

Can an Eye in Phthisis Be Rehabilitated? A Case of Improved Vision With 1-Year Follow-up

Phthisis bulbi implies, in clinical terminology, a shrunken globe, usually from ceased aqueous humor formation (phthisis meaning "wasting away"). The intraocular pressure approaches 0 mm Hg. As a consequence, the cornea becomes distorted and can develop edema and scarring, and the lens develops cataracts. Both seem to occur due to the lack of nutrition by the aqueous. In addition, edema can develop in the macula and the optic nerve head and vision suffers accordingly. Finally, cyclitic membranes and proliferative vitreal retinopathy can develop, resulting in total retinal detachment and scar formation. The cause of phthisis is often uveitis, either long-term or following trauma, surgery, or end-stage, heavily treated glaucoma.

Treatment of phthisis is generally considered hopeless. Aggressive treatment of intraocular inflammation in uveitis or avoiding too much cyclodestruction in glaucoma during the early stage with hypotony may delay or prevent the final collapse. In some rare instances of chronic cyclodialysis or severe cyclitic membranes, a surgical approach has been reported to be of some value.¹ In general, however, if the intraocular pressure is close to 0, the eye is collapsed, the cornea is edematous, the lens is cataractous, the ocular volume is reduced to half or one third of the normal volume, and the ocular layers are correspondingly thickened, no treatment has, to our knowledge, been effective in restoring vision.

Any proposed treatment of phthisis must be surgical, if for no other reason than to restore the clarity of the cornea. In this respect, standard corneal transplantation is

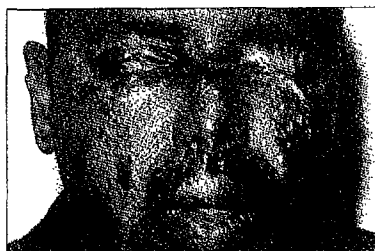


Figure 1. The patient suffered a severe alkali burn 9 months prior to surgery. Visual acuity was light perception OU.



Figure 2. The right eye seemed collapsed and the intraocular pressure was extremely soft by palpation. The distance from the corneal back surface to the retina was 15.5 mm.

ineffective, since the source of nutrition, a healthy flow of aqueous, is absent. With the advent of more advanced techniques for keratoprosthesis surgery and more long-term, postoperative stability, there may be new hope for eyes going into phthisis.² Initial attempts in this direction have been made in the past, but there was no success in restoring vision.³ In this report, we describe a patient with a chemical burn that led to a shrunken eye, no palpable intraocular pressure, an opaque cornea (due in part to the direct effect of the alkali), and an attached retina, who regained substantial vision following keratoprosthesis surgery and was followed up for 1 year.

Report of a Case. The patient, a citizen of Ghana, had alkali thrown in his face 9 months prior to the referral. The burn was extremely severe (**Figure 1**). Ophthalmic examination in April 1997 showed both eyes to be totally soft and collapsed. In the right eye, the upper eyelid margin was fused to the lower part of the cornea (**Figure 2**). Visual acuity was light perception only, with reasonably accurate projection. B-scan ultrasonography showed that the retina and choroid were attached and the vitreous cavity contained some low reflective debris and probably posterior vitreous detachment. An A-scan ultrasonogram showed a 15.5-mm distance between the posterior surface of the cornea and the anterior surface of the retina.

We tried keratoprosthesis, preceded by a buccal mucosal graft that was carried out by Jeffrey Green, MD. The keratoprosthesis surgery was done 2 months later, using a Dohlman-Doane type II device (Massachusetts Eye and Ear Infirmary, Boston).⁴ It was inserted into a 9.5-mm corneal graft from a frozen-stored donor eye in a previously described manner.⁵ After taking down the buccal mucosal graft to bare the cornea, the intraocular pressure was again evaluated by indentation and it seemed to be 0 mm Hg. The eye was opened with a 9.0-mm trephine and the lens remnants were excised. The ciliary body was totally scarred and atrophic. The optic disc and macula were inspected with an endoscope and a moderate vitrectomy was carried out. The graft-prosthesis combination was then placed in the opening and secured with sixteen 9/0 nylon sutures. A scleral patch with a central hole was threaded over the nub of the prosthesis for reinforcement, followed by reattachment of the hinged buccal mucosa so that it fully covered the cornea. Eight milligrams of dexamethasone sodium phosphate was injected through the lower eyelid.

Postoperatively, the mucosa overlying the keratoprosthesis broke up within 1 week (the goal is complete coverage for 2 months); however, later tissue retraction around the prosthesis nub was minimal. Topical medication consisting of 1% medroxyprogesterone acetate sus-

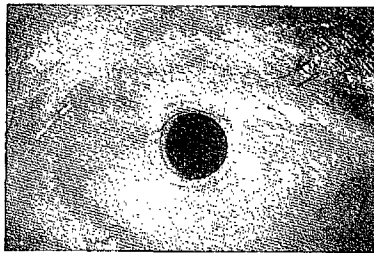


Figure 3. Appearance after buccal mucosal graft, followed up 2 months later by keratoprosthesis and vitrectomy. Visual acuity was 20/60 after a 1-year follow-up.

pension (to discourage tissue necrosis and melt) and an antibiotic was given 4 times daily, reduced to 2 times daily after 6 months.² One month postoperatively, the anterior chamber hardly exhibited any inflammatory reaction, only a rare cell; however, 40 mg of triamcinolone was injected once through the lower eyelid.

Visual acuity was 20/100 OU 1 week after the operation and gradually rose to 20/60 OU during the ensuing 5 months (**Figure 3**). This required a spectacle correction of +10.00 diopters (D) due to a miscalculation in the choosing of the prosthesis. The optic disc was difficult to see due to an overhanging vitreous veil, but the macula was without edema and showed no abnormalities. The visual field showed an enlarged blind spot and a suggestion of a nasal step, most likely due to a short period of high pressure soon after the burn. One year after the keratoprosthesis surgery the situation seems stable.

Comment. The definition of ocular phthisis is vague. Severe hypotony due to overfiltration after glaucoma procedures or extensive traumatic cyclodialysis is usually not labeled as phthisis. In such situations, the aqueous formation is grossly intact and therefore provides adequate nutrition to the cornea and the lens is provided, keeping these tissues transparent. If, on the other hand, the ciliary body is destroyed and the aqueous secretion has ceased, the damage is permanent and the cornea and the lens suffer accordingly. This state is labeled by most clinicians as "phthisis." Some ophthalmic pathologists further restrict the term *phthisis* to describe shrunken eyes that also show

generalized disorganization of the intraocular contents.⁶ In our case, the standard clinical definition of phthisis will be used—referring to irreversible cessation of the aqueous humor formation, near 0 mm Hg intraocular pressure, and a shrunken eye, without consideration of the status of the retina or other intraocular structures.

Our patient had the clinical appearance of a phthisical eye: no detectable intraocular pressure, the bulb collapsed to a much reduced axial length, a swollen and opaque cornea, and a ciliary body that looked totally atrophic on ophthalmic examination. No retinal detachment had occurred and there was no obvious macular edema (the optic nerve could not be well evaluated). The lack of macular edema might be attributable to a low residual, non-detectable, intraocular pressure. Palpation is a crude way of estimating the intraocular pressure but the only one available with this type of keratoprosthesis.

In general, phthisis seems to initially affect the cornea and the lens. Removal of a cataractous lens constitutes no particular clinical problem, but restoring corneal transparency is a more formidable challenge since standard transplantation is impossible. Technique and follow-up regimens for keratoprosthesis, however, have developed to a level where it seems reasonable to try it even in a phthisical situation. The follow-up time of our case is short (only 1 year), and long-term problems may still occur, as in many other keratoprosthesis cases. Still, our study does show that some vision can be restored, at least temporarily, in an eye with close to 0 mm Hg intraocular pressure and an intraocular volume reduced to less than half that of a normal eye.

Retinal detachment is a common end point in phthisical eyes, especially when chronic intraocular inflammation is prominent. In such cases, a keratoprosthesis has to be combined with methods to splint or reattach the retina—obviously a formidable task. Filling the vitreous cavity with a clear substance that exerts a mild, mechanical pressure outward may be a future possibility. The synthesis of a clear hydrogel, gly-

eryl methacrylate, has been interesting in this regard.⁷ This hydrogel was tested for lack of toxic reaction in the vitreous⁸ and in the cornea.⁹ It was also found that a dried pellet of this material placed in the eye of an animal could swell up, replace the vitreous, and exert a desired intraocular pressure.³ Our clinical experience with this hydrogel has so far been disappointing, however, because the hydrogel seems to be easily impregnated by proteins that render it opaque. Silicone oil can be used (D. R. C. Caldwell, MD, oral communication, November, 1994), which is optically clear, but does not exert a swelling pressure and is difficult to handle. Prevention or treatment of retinal detachment in phthisical eyes will clearly be the next frontier.

Our patient is, to our knowledge, the first report of a phthisical eye with severely opaque cornea who had substantial vision restored—at least during this short follow-up period.

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This study was funded by Saad A. A. Al-Rashed, Safat, Kuwait.

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