International Journal of Ophthalmology & Vision Research

Research Article

Benefits of Multidisciplinary Team Approach to Infant Keratoprosthesis

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Submitted: 22 June 2018; **Approved:** 05 July 2018; **Published:** 06 July 2018

Cite this article: Aquavella JV, Wozniak R. Benefits of Multidisciplinary Team Approach to Infant Keratoprosthesis. Int J Ophthal Vision Res. 2018;2(1): 008-010.

INTRODUCTION

Corneal opacities in infants and children pose unique management challenges. Penetrating Keratoplasty (PKP) has been used in order to clear the visual axis and prevent amblyopia, but has been historically associated with high rates of graft failure and other complications [1,2]. While there has been improving success of PKP in children following modifications in surgical technique, post-operative management and patient selection [3-5], there still remains a significant need for supplemental approaches to promote visual rehabilitation in this most vulnerable population, particularly in the cohort of infants and where PKP would be doomed to failure. Keratoprosthesis (KPro) implantation has become an important alternative to PKP in adult patients to treat a wide variety of severe corneal pathologies [6-8] and are appropriately performed by a solitary cornea surgeon.

The Infant Eye

However infants constitute a vastly different category of both subject and disease. The underlying biology and physiology systems are evolving and are little understood; the full ocular expression of genetic anomalies may result in dysgenesis with altered anatomy and malfunction of numerous ocular systems; active immune systems have a propensity for inflammatory sequelae; there is a reduced size of the globe and the ever presence of amblyopia potential.

Treatment Strategies

While the article by Fung et al, highlight the serious risks and complications associated with pediatric KPro implantation when performed under routine conditions, we believe that long term success with respect to device retention, visual acuity and complication rate can be achieved with an appropriate standardized surgical technique, post-operative management and most importantly, a multi-disciplinary dedicated team approach [9-12].

The team concept includes ophthalmic subspecialists in cornea, pediatrics, infant glaucoma, oculoplastic, vitreoretinal disease, and pediatric anesthesiologists. In addition to physicians, secondary support must involve nursing, operating room technicians, clinical technicians, surgical schedulers, program coordinators, and a variety of administrative support personal. The focused human and fiscal resources that must be assembled are difficult and complex to achieve for any institution. Yet are imperative if one is to provide an opportunity for visual rehabilitation in this population. Through a dedicated and coordinated effort within the keratoprosthesis team, all aspects of a child's visual development can be appropriately addressed and managed, providing the greatest potential for success [10-12].

METHODS

One of the first challenges is the decision to accept new patients. Given the low incidence of congenital as well as secondary cornea opacification and the very limited number of facilities electing to do this work, patients are often referred from distant locations within the USA as well as internationally. One single intake group must be organized to collect all data obtained by fax, telephone, email, and internet communications in the form of physician and hospital records as well direct communication from families. While the mother may be the first to notice the opaque ocular surface a variety of neonatologists, pediatricians, ophthalmologists and ophthalmic subspecialists may have conducted examinations and testing often under anesthesia. This data must be collected, collated and distributed to all team members who, following independent review, meet together on a regular basis to determine if a consultation is indicated. Communicators are in contact with the parents to explain the process; often physicians are available to answer specific questions by telephone, or email.

Once the decision has been made to arrange a consultation, travel arrangements must be in place, which often involve lodging, airline, visa, passports or exit and entry permits. The initial visit includes both an office visit followed by an examination under anesthesia with all necessary physician team members. At a minimum, this process requires physician evaluations from the cornea, retina and pediatric services as well as ancillary testing such as A- and B-scans. Based on the results of a complete examination, the physician team reconvenes to discuss the findings and appropriate next steps, which are then relayed to parents [10-12]. As with pediatric PKP, it is our experience that successful outcomes of pediatric keratoprosthesis are dependent on the severity of presenting disease. Peter's Anomaly, encompasses a spectrum of anterior dysgenesis manifestations that can range from mild corneal involvement to severe opacity, combined with malformation of iris, angle and lens. Co-existing glaucoma is also frequently encountered, as is posterior segment pathology. Furthermore, many patients have already undergone prior failed surgeries, further complicating the ocular anatomy and overall resilience. We select only cases not amendable to PKP. On occasion our decision is that nothing can be done (posterior dysgenesis, funnel detachment, intractable glaucoma, microphthalmos). Even if the team agrees that there is potential for useful vision, cases are not accepted unless there is evidence that the family is prepared to make the long term commitments necessary (travel, Exams under Anesthesia (EUAs), medication instillation, physician visits, expenses, etc.). In addition, for those patients who are not local to our institution, we require the prior agreement of local specialists to assist in monitoring the child and communicating with us as indicated. If there is lack of dedication to these principals no intervention is advised. Prior authorization with insurance companies must also be established. Our physicians accept all cases regardless of financial considerations, but institutional charges must be negotiated and in place.

Thus patient selection is critical when considering infant keratoprosthesis implantation. It has been our experience that disease without an autoimmune component (i.e. Peter's anomaly) portends improved long term success, with some patients retaining the device successfully with useful vision for over a decade (manuscript in preparation). However, the decision to move forward with a KPro can often be complex and difficult, thus we also provide the opportunity for multiple visits around the first examination under anesthesia in order to answer all questions and establish realistic expectations.

Indications for infant keratoprosthesis include PKP failure, anterior chamber dysgenesis, severe opacification, evidence of light perception and an anatomically well positioned retina. We have utilized the Boston type 1 for all pediatric procedures since 2003.

Surgical Procedure

The recommended minimal operative procedure consists of mobilization of conjunctiva and tenon's capsule posterior to rectus muscle insertions for 360 degrees, a 7-8 mm diameter cornea excision, extracapsular cataract extraction, and lysis of extensive anterior and posterior synechiae prior to suturing the KPro in place. We routinely use irradiated corneal donor tissue, and use interrupted 9-0 nylon sutures to secure the device in place. Once the KPro is implanted,

our vitreoretinal surgeons, who are comfortable working through the 3.2 mm KPro optic, perform a full pars plana vitrectomy as well as 360 degree retina periphery inspection and laser reinforcement when necessary. Implantation of an aqueous shunt (Glaucoma Drainage Device (GDD) by the pediatric glaucoma service is often performed as well. Once the vitrectomy is complete, the cornea surgeon returns to close the conjunctiva. We suggest a near total closure of Tenon's and conjunctiva over the optic, leaving a 1-2mm opening centrally. The conjunctiva will spontaneously retract in the majority of cases to the edge of the optic, providing excellent coverage over the corneal donor tissue. All patients receive a Kontour contact lens at the end of the case [11,12].

Post-operatively, we require extremely close follow up as well as intensive topical medical therapy including vancomycin, fluoroquinolone, and prednisolone. For all patients, compliance must be monitored, reinforced, and documented. Data must be collected from outside examinations and transcribed to the medical record. In order to provide the maximal chance of visual rehabilitation, patients are closely monitored for amblyopia, strabismus, glaucoma and retinal pathology. While gross refractive error may be estimated on the basis of axial length variations, confirmation with retinoscopy is performed and supplemented with automated devices when feasible. In our first series of 22 eyes from 17 patients aged 1.5-136 months, visual acuity ranged from light perception to 20/30 in the subset of verbal children, and all remaining infants demonstrated the ability to follow light, fingers and objects [12]. While visual outcomes can be variable, we continue to follow a subset of our original cohort who have retained the device for over 10 years with continued functional vision (manuscript in preparation). All data is collected under a standard IRB approved protocol.

DISCUSSION

Complications encountered vary from retroprosthetic membranes, glaucoma, retinal detachment, corneal melts and serious infections. In our initial work, retroprosthetic membranes formed in 5 eyes, additional surgery was required for pressure control in 3 patients and retinal detachment found in 2 patients [12]. However, we have now found that close monitoring of each patient allows for early intervention in order to avoid and minimize the consequences of complications. Surgical techniques such as the full conjunctival flap and full pars plana vitrectomy have in our experience reduced corneal melting and membrane formation, respectively. Additionally, implantation of a GDD at the time of initial surgery serves to temper pressure fluctuations and damage from glaucoma, thus the majority of children will receive GDD at the time of keratoprosthesis surgery. In general, unscheduled visits with availability of appropriate team members, must be accommodated to address all potential complications. All patients are provided with a dedicated keratoprosthesis phone number and email address in order to facilitate communication and planning. On occasion a simple scheduled EUA determines that extensive repairs are indicated so a 30 minute procedure becomes a several hour complex repair, involving both anterior and posterior segment specialists.

CONCLUSION

In the final analysis, the provision of useful vision is an enormous benefit during the early months of life, even if the progression of disease were to render the benefit transient. If all of the necessary management resources are not available the process is doomed to failure and should not be initiated. Even in the presence of appropriate resources the provision of useful vision remains a challenge, but one which is important to surmount. Thus the circumstances for the provision of care must be included in any description of potential benefit. And even then, the vast differences in the physiological response of individual children combined with the multiplicity of disease expression, and the fact that specific techniques and modalities are constantly being improved or developed render comparison difficult.

Not all infants and children can be given the opportunity for sight, but functional vision is an achievable goal [10-12].

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