Penetrating Keratoplasty in Children

Ken K Nischal MD, FAAP, FRCOphth
Director, Pediatric Ophthalmology, Strabismus and Adult Motility
Childrens Hospital of Pittsburgh USA
Professor of Ophthalmology
School of Medicine, Pittsburgh University
Vice Chair
Department of Ophthalmology, UPMC
Associate Medical Director
UPMC International

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Pediatric Keratoplasty in 30 Minutes

- Indications
- Penetrating Keratoplasty
- Deep Anterior Lamellar Keratoplasty
- Endothelial Keratoplasty
- Alternatives

Indications

- Tectonic
- Therapeutic
  - PKP
    - Congenital corneal opacification
    - Endothelial dystrophies in newborns
    - Full thickness scarring
  - ALK
    - Scarring (superficial)
    - Anterior stromal dystrophies
  - DALK
    - Stromal dystrophies
    - Mucopolysaccharidoses
    - Keratoconus
  - Endothelial Keratoplasty
    - Endothelial dystrophies
    - Endothelial cell failure
Peters Anomaly/Sclerocornea—Bad Terms?!

- Prefer Anterior Segment Developmental Anomaly
- Why?
- Clinico-pathologic Correlation of Congenital Corneal Opacification using UBM

### PRIMARY CORNEAL DISEASE

#### A DEVELOPMENTAL ANOMALIES OF CORNEA

- **i. Corneal dystrophies**
  - CHED, PPMD, CHSD, X-L ECD

- **ii. Cornea Plana (sclerocornea)**

- **iii. Corneal structural defects due to dermoids**
  - Isolated
  - Part of systemic disease e.g. Goldenhar, Linear sebaceous nevus

- **iv. CYP1B1 Cytopathy**

### SECONDARY CORNEAL DISEASE

#### A DEVELOPMENTAL ANOMALIES OF ANTERIOR SEGMENT

- **i. Kerato-Irido-Lenticular Dysgenesis KILD**
  - a. Iridocorneal adhesions only
  - b. Lens fails to separate from cornea
  - c. Mechanical e.g. due to PHPV
  - d. Lens separates but fails to form thereafter
  - e. Lens fails to form

- **ii. Irido-trabecular Dysgenesis**
  - a. Infantile glaucoma
  - b. Axenfeld-Rieger anomaly
  - c. Aniridia

### SECONDARY CORNEAL DISEASE CONT’D

#### B ACQUIRED CORNEAL DISEASE

- **i. Metabolic**
  - Mucolipidoses
  - Mucopolysaccharidoses
  - Cystinosis

- **ii. Trauma**

- **iii. Infectious Keratitis**
  - Bacterial
  - Viral
  - Protozoal
  - Fungal

- **iv. Non-infectious Keratitis**
  - Exposure
  - Neurotrophic
  - Interstitial e.g. Cogan’s disease
  - Steven-Johnson syndrome
  - Epidermolysis bullosa

- **v. Miscellaneous**
  - Keratoconus
  - Keratoglobus
  - Xeroderma pigmentosa
  - Endothelial failure

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**Intrasurgical Volumetric Imaging**

![Multiple adhesions (AAJ-174)](image)
Management Strategy

- Indications
- Pre-operative Assessment
- Operative Technique
- Postoperative Management
- Complications
- Alternatives

Child presents with Congenital Corneal Opacification

OCULAR

OPA + VEP/ERG + post USS + UBM [If possible at OPA]
+ Photograph

SYSTEMIC

geneticist

MRI

Develop. Paed.

Not possible awake

EUA + UBM

No Surgery

No Intervention

CLT

Surgical RX or Cyclodiode Laser
Under UBM Guidance

Surgical RX

No Corneal Tx

CLT

CLT

Visual Impairment Service

Glaucoma

Optical Iridectomy

Visual Rehabilitation
[CL + Glasses]

Corneal Tx

CLT

Histology

Removal of Sutures

Immuno.

GA

Trabeculectomy with Trabeculotomy with MMC or Ahmed Tube Implant with Scleral Donor with MMC
Pre-op assessment

Operative Technique for PKP

- Ehrlich, Rootman, Morin 1991 Can J Ophthalmol
- Flieringa ring - ESSENTIAL
- For PA / sclerocornea
  - oversize by 1mm
  - 4 x iridectomies
- Remove sutures in neonates by 6 weeks
What is Success?

Vision = 6/21 with CL

Vision = 6/180 with glasses

What is success?
What is success? Only Eye 20/2000 vision

Take Home Message

• Developmental vision more important than corneal clarity
• Peters anomaly/Sclerocornea NOT appropriate terms
• Primary corneal disease
• Secondary corneal disease
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**DALK**
Indications and Outcomes of Deep Anterior Lamellar Keratoplasty in Children

Samantha A. Harding, MRCPPh, Ken K. Ninchal, FRCPPh, Anjali Upponi-Paul, FRCS, Darren J. Pouler, MD

Purpose: To report our experience of deep anterior lamellar keratoplasty (DALK) in children.

Design: Retrospective case note review.

Participants: Nine patients (13 eyes) aged from 13 weeks to 14 years, 11 months at the Clinical and Academic Department of Ophthalmology, Great Ormond Street Hospital for Children National Health Service (NHS) Trust, London, United Kingdom.

Methods: A study of all pediatric patients undergoing DALK from February 2002 to October 2008 was undertaken. Deep anterior lamellar keratoplasty was attempted in 9 children (13 eyes); the procedure was successful in 11 eyes, and 2 eyes progressed to penetrating keratoplasty (PKP). One eye underwent repeat DALK. Preoperative examination included electroophthalmology, ultrasound biomicroscopy (UBM), and slit-lamp biomicroscopy.

Main Outcome Measures: Complications and visual acuity at last follow-up.

Results: Five patients had mucopolysaccharidoses (MPS), 3 patients had scarring presumed to be infectious, and 1 patient had keratoconus. Because of the failure of follow-up and loose sutures, 1 child with MPS had an epithelial resection, and the operation was repeated successfully. All grafts showed good graft clarity 10 to 60 months after grafting with visual acuities ranging from 0.28 to 1.0 logarithm of the minimum angle of resolution. Two children with nonspecific causes of scarring showed good visual acuities 24 to 51 months post-DALK. Two children who had conversion to PKP were lost to follow-up because they had moved abroad. In 4 of the 9 children with MPS, established techniques of DALK could not be performed because of excessive glycosaminoglycans (GAGs) in the stroma. Ultrasound biomicroscopy was used to guide trephination depth in the first instance. In 1 child with MPS, viscodissection was successfully used. All clinically diagnosed scars were histologically confirmed, and electron microscopy of corneal buttons confirmed the diagnosis in patients with MPS.

Conclusions: Deep anterior lamellar keratoplasty should be considered in children with MPS and partial-thickness scars. In MPS, viscodissection and the "big bubble" technique may not be useful if there are excessive GAGs in the stroma.

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DSAEK