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Diagnosis and Treatment of Congenital Cornea Opacity

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ABSTRACT

Congenital cornea opacity is a rare occurrence which results not only in blindness but which are associated with numerous related developmental challenges. Obtaining a specific clinical diagnosis has been difficult and most cases were attributed to generic categories of Peters Anomaly and Sclerocornea. Much progress has been made recently in identifying specific genetic defects which occur during the complex embryonic interactions resulting in the development of the infant eye. Traditional therapy has involved attempts to minimize amblyopia, optical iridectomy where feasible, therapy of associated glaucoma and cornea transplantation. The active infant immune system causing allograft rejection has been considered an impediment to cornea transplants in most cases. The small size of the globe contributes to technical surgical difficulties as well as a variety of post-operative complications.

Significant improvements in the design and implantation techniques of the keratoprosthesis have occurred over the past decade. The success of the Boston 1 device in treating otherwise inoperable cornea pathology was the stimulus for one group to begin implantation of these devices in infants afflicted with congenital cornea opacity. While this team has developed what they feel is a successful approach the procedure is considered controversial. Two publications report poor results when employing the traditional approach conducted by a single cornea surgeon as utilized in adults. They maintain that the technique should not be undertaken.

The institution advocating the procedure has developed a multidisciplinary team approach which it feels is essential to success in this infant population. Cornea, vitreo-retinal, glaucoma and oculo-plastic surgeons work together in evaluating prospective cases, performing the surgery, and administering post-operative care. They are supported by a dedicated group of nurses and technicians and a comprehensive administrative staff. Complications must be anticipated and aggressively addressed. Not all cases can be successful but it is possible to achieve long term results and the prospects for improvements in devices and availability of new drugs are incentives to continue this important work.

Keywords: Cornea opacity, Infant, Keratoprosthesis, Ocular embryology

INTRODUCTION

Congenital cornea opacity in infants and young children pose unique challenges with respect to both diagnosis and treatment. Blindness which is present during early infancy has been shown to have wide reaching implications [1-4]. Hearing is the only distance sense available in these instances. Sound in order to be useful requires tactile and motor interaction. Blind children require he presence of sighted individuals to assist them mediate with their environment. Subsequent sensory, motor, cognitive, social and language development are all adversely affected in congenital cornea opacity. Thus the provision of vision during infancy, even it was to be lost due to complications or disease progression, is an enormous benefit. The prevention of childhood blindness is an important aspect of the World Health Organization Declaration to eliminate preventable blindness [4].

Congenital Cornea opacity is rare, estimated to occur in approximately 2 per 100,000 births [4]. In the past the diagnosis has been characterized by clinical appearance such as Peters Anomaly or Sclerocornea [5-7]. A more complete clinical distinction recognizes etiologies of glaucoma, infection, congenital endothelial dystrophy, forceps delivery and metabolic aberrations. The work of Kenneth Nischal has been instrumental in our appreciation that the embryonic formation of the cornea and anterior chamber constitute a complex series of genetically controlled events [8]. We now distinguish both primary and secondary (mechanical) events

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relating to neonatal opacification of the cornea.

The initial recognition of a severe visual problem is most often made by the mother who becomes aware that normal iris coloration has been replaced by a white opacity. This is usually followed by a number of medical examinations involving, neonatologists, ophthalmologists, pediatric ophthalmologists and ultimately cornea surgeons. In mild or unilateral cases patching the good eye, creating a larger pupillary opening may be of some benefit. In cases of cornea opacity in the absence of other developmental anomalies (dysgenesis) [8] a cornea transplant may be recommended. While this procedure is associated with a high risk of subsequent graft failure as a result of the intense immune response present in infants and young children [9-11] many feel a single transplant should be attempted in these instances. It is important to realize that even a clear graft will not afford ambulatory vision if appropriate postoperative rehabilitation is not provided. In addition the integrity of the macula and the optic nerve must enable transmission of impulses to a healthy occipital cortex. However functional acuity with the ability to independently play and eat does not require demonstrated Snellen related competence. Other anomalies may present additional challenges to visual function regardless of the success achieved in removing the cornea opacity.

Initial indications for keratoprosthesis were limited to irreversibly blind adults with bilateral disease considered inoperable for standard cornea replacement techniques [12-14]. The introduction of the Boston 1 device dramatically altered the potential for intervention in adults [15-18]. The Flaum group in consideration of the success of this new device in adults in combination with the unfavorable prognosis for cornea transplantation in infants began consideration of keratoprosthesis surgery in this population [19]. The obstacles were formidable. The inflammatory response, technical difficulties in dealing with a small eye, dysgenesis, multiple compromised ocular systems and the ever present danger of amblyopia constitute significant impediments. The introduction of this technique for infants has been met with controversy [20,21]. One group of cornea surgeons has reported dismal results in 11 cases divided between 3 centers. They reported 5 cases with no light perception and complications including keratitis, endophthalmitis, glaucoma, device extrusion and retinal detachment. The conclusion was that this procedure should not be attempted [20].

The Flaum team has utilized a significantly different team approach rather than a procedure performed by a single cornea surgeon. They maintain that a multidisciplinary team of cornea, pediatric, glaucoma, vitreo-retinal and ocular plastic ophthalmic surgeons working with dedicated administrative, clinical and surgical support staff can achieve reasonable results [19]. While these focused human and fiscal resources are difficult to achieve in any institutional setting they are imperative if one is to deal with the many preoperative, operative and post-operative challenges. While complications do occur, some can be avoided, but all must be vigorously addressed in a timely fashion. They advocate lens extraction, parsplana vitrectomy and insertion of an aqueous shunt at initial surgery. Bandage contact lenses must remain in place and not infrequently require a partial lateral tarrsoraphy [19]. Communication with caregivers and referring physicians must be maintained so compliance with post-operative routines are insured, and frequent follow-up is essential. A thick conjunctiva flap can be fashioned at surgery to reduce surface melting [22]. Postoperatively prophylactic antibiotics and anti-inflammatory agents are essential [23]. Topical steroids as well as the recently available TNF alpha inhibitors are routine [24]. It is important to keep the bandage lens well hydrated and often moist chamber goggles are prescribed. Follow-up of one month, three months and after the first year every six months must be maintained.

Complications encountered include surface melting and retro prosthetic membrane [25,26]. Intraocular pressure must be monitored to avoid glaucoma; ultrasound examinations can detect retinal detachment which is an ever present danger. Thinning of donor tissue and retraction of the conjunctiva must be addressed with surgical repair as soon as they are noticed. Endophthalmitis in infants is related to noncompliance with antibiotic regimes [27]. The necessity of compounding antibiotics is a burden for care givers. To address this Flaum and others have reduced the dependence on compounded vancomycin and substituted Polytrim (trimethoprim/bacitracin available in commercial pharmacies).

Not all are candidates for this surgery. Micophthalmic eyes, Stevens Johnson and herpes simplex should not be attempted. Parents must be prepared for management over a long term so an informed consent understanding is an essential prerequisite. Arrangements must be made in advance for local ophthalmologists to monitor progress and communicate with the surgical team. A dedicated nurse coordinator is essential in maintaining communications with all.

In the final analysis functional acuity is an enormous benefit to future development and the capacity to provide this opportunity exists today and will undoubtedly improve in the future with the development of improved diagnostic techniques, devices, surgical procedures and drugs. It has been suggested that a means to control the immune inflammatory response is required but existent drugs have not been proven safe for utilization in infants and young children.

The importance of accurate diagnosis has been confirmed by a study indicating that eighty percent of these individuals with congenital disease retained their prosthesis while only 50% with acquired opacity retained the initial device. 44%

of those with congenital disease demonstrated improved vision while this dropped to 10% in those with acquired disease [22]. The convention of reporting keratoprosthesis results as short term less than 2 years, intermediate term from 2 to 5 years and long term 5 years or more [28]. It is to be noted that the Flaum group reports a cadre in which the device has been retain for periods of 10 to 17 years.

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