

Keratoconus and its treatment

An overview

Keratoconus can be defined as a progressive, non-inflammatory conical deformity of the cornea. It is characterised by corneal thinning and protrusion resulting in corneal distortion and decreased vision. Although most cases tend to be sporadic, studies indicate that the likelihood of a blood relative having keratoconus is as high as 10%. The incidence of the disease is relatively low and it occurs in all ethnic groups but with a slight male predominance¹.

Keratoconus is usually bilateral, but it is not uncommon to have an asymmetric presentation. The first eye affected typically suffers more severe consequences, while the second eye may not show any signs until years later, if at all². Symptoms of keratoconus, such as blurry or distorted vision, photophobia, halos around lights, and monocular diplopia, usually appear in the late teens. During the next 10 to 20 years, the condition generally worsens with intermittent periods of remission. In many cases, the degenerative process stops as a patient reaches their 40s. The end point of the disease can range anywhere from mild corneal irregularity, requiring little or no intervention, to severe corneal distortion and scarring, requiring surgery³.

Keratoconus can be classified by the degree of conicity as early or advanced, or morphologically by the shape of the cone. Early keratoconus usually manifests as a small island of irregular astigmatism in the inferior paracentral cornea. Meanwhile, advanced keratoconus is subdivided into three categories – nipple, oval or globus. The nipple form of keratoconus is comprised of a small, near-central ectasia 5mm in diameter or less. In this stage, it is common for the mid-peripheral area surrounding the base of the cone to retain a normal thickness and curvature². It is called the nipple form because there is sometimes an elevated fibroblastic nodule at the apex of the cone.

The most common shape in advanced keratoconus is oval, which is characterised by displacement of the corneal apex below the midline, resulting in an island of inferior mid-peripheral steepening. Many times, this steepening creates an area of normal or flatter-than-normal superior cornea 180° away⁴.

The globus form of keratoconus encompasses nearly three-quarters of the corneal surface. Unlike the nipple or oval forms of advanced keratoconus, the globus cone has no surrounding island of normal mid-peripheral cornea⁴.

Pathology

There has been much speculation on the cause of keratoconus, but the aetiology remains unclear. One theory is that it is

caused by a combination of increased lysosomal enzymes and reduced levels of proteinase inhibitors in the basal epithelium⁵. According to another theory, the primary pathology involves the degeneration of the basal cells of the corneal epithelium², while a different theory suggests the interleukin-1 system⁶. One recent hypothesis trying to explain the pathogenesis of keratoconus incorporates many of the recognised biochemical and molecular abnormalities.

The logic behind this theory is that keratoconic corneas have abnormal processing of free radicals and superoxides, as well as an excessive accumulation of destructive aldehydes or peroxy nitrates. Permanently damaged corneal cells undergo the process of apoptosis (programmed cell death), while reversibly damaged cells undergo wound healing or repair. As part of the wound healing process, various degradative enzymes and healing factors are upregulated. It is this upregulation process which somehow goes astray and results in corneal thinning and fibrosis⁷.

Keratoconus has been associated with several connective-tissue disorders such as Alpert's syndrome, Crouzon's syndrome, craniofacial dysostosis, Down's syndrome, Ehlers-Danlos syndrome, Marfan's syndrome, Osteogenesis Imperfecta, retinitis pigmentosa, and Turner's syndrome. It has also been linked to Axenfeld-Rieger syndrome, floppy eyelid syndrome, tapetoretinal degenerations (especially Leber's congenital amaurosis), Fuch's dystrophy, posterior polymorphous dystrophy, and granular and lattice dystrophies⁸. Patients with keratoconus tend to have a higher incidence of atopic conditions such as hay fever. In fact, a study by Zadnik et al found that 53% of keratoconic patients reported a history of atopy⁹. It has been hypothesised that when these patients rub their eyes to relieve the itching, they induce or enhance the degenerative process of keratoconus possibly through a mechanism of chronic keratocyte apoptosis¹⁰.

Signs and symptoms

Patients with keratoconus often complain

of decreased vision, photophobia, diplopia, visual distortion, asthenopia, and glare around lights. The things to look for are any young adult with irregular astigmatism, or a patient with myopic astigmatism whose spectacle prescription is changing more frequently than normal. Your suspicions should be heightened if this patient shows poor repeatability of the subjective refraction¹¹ or is not correctable to 6/6 in the phoropter.

Slit lamp findings on a patient with keratoconus include Fleischer's ring, Vogt's striae, corneal thinning, and Munson's sign. Fleischer's ring is a partial or complete annular line which demarcates the peripheral edge of the cone. It is found in about half of keratoconus patients and can vary in colour from yellow-brown to olive green. The ring is made from haemosiderin pigment in the basal epithelium². In cases of a subtle ring, a cobalt blue filter with diffuse illumination can be used to enhance its appearance.

Vogt's striae are vertical stress lines in the posterior cornea near the apex of the cone. They are seen as a series of sharp, whitish, vertical or oblique lines just anterior to Descemet's membrane. It is believed that they represent tension lines due to stretching of the stromal lamellae. Their presence can be confirmed by applying external pressure on the globe. The transient rise in intraocular pressure causes them to disappear¹².

In most cases of keratoconus, it is possible to actually see the corneal thinning in the inferior central region on slit lamp examination using an optic section and high magnification. This thinning will result in the displacement of the apex of the cornea below a hypothetical line bisecting the pupillary axis. The other common sign seen with a slight lamp biomicroscopy is Munson's sign, which is the bulging of the lower lid in downgaze and is typical of keratoconus patients.

Other tests which help to diagnose keratoconus include retinoscopy, keratometry and corneal topography. During retinoscopy, the red reflex in a keratoconic eye often demonstrates high amounts of irregular myopic astigmatism with a scissors motion. Sometimes, after dilation, keratoconic patients demonstrate a dark annular shadow surrounding the bright reflex at the apex of the cone. This shadow is caused by total internal reflection of light due to the conical cornea¹².

A patient with keratoconus will exhibit several features on keratometry. In many cases, the mires are oval due to the

amount of corneal astigmatism. They are usually distorted as well due to the irregular corneal surface. The central keratometric rings are frequently non-superimposable which suggests irregular corneal astigmatism, a hallmark of keratoconus¹³. Keratometry also reveals steepening, especially inferiorly. One quick way to check this is to observe the difference in K-readings in primary gaze versus upgaze. Most keratoconic patients will have a dramatic steepening of Ks in upgaze.

Although not vital to the diagnosis of keratoconus, corneal topography is fast becoming an invaluable tool because it provides information about the optical quality of the cornea beyond the central 3mm measured by keratometers¹³. In early keratoconus, topography reveals a characteristic pear-shaped elongation of the central mires mid-peripherally below the corneal midline. As the condition progresses, the steepening usually spreads nasally to include the inferonasal cornea. Advanced keratoconus can demonstrate rotational steepening above the midline along a superotemporal path. The last area to be affected by keratoconus is usually the superior nasal quadrant of the cornea⁴.

When inferior corneal steepening shows up on topography, make sure to rule out several things before coming to a diagnosis of keratoconus:

- **Deficient tear film.** Topographers work by reflecting light off the corneal surface, so areas of insufficient tear film can interfere with this process and make it appear as if there is corneal distortion. This effect can be minimised by instilling a drop of artificial tears before doing topography.
- **Pressure on the globe.** This is more of an issue if topography is done after applanation tonometry. In some cases, distorted mires from tonometry can be seen for as long as thirty minutes afterwards.
- **Incorrect fixation.** If the patient is not fixating at the centre, this will skew the rings and may make it appear as if there is an area of steepening or distortion.
- **Corneal distortion due to contact lens over wear.** Technically, it is possible to consider such factors as maximum axial curvature, maximum tangential curvature, and corneal toricity to differentiate between corneal warpage and keratoconus¹⁴. However, the best clinical way to do so is by repeating topography after the patient has not worn soft contacts for several days and gas permeable lenses for a couple of weeks. A patient suffering from corneal warpage will show signs of improvement after discontinuing contact lens wear, whereas a keratoconic cornea will not.

Treatment options

Treatment of keratoconus depends on the

severity of the condition. Initially, spectacles are successful in restoring vision. However, as the disease progresses, the patient will need contact lenses to provide optimal visual acuity. Lens options include soft (hydrogel) lenses, basic rigid gas permeable lenses, and specialty gas permeable lenses.

Unlike rigid gas permeable lenses, which help mask some of the corneal irregularity of keratoconus, soft lenses tend to drape over the cornea so they are used only in the early stages. In such cases, the lenses are usually toric lenses which are fit the same way you would any other patient with myopic astigmatism. Criteria to use to determine if soft lenses are acceptable is if they do not induce scarring and if best corrected visual acuity is 6/12 or better (this is usually the legal driving acuity).

When soft lenses no longer work, early keratoconic patients may achieve adequate acuity with large diameter RGP lenses. Since keratoconus patients often wear their contacts all day, it is important to use lenses with high oxygen permeability to minimise the risks of neovascularisation. Recommended lenses are the Boston XO, Fluorex 700, or the Fluoroperm 90 or 151. More advanced cases of keratoconus may require a lens designed specifically for keratoconus, such as the Soper, McGuire, or Rose K (see later).

If a keratoconic patient is intolerant of rigid lenses, the use of a rigid lens fitted over a hydrogel lens is sometimes a viable alternative. This concept is referred to as a piggyback fit and works well for many patients. The way this fitting technique works is that the patient is first fitted with a large diameter soft lens with proper centration and movement. Then, keratometry readings are taken of the 'new' corneal surface and a rigid gas permeable lens is fitted on top of the soft lens. Because the patient is wearing two lenses simultaneously, it is even more crucial with this fit to make sure that both lens materials are highly oxygen permeable³.

One significant complication of late keratoconus to watch for is corneal hydrops, which is stromal oedema caused by a rupture in Descemet's membrane. Symptoms of this include decreased vision, redness, photophobia and ocular pain. Treatment includes cycloplegia and a bandage – by either a contact lens or a pressure patch. In some cases, a hyperosmotic agent such as sodium chloride may help¹².

Once all contact lens options have been exhausted, surgery may be needed to obtain adequate vision. Surgical options include simple lamellar keratoplasty, epikeratophakia, thermokeratoplasty, or a penetrating keratoplasty⁸. When referring patients for surgery, the patient's vision should be poor enough that it interferes with their ability to work or drive. Like cataract surgery, there is no magic number at which the keratoconic patient needs to

undergo a corneal transplant. Often, it is based more on the patient's perception of vision. One patient may find that they cannot do their job with 6/9 acuity while another patient may be very satisfied with 6/18. If a patient has a very large area of thinning, if a stable contact lens fit is no longer possible, or if the patient has significant corneal neovascularisation, surgery may be performed earlier than otherwise indicated by visual performance alone.

Penetrating keratoplasty (corneal transplant)

The concept behind this surgical procedure is that clear vision is restored by replacing opaque host tissue with clear donor tissue. The following are the main steps in a corneal transplant¹.

1. In most cases, the donor tissue is harvested less than six hours after death. Corneas from children less than one year old are not usually used because they are floppy and likely to result in high corneal astigmatism. Corneas from donors over the age of 70 are also inappropriate because they have low endothelial cell counts.
2. Graft size is approximated before the operation using a variable slit beam. At the time of surgery, it is precisely calculated by trial placement of trephines with different diameters. In general, the ideal graft size is 7.5mm. Grafts larger than 8.5mm have increased risk of complications, while grafts which are too small can lead to excessive post-surgical astigmatism.
3. Donor corneas are prepared by trephining the previously excised corneoscleral tissue, endothelial side up in a concave Teflon® block. The ideal donor button is 0.5mm larger in diameter than the host opening. By slightly overlapping the tissues, it decreases the risk of post-operative glaucoma, enhances water-tight wound closure and helps prevent excessive post-operative corneal flattening.
4. The donor tissue is fixated to the host eye initially with four interrupted cardinal sutures. Closure is then completed with either interrupted sutures or continuous running sutures.
5. The anterior chamber is reformed with balanced salt solution.

Post-operative complications of a corneal transplant include anterior chamber collapse, iris prolapse, corneal epithelial defects, infection, glaucoma, wound leak, cystoid macular oedema and graft rejection¹. Nonetheless, the procedure is usually very successful^{15,16}, despite a long recovery period. In many cases, patients still have to wear spectacles and contact lenses after the surgery to achieve maximal visual acuity. Therefore, it may not be enough for the patient to be contact lens intolerant to recommend surgery. Adverse

recipient prognostic factors include severe stromal vascularisation, absence of corneal sensation, extreme thinning at the proposed host-graft junction, uncontrolled glaucoma, anterior synechiae, and active corneal inflammation.

Patient discussion

An important part of the treatment of all keratoconus patients should include patient counselling at the time of diagnosis. Some of the things which should be specifically discussed with keratoconic patients include:

- Spectacles may not fully correct vision because of corneal distortion caused by thinning. Even when spectacles work, the prescription will change more rapidly than in normal people. So, instead of it changing every year or two, it may change every six months or sooner
- The reason why contact lenses work and spectacles do not is because the contacts smooth out the cornea and fill in some of the irregularities. When starting the fitting process, it may take several visits to fine tune the prescription because of the irregular corneal surface
- The progression of keratoconus is unpredictable. Once properly fit, wearing contact lenses will not alter the clinical course of the disease. What the contacts will do is allow patients to make the most of their vision
- The possibility of needing a corneal transplant is around 10% or 20%. Therefore, contact lenses will provide a lifetime of good vision for four out of every five patients

Contact lens fits

Three of the more common gas permeable keratoconus fitting sets are the Soper Cone, McGuire, and Rose K. This article will focus on the Rose K, although the same principles can be applied to any fitting set.

There are a few soft lens designs to manage keratoconus, but they do not offer the same level of oxygen transmissibility and clarity of vision as RGP's and so are not widely recommended. Because keratoconus patients may need a corneal transplant in the future, it is important to avoid a lens which might increase corneal neovascularisation which, in turn, might make the patient more susceptible to graft rejection. These lenses should be considered only as a last resort for a patient unwilling to undergo surgery and intolerant of RGP's.

The other thing to consider when starting the fitting process with a patient with the nipple form of keratoconus is that if they actually do have the superficial fibroplastic nodule, they may be more limited in lens types. Since a rigid lens will erode the nodule, resulting in scarring and decreased vision, these patients often

require piggyback lenses, specialty keratoconus lens designs, or surgical excision of the nodule by manual superficial keratectomy or phototherapeutic keratectomy⁴.

The **Soper Cone** is a bicurve contact lens with a fitting philosophy based on sagittal depth. In this lens design, the vaulting effect of the lens increases as the base curve decreases for a given diameter. This change in the steep central posterior curvature is what is used to fit the cone. This design is based on the notion that apical bearing should be avoided in keratoconus because it will ultimately lead to corneal scarring.

The Soper design consists of 10 lenses, designated by the letters A to H. There are three groups for a given diameter/optic zone relationship¹⁷. A, B, C and D are designed for mild keratoconus, or K-readings of less than 48D in either corneal meridian. E, F and G are for moderate keratoconus or K-readings of 48D to 54D in either corneal meridian. H, I and J are for the advanced stage or K-readings of 54D or greater³.

The **McGuire keratoconic lens system** was introduced in 1978 and is a modification of the Soper design¹⁷. It consists of three diagnostic lens sets – each one formulated for the nipple, oval or globus type of keratoconus. The fitting philosophy is aimed at achieving a three point touch and is predicated upon the size of optic zone in relation to conical size. In this design, the optical zone sizes are varied from 6mm for the nipple cone to 6.5mm for the oval cone, and 7mm for the globus. Each lens also incorporates a series of four peripheral curves, which are blended together to create an almost aspheric relationship³. The secondary curve of the McGuire system is 0.5mm flatter than the central base curve. The third curve is 1mm flatter than the secondary curve. The fourth and final peripheral curve is 2mm flatter than the tertiary curve¹⁷.

Rose K is a lens design with complex, computer-generated peripheral curves based on data collected by Dr Paul Rose of Hamilton, New Zealand¹⁷. The lenses incorporate three peripheral systems – standard, flat and steep – in order to achieve the ideal edge lift of 0.8mm. It is available in base curves of 4.75 to 8.00 and diameters of 7.9 to 10.2. The design works by decreasing the optic zone diameter as the base curve gets steeper. Toric curves are available on both the front and rear surface as well as peripherally. Rose K lenses are traditionally made in the Boston ES material, but some laboratories do make them in the Boston XO material which provides the added benefit of increased oxygen permeability.

The Rose K set is manufactured by Nova Contact Lenses and David Thomas Contact Lens in the UK, and a unique software program is used to both cut and blend the multiple curves which

makes replacement lenses easy to reproduce¹⁷.

Suggested fitting protocol

One of the most important things to do when starting a keratoconus fit is to identify the morphological shape of the cone and to determine the stage of the condition. The shape is easily assessed when viewing the cornea with retro-illumination after dilation. The stage of the condition can be assessed by examining K-readings. If the mean K is less than 50D, the cone can be considered early stage. A mean K-reading from 50D to 55D is advanced, and one of greater than 55D is severe.

Prior to lens insertion, instill a drop of anaesthetic in each eye. You should do this even in previous contact lens wearers to decrease adaptation time, thereby decreasing chair time; otherwise, patients tear excessively which causes the lens to sit low and give abnormal fluorescein patterns. Apply the lens and let the patient sit in the waiting room for at least 20 minutes before evaluating the fluorescein pattern. As with all other lenses, examine the central area, the mid-peripheral area and the periphery. It is important that the lens is located centrally when you are evaluating it. If the lens lags down, use upward pressure on the lower lid to improve centration when judging fit.

In terms of determining the correct base curve, start with a base curve equivalent to the steeper of the two K-readings. Of course, the mires are often irregular in keratoconus so the K-reading may only provide a rough gauge of the trial lens. Once you have assessed the fit of this lens (it should be too steep), continue to flatten the base curve until you get the slightest amount of apical touch. If you find it hard to discern where the apex of the cone is, then the lens is too steep. The patient is generally most comfortable and will attain the best acuity when the weight-bearing forces of the contact lens are distributed evenly on the cornea – the so-called three point touch. This means that there should be minimal bearing (touch) at the apex of the cone, as well as an area of bearing between the periphery of the lens and the intermediate zone of the cornea. By making sure this ring of touch is incomplete, you permit freshly oxygenated tears to the central cornea when the patient blinks. By making sure the bearing is as little as possible at the apex of the cone, you will decrease the chance of scarring the cornea.

When selecting an optic zone diameter (pre-set with the Rose K lens), measure the pupil in average illumination and add 1mm or 2mm. At the same time, make sure that the optic zone fully covers the cone. If the cone is large and eccentric, it may be necessary to use a larger optic zone than you would solely based on pupil size, but in most cases accounting for the pupil

will cover both². If you do not fully cover the pupil, your patient will return complaining of decreased vision or glare and halos around lights.

The trial lenses for the Rose K fitting set come in an 8.7mm diameter, which works for most patients. When evaluating diameter, the upper edge of the lens should hit the tarsal plate of the upper lid. If it does not, measure from the top of the lens to the bottom of the upper lid, and add that number to the overall lens diameter of the lens.

When doing the over-refraction start with 1D steps initially, and then refine with 0.50D and 0.25D as you get closer to

the final prescription. Perform the final over-refraction in normal illumination to approximate normal light conditions and pupil size. If you find that you cannot obtain adequate vision with a spherical over-refraction, try a spherocylindrical one. Although the Rose K is available in both a front surface toric and back surface, using the front surface toric will make the fluorescein pattern easier to evaluate. If a toric lens is ordered, the lens will be made 0.3mm larger than the diagnostic lens and then a 0.3mm truncation is used along with 1Δ prism ballast to stabilise the lens.

The criteria for a successful keratoconic lens fit is no different than it is for a

standard fit. The lens needs to be comfortable for a patient to wear all day long and the vision and post-wear biomicroscopy need to be acceptable. The only difference is that acuity for the keratoconic patient may not be 6/6 even with contact lenses, but that is not a problem if the vision is satisfactory for the patient's needs.

All keratoconic patients should return for follow-up at least every six months to examine corneal surface integrity, evaluation of lens fit and for monitoring of changes in corneal topography. Remind them that keratoconus is a progressive disorder and there is no way to predict when and if it will start to progress again. Also inform patients that they need to attend immediately if they experience any symptoms such as decreased vision, photophobia, decreased lens comfort, decreased lens wearing time or increased difficulty inserting or removing the contacts. Any of these symptoms might indicate changes in the cornea which should be evaluated as soon as possible.

Common problems in keratoconus fitting

The single most common error in contact lens management of keratoconus is fitting too tight mid-peripherally. The goal should be to achieve a good stable fit, while allowing for enough movement to provide adequate tear exchange. Poor tear exchange can lead to poor oxygen transmission to the cornea and a pooling of metabolic debris underneath the optic zone, which often shows up as dimple veiling and stipple staining around the base of the cone. The latter problem can be alleviated by decreasing the optic zone diameter, flattening the existing secondary or peripheral curves, or by blending the junctions between the peripheral curves¹⁸. Conversely, a lens fit too flat will move excessively and feel uncomfortable. This can be fixed by steepening the base peripheral curves, which will decrease lens movement.

If coalesced staining of the apex of the cone is visible, there are usually two reasons – the lens is too flat or there are excessive deposits on the posterior lens surface. Apical staining ultimately leads to corneal scarring, so this should not be permitted. If the fit is too flat, it should be steepened. If the fit looks good, observe the lens alone in the slit lamp¹⁸. If the deposits are heavy, give the patient a new lens and begin aggressive enzyming. In some cases, it may be necessary to enzyme the lenses as often as once or twice a week to keep deposits down.

Sometimes, small bubbles under the lens will be seen, indicating a steep fit. This can usually be solved by flattening the base curve until you get the slightest apical touch, increasing the edge lift and/or reducing the diameter of the lens. If the bubbles persist, the lens can be fenestrated

Figure 1

A patient referred for surgery, who was fitted with speciality lenses instead. She now sees 6/7.5 OD, 6/6 OS

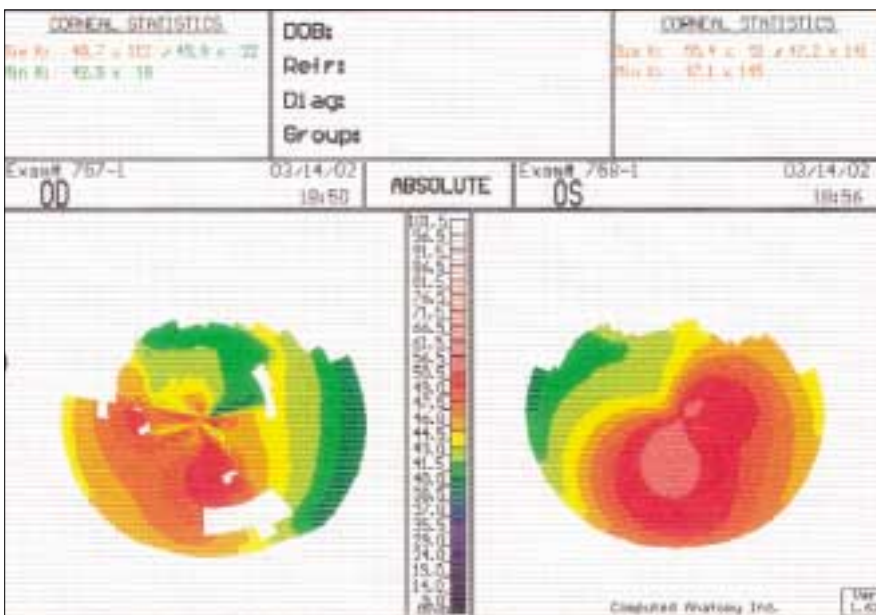
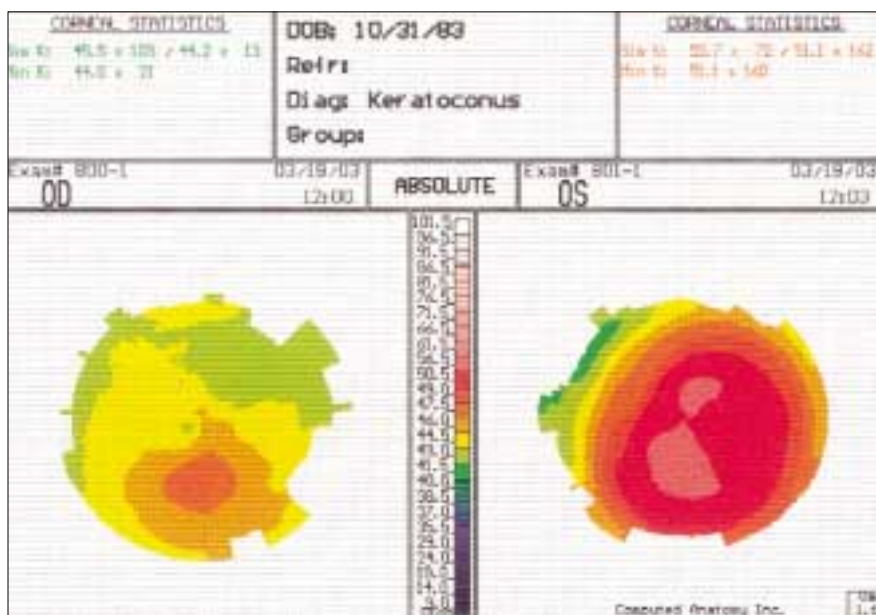


Figure 2

A 20-year old male referred for amblyopia. It turned out he had keratoconus and now sees 6/6 with speciality contact lenses



at the juncture of the optic zone and the secondary curve. Fenestration is not something you should routinely order on a lens, but you can do it as a last resort if all else fails.

If the lens is riding too low, centration can be improved by either increasing the diameter or steepening the base curve. If you decide to change the diameter, in most cases, the changes must be at least 0.3mm in order to see a clinical difference. If you see superior limbal staining or 3 and 9 o'clock staining, reduce the diameter and/or increase edge lift.

Conclusion

Changes in refractive error, such as an increase in astigmatism or a change in the axis, should raise the practitioner's index of suspicion of keratoconus. This is especially true with a young adult with myopic astigmatism who is no longer correctable to 6/6 at the phoropter. Before labelling a patient as amblyopic, take the extra time to perform keratometry or corneal topography to rule out keratoconus. Instead of subjecting the patient to countless hours of vision therapy, a contact lens fit may be more appropriate.

Although the standard treatment of keratoconus involves contact lens fitting, encourage patients to use spectacles as long as possible. Particularly in the early stages of keratoconus, there is no need to hurry patients into contact lenses if they are happy with their vision. A patient who has never worn 'hard' lenses before must be highly motivated to get through the adaptation period of RGPs. The best way to attain this motivation is to keep the patient in spectacles long enough so that they will notice the greatly improved vision obtained with RGP lenses. If the patient is rushed into RGPs and they do not notice the dramatic improvement in vision, they may not be so willing to continue.

Many practitioners shy away from keratoconic fits because they have heard that it is extremely difficult. While it is more challenging than fitting a spherical cornea with disposable soft lenses, the advances in medical technology and the quality of the keratoconic fitting sets make the process more bearable. Chair time and financial reimbursements aside, there is great satisfaction in helping someone who is visually impaired recapture a more normal lifestyle. **Figures 1 and 2** illustrate how specialty lenses can help patients achieve optimum vision.

References

For a full set of references, please email nicky@optometry.co.uk.