromal lamellar fibrils layer which are a result of fracture between stromal lamellae of anterior stroma. Similar biomechanical catastrophe is observed even in the cases of primary ectatic conditions like keratoconus which goes in to a chronic phase making the condition worsen with time because of this phenomenon called interfibrillar slippage which is thought to be the reason for observing Vogt’s striae more easily visible during the slit lamp examination for most of the ectatic corneas [12-14].

Correction of ectasia with contact lenses

A decreased visual performance with ophthalmic spectacle lenses if noted or when spectacle lenses doesn’t give a satisfactory visual outcome, there is a need for a trial of soft toric contact lenses to enhance the visual acuity of the eyes till the astigmatism is regular and is within the available prescription range. If it exceeds the range, a rigid gas permeable lens with a tri-curve design can be given as a trial. If vision, lens stability and comfort are still not achieved with a generalized RGP lens design, a specialty contact lens with a variable design can be used. Rose K, Rose K2 and Rose K2 NC comes with variable designs that can be fitted to the patients with a better comfort and vision than a traditional RGP lens.

Piggy back is another option to try, but due to the structural complexity, thickness and the complex maintenance, storage issues lead it to be an uncommon option. Hybrid lenses with a combination of silicone hydrogel and an RGP can be a mode of correction which are commercially available too (KeraSoft IC) which can allow to correct an astigmatism up to 12 D in a quarterly disposable modality that can ensure comfort, health and hygiene of the user [15].

In recent times the usage of corneal-scleral contact lenses like Rose K2 XL lenses and scleral lenses have been increased with some promising results in the category of enhancing vision if the previous smaller diameter designs are not giving sufficient vision to the patient, if the cost factor is not a barrier [16-18].

Cornea collage crosslinking with riboflavin

If the cornea is found to have a significant thinning or rapid changes in the topography, collagen cross linking with riboflavin is the first mode of treatment which is usually advised to improve the biomechanical strength and to delay the corneal changes. This is a 30-minute-long procedure which involves the exposure to ultra violet rays of 370 nm on the debrided central corneal epithelium of 6 to 7 mm diameter of riboflavin (vit B2) exposed epithelium to generate covalent bonds between stromal collagen fibrils. Recent advancements have made this procedure safer to endothelium by replacing dextran with HPMC and to increase comfort by adopting trans-epithelial cross linking by suing Benzaconium Chloride (BAK) or Ethylendiaminetetraacetic acid (EDTA) as a component making the Epithelium-On procedure possible [19-22].

Enhancing the riboflavin penetration by using a mild electricity, which can help the riboflavin penetrate deeper layers into the stroma which is referred as iontophoresis, in a lesser duration is explored with epi-on it will be more comfortable with [23].

Observations like using Pulsed oxygen during the treatment improved efficacy of postoperative outcome. The basic requirement to qualify for C3R was a minimum thickness of 400 microns which was tweaked by using a hypotonic saline to swell the corneal thickness of a person by which most of the patients who were diagnosed with a later stage of ectatic disorders can be benefitted with this novel treatment. The chances of reduction in the time duration of the procedure were also explored by using high intensity UV lamps increasing the UV exposure level to a safer level like 7 mw/cm² for 15 minutes and 18 mw/cm² for 5 minutes which are safer without affecting the endothelial cell density and limbal stem cell function [24].

Even after undergoing C3R procedure, for retaining the corneal shape most of the times, an RGP or Rose K series designs, or Kerasoft IC based on the severity of the condition can be indicated. The use of these speciality design lenses makes the wearer comfortable and gives a stable vision.

Intra-stromal corneal rings

A surgical intervention which was primarily invented for myopia control is later adopted for controlling the astigmatism because of ectasia and to improve rigidity of the cornea by implanting stromal rings which have varying sizes and thickness with single or dual segments. Most successful implants are done in patients with 450 microns corneal thickness and smaller mesopic pupil size. Patients who want to delay their corneal ectasia, who have contact lens intolerance and who wants to postpone penetrating keratoplasty procedure can undergo this procedure. These rings can effectively control the asymmetry in cornea and can able to give better unaided and best corrected vision.

Based on the nomograms followed in this procedure, smaller optical zone of 5 mm and a zone of 7 mm are implanted in the stromal layer of the peripheral cornea. Two segmented rings are usually inserted in each eye. Asymmetric rings can be used in which thicker segments are implanted in the steeper parts of the cornea and thinner segment in the other portion. Ring segments which are Allike are inserted in cases where the cone is centrally located, which can reduce the steepness of the cornea helping the uncorrected and best corrected vision to be improved. In mild ectatic cases, this procedure can help the patients regain their vision with spectacles itself. Recently few studies have concluded that a single segment implant can able to correct astigmatism and double segments can reduce the spherical ametropias effectively [25].

A study shows that a reduction of mean corneal power by 3.7 diopters...
and 3.3 diopters in first and second year after corneal rings implanted eyes. A combination of implanted stromal rings and C3R gives unchanged in the results of progression or regression even after 5 years of surgery which ascertain the stability in ectasia. Recently, new ways to make a tunnel for placing intra stromal rings were being explored using femto second technology, which makes the procedure easier and promising outcomes instead of manual tunnelling which can lead to unexpected complications, if the post-operative infections are controlled effectively [26].

After this procedure RGP or Soft contact lenses cannot be fitted to these corneas due to tunneling in the mid periphery and to correct the residual refractive error, only a spectacle lenses can be used.

**Corneal transplantation**

Patients who are not suitable for C3R or the Intrastromal rings due to lack of stromal thickness are advised for a corneal transplant procedure. Surgical procedures like penetrating keratoplasty, deep anterior lamellar keratoplasty, microkeratome assisted lamellar keratoplasty are now days more successful as the endothelium replacement is not necessary unlike in penetrating keratoplasty [27].

Various methods of separating Descemet’s membrane are available for a successful replacement with the donor tissue, however, recent studies show that there is a decent visual prognosis by following big bubble technique. Recent techniques of using the microkeratome which is useful as a substitute for the manual procedures and is expected to improve surgical precision and reproducibility in the end outcomes [28].

New research shows a combination of the microkeratome, automated lamellar therapeutic keratoplasty device to create a button from donor cornea, which can result in a perfect match of thickness to donor and receiver needs which can give you a better visual outcome is possible [29].

Scleral lenses, a promising and an emerging solution for the severe cases which allows the wearer to achieve good vision for a limited period if the patient is trained and educated about the proper care and maintenance in using them [30,31].

In the end, we need to keep in mind about the patient expectations, possible complications and tentative outcomes balanced with the patient conditions can help him significantly from the ectatic disorders.

**Conclusion**

Newer diagnostic tools can diagnose the ectatic conditions early and accurately, helping us for a prophylactic C3R which is an effective way of controlling the ectasia and helps us to achieve better vision with the help of contact lenses in the moderate stages. In severe ectatic conditions, a scleral lens trial can be given to see the improvement in vision. In cases where scleral lenses cannot improve the vision much, deep anterior lamellar keratoplasty can be performed and a contact lens to correct irregular astigmatism is indicated to achieve a better visual outcome.

**References**