

The Current Status of Infant Keratoprosthesis

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ABSTRACT

Since the release of the Boston I device twenty years ago a number of factors have contributed to its more widespread use as well as the concept of utilizing these devices to correct congenital cornea blindness. We currently recognize much of the complexity involved in the embryonic development of anterior segment features and the defects resulting from genetic anomalies. The technological aspects of performing surgery in these infant eyes combined with inherent propensity for inflammation have created an environment in which complications are not infrequent. Some have maintained that in view of this situation keratoprosthesis should not be contemplated in infants. Never the less a number of early benefits and careful planning combined with a multidisciplinary approach can achieve long term success.

Keywords: Cornea; Keratoprosthesis; Boston I; Congenital Cornea Opacity; Sclerocornea; Cataract

Introduction

The concepts of pediatric keratoprosthesis (KPro) and more recently infant keratoprosthesis are relatively new. Our first case was performed in 2003. Yet a close inspection reveals that while there are innovations, the general principles utilized are firmly based in historical events. Thus, we recall the team of Cordona, Castroviejo, and Devoe who pioneered the basic KPro techniques in the late 1950's as well as the research conducted by Claes Dohlman culminating in the design of the Boston Type 1 device [1]. If not for the early transactional success associated with this new device, first in end stage disease and then in a variety of less severe conditions, we would not have achieved the degree of success with adults which led to the implantation of these devices to address congenital cornea blindness.

Most cases of congenital cornea blindness until recently were grouped in clinical categories of Peters Anomaly or sclerocornea [2]. However, the work of Kenneth Nischal published in 2015 was important in advancing our understanding of the genesis of these diseases [3-5]. He proposed a clinical surgical classification which included glaucomatous disease associated with the presence of Haabs Strae, infectious conditions such as Herpes simplex, a dystrophic condition to include congenital hereditary endothelial dystrophy (CHED) and posterior polymorphous dystrophy (PPMD),

iatrogenic produced defects related to amniocentesis and forceps delivery and true developmental defects such as Peters Anomaly and sclerocornea. Primary Congenital Opacity can be further differentiated as follows: Those related to chromosome 20 defects which lead to CHED, PPHD, and some instances of congenial glaucoma. Isolated sclerocornea is characterized by anterior displacement of the limbal arcades, a visible anterior chamber and the absence of glaucoma. In complex sclerocornea the eye is microphthalmic with both glaucoma and cataract. While total sclerocornea presents with keratolenticular dysgenesis as well as anterior displacement of the arcades [3].

Secondary congenital cornea opacity has been described as including kerato-lenticular-irido dysgenesis in which the lens fails to separate from the cornea. This occurs during development and is thought to be related to defects in the Fox E3 chromosome. There is a form of mechanical secondary congenital cornea opacity in which the lens fails to form subsequent to its separation from the cornea, called primary aphakia. Other forms of secondary opacity are primary infantile glaucoma in which elevated pressure is the underlying cause of the opacity, aniridia, and Axenfield-Rieger disease. It is clear that determining the specific events underlying these conditions will require a significant amount of further investigation.

Over the past two decades changes in the design and utilization of the Boston 1 keratoprosthesis have combined to produce the observed significant improvements in device retention and reduction in complications [6]. These advances include the exchange of the diseased cornea with new donor tissue, the fenestration of the back plate, the placement of a bandage contact lens and the use of prophylactic antibiotics [7,8]. In addition, there is more attention to the prospect of elevated intraocular pressure and the recognition that glaucomatous disease when associated with keratoprosthesis is an aggressive condition which must be avoided if possible and treated vigorously when suspected. This usually takes the form of implantation of an aqueous shunt prior to prosthesis implantation, during the course of implantation, or following implantation as a secondary procedure.

There are a number of intrinsic and extrinsic impediments which must be considered prior to undertaking the implantation of a keratoprosthesis in infants and young children. The eyeball is small and general anesthesia requires the services of specially trained anesthesiologists. A vigorous immune/inflammatory response is characteristic so that even less invasive procedures such as penetrating keratoplasty are associated with a high rate of postoperative complications. Many of these eyes present with multiple pathology such as cornea opacity, cataract, glaucoma, anterior chamber dysgenesis as well as systemic comorbidity. Posterior segment pathology is not infrequent as well. Follow-up examinations require general anesthesia. And even simple tasks of instilling eye drops can be difficult for the parents to achieve. The recommended bandage contact lens may be difficult to maintain. A partial lateral tarsorrhaphy may help and wearing of moist chamber goggles will support hydration of the contact and thus assist retention. In the absence of a contact lens the goggles assist in maintaining hydration of the ocular surface and thus in preventing melting of the donor cornea and retraction of the conjunctiva [9].

Our initial efforts always utilized a post-operative antibiotic regimen of 14 mg/cc Vancomycin in combination with a fourth generation fluoroquinolone. Due to the fact that this strength of vancomycin requires that it be compounded in a hospital pharmacy combined with the prospect for antibiotic resistance current practice has substituted the use of Polytrim in place of the vancomycin. Thus, most have adopted a regimen of fluoroquinolone plus Polytrim. Nevertheless, a prophylactic antibiotic regime must be maintained indefinitely. In more than one instance delays in the availability of specifically compounded antibiotics has resulted in endophthalmitis [10]. Recently there has been some thought to the substitution of topical disinfection agents for the antibiotic regimen. We have recently incorporated a technique of covering the prosthesis with a thick conjunctival flap [11]. This reduces the incidence of melting of the donor cornea. There are those who feel the procedure should not be utilized in infants in view of the level of postoperative support necessary and the high rate of complications [12]. The fact that a multidisciplinary approach is necessary,

involving a number of ophthalmic specialists and support staff, is an impediment to the development of sites capable of providing the necessary resources. The rare occurrences of congenital cornea opacity limit the potential for interested ophthalmic surgeons to gain experience in the required techniques.

Factors favorable to the adoption of the procedure in infants include the almost immediate improvement in vision which is associated with numerous developmental advantages, even if the underlying improvement in vision were to prove to be of limited duration [13]. Thus, the critical importance of parental understanding of the associated difficulties and risks involved [13,14,15]. The infant population is noted for a strong immune response which is thought to be associated with the increased risk of postoperative melting and other inflammatory sequelae. In adults with preexisting severe ocular surface disease immunosuppression combined with total closure of a thick conjunctival flap for the first few weeks is thought to be effective. However, immunosuppression of infants is currently viewed as presenting an unacceptable risk. In the final analysis there are multiple genetic, ethical, developmental, and administrative aspects at work which have produced a variety of opinions. All of this aside from the devices technical and surgical aspects which must be considered. Yet we firmly believe in this initiative to provide functional acuity in cases where permanent blindness is the alternative.

References

1. DeVoe AG (1977) Keratoprosthesis. History, techniques, and indications. *Trans Sect Ophthalmol Am Acad Ophthalmol Otolaryngol* 83(2): 249-251.
2. Harris JK, Rao GN, Aquavella JV, Lohman LE (1948) Keratoprosthesis: technique and instrumentation. *Ann Ophthalmol* 16(5): 481-484.
3. Nouri M, Terada H, Alfonso EC, Foster CS, Durand ML, et al. (2001) Endophthalmitis after keratoprosthesis: incidence, bacterial causes, and risk factors. *Arch Ophthalmol* 119(4): 484-489.
4. Trief D, Marquezan MC, Rapuano CJ, Prescott CR (2017) Pediatric corneal transplants. *Curr Opin Ophthalmol* 28(5): 477-484.
5. World Health Organization Fact Sheet 273.
6. Fung SSM, Jabbar S, Harissi Dagher M, Tan RRG, Hamel P, et al. (2018) Visual Outcomes and Complications of Type I Boston Keratoprosthesis in Children: A Retrospective Multicenter Study and Literature Review. *Ophthalmology* 125(2): 153-160.
7. Gollamudi SR, Traboulsi EI, Chamon W, Stark WJ, Maumenee IH (1994) Visual outcome after surgery for Peters' anomaly. *Ophthalmic Genet* 15(1): 31-35.
8. Dohlman CH, Dudenhofer EJ, Khan BF, Morneault S (2002) Protection of the ocular surface after keratoprosthesis surgery: the role of soft contact lenses. *CLAO J* 28(2): 72-74.
9. Aquavella J, Wozniak R (2018) Benefits of Multidisciplinary Team Approach to Infant Keratoprosthesis. *Int J Ophthalmol Res* 2(1): 008-010.
10. Ramchandran RS, Diloreto DA, Chung MM, Kleinman DM, Plotnik RP, et al. (2012) Infectious endophthalmitis in adult eyes receiving Boston type I keratoprosthesis. *Ophthalmology* 119(4): 674-681.
11. Nass R, Frank Y (2010) *Cognitive and Behavioral Manifestations of Pediatric Disease*. Oxford University Press.

12. Fung SSM, Jabbour S, Harissi Dagher M, Tan RRG, Hamel P, et al. (2018) Visual Outcomes and Complications of Type I Boston Keratoprosthesis in Children: A Retrospective Multicenter Study and Literature Review. *Ophthalmology* 125(2): 153-160.
13. Payrau P, Moczar M, Moczar E (1969) Preparation of nonturgescible corneal transplants: initial surgical results. *Bull Soc Ophthalmol*.
14. Pasqualotto A, Proulx MJ (2012) The role of visual experience for the neural basis of spatial cognition. *Neurosci Biobehav Rev* 36(4): 1179-1187.
15. Orzolek Kronner C (2006) The impact of sight loss in social work practice. *J Soc Work Disabil Rehabil* 6(1-2): 157-177.

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