
Multidisciplinary approach in the management of keratoconus

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Introduction

Keratoconus (KCN) is a non-inflammatory corneal disease that may include stromal thinning, corneal protrusion, Fleischer's ring, Vogt's striae, increased nerve fibers, and rupture in Bowman's layer. Even though it has been well described in the literature for more than hundred years,¹ the etiology is still under study since KCN is manifested by a complex multifactorial interaction of both genetic and environmental factors that widely vary between individuals. There are sufficient evidences which show that KCN has a major genetic predisposition and there exists genetic heterogeneity rather than a single major gene-effect responsible for its development and progression.² However, etiology and pathogenesis of KCN have also been associated to other theories such as metabolic, immunological, endocrinological, ecological, etc.³ Indeed, KCN has also been linked to pro-inflammatory markers in tears, therefore it is through that might be initiated by a series of inflammatory events.⁴

One might say that the question of the underlying reason for KCN development remains unresolved right up to the present day. Independently of this, optometrists should know the current alternatives to manage KCN with a special interest in the compensations directly related with their competencies. The aim of this article is to give a general knowledge about the current alternatives in the management of KCN in the ophthalmology and optometry practice in order that professionals can recommend the best approach to their patients. For this purpose, we have conducted a bibliographic review of KCN in combination with the critical opinion from our ophthalmological and optometric specialists.

Detection and diagnosis

Screening of KCN has a great importance especially before performing refractive surgery procedures in which these are an aggravation factor for the acceleration of the ectatic process.⁵ Recognition of

changes in anterior corneal curvature by videokeratography is very important in the diagnosis of KCN considering that most cases never develop slit-lamp findings. However, videokeratography is not enough for the earliest detection, therefore latest elevation techniques for corneal characterization have shown nearly changes in the posterior surface and in the corneal thickness.⁶ Furthermore, new techniques to measure the epithelium thickness by high frequency ultrasound biomicroscopy first and later OCT epithelial mapping have derived in new indicators to increase de sensitivity and specificity for the detection of early KCN. These new screening indexes are based on epithelium thickness variations which are pronounced with increasing severity of KCN.⁷ Wave front aberrometry and biomechanical properties measured with the Ocular Response Analyzer have been also proposed with less impact than previously described⁵ while new instruments such as CorvisST need to be further studied to develop more comprehensive clinical applications.⁸

Optometric management

The first step in addressing KCN involves the optometrist work by means of fitting special contact lens (CLs) designs. Previously to decide the optimal treatment, a classification can be done based on morphology, disease evolution, ocular signs, and index-based systems.⁹ Table 1 shows the complexity degree of CLs fitting depending on these classifications. For early keratoconus, a soft or soft toric CL could be fitted but a rigid gas permeable (RGP) is the lens of choice as the disease progresses. Despite of the fact that a patient with KCN could be initially fitted with a soft contact lens (SCL), our recommendation is to start fitting an RGP lens because later will be more difficult to tolerate the RGP, and in this way the patient will get easily used to wear the RGP.

Despite this recommendation, SCLs could be an option for patients who are uncomfortable or cannot tolerate RGP. If this is the case, thicker lenses with

low water content should be used to neutralize the irregular astigmatism, especially those made in silicone hydrogel materials. In addition, large diameters of CLs will be recommended for severe apical displacements, a globus cone or a cone with large diameter.¹⁰

A better visual performance can be achieved with a RGP even though fitting process can be more complex than with SCLs. Indeed, RGP becomes the only viable option as the KCN progresses from early to moderate or severe stages. Traditional RGP lenses can be used in early keratoconus when the lenses usually rest on the apex of the cone and peripheral curves are needed for a successful fitting. There are three fitting philosophies based on corneal touches: apical clearance, apical bearing, and three point touch; the latter being the most widely accepted one.¹⁰ Three point touch pattern can be easily obtained with Intralimbal RGP lenses with larger overall diameters (10 mm) than traditional RGP lenses. These lenses are well tolerated, although tear fluid exchange is not as great as in traditional lenses. However, we rarely use intralimbal lenses at QVision, therefore we have omitted these in figure 1 which shows the CLs that we use more frequently. Other alternatives are to combine a SCL with a RGP lens (Piggy-back) or hybrid paralimbal contact lenses with a central RGP portion surrounded by a paralimbal hydrogel skin.

It is now commonly agreed that we have to avoid corneal touch anytime we can. In the last few years, a great progress has been reached with the new scleral CLs. These have the advantages of RGP lenses without corneal or limbal bearing that converts scleral lenses in the most adequate design to ensure corneal health. These lenses have a total bearing on sclera and the diameter is selected depending on the corneal topography, considering that more irregular corneas require larger diameters. The Scleral Lens Education Society classifies scleral lenses depending on bearing area and lens diameter. The lens rest partly on the cornea and partly on the sclera in corneo-scleral lenses, whereas mini-scleral (< HVID + 6 mm) or large-scleral lenses (> HVID + 6 mm) rest totally on the sclera. In our practice, we normally skip the corneo-scleral fit and we start directly with mini-scleral or also called semi-scleral which is our first choice in moderate and severe KCN.

To summarize, Figure 1 shows a diagram with some of the current alternatives that we commonly use in our clinical practice at QVision. This diagram has been built based on our experience and the preferences could differ with those of other practitioners.¹¹ As we have already mentioned, our first choice fitting a CL for KCN is a mini-scleral lens, this is used for advanced cases with a complete apical clearance in most situations. However, the results obtained with these lenses are not always optimal, specially in displaced ectasias. If the fitting fails with a mini-scleral lens, we choose a higher diameter or a scleral lens considering that a scleral indentation might be manifested and a suction cup will be required to remove the CLs from the eye.

In moderate and some severe cases, we use hybrid paralimbal CLs which combine the comfort obtained with a SCL and the optical quality achieved with an RGP. An apical clearance is also acquired with this lens but the area where both materials join may press the lens against the epithelium causing corneal fluorescein staining. In addition, we have learned from experience that if the fitting is not optimal in the first adaptation, it would be better to select another type of lens.

Finally, we use corneal RGP or a Piggy-back in early or moderate cases depending on the confortability that the patient obtains with the RGP lens alone. In last place figure 1 shows that we only fit custom soft CLs in incipient or early stages of KCN on which visual acuity is not highly affected by high order aberrations.

Ophthalmological treatments

Three techniques are currently used in the ophthalmological management of KCN which include *intracorneal ring segments (ICRS)*, *collagen cross-linking (CXL)* and *lamellar or penetrating keratoplasty*. ICRS are semicircular PMMA devices which, when implanted into the corneal stroma, induce a geometrical change in the central corneal curvature reducing the refractive error and the corneal aberrations, thus achieving better visual acuity results. Patients with severe keratoconus (decimal CDVA < 0.4) are the most adequate for ICRS with a high rate of success in terms of gain VA lines at six months, while poorer probabilities of

success are obtained for better CDVA.¹²

In terms of KCN progression, it is not clear if ICRS can be considered as a therapeutic option because even though patients with stable KCN do not progress after the intervention, progressive KCN before the intervention might present a regression after the surgery. In our KCN surgeries with ICRS we have not found regressions but our sample is not large enough for reliable conclusions about this.

The only clear way to stabilize the KCN is by means of CXL which consists in increasing corneal stiffness by irradiating with ultraviolet A (UVA) after it has been soaked with riboflavin (vitamin B), a photosensitizer.¹³ The combination of ICRS with CXL is potentially synergistic and even though previous studies reported that combining these two complementary interventions has been shown to be safe and effective, the ideal combined technique is not known at present.¹⁴

The last technique can be classified *in* Penetrating Keratoplasty (PK) and the more recent Deep Anterior Lamellar Keratoplasty (DALK). These are indicated as a last resort to poor visual acuity with CLs, CLs intolerance or inability to fit/wear contact lenses, and non-resolving corneal hydrops.¹⁵ DALK has as a great advantage the preservation of the host Descemet's membrane and endothelium with a decrease overall graft rejection episodes, including stromal and epithelial rejection.

Despite of the fact that ophthalmological interventions had been carried out, this does not mean an independence of using contact lenses in all cases. When a residual refractive error or an irregular astigmatism are presented after implantation of ICRS, fitting contact lenses becomes an option to improve visual acuity of patients who are not totally satisfied.¹⁶

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Table 1. Classification of Keratoconus and difficulty in contact lenses fitting.

Morphology	Cone Ø	Shape	Location	Quadrant	Difficulty
Nipple	≤5mm	Round	Central / Paracentral	Inf.-nasal	★
Oval	> 5mm	Oval	Paracentral / Peripheral	Inf.-temp.	★★
Keratoglobus	~9 mm		75% of the cornea		
Disease Evolution	Visual Acuity	Corneal Thinning	Corneal Scarring and Opacities	Other findings	Difficulty
Frustrate	~6/6 SPC	-	absent	-	★
Early	~6/6 SPC	mild	absent		★
Moderate	< 6/6 SPC ~6/6 CLs	significant	absent	Vogt's striae & Fleischer's ring; Irregular astigmatism 2 – 8 D	★★
Severe	<6/7.5 CLs	severe	present	Corneal steepening >5D and Munson's sign.	★★★

* SPC= Spectacle

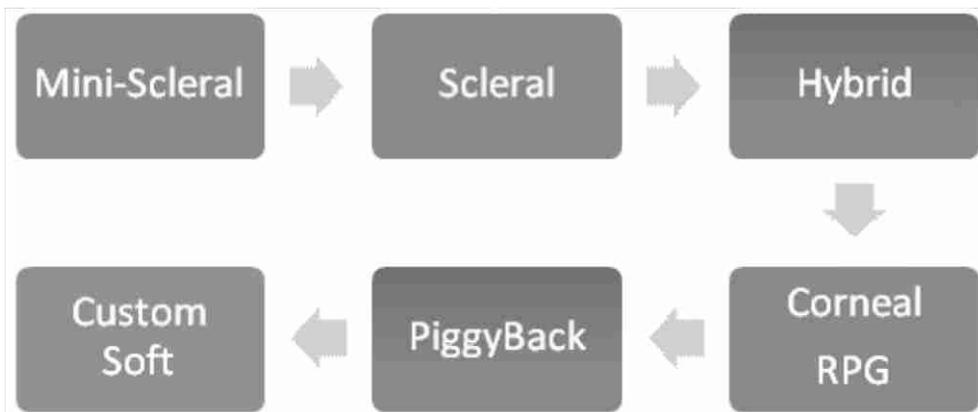


Figure 1. Preferred mode of contact lens correction based on the experience of QVision. Red boxes are lenses from RGP materials while blue boxes are soft lenses. The degraded boxes correspond to a combination of both.

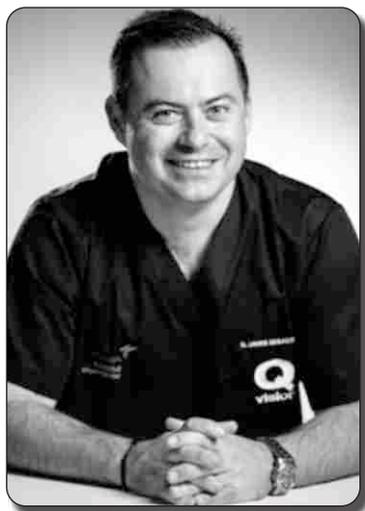
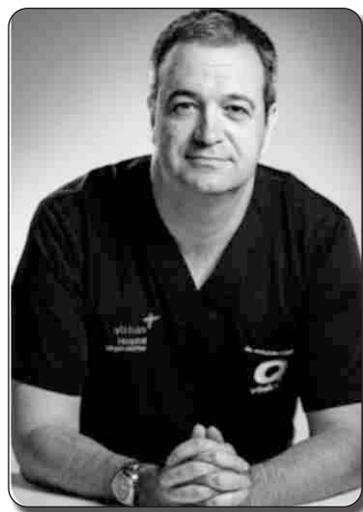


Environmental factors might be rubbing the eyes, wearing contact lenses, eye trauma, and ultraviolet exposure”.

Dr. Almudena Valero
Department of Cornea,
Refractive Surgery,
Presbyopia and Cataracts at QVision.

“New techniques of refractive surgery as Relex-SMILE are conducted trying to preserve the integrity of the cornea”.

Dr. Joaquín Fernández
Department of Refractive Surgery,
Presbyopia and Cataracts
at QVision.



“CLs do not slow the progression of KCN; indeed enhance its development if there is an apical touch”.

DOO. Javier Sebastián
Department of Clinical Optometry and Low Vision



“Some advantages of Scleral CLs are good centration and stability, stable vision, good comfort, less corneal compromise...”

DOO. Patrizia Salvestrini
Department of Clinical Optometry

“Keratoconus progression might be different depending on the type of contact lens fitted”

DOO. Elisa Hueso
Department of Clinical Optometry



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