

The role of keratoprostheses

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Corneal blindness is a major cause of global blindness, second only to cataract. For many patients, corneal transplantation could offer a second chance of sight. However, in some cases (eg, in patients with multiple graft failure, severe chemical burns and autoimmune diseases such as Stevens-Johnson syndrome and ocular mucous membrane pemphigoid) the ocular environment is too hostile for a corneal graft to survive. Keratoprostheses offer these patients the hope and prospect of visual rehabilitation.¹

In recent decades multiple keratoprostheses have been pioneered and developed.² Many have come and gone though and only two are principally used in clinical practice: the Boston Keratoprosthesis 'KPro' type I (Massachusetts Eye & Ear Infirmary, Boston, MA, USA) and the osteo-odonto keratoprosthesis also known as the 'OOKP' (originally described by Strampelli, and later modified by Falcinelli).² The Boston KPro type I is used for eyes with an adequate tear film and an intact blink mechanism. As such, the Boston KPro type I is an alternative to high-risk keratoplasty. The OOKP, on the other hand, is indicated if there is bilateral blindness from severe, end-stage ocular surface diseases in eyes with intact retinal and optic nerve function. The conjunctival and lid abnormalities make the eye unsuitable for conventional corneal transplantation or ocular surface reconstruction.¹

Other less common indications for Boston KPro type I reported in some case series included primary congenital glaucoma with concomitant corneal oedema, Aniridia, irido-corneal endothelial syndrome and gelatinous drop-like corneal dystrophy but the reported numbers are small.^{3–6} A second design, the Boston KPro type II, in which the optic is implanted through the closed eyelid, was designed for severe end-stage ocular surface disease.⁷ This was an attempt to replace OOKP so surgery can be done in a single stage with no

requirement for a suitable tooth but the retention and visual results are no match.

In most cases, OOKP surgery is performed only in one eye, with the other eye reserved as spare, in case the procedure fails in the first eye. Contraindications include age less than 17 years, smokers, phthisis bulbi, eyes with no light perception, and retinal detachment. Patients must have at least one good canine or premolar tooth, minimal gum disease, and, preferably, reasonably good dental hygiene. This would enable a suitable tooth, root and surrounding jaw bone to be harvested to create an adequate lamina. Patients are made aware of the severity of their condition and must fully understand the major surgery required and the potential for severe complications. Moreover, they must be prepared for the altered cosmetic appearance. Lifelong follow-up is required and patients must be highly motivated to comply with postoperative care.⁸

A systematic review done by Tan *et al*⁹ reported that the anatomical survival in all the OOKP studies was excellent, with a survival rate of >80% even at the 20-year time point. In all the studies, more than half of the patients achieved vision of better than 6/18. In three series visual acuity was better than 6/18 in >60% of eyes. The Boston KPro type I is indicated in cases with good tear function and ocular surface state; therefore, its results cannot be compared to those of OOKP studies because of differences in disease severity and ocular surface and tear film status.⁶ A recent study by Yaghouti *et al*¹⁰ using the Boston KPro type I and type II devices to treat SJS and severe chemical burns suggested poor anatomical and visual outcomes at 5 years (success rate zero). The most recent study by Lee *et al*⁷ using Boston KPro type II device reported one third of their patients maintaining 20/100 vision or better at the last follow-up visit (mean follow-up duration was 70 months) and they reported 50% device retention rate.

The most common long-term blinding complication in all keratoprosthesis types is glaucoma with all the difficulties to monitor and treat in those cases. Intraocular pressure is difficult to measure following all types of KPro.

Topical and systemic anti-glaucoma medications, aqueous shunt surgery, and cyclo-destructive laser are all potential management options. The reported retinal detachment rate after OOKP varied between 0 and 26%. The incidence of other sight-threatening complications such as endophthalmitis, is relatively low after OOKP (0–8%).⁹ While for Boston type I and II, the reported rate of endophthalmitis incidence was 11% and 14%, respectively¹¹ and retinal detachment risk after Boston type II as reported by Lee *et al* was 18.8%.⁷

There are some other complications reported after OOKP such as mucosal ulceration, lamina exposure, extrusion and resorption which can lead to loss of the eye.⁹ Complications common to Boston keratoprotheses include but are not limited to, retroprosthetic membrane formation (most common complication; reported range 13–60% of the cases), sterile keratolysis, infectious keratitis, and sterile vitritis. Device extrusion can occur when sterile keratolysis is not immediately addressed.¹²

Conclusion

Keratoprosthesis surgery is complex and requires meticulous care at each step to ensure the overall success rate and a careful lifelong follow-up commitment. A multidisciplinary team is needed to provide the best care for the patients. The literature lacks randomized controlled studies comparing different types of keratoprotheses for the different indications. However taking the current available evidence into account; we can conclude that Boston type I should be reserved for wet blinking eyes with no keratinisation neither bulbar nor tarsal, as an alternative to high-risk keratoplasty. We would not normally consider offering a Boston if the other eye is seeing, as we are converting what is usually a stable situation to one which can have complications causing loss of visual potential. OOKP is for dry keratinised surface with defective or absent lid or blink mechanism. Boston type II does not match the retention and visual results of OOKP.

Conflict of interest

The authors declare no conflict of interest.

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