EDITORIAL

Visual Rehabilitation in Corneal Blindness

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Blindness from corneal diseases remain a major ophthalmic health problem worldwide second only to cataract.[1] The spectrum of corneal blindness is quite different in developing and developed countries and encompasses a wide variety of infectious and inflammatory diseases. Although scarred corneas from preventable diseases like trachoma and malnutrition are rarely seen in developed countries, they remain prevalent in regions with poor health environment. On the other hand, autoimmune conditions such as Stevens Johnson syndrome (SJS) and ocular mucous membrane pemphigoid (MMP) as well as chemical injuries are seen worldwide and often present the most challenging situations for corneal specialists.

Eyes with severe corneal scarring in the context of ocular surface failure could just be able to distinguish light from dark and such a scenario is devastating when both eyes are affected. Blindness has not only a negative impact in the quality of life but also causes a considerably economic burden affecting the individuals, their families, and society. The main challenges from diseases causing corneal blindness are ocular surface scarring, dryness and stem cell deficiency, because such conditions prevent long-term survival of a corneal transplant. Thus, in these cases the only hope for visual restoration is to bypass the ocular surface and cornea with an artificial cornea or keratoprosthesis (KPro).[2,3]

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Over the past five decades multiple KPros have been pioneered and developed but most of them just had temporary existence, reflecting the complexity and challenges in the pursuit of a successful artificial cornea. Currently only two principal KPro devices are used in clinical practice: The Boston KPro type I (Massachusetts Eye & Ear infirmary, Boston, MA, USA) and the Osteo-odonto-keratoprosthesis known as "OOKP" (originally described by Strampelli and later modified by Falcinelli).[2,4]

The Boston type I is a synthetic KPro and is the most common device implanted worldwide.[5]The current design of the Boston type I KPro is composed by a front plate with an optical polymethyl acrylate (PMMA) stem and a titanium back plate. A donor cornea is sandwiched between the plates and it is the carrier cornea button which is sutured onto the eye. The presence of a wet ocular surface is paramount when considering Boston type I KPro implantation: both adequate tear production and intact blinking mechanism are necessary. As such this KPro is primarily an alternative to high-risk penetrating keratoplasty. Other indications for Boston type I KPro include chemical injuries, primary congenital glaucoma with corneal decompensation, aniridia, irido-corneal syndrome and gelatinous drop-like dystrophy.

The Boston type II KPro is another design in which the optic is implanted through the closed eyelid. This device is indicated in end-stage ocular disease with dryness and or adnexal abnormalities.[6]However careful patient selection must be done as severe dryness and contracted lid fornices are accompanied by chronic inflammation which implicates high risk of failure for both Boston 1 and 2 KPro.[5,6] Hence in patients with advanced stage of Stevens Johnson syndrome, chemical injuries or ocular pemphigoid, only biological KPro scan provide long-term survival.

The OOKP and the tibial bone osteo-keratoprosthesis are the two well-known biological

KPros. Despite both have reported favourable anatomical and functional success, the OOKP have proved to have achieved better long-term outcomes.[7]The OOKP is a two-stage procedure and uses the patient's own tooth and alveolar bone to create a frame (osteo-odonto-lamina) that carries a PMMA optical cylinder. After its initial preparation, the lamina is implanted in a submuscular pocket for nourishment and growth of surrounding connective tissue. At the same time the ocular surface is covered by a buccal mucosa; graft (stage 1). Three to four months later the lamina is retrieved and then sutured onto the sclera after central cornea trephination and removal of iris, lens, and anterior vitreous (stage 2). The buccal mucosa that was initially reflected will now cover all the lamina except the central optic cylinder through which patients see. The unique autologous biological composition confers the OOKP excellent bio-integration and no immunological rejection.

The Boston type I and type II KPro are both one-stage procedure, however the OOKP is a more complex multistage procedure and is available only in a handful of centres around the world. In general patients who are considered for a KPro should have bilateral blindness and commit to lifelong follow-up. They need to be highly motivated to comply with postoperative care and particularly in the case of the OOKP must be prepared for the altered cosmetic appearance. Bypassing an opaque cornea improves vision with all types of KPro, but the amount of improvement is determined by the status of the retina and optic nerve. It is not meaningful to compare the anatomical retention outcomes between the Boston type I and the OOKP as the indications for each KPro are different. But despite better results due to modifications in its design and improvements in post-operative management with the Boston type I, the long-term anatomical retention rate (five or more years) in SJS and severe chemical burns remains below 50%.[8] Similar retention rate is also achieved by the Boston type II KPro which has comparable indications to the OOKP.[6] In contrast the reported long-term anatomical retention rate from the OOKP in all studies is above 80% (even at the 20 year time point) with more than half of the patients achieved vision better than 6/18.[2,9] Thus, in such truly challenging cases of corneal blindness the OOKP has proved the most effective in restoring sight and the most endurable Keratoprosthesis.[2,4,7]

Complications are not uncommon in keratoprostheses. The most common long-term blinding complication of all KPros is glaucoma, which can affect two thirds of cases and be a pre-existing condition in more than one third.[5-7] In addition to the challenges of estimating intraocular pressure in KPro eyes, topical medications are not absorbed through buccal mucosa in OOKP eyes and not all patients can tolerate oral acetazolamide. Glaucoma in KPro eyes usually requires surgical implantation of tubes which have variable results but commonly fail in the long-term. Endophthalmitis is the most feared complication in KPro and has been reported in less than 20% of cases, being lower in the OOKP (0-8%).[5-7] Other complications seen are retroprosthetic membranes, retinal detachment, keratolysis in Boston type I and lamina exposure in OOKP due to buccal mucosa defects.

In summary, patients considered for a KPro must be carefully assessed for an appropriate decision whether this treatment should be offered as well as the type of device required for each case. They also should be aware that long-term success is not guaranteed, and that the treatment is not reversible in the case of OOKP. Future improvement of the ophthalmic use of stem cells may allow to regenerate the ocular surface restoring corneal transparency or providing appropriate environment for a corneal graft. But in the meantime, for the most challenging cases of corneal blindness, keratoprosthesis is the only hope for visual rehabilitation.

Conflict of Interest

Authors declared no conflict of interest

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