Management of advanced corneal ectasias

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ABSTRACT
Corneal ectasias include a group of disorders characterised by progressive thinning, bulging and distortion of the cornea. Keratoconus is the most common disease in this group. Other manifestations include pellucid marginal degeneration, Terrien's marginal degeneration, keratoglobus and ectasias following surgery. Advanced ectasias usually present with loss of vision due to high irregular astigmatism. Management of these disorders is difficult due to the peripheral location of ectasia and associated severe corneal thinning. Newer contact lenses such as scleral lenses are helpful in a selected group of patients. A majority of these cases requires surgical intervention. This review provides an update on the current treatment modalities available for management of advanced corneal ectasias.

INTRODUCTION
The term corneal ectasias includes a group of conditions characterised by progressive thinning, bulging and distortion of the cornea. The commonly encountered ectatic disorders of cornea are keratoconus, pellucid marginal degeneration (PMD), Terrien's marginal degeneration (TMD), keratoglobus, postrefractive surgery ectasia (photorefractive keratectomy (PRK), radial keratectomy (RK) and laser in situ keratomileusis (LASIK)) and postkeratoplasty ectasia.[1] The management of these cases is a major challenge for corneal surgeons because it affects visual acuity (VA) and decreases corneal strength. Thus the aim of the treatment in such cases includes improving VA and simultaneously providing tectonic support to the cornea.[2]

The management of early cornea ectasia is well established in literature, but the management of advanced corneal ectasias (ACEs) is still challenging since there are no definitive guidelines. Newer generation contact lenses (CL) may be useful in some of these cases, but a majority of these cases require surgical intervention. Penetrating keratoplasty (PKP) was the most commonly performed surgical procedure for such disorders in the past. However PKP is associated with a higher risk of complications such as corneal neovascularisation, graft rejection, induced astigmatism and glaucoma.[3–5] Newer surgical techniques and modification of existing techniques over the last decade have minimised these limitations.[3–5] This article provides an update on the currently available treatment options for advanced corneal ectasia.

DEFINITION
The term advanced corneal ectasia is well defined in cases with keratoconus, but in other disorders such as PMD or TMD there are no defining criteria. Keratoconus is a non-inflammatory, bilateral corneal ectasia characterised by gradual thinning and protrusion of the cornea.[4] The severity of the keratoconus was classified by Krumeich *et al* based on the mean keratometry readings on the anterior map of corneal topography, thickness at the thinnest location and the refractive error of the patient.[6] According to this classification advanced keratoconus is considered as: keratometric reading (Km) of >55 D, refraction not measurable, central corneal scarring and corneal thickness <200 μm at the thinnest location.[6] Buxton *et al* have defined advanced keratoconus as Km >52.00 D but <62 D and severe keratoconus as Km >62.00 D.[7] PMD is a bilateral, peripheral corneal ectatic disorder characterised by a band of thinning cornea 1–2 mm in width, typically the inferior cornea, extending from the 4 o'clock position to the 8 o'clock position.[8 9] TMD is a bilateral peripheral corneal ectasia primarily affecting the superior cornea and characterised by high irregular astigmatism, thinning, inflammation, vascularisation and lipid deposition.[10] Keratoglobus is a bilateral corneal ectatic disorder characterised by globular protrusion of the cornea due to generalised thinning, most marked in the periphery. It is usually present at birth and these corneas are prone to rupture after minimal trauma or even spontaneously.[11] Corneal ectasia after surgery is characterised by progressive corneal thinning and irregular astigmatism.[12] These changes have been described as secondary keratoconus by some authors. It has been reported in literature following LASIK, RK, PRK, PKP and deep anterior lamellar keratoplasty (DALK).[13–19]

CLINICAL FEATURES
Patients with advanced corneal ectasia present with distorted VA due to irregular astigmatism or corneal scarring.[20] Most of the patients are already diagnosed at an early stage and gradually progress to an advanced stage following natural course of the disease or failure to halt the progression. The important clinical features with differentiating points are given in table 1.[20]

INVESTIGATIONS
Investigations are required for diagnosis, documentation of progression and planning of treatment.

Pachymetry
Corneal thickness measurement is important for the diagnosis and management of advanced corneal ectasia. Ultrasonic pachymetry remains the standard method for corneal thickness measurement.[21] A nine point pachymetry should be done to map the entire cornea. The advantage is that it is simple and cost-effective, but the limitation is the risk of

Table 1 Differentiating clinical features of corneal ectasias[20]

Characteristics	Keratoconus	PMD	Keratoglobus	TMD
Frequency	Most common	Less common	Rare	Rare
Laterality	Usually bilateral	Bilateral	Bilateral	Usually bilateral
Age at onset	Puberty	20–40 years	Usually at birth	20–40 years
Thinning	Inferior paracentral	Inferior band 1–2 mm wide	Maximum in periphery	Superior cornea
Protrusion	Thinnest at apex	Superior to band of thinning	Generalised	None
Iron line	Fleischer ring	Sometimes	None	None
Scarring	Common	Only after hydrops	Mild	Less common (vascularisation and lipid deposition seen)
Striae	Common	Sometimes	Sometimes	None

PMD, pellucid marginal degeneration; TMD, Terrien's marginal degeneration.

corneal injury and transmission of infection as it is a contact procedure.[21 22] Newer investigational modalities like Scheimpflug imaging and anterior segment optical coherence tomography (ASOCT) are more accurate non-contact methods and are extremely helpful in these cases.[23 24]

Corneal topography

Various tools such as videokeratography (VKG, Placido disc based principle), Orbscan II (slit-scanning) and Pentacam (Scheimpflug imaging) can be used for topographical assessment of the cornea. VKG is useful for surface topography but a major limitation is that peripheral corneal and posterior curvature mapping is either not possible or is less accurate in cases with advanced ectasia.[21–23] Compared with VKG, Orbscan II gives a better pachymetric map, but repeatability and posterior curvature mapping is still less than accurate.[21–23] Pentacam has the advantage of providing precise corneal thickness, and posterior elevation with good repeatability.[23] Hence Pentacam is the preferred modality by most of the corneal surgeons.

Anterior segment optical coherence tomography

The newer generation ASOCT provides real-time high-resolution cross-sectional mapping across any meridian of choice in ectatic corneal disorders, hence it is an extremely useful tool.[24] In addition to corneal thickness values it gives details about anterior segment structures. It is a useful post-operative tool for evaluating corneal graft, graft host junction and graft interface.[25] ASOCT is also useful for the diagnosis and management of complications such as corneal hydrops.[26 27]

COMPLICATIONS

Complications of ACE include corneal hydrops, corneal perforation and corneal vascularisation.[20] Corneal hydrops is characterised by corneal oedema due to seepage of aqueous humour through a tear in the Descemet's membrane (DM).[1] Spontaneous corneal perforation or perforation after trivial trauma is commonly seen in cases with keratoglobus and TMD.[10]

Corneal hydrops has been reported with keratoconus, PMD, TMD, keratoglobus and post-LASIK ectasia.[1] A tear in DM, with subsequent seepage of aqueous humour into the corneal stroma is the underlying mechanism in all cases. If not treated, resolution usually takes a long time and occurs by endothelial sliding over a period of 2–4 months.[1] Medical management consists of topical hypertonic drops, topical steroids, prophylactic antibiotic drops and antiglaucoma medications. However persistent oedema can cause complications such as corneal neovascularisation, infection and corneal perforation. Surgical intervention is often performed to shorten the duration of the disease.[1 27] Intracameral injection of air/isoexpansile gases is the most commonly performed procedure. In the presence of a large DM detachment or stromal clefts, ASOCT guided intrastromal drainage with stab incisions, compressive sutures and even penetrating keratoplasty may have to be performed.[1 27]

MANAGEMENT

Management of ACE is challenging. Various treatment options, non-surgical and surgical have been tried, which are briefly discussed below.

Non-surgical management

Spectacle correction in ACE has no or minimal role. These may be prescribed to selected patients who are intolerant to CL and are not willing to undergo any surgery. Corneal rigid gas permeable (RGP) CL are hard CL that have the advantage of masking corneal irregularities, thus providing a regular anterior refractive surface. These are often the initial lenses to try in cases of corneal ectasia. RGP lenses usually rest on the apex of the cone; so to fit RGP lenses in keratoconus, lenses that have a tricurve or more peripheral curves are used. There are three different types of CL fitting philosophies followed in fitting of these lenses; apical clearance, apical bearing or three point touch. In apical clearance fitting there is no bearing or touch in the apical area and the lens bearing in the periphery. Advantages are reduced risk of scarring, whorl keratopathy and erosions; the limitation is tightening at the periphery can hamper tear exchange and the edge of the lens can come into the visual axis, especially in cases with advanced ectasia. In apical bearing fitting, the optic zone of CL touches the apex of the cone. The advantage is better quality of vision but the problem is there can be heavy bearing on the cornea resulting in corneal scarring and intolerance over long-term use. In three point touch, fitting the lens bearing is shared between the apex and the midperipheral cornea which minimises the risk of apical scarring. These lenses provide good vision, better comfort and prolonged wearing time and are hence the most preferred type of lenses.[28] Hybrid CL contain an RGP centre with a soft skirt. New-generation hybrid CL provide higher oxygen permeability and greater strength of the RGP/hydrogel junction. These lenses are fitted with no or minimal apical touch in the central cornea. The lenses can be fitted on cones of any severity but the problem with these lenses is they can cause hypoxia-related changes such as vascularisation and central corneal clouding.[28] Newer generation CL such as Rose K, scleral lenses, prosthetic replacement of the ocular surface ecosystem (PROSE) and Boston ocular surface prosthesis (BOSP) have shown promise in some studies.[28–34]

Rose K Lenses (Rose K, Rose K2 XL and Rose K2 IC) are multicurve lenses with a small optical zone which snugly fits over the cone. The Rose-K CL provides greater comfort, better quality of vision and requires less chair time in cases with keratoconus.[29 30 33] The Rose K2 IC is a large diameter, intralimbal lens that can be used for large or oval cones.[28] *Scleral Lenses* rest on the sclera and do not touch the cornea and limbus, leaving a clear area between the CL and the cornea. The advantages are good centration, stability and improved VA. The *PROSE* is a non-fenestrated scleral CL that is filled with fluid prior to insertion in the eye. Treatment has a high success rate when measured by the ability to achieve satisfactory fit and impact on VA.[31 34] PROSE treatment can be an alternative to PKP for patients with corneal ectasia who are CL intolerant.[34] The *BOSP* is a fluid-filled scleral CL. These lenses rest on the sclera and do not touch the cornea. There is a constant pool of tears over the cornea, which acts as a liquid corneal bandage and avoids any friction between the posterior surface of the CL and the corneal apex.[33] In addition these lenses mask corneal surface astigmatism and improve best-corrected VA.[33] Thus these lenses are extremely useful in patients with advanced ectasia where the patients are intolerant to CL, or immediate surgery is not possible, or when surgery is refused by the patient. These lenses have also been found to be useful in various ocular surface disorders such as Stevens-Johnson syndrome, Sjogren syndrome graft versus host disease, chemical injuries, dry eyes, limbal stem cell deficiency, ocular cicatricial pemphigoid, exposure keratitis and toxic epidermal necrolysis.[33] The limitation of the use of scleral lenses is high cost, reduced tear exchange and difficult insertion-removal, which requires considerable practice.[28 32–34] Table 2 summarises the outcome of these lenses in cases with corneal ectasias. Overall, studies have shown a good outcome with these lenses.[28 29–34]

Surgical management

Most cases with ACE require surgery for visual rehabilitation as well as for improving corneal strength. Cases with corneal perforation require urgent surgical intervention. Prophylactic surgery is indicated in cases with TMD when perforation is imminent due to extreme thinning.[10 20]

There are many challenges in the surgical management of ACE. First, due to the involvement of the paracentral and peripheral cornea, a large graft with increased proximity to the limbus is often required which increases the chances of graft rejection. Second, extreme corneal thinning extending over a very wide area makes suturing difficult and increases the chances of intraoperative DM perforation. Moreover, recurrence of the original disease can complicate the long-term outcome.[35 36] Lastly, all these procedures are technically difficult with a steep learning curve.[20]

Intrastromal corneal ring segment

Intrastromal corneal ring segment (ICRS) is a method of improving CL tolerance and best-corrected VA for patients with corneal ectasia having a clear cornea.[37] ICRS are commonly indicated for cases with moderate keratoconus. Recently, Intacs SK (SK—severe keratoconus) has been introduced for use in more severe forms of corneal ectasia. It has two significant design modifications—a smaller inner diameter of 6.0 mm compared with 6.8 mm of the standard Intacs; and an elliptical cross section compared with a hexagonal cross section of the standard Intacs.[38] Since their introduction ICRS have been used in cases of ACE with favourable outcomes (table 3) and without any significant complications.[37–42]

Large diameter penetrating keratoplasty

A conventional large diameter PKP is performed in cases with ACE so as to include the thinned out periphery.[43 44] In cases with PMD large eccentric PKP has been described.[43] The problems with such a graft is an increased risk of rejection due to proximity to the limbus. A decentred graft can induce severe postoperative astigmatism and higher incidence of rejection, because of proximity to the limbus (table 3).[43 44]

Lamellar keratoplasty

Crescentic lamellar keratoplasty (LK) is a surgical technique where a crescentic corneal transplant is performed to reinforce the area of thinning. It is one of the earliest surgical procedures reported by Schanzlin *et al*[58] Few case reports have described the use of this technique in management of spontaneous perforation in cases of PMD.[59] *Compressive C-shaped LK* is a peripheral LK technique described by Cheng *et al*[45] in cases with PMD/TMD. A 'match and patch' lamellar graft procedure is performed. Precise dissection of the lamellar recipient bed is done to achieve vertical margins and an even stromal bed depth. A lamellar donor undersized by 0.25–0.5 mm is then sutured to the recipient bed. Suturing a narrower donor onto a wider recipient bed results in flattening and reduction of ectasia.[45] *Modified deep lamellar keratoplasty* is a technique described by Shi *et al*[46] where the initial four-fifths of the corneal lamellae

Table 2 Outcome of various contact lenses in advanced corneal ectasia

Author	Type of CL	Type of study	Indications	Sample size	Outcome
Gupta *et al*[29]	ROSE K versus Soper lenses	Randomised control trial	Keratoconus	n=60	ROSE K cases; significantly better glare acuity and contrast sensitivity BCLCVA—no significant difference
Fernandez-Velazquez [30]	Kerasoft IC versus ROSE K	Retrospective	Keratoconus PMD	n_1=94 (KC) n_2=77(ROSE K)	BCLCVA—no significant difference
Baran *et al*[31]	PROSE	Retrospective	Corneal ectasia	n=59	Satisfactory fit and significant improvement in visual acuity
Lee *et al*[34]	PROSE	Retrospective	Keratoconus Post-PKP	n_1=25 n_2=18	Improvement in visual acuity—88% and OSDI score—79% of cases
Rathi *et al*[33]	BOSP	Retrospective	RGP lenses failure and cases of corneal ectasia	n=23	Significant improvement in visual acuity

BCLCVA, best contact lens corrected visual acuity; BOSP, Boston ocular surface prosthesis; CL, contact lenses; KC, keratoconus; OSDI, Ocular Surface Disease Index; PKP, penetrating keratoplasty; PMD, pellucid marginal degeneration; PROSE, prosthetic replacement of ocular surface ecosystem; RGP, rigid gas permeable.

Table 3 Outcome of various surgical techniques in advanced corneal ectasia

Author	Surgery	Type of study	N	Indication	Outcome	Complications
Rodriguez *et al*[39]	Intacs SK	Retrospective longitudinal case series	6	Post-LASIK advanced ectasia	Average Km reduced from 53.80 ±6.30 D to 48.57±5.00 D	No significant complications
Khan *et al*[40]	Intacs SK	Case series	31	Moderate to severe keratoconus	Average Km reduced from 52.07 D to 46.15 D for K1 and from 57.9 D to 51.2 D for K2	Segment extrusion
Sansanayudh *et al*[41]	Intacs SK	Retrospective non-randomised study	10	Advanced keratoconus	Significant improvement in visual and aberrometric outcome	No significant complications
Fahd *et al*[42]	Intacs SK	Case series	24	Moderate to severe keratoconus	Reduction in myopia and astigmatism Improvement in BCVA Average Km of 52.03 ±4.49 D	No significant complication
Kotb and Hantera[38]	Intacs SK	Prospective, non-comparative	37	Keratoconus, Krumeich's stage II—31 eyes, Krumeich's stage III—6 eyes	Significant improvement in UCVA at 6 months	Bullous keratopathy Overlapping of distal end of ICRS
Varley *et al*[43]	Large diameter PKP	Prospective non-comparative	11	PMD	Mean BCVA 20/30 Average keratometric astigmatism —2.46 D	Graft failure Retinal detachment Bacterial corneal ulcer
Speaker *et al*[44]	Large diameter PKP	Case series	15	Keratoconus and PMD	1 graft failure	9/5 graft rejection
Cheng *et al*[45]	Compressive C-shaped LK	Retrospective, non-comparative, interventional case series	4	PMD/TMD	No recurrence Stable astigmatism ranging from 0 D to −2.75 D BCVA ≥ 20/40	Cataract progression
Shi *et al*[46]	Modified deep LK	Prospective	65	Keratoconus (K> 60 D)	No detectable corneal graft rejection Average BCVA—20/25 at 1 year	Interface fluid-9 cases
Rasheed and Rabinowitz[47]	Simultaneous LK with PKP	Retrospective, interventional case series	5	PMD	No graft rejection BCVA ≥ 20/40	Increased IOP PSC
Jones and Kirkness[48]	LK followed by secondary PKP	Case report	2	Keratoglobus	The right and left eye BCVA 6/60 and 6/18, respectively	Right eye graft decompensation
Vajpayee *et al*[49]	'Tuck in' LK	Retrospective	4	Post-PKP corneal ectasia	Mean Km decreased from 59.67 D to 43.50 D, Significant improvement of BCVA from mean of 0.05 to 0.34	No significant complication
Kaushal *et al*[50]	'Tuck in' LK	Prospective interventional case series	12	Keratoconus with PMD (n=8) Keratoglobus (n=4)	All patients had BCVA > 20/80 Mean Km decreased from 57.54 D to 46.36 D, Mean spherical equivalent decreased from −7.8 D to 1.23 D	No significant complication
Busin *et al*[51]	Tissue excision and corneal tuck	Prospective interventional case series	3	Extremely advanced PMD	Stable resolution of the ectasia in all cases BCVA—20/30 to 20/50	No significant complication
Reinhard and Sundmacher[52]	Sclerokeratoplasty	Case report	2	Keratoconus with peripheral ectasia	Better contact lens fitting Improvement in BCVA	No significant complication
Kanellopulous and Pe[53]	Corneoscleroplasty	Case report	1	Keratoglobus	BCVA 20/50	No significant complication
Maccheron and Daya[54]	Wedge resection	Retrospective	7	PMD	Average reduction in keratometric cylinder of 9.1 D Improved UCVA, BCVA, keratometric cylinder, and spectacle or CL tolerance	No significant complication
Busin *et al*[55]	Combined wedge resection and bevelled penetrating relaxing incisions	Prospective, non-comparative, interventional case series	10	PMD	BCVA ≥ 20/40 in 8 of 10 cases Keratometric astigmatism reduced from 15.1 D preoperatively to 4.6 D postoperatively	No significant complication
Cameron[56]	Lamellar crescentic resection	Prospective	5	PMD	BCVA ≥ 20/40 in four of the five eyes.	Inferior pannus, loose sutures, Recurrence
Javadi *et al*[57]	Lamellar crescentic resection	Prospective	15	PMD	BCVA of 20/40 in 71% of cases Mean astigmatism of 16.00 D at 6 weeks and 4.30 D 2 years postoperatively	No significant complication

BCVA, best corrected visual acuity; CL, contact lenses; ICRS, intrastromal corneal ring segment; IOP, intraocular pressure; Km, keratometric reading; LASIK, laser in situ keratomileusis; LK, lamellar keratoplasty; PKP, penetrating keratoplasty; PMD, pellucid marginal degeneration; PSC, posterior subcapsular cataract; SK, severe keratoconus; TMD, Terrien's marginal degeneration; UCVA, uncorrected visual acuity.

(diameter of 7.75 mm) are removed using a vacuum trephine followed by lamellar stromal dissection with a diamond knife leaving 2 mm (in diameter) of DM at the top of the cone. A corneal graft (8.0 mm in diameter) is then sutured to the recipient.

Lamellar keratoplasty with penetrating keratoplasty

PKP has the advantage of excellent visual outcome but the disadvantages are the need for a large graft, proximity of the graft to the limbus and high post-PKP astigmatism. LK has the advantage of providing tectonic support and preserving the host endothelium but, interface haze and quality of vision is a concern. Several authors have attempted to combine both the procedures so that a large diameter lamellar graft can provide tectonic support to the weakened peripheral host cornea while a central small diameter full thickness graft can provide excellent VA.[47 60 61]

Simultaneous peripheral crescentic LK and central PKP: LK is followed by central PKP.[47] The lamellar transplant restores normal thickness to the peripheral thinned cornea and enables good edge-to-edge apposition at the time of PKP. This reduces the risk of high post-PKP astigmatism and allows for selective suture removal and astigmatic keratotomy to reduce any residual astigmatism. The advantage is, performing the two procedures in the same sitting helps avoid the need for two separate corneas as in cases of sequential LK followed by PKP. The limitation of this surgery is its technical difficulty.

Tectonic LK followed by secondary PKP: A tectonic LK is followed by a secondary PKP usually after 6 months.[61] In this technique, the host cornea is first trephined to the depth of the anterior stroma within the limbus. The lamellar dissection technique is used for tunnelling into the sclera under the limbus to preserve stem cells. The host corneal epithelium is completely debrided, and a donor corneoscleral button, denuded of its endothelium, is sutured on top of it with interrupted sutures. Six months later, a central PKP is performed.[48]

'Tuck in' lamellar keratoplasty

'Tuck in' LK is a special technique of LK for cases of advanced peripheral corneal-thinning disorders like PMD, keratoglobus, or cases with a combination of keratoconus and PMD.[50 49] The surgery involves central anterior stromal lamellar resection followed by creation of a peripheral intrastromal pocket circumferentially in the corneal periphery up to a point 0.5 mm away from the limbus. The donor tissue is prepared such that it has a central full-thickness graft with a peripheral partial-thickness flange of about 2.5–3 mm. The flange of the donor lenticule is tucked into the intrastromal pocket of the host and the graft is sutured to the host. In cases of PMD, only an inferior 180° tucking is done. The central full-thickness graft provides tectonic support to the central cornea while the thin peripheral flange tucked into the intrastromal pocket integrates into the host and provides tectonic support to the peripheral cornea. Moreover, there is no damage to the recipient's limbal stem cells as dissection of the limbal region is avoided, which subsequently promotes healing of the epithelium at the graft-host junction. Combined tissue excision and corneal tuck was described by Busin *et al*[51] in advanced PMD where partial excision of the ectatic part and tucking of the residual thinned corneal lips is done.

Corneoscleroplasty

Lamellar horseshoe-shaped sclerokeratoplasty is a technique where a large 14-mm corneoscleral lamellar allograft is done to support corneal thinning and scleral thinning.[52] Kanellopolous and Pe[53] reported the use of a corneoscleral rim over the thinned corneal periphery of a patient with keratoglobus that acted as a buttress while avoiding any manipulation of the central visual area.

Wedge resection

When ectasia is confined to a small sector of the periphery a *wedge resection* can be performed to reduce astigmatism.[62–64] This technique has several advantages over a corneal graft: preservation of normal central cornea, no risk of rejection or interface haze, better wound strength and shorter visual rehabilitation period.[62–64] However, postoperative unstable astigmatism is an issue due to persistent tension at the sutured wound. Various modifications have been described to improve the outcome of wedge resection. Maccheron and Daya[54] have reported wedge resection followed by complete (limbus to limbus) or partial host deep lamellar dissection to enable closure by mobilising the host anterior lamellar cornea for cases of PMD. Busin *et al*[55] have reported corneal wedge resection combined with paired, opposed clear corneal penetrating relaxing incisions for the treatment of PMD. After crescent-shaped wedge excision of the thinned area inferiorly, the anterior chamber is filled with viscoelastic, and a 3.2-mm metal keratome is used to create two bevelled, penetrating, clear-corneal keratotomies, one at each of the topographic steep axes, immediately anterior to the limbus. The relaxing incisions prevent the astigmatic drift seen following wedge resection.[55] *Lamellar crescentic resection* is similar to wedge resection where the inferior thinned area is excised followed by reapposition of normal-thickness stroma with multiple interrupted 10-0 polypropylene sutures.[56 57]

Outcomes: Table 3 summarises the outcome of these procedures.[43–64]

PREVENTION OF ADVANCED ECTASIA

Collagen cross linking

Ultraviolet-A/riboflavin-mediated collagen cross linking (CXL) increases the biomechanical strength of the cornea through the formation cross links between collagen fibres. CXL is done mostly in early cases of corneal ectasia with a central corneal thickness >400 microns. Recently CXL has been reported in thinner corneas with modifications such as the use of a hypo-osmolar riboflavin solution.[65–68] However, at present the evidence regarding the efficacy and safety of CXL in thin corneas, is limited.

Mid-stromal isolated Bowman layer transplantation

This is a new surgical technique to reduce and stabilise ectasia in eyes with advanced keratoconus. Mid-stromal transplantation of an isolated Bowman layer graft leads to stabilisation of ectasia by the Bowman layer itself, as well as through the wound-healing effect between the host stroma and the Bowman layer graft.[69 70] In this technique an isolated Bowman layer 9.0–11.0-mm in diameter is dissected from the anterior stroma over 360° using a McPherson forceps and a custom-made stripper. This Bowman roll is submerged in 70% ethanol to remove remnant epithelial cells. A mid-stromal pocket up to the limbus over 360° is created under air using the manual dissection technique. The Bowman roll is inserted into the stromal pocket using a special glide. Then it is unfolded and centred by manipulating the graft with a cannula and a balanced salt solution. Van Dijk *et al* have reported reduction and stabilisation of corneal ectasia in two published series (one study involving 20 eyes and the other involving 10 eyes of progressive, advanced keratoconus) following successful mid-stromal isolated Bowman layer transplantation.[69 70] The same author further evaluated the role of this technique to stabilise ectasia, to postpone surgery and to enable continued daily CL wear, in 22 eyes with advanced keratoconus.[71] Reduction and stabilisation of corneal ectasia could be achieved in 20 eyes with this technique. Two surgeries were complicated by an intraoperative perforation of DM. Thus early results, using this new technique, are encouraging. However, long-term studies are needed to establish the safety and efficacy of this procedure. At this point it can be said Bowman layer transplantation could become a supplementary treatment option in the management of advanced keratoconus to postpone PK or DALK and to minimise the risk of long-term complications.[69–71]

CONCLUSIONS

Management of advanced corneal ectasia is a challenge for any corneal surgeon. Over the last decade a number of new surgical techniques have evolved with variable success. The evidence supporting the safety and efficacy of these techniques is inadequate but these techniques are extremely helpful in difficult cases. Prevention of progression at an early stage of disease is the best way to deal with these disorders. Modifications in collagen cross linking and mid-stromal transplantation of Bowman layer may be promising tools for prevention of such disorders in future.

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