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Case Report

Limbal Stem Cell-Sparing Corneoscleroplasty with Peripheral Intralamellar Tuck: A New Surgical Technique for Keratoglobus

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Keywords

Keratoglobus · Corneoscleral graft · Limbal stem cells

Abstract

Purpose: To describe the technique of limbal stem cell-sparing corneoscleroplasty for the management of advanced keratoglobus. ***Methods:*** A patient with bilateral advanced keratoglobus, with best-corrected visual acuity of 20/400 in the right eye and 20/200 in the left eye, underwent limbal stem cell-sparing corneoscleroplasty of the right eye. Initially, a 360-degree limbal incision with 200-µm depth was created, followed by a sublimbal tunnel dissection into the sclera, in order to conserve stem cells. Next, a limbus-to-limbus lamellar keratectomy at 200-µm depth was performed. Meanwhile, a donor corneoscleral button with preserved endothelium of the central 8 mm was fashioned. Prior to suturing the donor corneoscleral graft using a modified suturing technique to cover its scleral component, a full-thickness trephination of 8-mm diameter was completed in the central host cornea. ***Results:*** Reepithelialization occurred within the first week. No episodes of rejection, intraocular pressure spikes, or epithelial breakdown were observed postoperatively. At the 6-month follow-up, the patient had 20/70 best-corrected vision and a smooth cornea with regular astigmatism on topography. ***Conclusion:*** Limbal stem cell-sparing corneoscleroplasty is a single-step

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technique for restoring the structural integrity of the cornea in advanced keratoglobus while preserving the host limbal stem cells.

Introduction

Keratoglobus is a rare form of a noninflammatory corneal ectasia that is characterized by bilateral limbus-to-limbus thinning of the cornea, mainly involving the periphery [1]. The advanced thinning causes the cornea to bulge forward and leads to a risk of perforation following minimal trauma, which is even reported to occur spontaneously [1–3]. High myopia and irregular astigmatism that fail to correct with spectacles lead to poor vision. In addition, the corneal shape typically precludes contact lens fitting [1]. As such, surgical management is indicated to improve vision and/or avoid impending corneal rupture. However, owing to the advanced peripheral corneal thinning present at the time of operation, surgery is extremely challenging [4, 5].

Consensus on an optimal method of management does not exist. Lamellar keratoplasty techniques, including epikeratoplasty, risk developing problems at the interface between the host bed and the graft [5–9]. Moreover, central dissection of a thin host bed during anterior lamellar keratoplasty might be difficult or even impossible to perform. Full-thickness keratoplasty techniques, on the other hand, avoid interface problems, but do have a risk of endothelial rejection [10, 11]. Conventional penetrating keratoplasty have wound-healing problems due to the disparity in thicknesses between the graft tissue and the host bed [12, 13].

Nevertheless, the surgical aim in the management of keratoglobus should be to restore corneal and scleral anatomical integrity by replacing or supporting weak corneal tissue, both peripherally and centrally, while maintaining central corneal optical clarity. Jones and Kirkness [14] described a 2-stage procedure that combines an initial lamellar keratoplasty to provide the structural support needed for a secondary full-thickness penetrating keratoplasty. Alternatively, a single-step technique can be performed by suturing a large corneoscleroplasty over a peripheral lamellar scleral preparation, but disruption of the peripheral limbal stem cells leads to delayed reepithelialization postoperatively [15].

A single-step technique that improves peripheral structural integrity, provides a clear visual axis, and preserves the limbal stem cells might be more acceptable to the patient and surgeon, especially when access to good-quality grafts might be limited. Here we describe a modified surgical technique of limbal stem cell-sparing corneoscleroplasty for the management of advanced keratoglobus, which consists of a 1-stage full-thickness central penetrating keratoplasty and peripheral lamellar corneoscleral tucking.

Materials and Methods

Case Presentation

A 45-year-old man known to have bilateral keratoglobus with poor vision and contact lens intolerance was referred to the Beirut Eye Specialist Hospital for management of his eye condition. History was negative for any previous corneal surgery, complaints of allergic eye disease, or evidence of joint hyperextensibility or collagen vascular disease.

At the initial visit, uncorrected visual acuity was counting fingers at 2 m in both eyes and best-corrected visual acuity (BCVA) was 20/400 in the right eye (with a refraction of –11.25, +4.75, axis 140°) and 20/200 in the left eye (with a refraction of –9.50, +3.75, axis 10°). On slit-lamp examination the patient had bilateral central and peripheral corneal thinning with a forward bulge typical of keratoglobus, in addition to mild central corneal scars (Fig. 1a, b). Corneal topography (Pentacam; Oculus Optikgerate GmbH, Wetzlar, Germany) was done to assess keratometry and thickness of the cornea at various points (Fig. 2a–f). Simulated keratometry was 49.5/59.8 dpt at 81° and 51.2/54.3 at 110° in the right and left eye, respectively (Fig. 2a, d). Pachymetric maps revealed a central thickness of 275 μm in the right eye and 321 μm in the left eye; the thinnest areas were 211 and 203 μm in the right and left eye, respectively (Fig. 2b, e). Bilateral peripheral corneal thinning was evident clinically and in the topographic maps (Fig. 1, 2c, f).

Surgical Technique

A demonstration of the surgical technique can be seen in the online supplementary video (see www.karger.com/doi/10.1159/000471789). Surgery was performed under general anesthesia. After the patient was scrubbed and draped in a sterile manner, a lid speculum was used to open the eyelid and expose the eye. A calibrated diamond knife set at 200-μm depth was used to create a 360-degree circumferential incision at the inner edge of the limbus (Fig. 3a). At a depth of 200 μm, a lamellar dissection using a blade-57 knife was performed to extend the primary incision about 1 mm beyond the limbus into the sclera 360° circumferentially in an effort to preserve the limbal stem cells (Fig. 3b, c). After creating the 360-degree corneoscleral pocket, an 8.5-mm well was placed centrally on the cornea and filled with 20% ethanol alcohol to prevent damage to the host limbal stem cells. After 50 s of exposure, the alcohol was absorbed with a small sponge, and the epithelium was debrided using a dry polyvinyl alcohol sponge (Merocel; Medtronic, Inc.). The cornea was thoroughly rinsed with a balanced salt solution. Lamellar dissection starting from the periphery of the cornea was carried out to perform a limbus-to-limbus lamellar keratectomy at 200-μm depth (Fig. 3d).

The donor corneosclera obtained from the eye bank was prepared as follows: after measuring the diameter of the peripheral circumference of the dissected host limbal pocket using a manual caliper, the donor corneoscleral tissue was shaped through a peripheral sclerectomy to fit the size of the host corneolimbal bed. Holding the donor tissue with a Pierse Hoskins forceps (DTR Medical Ltd.), the margins of the donor tissue were trimmed circumferentially using a scleral scissors to create a tapered edge for better stability and apposition into the pocket. A manual trephine of 8.00 mm was centered and used to make an initial partial thickness groove from the endothelial side of the donor graft. The endothelium outside of the central 8.00-mm zone was then peeled off.

After preparing the donor tissue, a manual trephine set at 8-mm diameter was used to partially punch out the central host cornea. Viscoelastic was then injected into the anterior chamber. A manual Katzin curved corneal transplant scissors (Bausch & Lomb) was used to complete the central full-thickness trephination (Fig. 3e, f).

The donor corneoscleral graft was then placed into the host pocket, and the dissected host limbal area was repositioned and sutured over the donor corneoscleral graft using 10-0 nylon sutures (Fig. 3g). In order to ensure a proper apposition of the trimmed peripheral rim of the donor sclera to the outer circumference of the host limbal pocket, a modified suturing technique was practiced: the needle was passed radially at partial thickness through the donor peripheral sclera and then into the host limbal pocket to exit through the host sclera; it was then passed through the peripheral edge of the host limbus before securing and burying the knot (Fig. 3h, i).

After completing 16 interrupted sutures, it was noted that between 2 o'clock and 3 o'clock the posterior flap of the host pocket did not appose the stromal face of the donor button at the edge of the donor endothelium (Fig. 3j). In order to appose donor and host tissue under appropriate tension, a peripheral full-thickness paracentesis incision was created between 4 o'clock and 5 o'clock. The long-curved needle of one end of a 10-0 double-armed Prolene suture was then passed through the paracentesis incision into the anterior chamber to reach the edges of the fish-mouthed unapposed posterior flap of the host pocket, before being passed upward to exit the surface of the donor corneal tissue. These steps were repeated, such that the long-curved needle at the other end of the double-armed Prolene suture would hook another area in the posterior flap of the host pocket before it is passed upward to exit the surface of the donor corneal tissue anchor. Knots were then secured (Fig. 3k, l).

At the end of the procedure, an air bubble was injected into the anterior chamber to enhance the adhesion of the posterior flap of the host pocket to the graft tissue. A 1.0-mL subconjunctival injection of dexamethasone (2 mg) and gentamicin (20 mg) was given, and a soft bandage contact lens was fitted. Postoperatively, the patient received topical prednisolone acetate 1% eye drops and moxifloxacin 0.5% eye drops 6 times daily, as well as preservative-free artificial tears. Topical prednisolone was tapered gradually over a period of 3 months.

Results

At day 1 postoperatively, visual acuity was counting fingers at 2 m, the graft was well centered, and the interface was clear with a subtotal epithelial defect. The anterior chamber was well formed, and the digitally assessed ocular pressure was normal. At 1 week postoperatively, the epithelial defect had completely healed and the graft was clear. At 1 month postoperatively, the corneal suture that was placed to appose the posterior flap of the host pocket to the donor corneal tissue was removed. At the 3-month follow-up, BCVA was 20/400 in the right eye, and all interrupted sutures were removed. At the 6-month follow-up visit, BCVA was 20/70 (with a manifest refraction of –2.24, +5.00, axis 180°). Corneal topography at 6 months postoperatively demonstrated high regular astigmatism with a central corneal pachymetry of 629 μm, a simulated K reading of 49.5 dpt, and an anterior chamber depth of 5.75 mm (Fig. 4). There was no clinical sign of peripheral iridocorneal synechia. Good apposition of the posterior flap of the host pocket to the donor tissue was maintained all throughout the follow-up period (up to 6 months postoperatively) even after removal of the fixating sutures. Central corneal clarity as well as epithelial integrity was maintained up to the last follow-up visit.

Discussion

Surgery in keratoglobus is necessary to improve vision in patients with spectacle and contact lens intolerance, and to prevent impending perforation [1]. Several surgeries, broadly categorized into full-thickness and lamellar keratoplasty procedures, have been proposed. Lamellar procedures with or without modifications that preserve the limbal stem cells include epikeratoplasty [1], central lamellar keratoplasty with peripheral intralamellar tuck [6, 8], limbal stem cell-sparing lamellar keratoplasty [5], and use of a corneoscleral rim over the thinned corneal periphery [16]. Full-thickness procedures include conventional penetrating keratoplasty [12, 13], corneoscleroplasty [15], and a 2-step tectonic lamellar keratoplasty followed by secondary penetrating keratoplasty [14]. Despite the lack of consensus on an optimal surgical method, surgery should aim to restore both the corneal and the scleral anatomy while providing the best optical clarity for vision.

The advanced corneal thinning in keratoglobus, which may be up to one-fifth of the normal corneal thickness [1], poses challenges even to the most skilled corneal surgeons. As such, anterior lamellar techniques can be very difficult to perform and might have suboptimal outcomes due to the inevitable problems associated with the graft-host interface that would degrade vision [11]. Conventional penetrating keratoplasty is technically also difficult due to the disparity in peripheral graft-host thickness, which precludes adequate wound closure postoperatively [12, 13].

Perhaps corneoscleroplasty with maintenance of the angle drainage and the 2-step tectonic lamellar keratoplasty with secondary penetrating keratoplasty most effectively deals with the issue of restoring corneal and scleral structural integrity while maintaining a clear visual axis [15, 14]. Corneoscleroplasty as described by Burk and Joussen [15], however, fails to spare the limbal stem cells and is associated with poor reepithelialization postoperatively. On the other hand, the 2-step approach described by Jones and Kirkness [14] preserves the limbal stem cells but requires 2 procedures separated by time. A single-step technique that improves peripheral structural integrity, provides a clear visual axis, and preserves the limbal stem cells might be more acceptable to the patient and to the surgeon, especially when access to good-quality grafts is limited.

The current method of limbal stem cell-sparing corneoscleroplasty spares the trabecular meshwork structures, angle structures, and the limbal stem cells by creating a lamellar pocket that is dissected into the peripheral sclera. In our patient, the corneal epithelium was reepithelialized within the first week. In addition, there were no episodes of corneal edema, intraocular pressure spikes, or epithelial breakdown observed up to 6 months postoperatively. Our modified suturing technique might have played a favorable role in keeping the peripheral host-donor junction apposed, which also facilitated the rapid reepithelialization. Special attention should be exercised during suturing of the corneoscleral button, in order to appose the inner host-donor interface at the endothelial junction level. In the case of “unapposing” donor-host tissue at the endothelial junction, a long double-armed Prolene suture can be passed as described earlier.

Overall, the individual steps of this modified technique are not difficult to perform by a corneal surgeon. Moreover, the risk of endothelial corneal rejection is theoretically similar to that of a full-thickness penetrating central corneal transplantation. Besides, the use of current immunosuppressive agents is effective in preventing rejection, if it occurs [10].

In summary, this new modified limbal stem cell-sparing corneoscleroplasty seems to be a satisfactory single-step technique for restoring the structural integrity of the cornea in keratoglobus while preserving the limbal stem cells and without affecting central optical clarity.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

This study did not receive external funding. None of the authors has any proprietary, commercial, or financial interest in any of the products mentioned.

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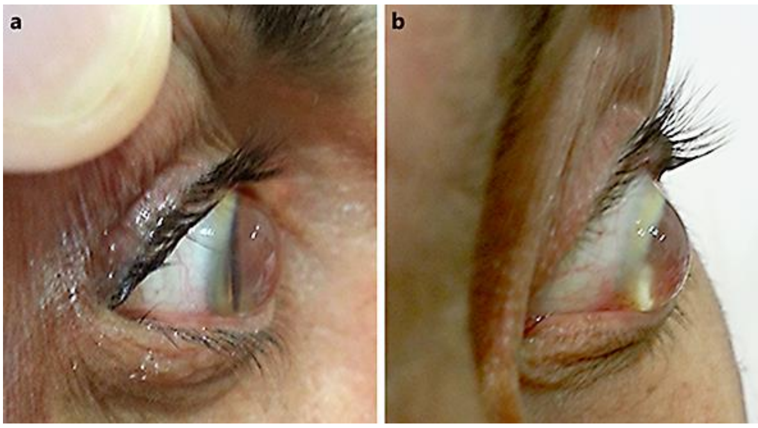

Fig. 1. Slit-lamp photograph of the right eye (**a**) and the left eye (**b**) showing marked peripheral and central corneal thinning with forward bulging consistent with keratoglobus.

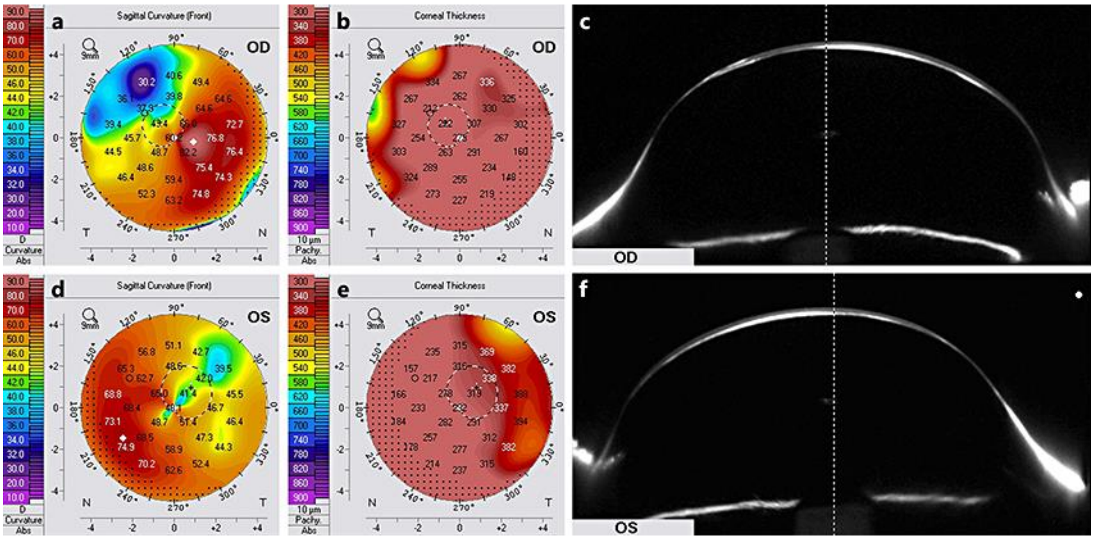

Fig. 2. Corneal topography displaying the preoperative sagittal curvature, pachymetric, and anterior segment cross-section maps of the right eye (**a–c**, respectively) and the left eye (**d–f**, respectively). Notice the marked peripheral and central corneal thinning.

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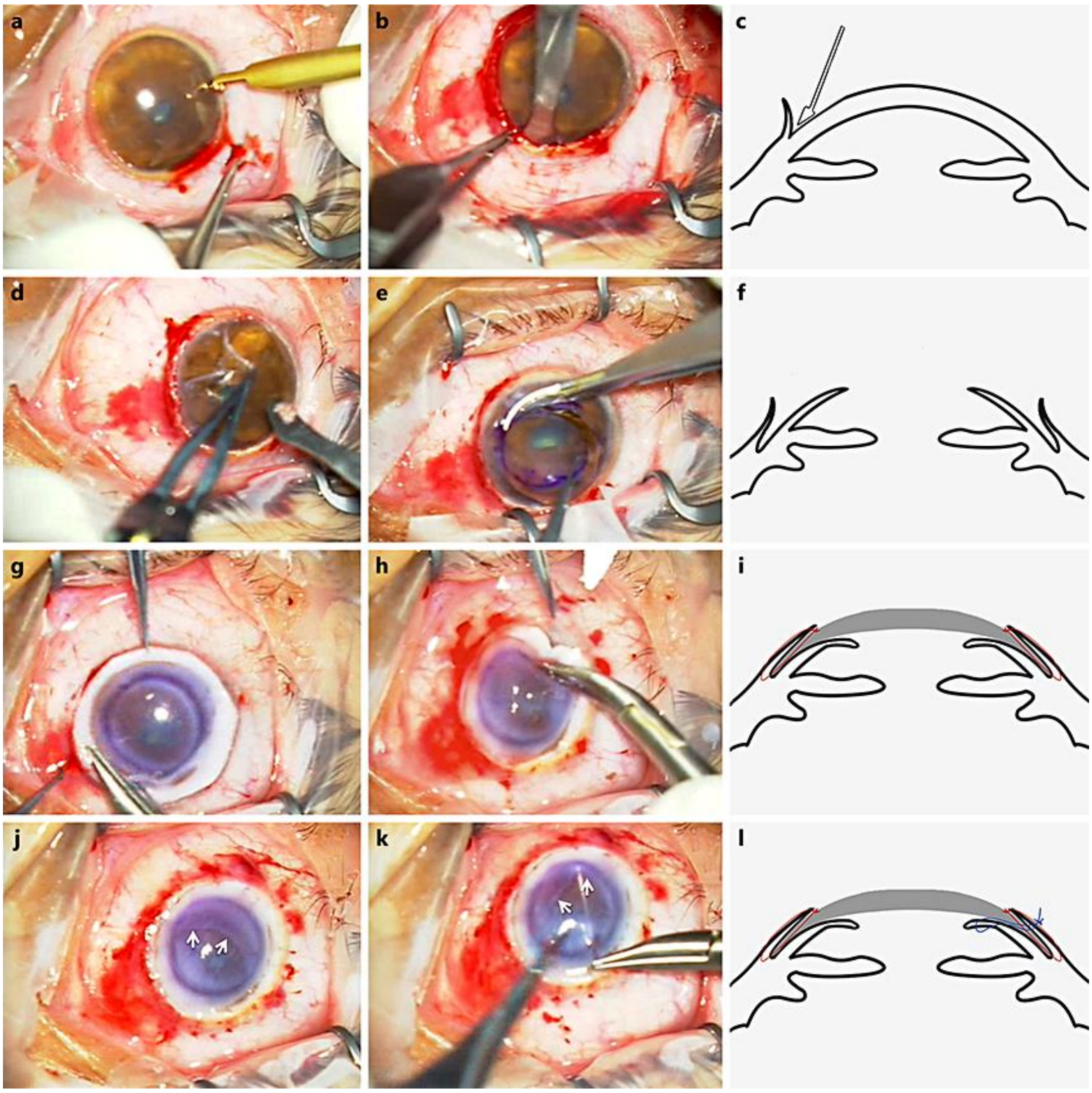

Fig. 3. Intraoperative photographs of the right eye displaying the surgical steps: **a** 360-degree incision with 200-µm depth using a diamond knife over the cornea, 1 mm from the limbus. **b** Using a blade-57 knife, a 360-degree lamellar dissection under the limbus into the sclera was performed at 200-µm depth. **c** Schematic diagram portraying steps **a** and **b**. **d** Limbus-to-limbus lamellar keratectomy at 200-µm depth starting from the periphery of the cornea. **e** Using a Katzin curved corneal transplant scissors a central 8-mm full-thickness trephination was completed. **f** Diagram depicting the resulting corneal anatomy after steps **a–e** were performed. **g**, **h** Suturing of the donor corneoscleral graft into the host pocket using a modified suturing technique as described in the text. **i**, **j** Note the fish-mouthed unopposed posterior flap of the host pocket (arrows) after sixteen 10-0 nylon sutures were placed. **k** The long-curved needle, at one end of a 10-0 double-armed Prolene suture, was passed through a paracentesis incision to appose the donor and host tissue under appropriate tension. **l** Schematic diagram illustrating the final anatomy at the end of surgery.

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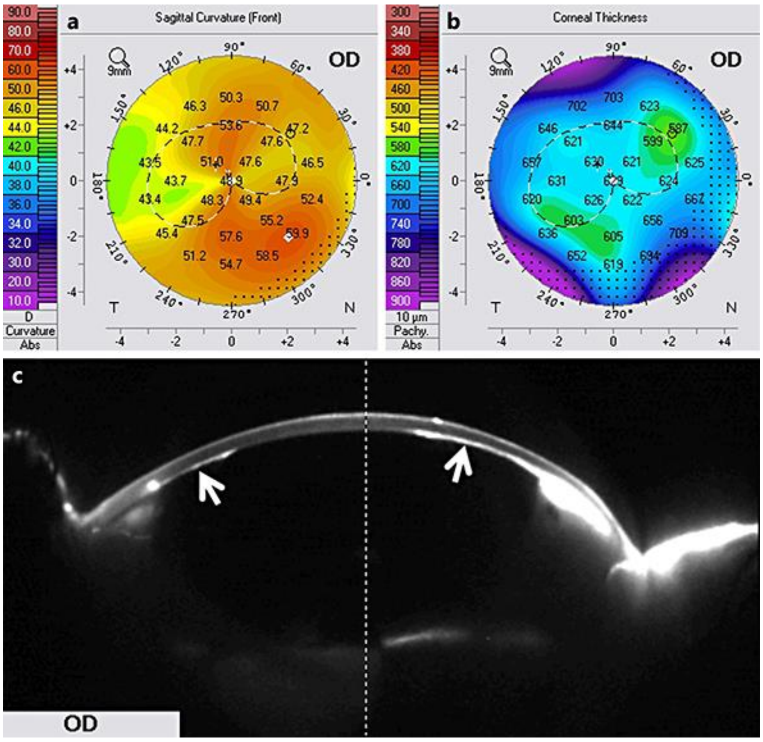

Fig. 4. Corneal topography of the right eye at 6 months postoperatively. **a** Sagittal curvature map, showing a high regular astigmatism. **b** Pachymetric map. **c** Anterior segment cross-section map. Note the good apposition of the host endothelial tissue to the corneoscleral graft (arrows).