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A REVIW ON ECTATIC CORNEAL DISEASE- KERATOCONUS

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Abstract:

Normally, the cornea has a dome shape, like a ball. Sometimes the structure of the cornea is just not strong enough to hold this round shape and the cornea bulges outward like a cone. This condition is called keratoconus.[1,2] This abnormal shape prevents the light entering the eye from being focused correctly on the retina and causes distortion of vision. This is occur in one or both eyes and often begins during a person's teens or early 20s. Treatment for keratoconus depends on the severity of your condition and how quickly the condition is progressing. Mild to moderate keratoconus can be treated with eye glasses or contact lenses. In some people the cornea becomes scarred or wearing contact lenses becomes difficult.[2,4] In these cases, surgery might be necessary. Keratoconus also is associated with over exposure to ultraviolet rays from the sun, excessive eye rubbing, a history of poorly fitted contact lenses and chronic eye irritation [5,6]

Keywords: *keratoconus, Corneal topography, Corneal cross-linking, Hybrid contact lenses Corneal ring implants, keratoplasty.*

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INTRODUCTION:

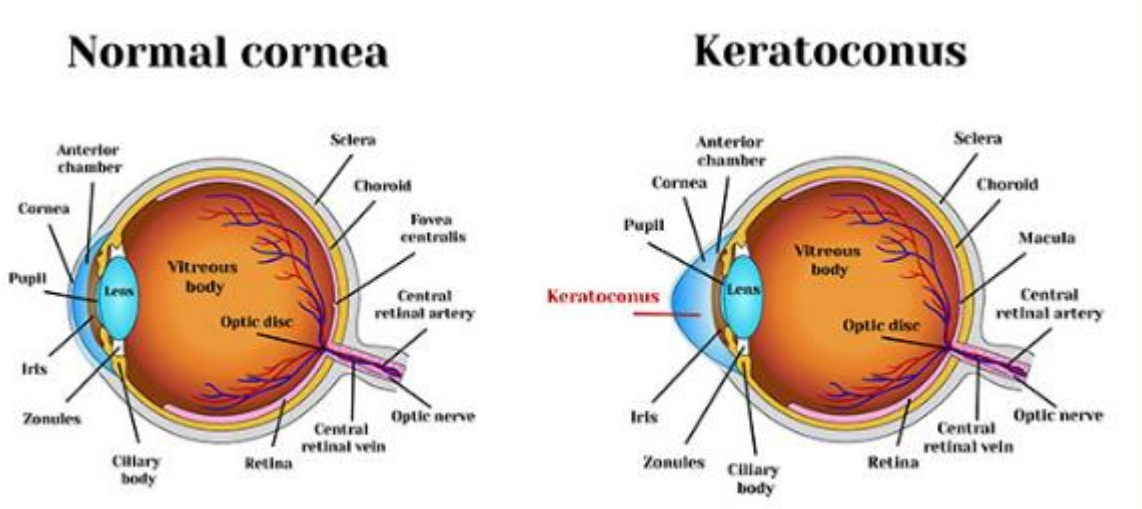
Keratoconus is a bilateral but typically asymmetric, non-inflammatory corneal ectasia characterized by progressive corneal thinning, conical protrusion, scarring, and decreased biomechanical strength of the cornea [1-6]. The result is distorted and subnormal vision [7,8]. As keratoconus progresses, the cornea bulges more and vision may become more distorted. In a small number of cases, the cornea will swell and cause a sudden and significant decrease in vision. The swelling occurs when the strain of the cornea's protruding cone-like shape causes a tiny crack to develop. The swelling may last for weeks or months as the crack heals and is gradually replaced by scar tissue. If this sudden swelling does occur, your doctor can prescribe eye drops for temporary relief, but there are no medicines that can prevent the disorder from progressing [9,10]]. Which is occur in one or both eyes and each eye may be affected differently. Eye glasses or soft contact lenses may be used to correct the mild nearsightedness and astigmatism that is caused by the early stages for keratoconus. In a few cases, a corneal transplant is necessary. However, even after a corneal transplant, eyeglasses or contact lenses are often still needed to correct vision [11,12].

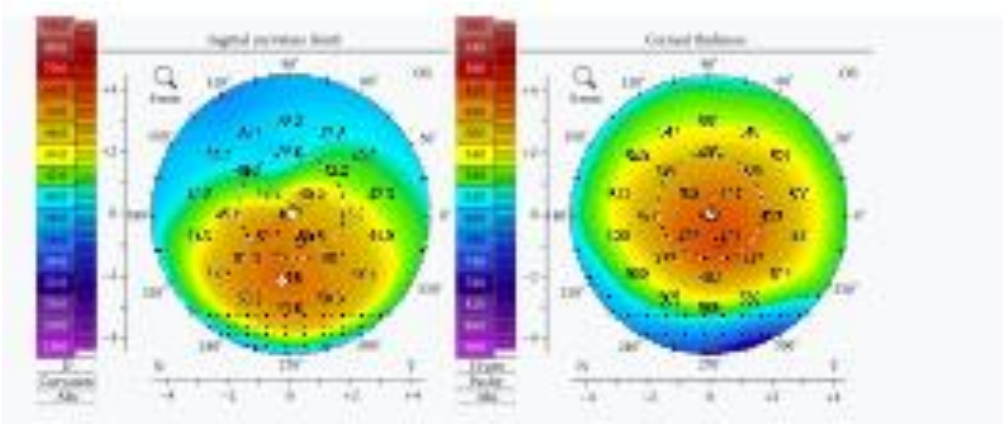
CAUSES

The exact cause of keratoconus is unknown. Both genetic and environmental factors may play a role in the development of keratoconus.. The genetic factors involve abnormalities in the structure of collagen, which result in a weak and flexible cornea [13,14]. Keratoconus is more common in people with Down syndrome, Marfan syndrome, and Leber congenital amaurosis, and certain genetic conditions. In these cases, the cause depends on the specific condition. Environmental factors may include living in sunny, hot areas of the world, while eye-rubbing is a major behavioral factor in the disease. Malfunctioning enzymes that normally help maintain the health of the cornea may play a role.[15,16] All of these factors contribute to the main problem in keratoconus, which is the defective collagen structure that results in thinning and irregularity of the cornea. Keratoconus occurs more frequently in patients with atopy (asthma and eczema) or severe ocular allergies. It may also be linked to hormonal factors because it is more frequent during puberty and also may progress during pregnancy. And long term rigid contact lens wear also a risk factor [17,18].

PATHOGENESIS

Once initiated, the disease normally develops by progressive dissolution of Bowman's layer which lies between the corneal epithelium and stroma. As the two come into contact, cellular and structural changes in the cornea adversely affect its integrity and lead to the bulging and scarring characteristic of the disorder.[19,20]keratoconic corneas show signs of increased activity by proteases, a class of enzyme sthat break some of the collagen cross-linkages in the stroma, with a simultaneous reduced expression of protease inhibitors. Other studies have suggested that reduced activity by the enzyme aldehyde dehydrogenase may be responsible for a build-up of free radicals and oxidising species in the cornea. Whatever the pathogenetical process, the damage caused by activity within the cornea likely results in a reduction in its thickness and biomechanical strength [21,22].





DIAGNOSING KERATOCONUS

Your ophthalmologist may use a number of tools to make a diagnosis of keratoconus, including:

A slit lamp, which combines an intense light source with a microscope to examine your eyes

A **keratometer** to measure the curvature of the cornea

Corneal topography, it creates a 3D model of the cornea to detect any subtle changes. Which an automated instrument projects an illuminated pattern onto the cornea and determines its shape from analysis of a digital image. The topographical map reveals distortions or scarring in the cornea, with keratoconus revealed by a characteristic steepness of curvature which is usually below or around the centre of the cornea. The topography record of the degree and extent of the deformation is used for assessing its rate of progression. Unilateral cases tend to be uncommon. Sometimes it's a mild condition in the better eye, below the limit of clinical detection. It is common for keratoconus to be diagnosed first in one eye and not until later in the other. At our hospital, we use a very sensitive pentacam occulyser which records data from 25,000 points on the cornea [23-25].

Different stages corneal topography:

Stage 1: Eccentric steepening Induced myopia and/or astigmatism of ≤ 5.0 D K-reading ≤ 48.00D

Vogt's lines, typical topography

Stage 2: Induced myopia and/or astigmatism between 5.00 and 8.00 D K-reading ≤ 53.00 D, Pachymetry ≥ 400 µm

Stage 3: Induced myopia and/or astigmatism between 8.01 and 10.00 D K-reading > 53.00 D, Pachymetry 200 to 400 µm

Stage 4: Refraction not measurable K-reading > 55.00 D Central scars, Pachymetry ≤ 200 µm.

Corneal topography of stage II keratoconus [26,27]

TREATMENT

Keratoconus is a known progressive disorder. All modalities of treatment whether it is glasses, contact lenses, all correct the refractive error secondary to the cone formation; they do not treat the keratoconus. Reason tells us that anything which is progressive first needs to be stopped and for that there is only one treatment that is available which is C3R or CXL. Corneal collagen crosslinking (C3R/ CXL) is a well-accepted and welcome addition to treatment options for keratoconus. Before C3R was approved, treatment options to rehabilitate vision were quite limited, mostly to contact lenses and when they failed one needed to undergo a penetrating keratoplasty. [28]

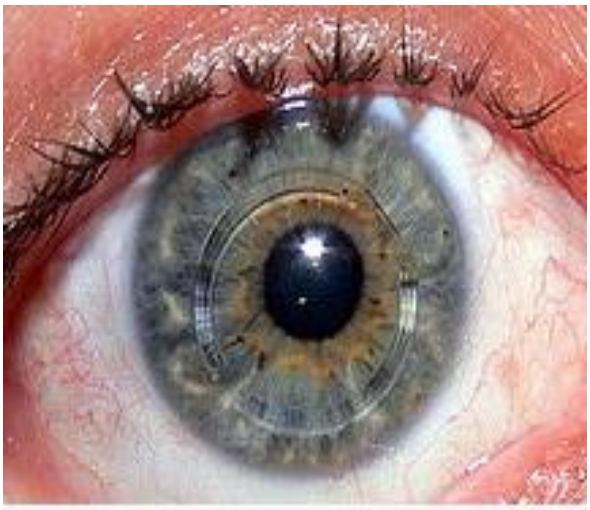
Some of the treatments recommend for karatoconus

Contact lenses

Rigid (hard) or hybrid contact lenses may be prescribed and are particularly effective in treating keratoconus for a period of time. These are made from a special material that allows the contact lens to mask the abnormal shape of the cornea and improve vision. However, contact lenses do not stop the condition from progressing and will eventually become ineffective.

Corneal ring segments

This is a surgical option involving the insertion of clear plastic segments into the cornea. These segments are designed to reshape the front surface of the eyeCorneal ring segments are reserved for advanced cases of corneal ectasia, where the patient's vision is not correctable with glasses or contact lenses.[29]



A pair of Intacs after insertion into the cornea

A small incision is made in the periphery of the cornea and two thin arcs of polymethyl methacrylate are slid between the layers of the stroma on either side of the pupil before the incision is closed by a suture.[58] The segments push out against the curvature of the cornea, flattening the peak of the cone and returning it to a more natural shape. The procedure offers the benefit of being reversible and even potentially exchangeable as it involves no removal of eye tissue, thus correcting refractive errors caused by keratoconus

Corneal collagen cross-linking

This treatment can effectively stop the progress of keratoconus. It involves a combination of collagen and riboflavin (vitamin B2), which are activated by ultraviolet light to significantly strengthen the rigidity of the cornea.

First, the top layer of the cornea (epithelium) is gently removed. The cornea is then saturated with collagen and riboflavin, and UV light applied to activate the solution. This causes the collagen strands to bond across the cornea and strengthen it. Early treatment with collagen cross-linking can slow or sometimes even stop progression of keratoconus.[30] The procedure takes approximately one hour and is performed as an outpatient procedure in the clinic. Patients usually experience some mild discomfort in the immediate post-operative period.

Following treatment, the patient is fitted with a contact lens that stays in place for up to three days. Antibiotic drops are applied to the treated eye until the surface of the eye has healed. This is followed by steroid drops for approximately 5 to 6 weeks.

Corneal transplantation (keratoplasty)

This will only be suggested if all other treatments options have been exhausted. About 10–20% of patients eventually require corneal transplantation.

There are two types of corneal transplants – partial-thickness and full-thickness (or penetrating). It is usually the latter that is recommended for patients with keratoconus [31].

A corneal transplant is a complex procedure and requires admission to a day surgery. It is generally performed under local anaesthetic, with the option of a sedative. During the procedure, your surgeon will cut out the abnormal section of cornea and replace it with donor cornea, which will be stitched into place. The stitches will be removed at a later date. Your own corneal cells will gradually grow and fuse to the donor tissue. A full recovery can take up to a year.

RELATED DISORDERS

Several other corneal ectatic disorders also cause thinning of the cornea:[30]

- Keratoglobus is a very rare condition that causes corneal thinning primarily at the margins, resulting in a spherical, slightly enlarged eye. It may be genetically related to keratoconus.
- Pellucid marginal degeneration causes thinning of a narrow (1–2 mm) band of the cornea, usually along the inferior corneal margin. It causes irregular astigmatism that, in the early stages of the disease can be corrected by spectacles. Differential diagnosis may be made by slit-lamp examination.
- Posterior keratoconus, a distinct disorder despite its similar name, is a rare abnormality, usually congenital, which causes a non progressive thinning of the inner surface of the cornea, while the curvature of the anterior surface remains normal. Usually only a single eye is affected.
- post lasik ectasia is a complication of lasik eye surgery.

CONCLUSION:

It is therefore difficult to say exactly how much the treatment helped. None of the studies reported the risk of eye sight getting worse but, on average, treated eyes had better vision about 10 letters better as compared to untreated eyes. Corneal collagen cross-linking (CXL) seems to be accepted worldwide as a breakthrough treatment in the management of keratoconus Higher-quality studies are needed before an appropriate metaanalysis can be conducted to

confirm the importance of this treatment

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