Keratoplasty in Infants and Children

An Essay Submitted for partial fulfillment of M.Sc. degree in Ophthalmology

by

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Abstract

Corneal transplantation in a pediatric patient population poses special challenges that are not encountered in adults. The surgical procedure is technically more complex because of the small size of the globe, reduced rigidity of the cornea and sclera, and positive vitreous pressure with frequent anterior displacement of the lens-iris diaphragm. Severe inflammatory reaction, secondary glaucoma and immunological graft rejection are also much more likely to occur after surgery. Postoperative visual rehabilitation and amblyopia therapy are crucial but complex in a pediatric patient population. The combination of a dedicated multispeciality transplant team and the education and cooperation of the patient's family are imperative for a successful outcome.

Key Words:

Indications, Preoperative Evaluation, Surgery, Postoperative Care, Postoperative Complications, Optical Correction and Amblyopia Management, Visual outcome and five-year view.

بسم الله الرحمن الرحيم

ربنا لا تؤاخذنا إن نسينا أو أخطأنا
ربنا ولا تحمل علينا إصرا كما حملته
علي الذين من قبلنا ربنا ولا تحملنا
ما لا طاقة لنا به و اعف عنا واغفر
لنا وارحمنا أنت مولانا فانصرنا
علي القوم الكافرين

(آية ٢٨٦ سورة البقرة)

To…

my parents

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List of abbreviations

ACE	Aphakic corneal edema.
AK	Astigmatic keratotomy
ALK	Anterior lamellar keratoplasty
BM	Bowman's membrane
BSS	Balanced salt solution.
CCO	Congenital corneal opacification
CHED	Congenital hereditary endothelial dystrophy
CHSD	Congenital hereditary stromal dystrophy
CL	Contact lens
DALK	Deep anterior lamellar keratoplasty
DLEK	Deep lamellar endothelial keratoplasty
DM	Descemet's membrane
DMEK	Descemet's membrane endothelial keratoplasty
DSEK	Descemet's stripping with endothelial keratoplasty
FSEK	Femtosecond enabled keratoplasty
HLA	Human leukocyte antigen
ICE	Iridocorneal Endothelial
ICG	Indocyanine green
Intacs	Micro-Thin Prescription Inserts
IOL	Intraocular lens
IOP	Intraocular pressure
IV	Intravenous
KC	Keratoconus
KILD	Kerato-irido-lenticular dysgenesis
KP	Keratoplasty
LASIK	Laser in situ keratomileusis.
LK	Lamellar keratoplasty
LRK	Lamellar refractive keratoplasty
MPS	Mucopolysaccharidoses
OAV	Oculo-auriculo-vertebral
PA	Peters anomaly
PAS	Peripheral anterior synechiae
PCE	Pseudophakic corneal edema
PCG	Primary congenital glaucoma
PEDIG	Pediatric eye disease investigator group
PK	Penetrating keratoplasty
PKG	Post keratoplasty glaucoma

PLK	Posterior lamellar keratoplasty
PPMD	Posterior polymorphous dystrophy
PRK	Photorefractive keratectomy
RK	Radial keratotomy
UBM	Ultrasound biomicroscopy
UVA	Ultra violet A
VA	Visual Acuity
VEGF	Vascular endothelial growth factor
VKC	Vernal keratoconjunctivitis

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Introduction

Corneal blindness remains a leading cause of childhood visual impairment, particularly in developing countries **(Zeidan et al., 2007 & Muhit et al., 2007)**.

Providing a clear visual axis during the critical period of visual system maturation is essential to prevent amblyopia that could otherwise unfavorably impact a child's visual development. Although once contraindicated in children, penetrating keratoplasty is now the treatment of choice for a carefully selected group of infants and children with corneal opacities due to a variety of etiologies **(Sharma et al., 2007)**.

Pediatric keratoplasty differs from adult keratoplasty in many aspects. In pediatric grafts the surgery is technically more difficult, postoperative care is more challenging, timing of suture removal is controversial and graft rejection is more common. Furthermore, amblyopia is a major concern in pediatric patients, adversely affecting the visual prognosis (**Cosar et al., 2003)**.

Over the past two decades, a better understanding of wound healing of corneal transplants in infants and children and modifications of surgical techniques have contributed to a substantial improvement in the prognosis of pediatric penetrating keratoplasty (PK) **(Brown et al., 1983, Stulting et al., 1984 & Dana et al., 1995)**.

Acquired corneal scars, corneal decompensation, older children, and phakic eyes had the best prognosis. Corneal perforations, active inflammation or infection, and infants with multiple ocular anomalies had the poorest prognosis. Children undergoing combined procedures did less well than those undergoing a single- or two-staged procedure **(Cowden et al., 1990)**.

The indications are classified into three groups: congenital, acquired non-traumatic and acquired traumatic. Peters' anomaly, keratoconus, herpes simplex keratitis, penetrating trauma and congenital dystrophies are the most common **(Waring et al., 1977 & Patel et al., 2005)**.

Preoperative assessment of pediatric patients includes examination under anesthesia, observation with slit lamp, intraocular pressure measurement, gonioscopy, indirect ophthalmoscopy if the media are clear and electrophysiological studies as well as B scan ultrasonography if there is media opacity. Visual acuity is measured by Teller acuity cards in the pre-verbal child, Sheridan-Gardner single letter matching, Kay picture cards or tumbling E charts in young children and Snellen chart in older children **(McClellan et al., 2003)**.

At each postoperative visit, visual acuity is measured and slit lamp biomicroscopy is performed to assess the condition of the graft. Intraocular pressure is measured by either Goldmann applanation or Schiotz tonometry. Cycloplegic refraction is used to assess all postoperative refractive errors. Any sign of graft rejection is treated promptly with high dose topical steroids and in some cases, a short course of oral steroids occasionally subconjunctival or sub-tenon's depot steroids

are used to manage rejection **(McClellan et al., 2003)**. http://bjo.bmj.com/content/81/12/1064.full - fn-1

Graft failure following PKP is very common and usually occurs in the first year. The poorer prognosis for sclerocornea is probably related to the higher incidence of major anatomical alterations in this condition. Angle anomalies resulting in primary glaucoma and often buphthalmos occurs in almost half of the eyes with sclerocornea. Complications in eyes with primary glaucoma (because those usually have thinner corneas) are from stretching of the eye. The surgical procedure in most eyes is complicated by a phenomenon almost unique to infants that is massive outpouring of fibrin upon entering the anterior chamber, promoting broad anterior synechiae, resulting in postoperative glaucoma. The management of postoperative glaucoma is unsatisfactory since medical treatment frequently fails. When surgery is required to lower the intraocular pressure, cyclocryotherapy is the preferred procedure in this patient group **(Frueh et al., 1997)**.

Sutures in a vascularised infant cornea incite an inflammatory reaction which will lead to graft rejection unless the sutures are removed in the first postoperative weeks. This inflammatory response to sutures is more frequent in eyes with the oval shaped corneas of sclerocorneas where the suture tracks approximated or run through the conjunctiva **(Brown et al., 1983)**.

Finally penetrating keratoplasty for infants and children has the potential for excellent long term results and should be performed in the first weeks of life in both bilateral and unilateral cases **(Frueh et al., 1997)**.

Aim of the Work

The aim of the work is to highlight the importance of this special and peculiar surgical technique in such critical age group of patients with its different precautions and modifications. This will include: indications, preoperative evaluation, timing of surgery, surgical technique as well as postoperative follow up.

Chapter 1

Indications

Overview

Nowadays, keratoplasty is considered one of the world's most widely practiced human organ and tissue transplantation methods. Compared with the adult's cornea, the child's cornea is thinner; less rigid and more elastic, presenting a wide range of challenges pre-, intra- and post-operatively. Both rejections and complications are more commonly seen in pediatric keratoplasty, leading to a higher failure rate. Prior to 1970, corneal transplantation was considered to be contraindicated because of a less firm sclera and the forward displacement of the lens-iris diaphragm in pediatric anterior segment surgery, but with the improvement of surgery techniques in the last 20 years, the success rate of pediatric keratoplasty has increased tremendously **(Stulting et al., 1984, Dana et al., 1995 & Aasuri et al., 2000)**.

Indications for pediatric penetrating keratoplasty can be classified under three broad categories: congenital, acquired non-traumatic and acquired traumatic corneal opacities. Congenital opacification of the cornea can result from neurocrestopathies or mesodermal dysgeneses, such as Peters' anomaly, sclerocornea, anterior staphyloma/keratectasia and infantile glaucoma. Other congenital conditions include corneal dystrophies, mucolipidoses, mucopolysaccharidoses and congenital corneal tumors. Most acquired non-traumatic corneal opacities are caused by infectious keratitis and keratoconus, while penetrating or blunt trauma,