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Case Report

Transscleral Cyclophotocoagulation for the Treatment of Uncontrolled Glaucoma in a Boston Keratoprosthesis Type II Patient

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Keywords
Boston keratoprosthesis type II · Transscleral cyclophotocoagulation · Glaucoma · Intraocular pressure · Scleral pressure

Abstract
Postoperative endoscopic cyclophotocoagulation (CPC) for the treatment of glaucoma in patients with Boston keratoprosthesis type II (BKPro II) was first described in 2017 by Poon et al. (*Endoscopic cyclophotocoagulation for the treatment of glaucoma in Boston keratoprosthesis type ii patient. J Glaucoma. 2017 Apr;26(4):e146–9*). As we do not have this device, we present a case of transscleral CPC (TSCPC), in a BKPro II patient who had graft versus host disease and developed uncontrolled glaucoma. We dissected plane by plane to expose the bare sclera and performed the procedure as traditionally described. We concluded that this is a safe, controlled, and effective option in this patient population where the glaucoma treatment options are very limited. To the best of our knowledge, this is the first case report to describe the surgical technique of TSCPC in a BKPro II patient.

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Introduction

Long-term glaucoma is the most consequential and severe complication after any type of keratoprosthesis. The prevalence of glaucoma found before Boston keratoprosthesis (BKPro) surgery has been reported in several outcome studies to be between 36% and 76% [1–5]. The rate of de novo glaucoma appearing after BKPro surgery has been reported to range from 2% to 50% [1, 5–7]. Furthermore, Crnej et al. [8] described glaucoma progression of 21% annually after BKPro implantation. Glaucoma is usually most advanced and can rapidly lead to total irreversible blindness in particularly vulnerable cases like chemical burns and autoimmune diseases, for which BKPro type II (BKPro II) may be implanted.

High intraocular pressure (IOP) is currently the only cause of glaucomatous neuropathy we address; that is, despite knowing that the disease is multifactorial, novel treatments have yet to be adopted in clinical practice. To make things more difficult, the only feasible way to measure IOP in clinical practice is digital palpation of the sclera, which gives only a rough estimate of the pressure. Moreover, glaucoma treatment for BKpro II patients is limited to oral carbonic anhydrase inhibitors and surgery, either using glaucoma drainage devices or transscleral or endoscopic cyclophotocoagulation (CPC) [1, 6, 9, 10] as the penetration of topical glaucoma medications through complete tarsorrhaphy is known to be extremely low.

There is a paucity of data on CPC in this patient population. To the best of our knowledge, this is the first case report to describe the surgical technique of transscleral CPC (TSCPC) in a BKPro II patient.

Case Report

A 64-year-old man with graft versus host disease after an allogeneic bone marrow at 45 years of age was referred to our hospital for evaluation. Since the transplant was performed, he had undergone various types of surgery including eyelids surgery, phacoemulsification, lens implantation in both eyes, and many penetrating keratoplasties, again in both eyes. At the age of 63 years, he had his right eye eviscerated due to various episodes of infectious keratitis.

On his first visit, his visual acuity (VA) was light perception in his only eye, and slit-lamp examination showed a failed penetrating keratoplasty with vascularization and areas of thinning. A B-scan detected no abnormalities in the posterior pole. IOP was 9 mm Hg, and Schirmer's test revealed a very dry eye (0 mm). At that point, the patient was on moxifloxacin qid, dexamethasone bid, and doxycycline 100 mg tid.

His hematologist started him on immunosuppressive therapy (tacrolimus) to control the basal inflammation. An aphakic BKPro II was implanted with no complications. The intraocular lens as well as part of the iris was removed. Four days after surgery, his VA was 20/20 with no correction. Funduscopic examination showed a 0.3 cupping of the optic nerve and IOP estimated by digital palpation at approximately 15–20 mm Hg. During the follow-up, systematic optic nerve photographs were taken, and visual field (VF) tests and retinal nerve-fiber layer optical coherence tomography were performed (shown in Fig. 1).

Six months after surgery, the retinal nerve-fiber layer optical coherence tomography measurement detected a minimal defect in the temporal quadrant, and fundus examination showed a slight increase in optic nerve cupping (0.5). Two months after these early signs, the VF confirmed a rapid deterioration (shown in Fig. 2), and hence, the patient was put on oral acetazolamide 125 mg tid and brimonidine plus timolol tid and scheduled for CPC.

The surgery was performed under general anesthesia. The complete tarsorrhaphy was opened carefully with an electrosurgical scalpel, and the incision was made using the initial tarsorrhaphy scar and in a "U" shape with respect to the optical stem (shown in Fig. 3).

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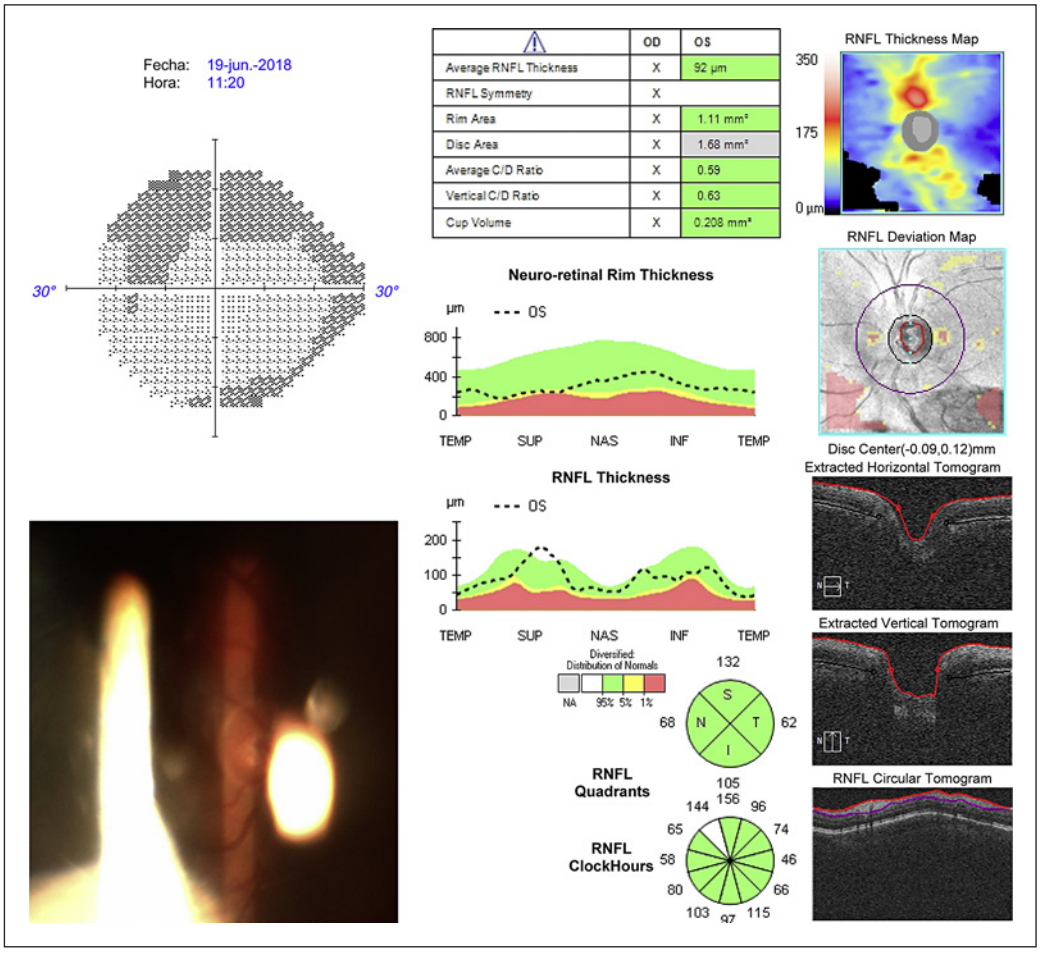

Fig. 1. VF, RNFL OCT images, and optic nerve photographs immediately after BKPro II implantation (June 2018). RNFL OCT: retinal nerve fiber layer optical coherence tomography.

We carefully dissected plane by plane down to the sclera exposing the limbus. The ocular surface was exposed, and the wound held open with 2/0 silk traction sutures. TSCPC was performed in the lower hemisphere (180°) with an Iris Medical OcuLight SLx laser system (Iridex, Mountain View, CA, USA) (shown in Fig. 4), applying 15 shots with the exposure duration set at 2 s and the power at 2,200 mW. We closed the surgical wound using 5/0 Vicryl sutures for the subcutaneous tissue and 6/0 nylon sutures for the skin. Finally, to avoid post-operative inflammatory response, peribulbar triamcinolone was injected.

After the surgery, the IOP remained low and stable, but we continued oral acetazolamide 125 mg bid. Despite all efforts, as described in the literature, the glaucoma progressed. Therefore, we decided to conduct another TSCPC session, treating the upper nasal quadrant (90°), 8 months after the first one. This second session together with the additional administration of subcutaneous adalimumab 40 mg every 15 days seems to have brought the glaucoma under control. Subjectively, the patient reports that his eye stabilized after this intervention, voicing clearer vision and no more VF loss. He remains stable after 3 years of follow-up. Currently, he is on adalimumab 40 mg every 15 days, oral acetazolamide 125 tid, topical moxifloxacin bid, topical vancomycin bid, and topical prednisolone bid. His VA is 20/20, and the IOP is approximately 10–15 mm Hg as assessed by digital palpation. Results from recent assessments show marked damage but no further progression.

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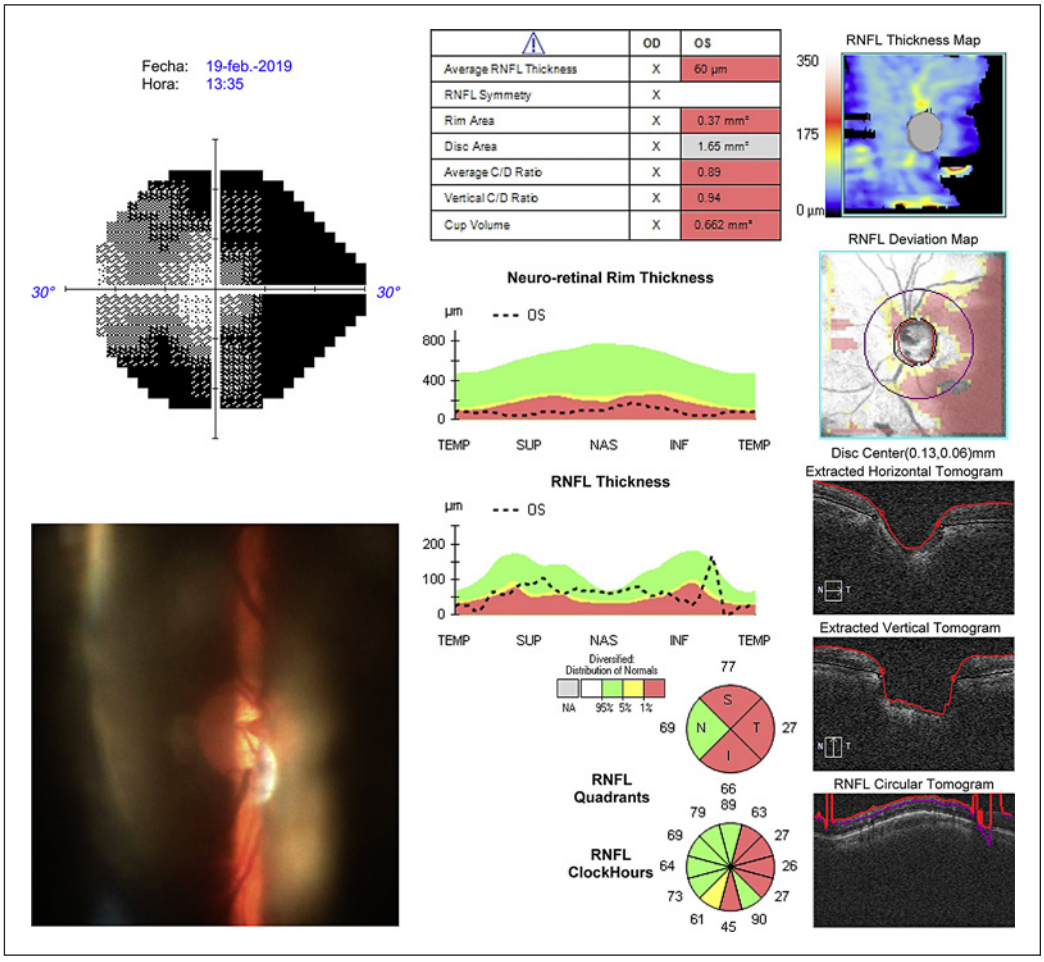

Fig. 2. VF, RNFL OCT images, and optic nerve photographs showing rapid deterioration 8 months after BKPro II implantation (February 2019). RNFL OCT: retinal nerve fiber layer optical coherence tomography.

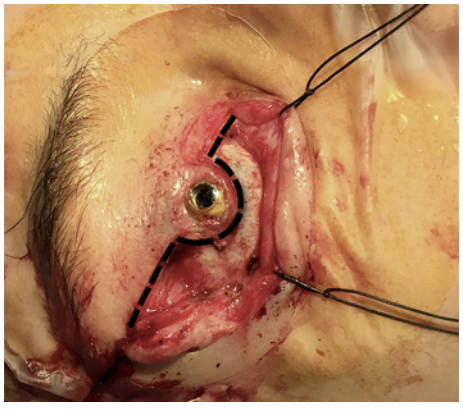

Fig. 3. Surgical incision using the initial tarsorrhaphy scar in a "U" shape with respect to the optical stem.

Discussion

BKPro patients show a high prevalence of glaucoma. These rates are mainly explained by the underlying levels of inflammation due to preexisting autoimmune diseases or other

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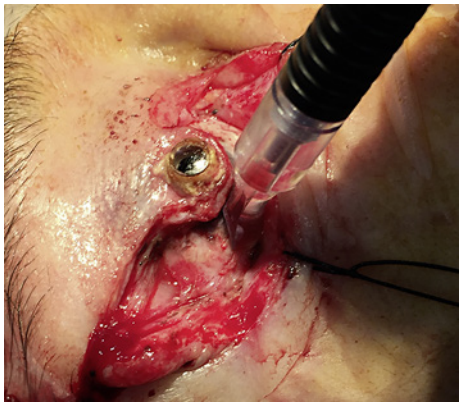

Fig. 4. CPC being performed in the lower hemisphere.

conditions, and this is especially true in the case of BKPro II carriers who have very severe ocular surface diseases. As mentioned, up to 76% of the patients [1–5] present glaucoma before BKPro implantation, and up to 50% of those who do not then develop de novo glaucoma [1, 5–7]. Glaucoma in KPro carriers may be due to several causes, among them: angle closure (especially in temporal quadrant), peripheral synechiae, aphakia, crowding and scleral rigidity produced by the back plate, hypertensive spikes, response to steroids, and mainly, as mentioned, due to inflammation [11]. This multifactorial glaucoma represents the main reason for rapid and irreversible loss of vision after implantation of this device [2].

IOP measurement continues to be the main bone of contention in BKPro patients. To date, there is no validated method to measure IOP reliably in BKPro patients, transpalpebral digital palpation of the sclera being considered the gold standard for IOP calculation in both types of BKPro. This method provides only a rough estimate, but some studies have reported that when performed by experienced doctors, it does accurately detect high IOP [12].

To control an undesired rise in IOP after BKPro implantation, we have several options. In this report, we focus on TSCPC, which is one approach for treating BKPro patients with glaucoma.

Success rates described are variable, ranging from 36.7 to 93.8%, and rates of hypotensive response range from 12.3 to 66% [13]; these seeming to be related to the total amount of energy administered. Regarding the complications described, again, an association has been observed with the total energy applied, with feared complications like hypotony, phthisis bulbi, and loss of vision.

The role of adjunctive TSCPC in 18 patients with BKPro I was reviewed by Rivier et al. [14]. The outcomes of that study revealed that glaucoma refractory to Ahmed glaucoma valve or medications could be managed effectively with TSCPC. Approximately two-thirds of the patients treated recovered normal IOP with a single procedure, a third required additional laser treatment, and 11% more than two sessions.

In another study recently published by a Canadian group [10], TSCPC controlled glaucoma in 61% of the patients with BKPro I treated but did not reduce the number of glaucoma medications. Again, two-thirds of the patients achieved normal IOP after a single treatment. In this study, despite TSCPC, glaucoma progressed in 39% of cases. The failure to control IOP using TSCPC could be attributed to treating end-stage and incoercible glaucoma and anatomical changes of the ciliary body that may occur before or after KPro surgery.

The fact that direct visualization of the target could reduce the amount of energy to which the surrounding tissues are exposed led to the development of endoscopic CPC (ECP). The ciliary processes can be visualized and treated 270–360° by a limbal or a pars plana entry,

and less energy is required to achieve shrinkage and whitening of the processes. Success rates of 43–95% have been reported, with less serious associated complications [15]. The main disadvantage of ECP is the risk associated with penetrating surgery.

In a paper published in 2017, Poon et al. [6] described for the first time the surgical technique of ECP in a BKPro II carrier through incisions made in the eyelids. In this case, the power was set at 2,500 mW, and the ciliary body was treated for 240°. This approach represents a minimally invasive procedure.

As we did not have a suitable endoscopic device, we decided to perform TSCPC. In the literature, it has been mentioned that opening the tarsorrhaphy and dissecting the eyelids down to the bare sclera can be challenging because of the loss of normal tissue planes that may occur after the union of ocular surface and eyelid submucosal tissues [6, 16]. We performed careful plane-by-plane dissection until we reached the sclera and encountered no major difficulties with this meticulous approach. We recommend using a very low-energy electrosurgical scalpel to control the dissection and thereby not damage the globe or the overlying tissues. Another useful maneuver is to hold the eyelids back with a 2/0 silk suture to allow wide exposure of the sclera and obtain a comfortable view to complete the TSCPC. When closing, we recommend also doing this carefully, ensuring that no gap is left around the stem in order to avoid potential infections.

Glaucoma drainage devices have demonstrated active flow in previous studies [1] in patients with BKPro, especially BKPro I. In BKPro I carriers, the subconjunctival plate creates a space surrounded by the filtering bleb where aqueous humor accumulates, mainly when located posteriorly where the tissues seem to be more permeable. But what happens in BKPro II eyes? Another idea that encouraged us to perform TSCPC is that despite other authors describing the placement of an Ahmed valve when preoperatory glaucoma is detected in BKPro II [16, 17], we have doubts about the drainage function of this valve in the absence of conjunctiva. Specifically, the subconjunctival space collapses after a BKPro II implant, and the corresponding structure is completely removed; subsequently, the Ahmed plate is in direct contact with the eyelid tissues which have a notably different tension and rigidity to those of the conjunctiva. For these reasons, we hypothesize that there might not be sufficient differential pressure between the anterior chamber and subconjunctival space, and this might mean that the valve is unable to remain open. Furthermore, we believe that in the long term and in eyes with a filtering bleb, it may be doomed to failure because of the lack of conjunctiva and the significant inflammation in BKPro eyes leading to major fibrosis of the hypothetical device capsule. We conclude that further studies need to be conducted to assess the real effect on IOP regulation when implanting these valves in BKPro II patients.

Finally, we would like to highlight two other issues. First, as has been reported, glaucoma in these BKPro patients is extremely destructive. Therefore, based on our experience, we recommend not delaying surgical treatment in BKPro II carriers and being very aggressive to minimize the damage to the optic nerve. Second, it is also essential to underline the role of immunosuppressants (in particular, tumor necrosis factor-α inhibitors) when controlling basal inflammation in eyes with these conditions. These drugs have shown [18] to be considerably neuroprotective of retinal ganglion cells and in this way, presumably, reduce the risk of glaucoma development and progression. Therefore, we suggest considering such treatment in BKPro patients can likely present late glaucoma. We conclude that TSCPC may be a safe, controlled, and effective procedure in BKPro II patients when ECP is not available.

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Statements of Ethics

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. The research was conducted in accordance with the tenets of the Declaration of Helsinki. Case reports do not require approval number by the Institutional Review Committee of the Cruces University Hospital.

Conflict of Interest Statement

The authors have no conflicts of interest to declare. No nonfinancial relationships influenced the writing of the manuscript.

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Authors Contributions

Ana Orive Bañuelos: corresponding author; design of the work; acquisition, analysis, interpretation, and draft and revision for intellectual content; Begoña Arana Larrea: acquisition, analysis, and interpretation of the work; Alja Crnej MD: analysis, interpretation of the work, and draft and revision for intellectual content; Ana Arce Soto: analysis and interpretation of the work; Noelia Andollo Victoriano: interpretation of the work and final approval of the version to be published; and Jaime Etxebarria Ecenarro: analysis, interpretation, draft and revision for intellectual content, and final approval of the version to be published.

Data Availability Statement

Data and additional information are available upon request via corresponding author's email. Multiple assessments (VFs, funduscopic photographs, optical coherence tomographies, and VA recordings) were performed during patient follow-up. Due to space limitations, only the most significant ones have been provided.

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