

Controversy in the Management of Congenital Cornea Opacification



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Short Communication

The management challenges associated with congenital ocular disease in infants have long been recognized. Depending on the degree of microphthalmos, vascularized cornea opacity, iris and lens dysgenesis, buphthalmous, and glaucoma many of these infants have been declared to be irreversibly blind.

Penetrating keratoplasty has been attempted in some milder cases where the pathology is limited to the cornea in an effort to avoid subsequent amblyopia but pediatric corneal transplants are associated with a high risk of graft failure and complications [1,2]. In the more severe cases standard cornea transplantation is contraindicated.

In the adult population, since the introduction of the Boston I device, keratoprosthesis is being increasingly utilized as an important alternative to cornea transplantation as well as the newer endothelial replacement techniques. Infants however constitute a significantly different category with anatomic and biological systems evolving, active immune response, and the absence of a clear understanding of the underlying diagnosis [3].

The recent publication of a small series of pediatric keratoprosthesis procedures performed by a single surgeon under standard conditions highlighted serious and irreversible complications with the authors' conclusion that the procedure should not be performed [4].

Yet one group has reported a level of visual restoration utilizing a combination of multidisciplinary approach, specialized surgical techniques, intense preoperative evaluation and a comprehensive postoperative management style [5-7]. While we await their long term results, it is clear that a useful level of acuity can be preserved in some cases. In view of these diverse opinions the question has been raised: should this work continue?

It is clear that the provision of some level of vision, even if transient, is associated with a significant positive impact on infant development. The Texas School for the Blind and Visually Impaired has categorized these beneficial effects

in areas of sensory development, motor development, self-concept, cognitive development, social development, language development, and experience [8].

The early visual experience in newborn infants has been studied and shown to have an important role in the development of spatial cognition [9]. Severe visual impairment however has a profound negative effect on the development of bilateral coordination. And childhood blindness can be said to have a negative effect on growth, development, social and economic opportunities [10].

What then are the negatives? Since these cases are rare and the techniques specialized families must travel long distances to centers where care is available, cases must be screened for the parents ability to maintain years of frequent examinations (many under anesthesia), they must have the means of providing the intense postoperative management. Drops must be instilled frequently at first but some are necessary for life, local eye physicians must be available for consultation, and a variety of potential complications must be avoided, or if present recognized and treated.

Should the children of parents committed to providing this necessary intensive care be denied not only the potential for useful vision but the positive effects on development as well? Should we not expect that over time those pioneers who provide care to this population will become more proficient and that new discoveries and improvements will result in a reduction of complications as well as an increase in the number of providers willing to participate?

With advances in medicine and science we anticipate that a larger percentage of these unfortunate cases will become amenable to improved visual restoration and preservation techniques based on advances in the currently available technology.

Thus in the final analysis, despite the recently reported experiences of our colleagues, and in view of our strong belief

that significantly different preoperative, operative, and post-operative approaches do result in the provision of useful vision, the continued treatment of infants afflicted with severe congenital corneal opacity must continue.

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