

Pediatric Keratoplasty: Strategies to Optimize Outcomes

Pediatric keratoplasty can have a profound impact on a child's life. Full-thickness corneal transplantation in the pediatric population is more challenging and has a higher complication rate than in adults. Caring for children with corneal disease requires a multidisciplinary approach including the pediatrician, pediatric ophthalmologist, and cornea specialist, among others. In some complex cases, glaucoma, retina, and oculoplastics specialists may also be involved. The child's caregivers are particularly important members of this team, and they need to have realistic expectations about the outcomes, challenges, and long-term care that is required.

Careful preoperative, intraoperative, and postoperative protocols for pediatric penetrating keratoplasty (PK) are essential in reducing the risk of surgical complications in children. Here, we describe the modified surgical technique we use in young patients.

Indications

Pediatric corneal disease can be divided into congenital and acquired pathologies. In the United States, congenital corneal disease is the most common indication for pediatric PK, but in some countries, infections and scarring are more common indications.¹ It is important to consider the reason for surgery as well as associated ocular pathology when counseling the family.

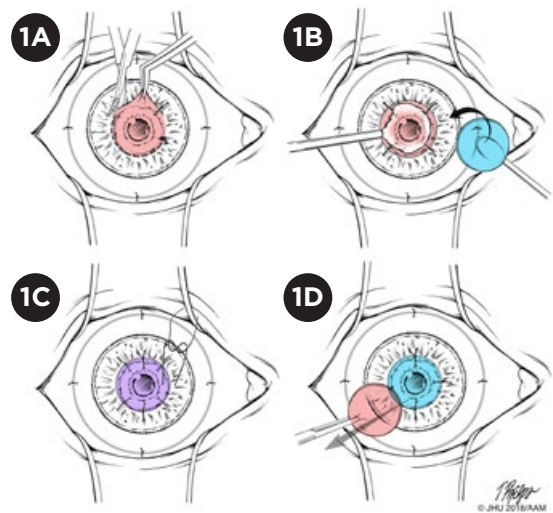
Isolated acquired corneal scarring or ectasia has the most favorable prognosis, while congenital disease, especially when associated with glaucoma, has the poorest prognosis.²

Preparation

When considering surgery, the physician should screen for any social circumstances that would preclude proper postoperative management. Caregivers need to be prepared for the extensive ophthalmic care required for pediatric transplant recipients, including frequent clinic visits, long-term eyedrop use, amblyopia treatment, and multiple trips to the OR for exams, suture removal, and possibly other surgical interventions.

It is important to discuss realistic expectations about outcomes—in particular, that vision will most likely be closer to 20/200 than 20/20—as well as the lifelong risk of transplant rejection and infection.

Evaluation. Children with congenital corneal abnormalities should undergo a systemic evaluation, and they often require genetic testing. For children with additional systemic concerns, the ophthalmologist should coordinate



SURGICAL TECHNIQUE. (1A) Suturing the host cornea (pink) back on to itself during dissection. (1B) Placing the donor cornea (blue) on a viscoelastic interface over the host cornea. (1C) Placing the cardinal sutures. (1D) Removing the host cornea only after the first three donor corneal sutures have been placed. (See text for further detail.)

with the pediatrician and other specialists to minimize the number of exams under anesthesia.

An ocular exam under anesthesia should be considered preoperatively to identify other eye abnormalities that might preclude surgical success or require additional intervention. Ultrasound biomicroscopy or anterior segment optical coherence tomography can help with surgical planning in cases with a limited view of the anterior chamber. Other assessment techniques, including B-scan, visual evoked potentials, and corneal topography, may also be helpful.

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Timing. The timing of surgery is controversial. Earlier intervention is better in terms of amblyopia management; however, the risks of general anesthesia are higher for infants, especially those under the age of 3 months. Graft survival is generally better for older children, although this may be related to differences in indication rather than to the timing of surgery.²

Surgical Technique

General anesthesia is required. Supplemental retrobulbar anesthesia may be added to reduce general anesthesia requirements and to help with early postoperative pain. Pediatric patients are more likely than adults to experience positive posterior vitreous pressure following retrobulbar injection. The use of a Honan balloon prior to surgery, pilocarpine 1% or 2%, and/or reverse Trendelenburg positioning during surgery may be helpful in reducing the risk. Preoperative intravenous mannitol may be considered, depending on the child's weight and systemic health, and should be given as an infusion over at least 15 minutes rather than as a bolus.

Following is our preferred surgical approach for PK in children (Fig. 1). It is a modification of the "Price graft-over-host" technique.³

1. Pediatric sclera is less rigid than adult sclera, and we recommend the use of a Flieringa ring to stabilize the iris-lens diaphragm. Moreover, because the sclera is thinner in children, the surgeon must be careful not to perforate the globe.
2. The donor tissue is trephined in the same manner as for an adult PK. Use of tissue from donors over the age of 4 years is recommended, as tissue from younger donors is more difficult to manage during surgery.
3. We routinely perform at least one peripheral iridotomy because children are more likely than adults to develop postoperative angle closure.
4. We mark the center of the cornea and use an eight-pronged radial keratotomy marker to mark the cardinal meridians. We then use a vacuum or handheld trephine to incise the cornea to approximately 50% to 75% of its depth. We prefer a smaller graft, as

it is possible that the child will need multiple surgeries during his or her lifetime. For infants, we typically use a 6-mm host trephine with a graft trephine that is 0.5 mm larger. For older children, a 7-mm host trephine with a graft trephine 0.5 mm larger is usually appropriate.

5. Using a 15-degree blade, the surgeon incises the cornea along the trephination incision and injects viscoelastic into the anterior chamber.

6. We then proceed with a modified technique for removal of the host cornea to reduce the risk of lens extrusion or expulsive hemorrhage. This technique requires cutting the host tissue with corneal scissors in the same fashion as with an adult transplant. However, as each quadrant is cut, a suture is placed in the host cornea approximately 45 degrees from the cardinal positions (Fig. 1A).

7. Once the host cornea is completely separated from the host bed and held in place with four 10-0 nylon sutures, it is covered with a cohesive viscoelastic, and the donor tissue is placed on top of the viscoelastic (Fig. 1B).

8. Three cardinal sutures are used to secure the donor tissue to the host bed (Fig. 1C).

9. The host corneal sutures are then cut and the host cornea is gently removed from under the donor tissue through the area where the last cardinal suture will be placed. During this process, the surgeon should take care to maintain a layer of viscoelastic between the host and donor corneas (Fig. 1D).

10. The donor cornea is then sutured using 16 interrupted 10-0 nylon sutures, with all the knots buried. A running suture is contraindicated in pediatric patients, since such sutures loosen more quickly.

11. Finally, we typically inject dexamethasone (Decadron) and cefazolin subconjunctivally and then apply prednisolone 1% and gentamicin ophthalmic drops. If the child is monocular, we place a clear shield on the eye; if binocular, we apply erythromycin ointment and a patch and metal shield.

We recommend oral acetaminophen for postoperative pain control but find that children typically do not complain

of pain or appear uncomfortable (although they usually dislike the shield).

Postoperative Management

Suture removal starting as early as 3 weeks after surgery is indicated in pediatric transplants to reduce the risk of corneal neovascularization. With infants, all sutures should be removed by 3 months postoperatively.

Children tend to have a strong inflammatory response following PK; thus, a topical steroid should be administered frequently and tapered slowly. In patients with complex surgery, especially in combination with glaucoma, cataract, or retinal surgery, a short course of oral steroids may be considered if there are no systemic contraindications.

Compared with adult patients, children more commonly experience graft rejection, infection, and glaucoma following PK.¹ For this reason, caregivers should be taught how to perform a penlight exam, which should be done daily. They should also be educated about signs such as fussiness, photophobia, and tearing that might indicate complications.

Ongoing Follow-up

Corneal transplantation can lead to profound improvements in a child's vision and quality of life, but even a clear graft does not ensure clear vision. Amblyopia therapy must be initiated as soon as possible and is usually managed in conjunction with the pediatric ophthalmologist. Information about services such as low vision aids for school and home should be provided to families. Finally, it is important to work with the patient's pediatrician and other specialists to help ensure proper development.

1 Trief D et al. *Curr Opin Ophthalmol*. 2017; 28(5):477-484.

2 Karadag R et al. *Am J Ophthalmol*. 2016;171:95-100.

3 Loden JC, Price FW Jr. *J Cataract Refract Surg*. 1998;24(6):736-738.

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