THE EMERGING ROLE OF EXCIMER LASER SURGERY IN KERATOCONUS

An essay

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Abstract

Patients with keratoconus are typically frustrated with their visual limitations that negatively impact their lifestyle. These patients often have exhausted their options with glasses and contact lenses, leading them to investigate surgical options.

In addition, these patients are typically otherwise healthy young adults at the prime of their professional and personal lives, and often have expectations similar to those searching for elective vision correction.

Previously, there was an old concept that keratoconus and formefruste keratoconus are contraindications for excimer laser surgery but with the development of corneal collagen cross linking a new concept appeared “why not to use excimer laser for correction of the refractive error after stabilization of the ectatic condition? “And so we can provide an enhanced lifestyle for these patients while maintaining a high level of safety.

Key Words:

Diagnosis of Keratoconus, Corneal Collagen Cross Linking, Formefruste Keratoconus, Excimer Laser Surgery and Keratoconus

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Introduction

The term Keratoconus, which was first described in detail in 1854, is derived from the Greek words *Kerato* (cornea) and *Konos* (cone).

It is one of the most common primary ecstasias .

Keratoconus is a bilateral, asymmetric corneal degeneration which is characterized by localized corneal thinning which leads to protrusion of the thinned cornea. Normally, this corneal thinning occurs in the inferior-temporal as well as the central cornea .However, superiorly located corneal thinning has also been described.

The corneal protrusion usually follows the corneal thinning, leads to high myopia and irregular astigmatism thus affecting the quality of vision. Keratoconus usually becomes clinically apparent during the second decade of the life, mostly during puberty, although earlier onset and later onset have also been described, and it typically progresses until the fourth decade of life, when it usually stabilizes**. (Rabinowitz, 1998)**

The age of incidence of keratoconus has been estimated to be between 5 and 23, and the prevalence in the general population to be 5.4 out of 10000 subjects. **(Krachmer et al., 1984)**

It would not also be surprising to expect an increase in the incidence and prevalence rates of this disease over the next few years with the current wide spread use of corneal topography leading to improved diagnosis.

A recent study has determined that 50% of non-affected eyes of subjects with unilateral keratoconus would develop the disease within 16 years**. (Rabinowitz et al., 2003)** Both genders and all ethnicities are likely to be affected by keratoconus. **(Romero-Jiménez et al., 2010)**

The pathogenesis of keratoconus is unknown and it is most likely multifactorial.Several studies have reported a strong association between eye rubbing and the development of keratoconus. Also contact lens wear is another form of corneal micro trauma associated with keratoconus. **(Weed et al., 2008)**

The genetic basis of keratoconus has been studied through linkage mapping and mutation analysis to reveal its molecular basis and pathogenesis. Mapping studies have identified a number of loci for autosomal-dominant inherited keratoconus: 20p11-q11, 16q22.3-q23.1, 3p14-q13, 2p24, 15q22.32-24 and 5q14.3-q21.1. Other potential loci have also been reported indicating genetic heterogeneity. **(Espandar and Meyer**,**2010)**

The hereditary pattern is not predictable although the strongest evidence of genetic involvement is a high concordance rate in monozygotic twins.

(Edwards et al., 2001) A positive family history has been reported in 6-8% of the cases and its prevalence in first-degree relatives is 15-67-times higher than that in the general population**. (Wang et al., 2000)**In addition, unaffected first-degree relatives have a higher rate of abnormal topography compared with the general population**. (Rabinowitz et al., 1990)**

Keratoconus may be associated with many ocular conditions as retinitis pigmentosa, infantile retinal degeneration (Leber's congenital or infantile amaurosis), blue sclera, microcornea, aniridia, corneal degeneration, congenital cataract, ectopia lentis, lenticonus, macular coloboma, retinal dysplasia and floppy eyelid syndrome.

The presence of keratoconus (KC) has been confirmed clinically and histologically in corneas with lattice-granular (Avellino) dystrophy, with

posterior polymorphous dystrophy, with coincident features of posterior polymorphous dystrophy and essential iris atrophy, and with Fuch's endothelial dystrophy. **(Elder MJ., 1994)**

Keratoconus commonly develops as an isolated condition, although it has also been described in association with many syndromes and diseases such as Down's syndrome, Ehlers-Danlos syndrome, osteogenesis imperfecta, mitral valve prolapse, and other connective tissue diseases. **(Romero-Jiménez et al., 2010)**

Diagnosis of Keratoconus

The ocular symptoms and signs of keratoconus vary depending on disease severity.

At incipient stages, also referred to as subclinical or forme fruste, keratoconus does not normally produce any symptoms and thus can pass unnoticed by the patient and practitioner unless specific tests (i.e. corneal topography) are undertaken for diagnosis .**(Arntz et al.,2003)**

Presentation is usually with impaired vision in one eye caused by progressive astigmatism and myopia. The patient may report the need for frequent change of his spectacles or decreased tolerance to his contact lenses. **(Kanski, 2007)**

Disease progression is manifested by a significant loss of visual acuity which cannot be compensated for with spectacles or contact lenses. Therefore, eye care practitioners should be suspicious about the presence of keratoconus when a visual acuity of 6/6 is difficult to achieve with increasing against-the-rule astigmatism.

Near visual acuity is generally found to be better than expected from the refraction, distance visual acuity and age of the patient. **(Rabinowitz, 1998)**

External signs:

Munson's signs

Rizzuti phenomenon

Slit-lamp findings:

Stromal thinning

Posterior stress lines (Vogt's striae)

Iron ring (Fleischer ring)
Scarring: epithelial or subepithelial
Signs detected by retroillumination:
Scissoring on retinoscopy
Oil droplet sign* ("Charleaux")
Signs detected by photokeratoscopy:
Compression of mires inferotemporally (egg-shaped mires)
Compression of mires inferiorly or centrally
Videokeratography signs:
Localized increased surface power
Inferior-superior dioptric asymmetry (I-S)
Relative skewing of the steepest radial axes (SRAX) above and below the horizontal meridian
Videokeratography indices:
Keratometric (K) value greater than 47.2
I-S value greater than 1.4
KISA% (K= K-reading, I-S = Inferior – Superior Steeping, A= Astigmatism) greater than 100

Table1: Signs of KC

The appearance of “scissor” shadows while performing retinoscopy suggests the development of irregular astigmatism.

The Charleux oil drop that is observed by backlighting the mydriatic pupil also poses a warning sign. (Fig. 1)

(Rabinowitz, 1998)

In moderate and advance cases of keratoconus, a hemosiderin arc or circle line, commonly known as Fleischer's ring, is frequently seen around the cone base. This line has been suggested to be an accumulation of iron deposits from the tear film onto the corneal epithelium as a result of severe corneal curvature changes induced by the disease and/or due to modification of the normal epithelial slide process.

(Romero-Jiménez et al., 2010)

Another characteristic sign is the presence of Vogt's striae (Fig.2), which are fine vertical lines produced by compression of the Descemet's membrane, which tend to disappear when physical pressure is exerted on the cornea digitally**.(Li et al.,2004)** or by gas permeable contact lens wear**.(Davis et al.,1993)**

The increased visibility of corneal nerves (Fig.3) and the observation of superficial and deep corneal opacities (Fig.4) are also common signs, which can be present at different severity stages of the disease**.(Li et al.,2004)**

Munson's sign, a V-shape deformation of the lower eyelid when the eye is in downward position, and Rizzuti's sign, a bright reflection of the nasal area of the limbus when light is directed to the temporal limbal area, are signs frequently observed in advanced stage.

Breaks in Descemet's membrane have been described in severe keratoconus, causing acute stromal oedema, known as hydrops and sudden vision loss and significant pain. **(Thota et al., 2006)**

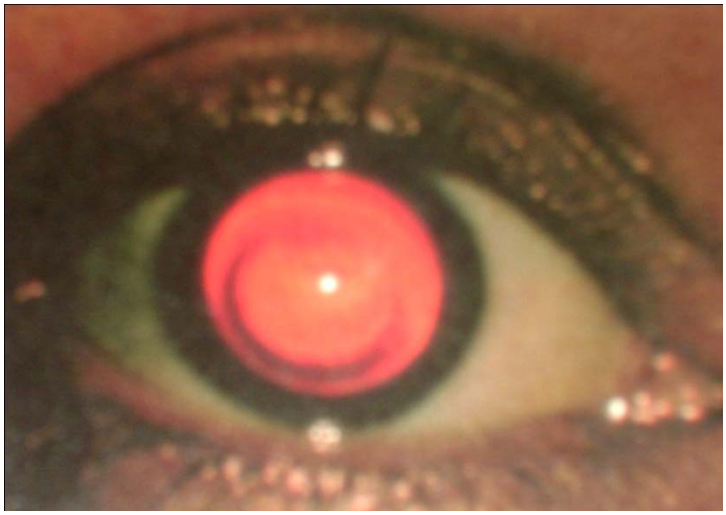

Figure 1: Oil droplet sign **(Kanski, 2007)**

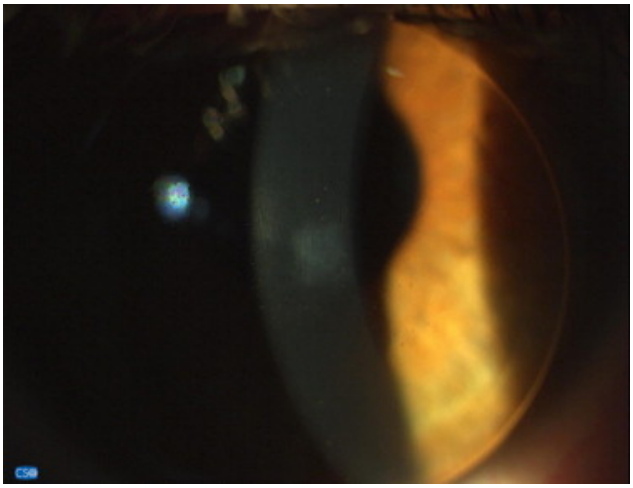

Figure 2: Vogt's Striae sign. Vertical lines in Descemet's membrane are noted. **(Romero-Jiménez et al., 2010)**

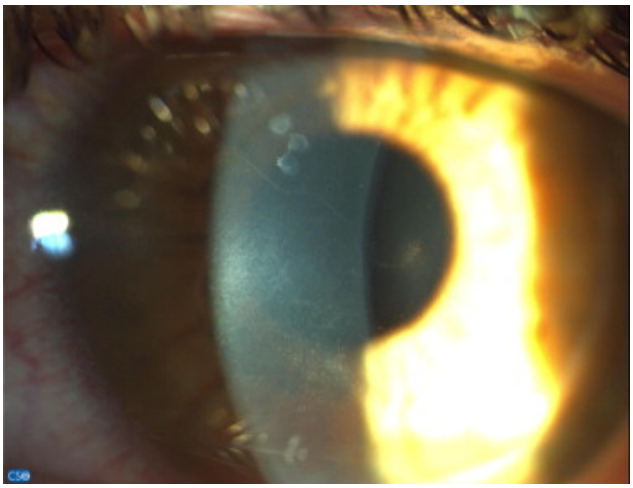

Figure 3: Increased visibility of corneal nerves in keratoconus.

(Romero-Jiménez et al., 2010)

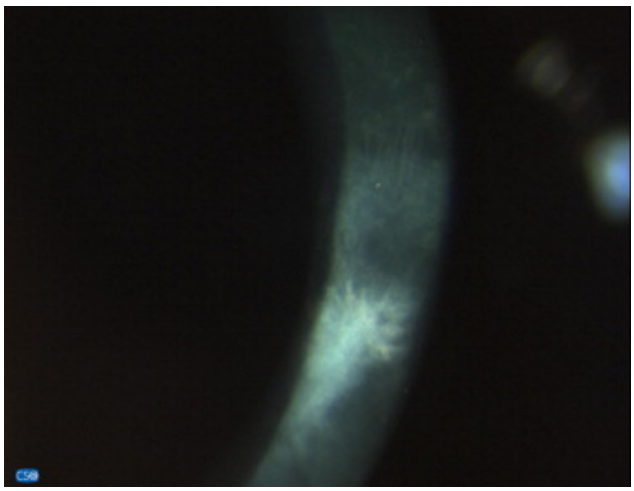

Figure 4: Significant corneal scarring induced by contact lens wear.

(Romero-Jiménez et al., 2010)

Topographic finding

The topographic findings are usually the same in both eyes of the patient, although it may be more advanced in one eye relative to the other. (**Rabinowitz et al., 1993)**

The majority of patients have central steepening extending into the periphery. In this group, corneal steepening is usually confined to one or two quadrants (Fig. 5).
(Tang et al., 2005)

A smaller group of patients has central topographic alterations. Many central cones have a bow-tie configuration similar to that found in naturally occurring regular astigmatism. In the KC patient, however, the bow-tie pattern is asymmetric, with the inferior loop being larger in the majority of cases (Fig. 6)**. (Burns et al., 2004)**

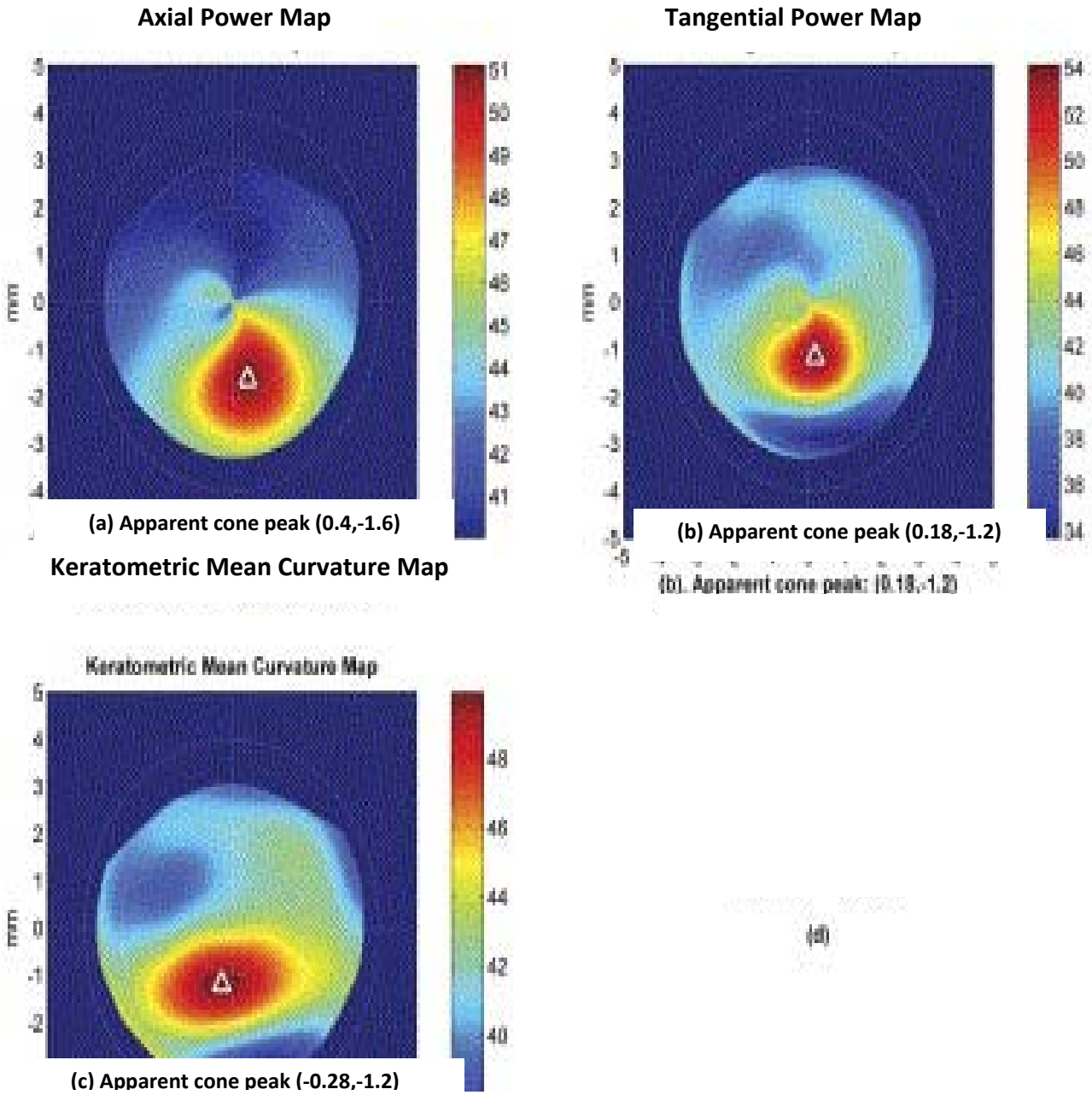

Figure 5*:* Corneal topography of KC

(Tang et al., 2005)

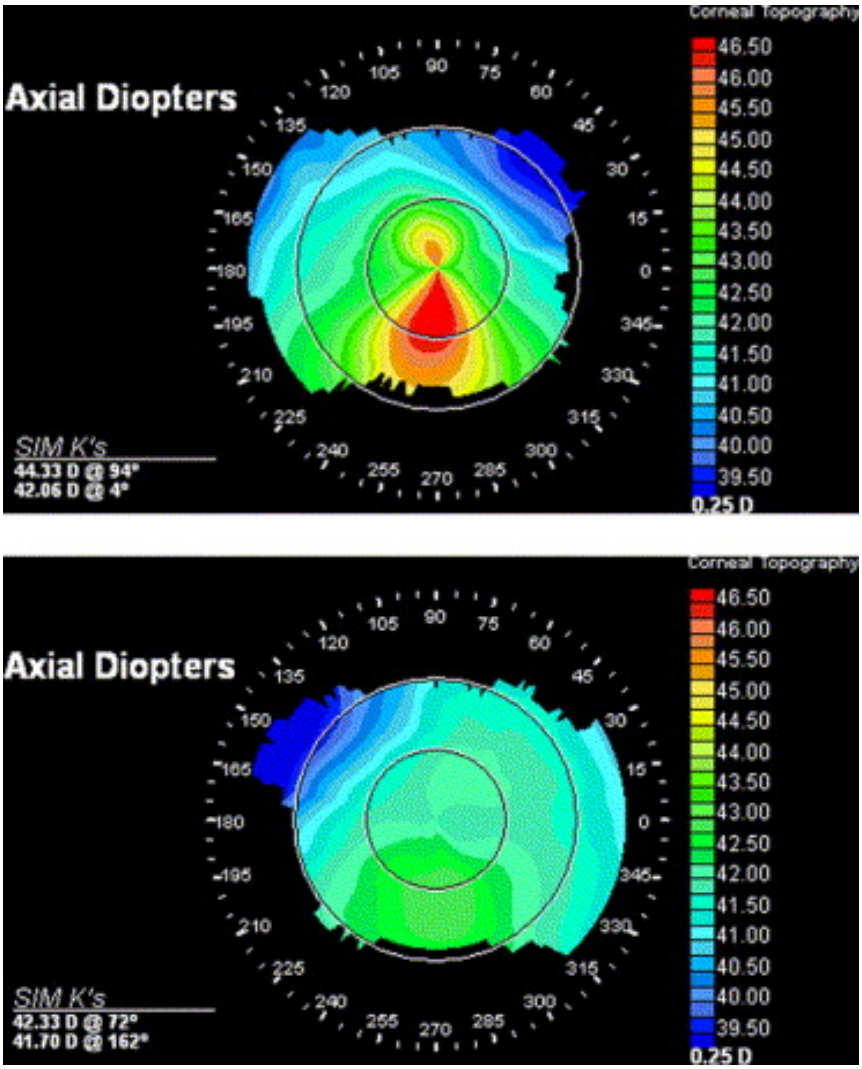

Figure 6*:* Corneal topography. (Top) shows a distinct asymmetric bow-tie pattern typical for KC. (Bottom) shows faint vertical asymmetry (KC suspect pattern). For both maps, the same color scale (steps of 0.25 D) was used.

(*Bühren* et al., 2007)

In contrast to patients who have with-the-rule astigmatism, in KC the steep radial axes above and below the horizontal meridian appear skewed, giving the bow-tie a lazy-eight configuration. Another pattern found in central cones is more symmetric steepening without a bow-tie appearance.These peripheral and central cones probably correspond roughly to the oval-sagging and nipple-shaped cones described by Perry et al. **(Perry et al., 1980)**

Another topographic finding is the presence of an area of increased corneal power surrounded by concentric areas of decreasing power. **(Wilson et al., 1991)**

Similar patterns have been noted in clinically normal family members of patients with KC and in the clinically normal fellow eye of patients with unilateral KC. (**Rabinowitz et al., 1990)**

Videokeratography-derived quantitative indices

The use of quantitative videokeratography-derived indices potentially represents a more reproducible way of quantifying KC and its early phenotypes. It may also enable us to determine an accurate transition from normal to suspect and subsequent KC topography and designate topographic criteria for KC suspects who have minimal overlap with normal population. It also provides more objective information than simple visual interpretation**. (Smolek and Klyce, 1997)**