Pediatric Corneal Transplant Surgery: Challenges for Successful Outcome

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Current Concepts in Pediatric Corneal Transplant Surgery

Visual rehabilitation of pediatric corneal blinds is a major challenge to corneal transplant surgeons. Penetrating keratoplasty is the only way to restore vision and prevent irreversible blindness due to amblyopia. Performing penetrating corneal grafts in children poses difficulty in evaluation, technical difficulties during surgery and problems during follow-up. Younger children do not cooperate for proper slit-lamp examination and need to be examined under general anaesthesia. In addition, corneal surgeon encounters problems of intraoperative positive pressure and difficulty in suturing during corneal transplant surgery. During follow up, allograft rejection, post penetrating keratoplasty astigmatism and post penetrating keratoplasty glaucoma are more frequent in pediatric group as compared to adult recipients. In case the graft is successful, the child requires rigorous treatment for amblyopia. Parents need to be counseled before surgery regarding possible visual outcome and chances of obtaining clear graft.

Indications of Penetrating Keratoplasty

Indications of penetrating keratoplasty may be grouped into congenital corneal opacities and acquired corneal opacities. Among the congenital causes Peter□s anomaly, congenital hereditary endothelial dystrophy(CHED), posterior polymorphous dystrophy, sclerocornea, dermoid and mucopolysaccharidosis are common indications for performing surgery (Fig.1).

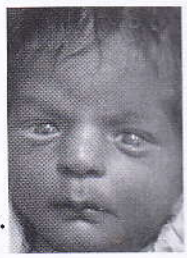

Fig 1. Bilateral congenital corneal opacity.

Fig 2.Bilateral acquired corneal opacities.

1, 2 Of the acquired causes traumatic corneal opacities, infectious keratitis, keratoconus, post cataract surgery corneal edema, non-penetrating corneal edema, are the main indications.1 Corneal edema due to endothelial cell decompensation in patients with buphthalmos has been successfully treated by penetrating keratoplasty.3 In cases of traumatic corneal scars the visual outcome depends upon the extent of injury to the posterior segment. The occurrence of indirect optic nerve injury, choroidal rupture and retinal detachment may limit the visual outcome after penetrating keratoplasty. In developing countries corneal opacities resulting from healed infective keratitis (bacterial, viral and fungal) constitute a major group of indications among the acquired causes (Fig.2).

4 Corneal opacities following keratomalacia is another frequent indication in developing countries (Fig.3).

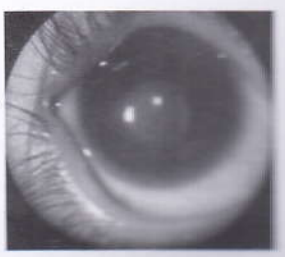

Fig 3.Unilateral acquired corneal opacity.

5 Congenital corneal opacities are usually bilateral, where as acquired corneal opacities are mostly unilateral.

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Age at the time of Penetrating Keratoplasty

Recent studies have shown that penetrating keratoplasty in children should be performed at the earliest to prevent irreversible amblyopia. In neonates with congenital corneal opacities, penetrating keratoplasty is advocated as soon as the child is fit for general anaesthesia. In case of acquired corneal opacities the waiting period for penetrating keratoplasty should be minimized. In case the waiting list for penetrating keratoplasty is long, the child is given priority over the adults. Penetrating keratoplasty in neonates and very young children is technically difficult and the risk of graft failure is high. In a study children having undergone unilateral cataract surgery before the age of four months had better visual outcome as compared to those after four months. The study indicates that penetrating keratoplasty in children should be performed early to have better visual outcome. The youngest neonate reported to have undergone successful penetrating keratoplasty for large corneal perforation is, a 34 week post conception infant weighing 1 lb.

Neonates due to immunological immaturity are less predisposed to graft rejection. The immune system in neonates develops very early in gestation and is fully developed by birth. Neonatal immune system has characteristically dominance of T suppressor cells and qualitatively less functional B cells. It has been well documented that the anterior chamber is a privileged site for transplantation. This is mainly due to the absence of blood vessels in the cornea and non access to lymphatic system. This makes antigen presentation, which is the part of the afferent limb of rejection, ineffective. Clinical studies have also shown that in neonates with isolated corneal opacities, corneal transplantation in the neonatal period resulted in better prognosis in terms of both graft clarity and vision improvement.6 The success of these grafts has been attributed to both neonatal immune tolerance and clear visual axis during the most initial period of visual development. However, at 6 months of age, the immune system is fully developed and hyperactive and corneal grafts performed at that age are at higher risk for graft rejection.

It is widely accepted that the penetrating graft in a child should be performed at the earliest. But how early it should be, remain a question unanswered. Neonates with unilateral corneal opacity may be undertaken for penetrating keratoplasty at 2 months of age. While neonates with bilateral corneal opacities, the first eye may be operated at 10 to 12 weeks of age and the second eye may be taken 6 to 8 weeks later. Contrary to the conventional rule in adults, according to which we usually operate the eye with poor vision first, in neonates if bilateral surgery is required we operate eye with better potential first. This is done to decrease the chances of developing amblyopia in the better eye.

Evaluation of Infants or Neonates with Congenital Corneal Opacities

Detailed examination of infants and neonates with congenital corneal opacities is essential to plan treatment. The visual acuity must be ascertained. Response to light stimulus, fixation at light source and following of the movement of illuminated object or light source are helpful. Responses are to be observed carefully and even parents should be demonstrated these tests. Detailed personal history, obstetric history and family history in a child with congenital corneal opacity is recorded. Detailed ocular examination may not be possible in the consultation chamber as the neonates and infants will never be steady and fixate to allow detailed slit lamp bio-microscopy. This part of the examination is better performed under general anesthesia. The amount of information we get on detailed examination under general anaesthesia helps us in decision making. Under general anaesthesia, the detailed anterior segment examination, measurements of corneal diameters (both horizontal and vertical) and intraocular pressure readings are recorded. Details of corneal opacity, whether central or peripheral, localized or diffuse, are recorded. Peripheral corneal opacities occur in partial sclerocornea and peripheral corneal ulcers. Central corneal scars may occur in Peter□s anomaly. Perforated corneal ulcer may require therapeutic penetrating graft. Direct and indirect

ophthalmoscopy to visualize retina is performed details of retina, macula and disc are recorded.

Investigations

Ultrasonography, A scan and B scan are performed to evaluate vitreous and retina status. In case eye is microphthalmic there is always possibility of associated ocular anomalies. The ultrasound biomicroscopy (UBM) gives more details of intra ocular pathologies. UBM is of special importance in patients with corneal opacity and associated glaucoma. Configuration of anterior chamber angle, details of angle structures and ciliary body are better delineated by UBM. In patients with anterior staphyloma, UBM may provide correct position of the iris and details of iris incarceration or iris adhesions. UBM has been of immense value on studying the structural alterations in pathological conditions including sclerocornea, Peter□s anomaly, aniridia and ocular trauma. The status of lens and the integrity of the posterior capsule should be evaluated. In case the cataract is present, an additional surgical procedure of cataract extraction and post chamber IOL implantation along with penetrating keratoplasty should be planed.

Pediatric Keratoplasty Constraints

Pediatric penetrating keratoplasty poses a challenge to corneal surgeons. The major constraints to perform corneal transplants in neonates include technical difficulties due to small eyes and positive posterior pressure during surgery. In severe cases of sclerocornea ,the distinction between the sclera, limbus and cornea is obliterated and decision on the size of graft and centration becomes difficult. Low scleral rigidity cause extreme positive posterior pressure, resulting in a forward bulge of iris lens diaphragm that makes the surgery difficult.7 At times the positive pressure is extremely high and may cause extrusion of lens and loss of vitreous. Associated ocular abnormalities i.e. cataract, glaucoma and microphthalmia make the surgical procedure complicated and increase the operating time significantly. In the immediate post operative period severe inflammation is observed. Post surgery evaluation is difficult and most of the times, examination under general anaesthesia is needed. In case the surgery is delayed, immaturity of the visual system leads to amblyopia.

Preparation before Corneal Transplant Surgery

The aim of corneal surgeons is to attain a clear visual axis and prevent amblyopia by performing corneal transplant at an early age. Corneal transplants may be performed as soon as the child is fit for general anaesthesia. Corneal opacification should not be considered in isolation. There is always a possibility of associated eyelid and adnexal abnormalities. These abnormalities should be first corrected so that the graft surface following surgery is well protected and the integrity is not affected. Raised intraocular pressure should be controlled and brought to normal range either with medical treatment or surgical (glaucoma filtering surgery) intervention. Associated posterior segment anomalies, including retinal detachment or vitreous hemorrhage should be evaluated and treated. Child should be examined by a pediatrician before surgery and cleared for surgery under general anaesthesia.

Donor Tissue

Exact age matching between the donor and the recipient may not be possible, however excellent grades of tissue from younger donor should be used. Donor□s age between 4 to 30 years is best suited for children. Donor corneas from younger than 4 years are relatively difficult to handle during surgery and subsequently. Donor cornea from infants and very young children have steeper cornea and may result increase corneal curvature of the graft.8, 9 Unusually high myopia (60 D) has been reported from steeper donor cornea from young donors. This makes amblyopia treatment difficult. The donor cornea for corneal transplants in children should have endothelial cell count close to 3000.

Surgery

Extremely high positive posterior pressure is a major intro-operative problem encountered during corneal transplant surgery in Infants and neonates. Although it is impossible to eliminate the positive posterior pressure, however every effort should be made to keep it minimum. Digital pressure or application of Honan□s balloon is another good option to keep posterior pressure down. External pressure on the globe due to speculum should be avoided. Flieringa□s ring should be applied in every case. In case of smaller palpebral aperture lateral canthotomy reduces posterior pressure. Use of pre-op mitotic drops or intra cameral miotic may be useful to keep iris lens diaphragm behind. An experienced anaesthetist may be asked to keep the neonate at deeper plane of anaesthesia. A non depolarising muscle relaxant (NDMR) reduces the risk of movement and contraction of extraocular muscle. Keeping head at a higher level than feet (15 degree anti-Trendelenburg position) may be helpful. Anaesthetist may be requested to hyperventilate the child in case posterior pressure is extreme. Hyperventilation decreases posterior pressure and vitreous pressure by reducing the central venous pressure and choroidal venous blood volume.

A pre placed mattress suture is helpful in securing the graft immediately and pushing the iris lens diaphragm behind by injecting visco elastic substance.10 The number of mattress sutures used may be 1 or 2 depending upon the requirement. These sutures if needed can also be applied after trephination of host cornea. Use of 8 □0□ silk or monofilament to place cardinal sutures in case of extreme positive pressure is another good option. It secures the graft and one can inject viscoelastic substance to push iris lens diaphragm back. A cohesive viscoelastic substance i.e. healon GV or healon 5 may be used to keep positive posterior pressure down and allow suturing.

Surgical procedure should be completed in a shortest possible time. After punching the donor tissue all the instruments required for recipient trephination and donor button suturing should be kept ready. Neonates or infants may be given intravenous mannitol 20% (0.5 to 1.5 gm / KBW) to reduce the vitreous volume and thus decreasing the positive posterior pressure. At times the positive posterior pressure is extremely high and it may not be possible to suture the graft unless we reduce it. Some of the surgeons have advocated placement of flat instrument (lens spatula) over the iris to prevent lens extrusion and vitreous loss. We routinely leave the recipient corneal button attached at 3 □0□ clock position and do not excise it completely. We place donor button in the recipient opening and start suturing. After securing donor corneal button with 4 cardinal sutures we excise the host corneal button and continue suturing. This intact recipient corneal button in situation of extreme positive posterior pressure is put back on the recipient open. Few more cardinal sutures are applied and posterior pressure is reduced. Then the donor button is sutured. We have found this simple method extremely useful in combating high posterior pressure (Nirankari and Sharma, unpublished data).11 In case positive pressure is extreme and passing of 10 □0□ nylon is difficult and it is not holding, it is wise to use 8 □0□ nylon / silk suture and replace these sutures after the suturing in complete. Pars plana vitrectomy before trephination has been advocated to prevent posterior pressure for patients who are at higher risk for developing extreme positive posterior pressure during penetrating keratoplasty.

Size of the Graft

In a large multicentric study the average graft size was 7.1 mm diameter. Graft size may be determined according to the diameter of the cornea. For a normal sized cornea (10.5 mm) 7.5 mm diameter graft should suffice. However in case of Micro-ophthalmia/Microcornea graft size may be decreased according to the diameter of the cornea. Placing a normal sized graft (7.5 mm) in these eyes brings host graft junction very close to the limbus. This may predispose the graft to allograft rejection and its failure. Use of

small diameter grafts in otherwise normal cornea may have several disadvantages. This may result in higher astigmatism. With the use of small grafts the number of viable endothelial cells decreases significantly. In case the graft size is reduced from the size 8 mm to 6 mm diameter the number of viable endothelial cells on the graft decreases by 44%. Thus smaller grafts will be predisposed to graft failure as the redistribution of endothelial cells will result in further lowering to final cell count well below 1000 per mm2. It is advisable to use over sized donor corneal button (0.5 mm) routinely. In a study 1 mm over sizing for pediatric case has been advocated to decrease incidence of peripheral anterior synechia.12 This option should be used with extreme caution as this may cause difficulty in suturing of the donor button and in malaposition of the host-graft junction. In addition, if the donor happens to be young less than 2 years of age, the donor cornea is steeper and 1 mm over sizing will result in high myopia, and amblyopia therapy may be difficult.

Alternatives to Penetrating Keratoplasty

In children with corneal opacities partially obscuring the visual axis, gas permeable contact lens may be tried.13 Parents are usually apprehensive of gas permeable contact lenses. They should be adequatetly counseled and risk of developing amblyopia in absence of wearing rigid gas permeable contact lens should be explained. Majority of the patients suffering from opacities resulting from traumatic corneal lacerations can be managed by fitting rigid gas permeable contact lenses. (Fig 4.)

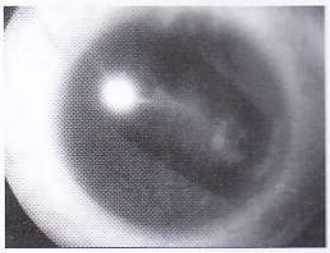


Fig 4.Healed corneal laceration for RG P contact lens fitting.

13 Some of the children are co-operative for fitting and monitoring during the follow-up. Children usually learn quickly how to insert or take out rigid gas permeable contact lenses. They take care of contact lenses as per directions.

In case opacification is central and peripheral cornea is clearer one can evaluate the child for auto rotational keratoplasty.14 The significant advantage is that the risk of allograft rejection is eliminated and chance of graft success is enhanced. However, post keratoplasty astigmatism and other problems of operating up on neonates / infant remain unchanged.

In case corneal opacity is central and larger part of the inferior cornea is clear, one can consider optical iridectomy.15 Optical iridectomy performed on the superior half of the iris does not serve any purpose as large part of it will be covered by the eyelids. Optical iridectomy is best performed in the lower nasal quadrant however one can opt for lower temporal in case opacity is extending into the lower nasal quadrant. In our experience patients are not happy with an optical iridectomy. Most of the times it does not provide an adequate vision to prevent amblyopia Infants and children with superficial corneal disease may be evaluated for superficial keratectomy. Most of the children suffering from pannus or conjunctivilization due to partial limbal stem cell deficiency can be treated with superficial keratectomy. Once a dissection plane is reached, it is extremely easy to remove the superficial corneal tissue. Children suffering from vernal ulcer and plaque formation need epithelial debridement and plaque removal in addition to medical treatment. The objective should be to achieve smooth and transparent corneal surface. To promote epithelization, bandage contact lens may be applied. Children suffering from chemical eye injury and having conjunctivilization due to partial limbal stem cell deficiency need superficial keratectomy with amniotic membrane transplant (Fig 5).

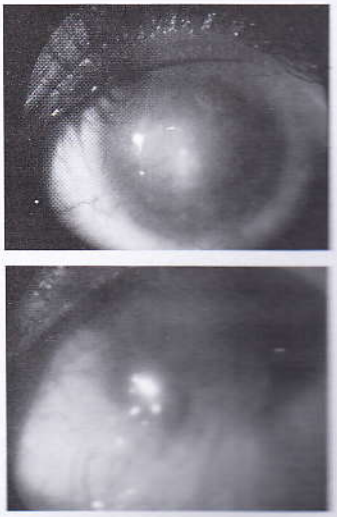


Fig 5.Chemical Eye Injury
Before amniotic membrane transplant
After amniotic membrane transplant

In case of partial thickness corneal opacities or in condition where endothelium is healthy, lamellar corneal surgery, lamellar keratoplasty or deep anterior lamellar keratoplasty may be advised. In case of superficial corneal opacities due to healed bacterial or fungal corneal ulcers one should consider lamellar keratoplasty. Corneal endothelium is healthy in these cases. Depending upon the depth of involvement of cornea one may chose to perform either lamellar keratoplasty or deep anterior lamellar keratoplasty.16

Advantage of DALK / LK are that all the intra-operative problems (extreme positive posterior pressure, danger of extrusion of lens and difficulties of suturing peripheral anterior synechia) may be avoided. In addition, risk of allograft rejection in DALK / LK is significantly less as compared to penetrating keratoplasty and post PK astigmatism is lower with visual rehabilitation being faster. These procedures may not be suitable for children with corneal opacities and deep corneal vascularization. Patient developing corneal opacities following recurrent herpes simplex keratitis are also a not suitable candidate either LK or DALK, as they may develop recurrence of HSK infection from residual corneal tissue. Cornea has been documented as site of viral latency and recurrence that may occur from corneal tissue alone. Limbal dermoids are best treated with LK / DALK depending upon the level of involvement (Fig 6).

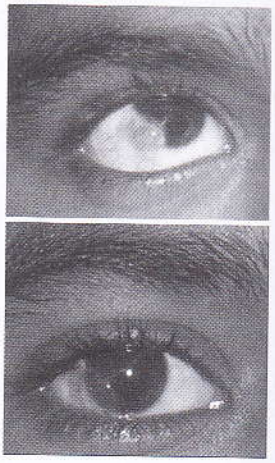


Fig 6.(a) Limbal dermoid pre-operative
(b) Limbal dermoid post surgery, DALK.

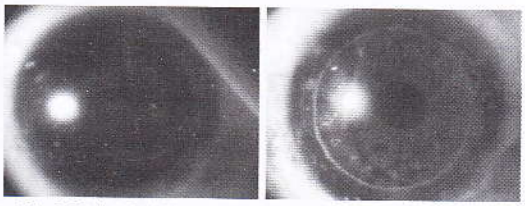


Fig 7.(a) DALK for multiple stromal foreign bodies (Pre op)
(b) DALK for multiple stromal foreign bodies (Post op)

17 We have treated a child suffering from corneal scaring due to multiple intrastromal foreign bodies in the right eye following cracker injury by performing DALK (Fig 7).

We have also treated corneal perforation with anterior staphyloma by performing DALK (Fig 8).

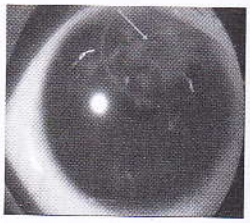


Fig 8.DALK for chronic corneal perforation with anterior staphyloma (Arrow corneal perforation)

High Risk Penetrating Keratoplasty

Presence of deep corneal vascularization in two quadrants or more predisposes the corneal grafts to higher risk of allograft rejection. Children having undergone penetrating keratoplasty for herpes simplex keratitis are at higher risk of graft rejection, recurrence of disease and failure. Children should be put on oral acyclovir prophylaxis. Children undergone regrafting are at higher risk of allograft rejection due to prior sensitization. Patients having corneal opacities in association with limbal stem cell deficiency and ocular surface disease are also at higher risk of developing graft failure due to allograft rejection. Children suffering from corneal opacification and associated ocular surface disease should undergo limbal stem cell transplant and amniotic membrane transplant prior to penetrating keratoplasty. Patients with severe ocular surface disease due to chemical eye injury, Steven□s Johnson Syndrome may undergo deep lamellar keratoplasty. Deep lamellar keratoplasty has a lower risk of allograft rejection.

Perioperative Care

Success of corneal transplant surgery in children is determined by meticulous peri-operative care. Thorough examination of child including slit lamp biomicroscopy should be performed. During the follow-

up visits parents should be explained danger signs and should be asked to report immediately as soon as the danger signs appear. The younger children and infants do not communicate their symptoms. However in case the child is irritable or crying without any obvious reason, he should be brought for eye check-up. The first examination is usually at 24 hours interval. Infants or children should be examined daily for one week and an alternate day for two weeks. After that once weekly examination is carried out. We examine the status of the graft clarity, wound integrity, epithelial healing, intraocular inflammation and intraocular pressure every visit. In case child develops any problem child may be examined early to the start treatment.

Complications:

Early

The common problems encountered in the first few days are disruption of host graft junction, fibrinous reaction, delayed epithelization and high intraocular pressure . Infective keratitis although rare (5%) may occur and needs intensive topical antibacterial treatment. Fibrinous exudative reaction should be treated with topical and systemic steroids. Delay in epithelization may be secondary to severe uveitis or high intraocular pressure. In these cases severe uveitis is treated with topical or systemic steroids. Once uveitis or high intraocular pressure is controlled epithelial defect heals completely. Rarely endophthalmitis following penetrating keratoplasty may occur.

Intermediate

Neonates and infants need to be examined under general anaesthesia at around 3 weeks even if they have no visible problem. Wound healing in neonates and infants is fast and sutures become loose. At 3 to 4 weeks loose suture should be removed. At 6-8 week complete healing occurs and sutures may be removed. Children aged between 2 to 4 years may be observed for loose suture. The loose suture should be removed immediately. Children above 6 years behave more or less similar to adults and selective suture removal should be performed.

Late Complications

Allograft rejection, suture related infections, recurrence of disease (HSK, keratoconus), post keratoplasty astigmatism and post keratoplasty glaucoma may occur. Suture related complications can be prevented by immediate removal of loose sutures. Loose suture may cause an epithelial defect, secondary inflammation, graft vascularization and trigger allograft rejection. Post keratoplasty astigmatism is higher with PKP as compare to lamellar procedure (LK/DALK). Selective suture removal can be done to reduce astigmatism in children aged 6 years of more.

Allograft Rejection

Parents should be explained that allograft rejection might occur any time after surgery. Studies have shown that 30% to 70% pediatric grafts fail within first six months and 65% to 85% within first year. Neonates and infants should be closely monitored for development of allograft rejection. Children are unable to complain about the symptoms and usually present late for treatment. Parents should be educated to bring the child for examination on observing any redness, discomfort, and opaqueness in the graft or decrease in vision. On every visit slit-lamp examination should be done to detect early signs of allograft rejection. Infants and children may not present with the characteristic signs of allograft rejection. Graft edema even in the absence of keratic precipitates should also be treated as allograft rejection. Intra ocular pressure should be monitored in these cases. Once allograft rejection is diagnosed, the child is put on prednisolone acetate (0.1%) eye drop every one hour and atropine ointment twice daily. In addition oral prednisolone (1 mg/kbw) should also be started. In recent studies topical cyclosporine A (2%) has been

found successful in treating graft reaction.18 It has also been used to prevent graft rejection in high risk cases. Cyclosporine A being lipid soluble, topical cyclosporine A (2%) need to be prepared in castor oil in the hospital pharmacy. At times young children may not tolerate it, as it causes significant ocular irritation. It has also been reported to cause persistent epithelial defects and delayed epithelial healing. Recent studies have shown that topical cyclosporine A prepared in the aqueous solution is also effective and can be prepared in preservative free artificial tear solution. Topical cyclosporine A 0.05% (Restasis, Allergan) has been found effective in treatment and prevention of graft rejection in high risk cases and reported to be equally effective in concentrations ranging from 0.05% to 1%.. The formulation of the of the drug is such that it releases large number of microdroplets in the tear film.

Glaucoma

Treatment of glaucoma prior to or after penetrating keratoplasty includes medical treatment, trabeculectomy with adjuvents and/or trabeculotomy. Treatment of refractory glaucoma is a real challenge as it will not only damage the optic nerve but also corneal graft resulting graft failure. Glaucoma drainage implant procedures have shown encouraging results in the treatment of refractory post penetrating keratoplasty glaucoma.19 Several authors have reported favorable results of pediatric penetrating keratoplasty following control of IOP using glaucoma drainage implant procedures. Although conventionally glaucoma is first controlled and only then is penetrating keratoplasty performed. At times due to the risk of development irreversible amblyopia glaucoma implant surgery may be combined with penetrating keratoplasty to provide early visual rehabilitation and preventing the development of amblyopia.

Treatment of Amblyopia

Amblyopia treatment should be started as early as possible following surgery. Cycloplegic refraction should be done and glasses should be prescribed. Parents should be explained that the normal eye of the child need to be patched so that he uses the operated eye. The schedule for patching may be the same as used for standard amblyopia treatment. In children upto 2 years (2:1), 2 to 3 years (3:1), 3 to 4 years (4:1), 4 to 5 years (5:1) and 6 years or above (6:1) should be used.

Outcome

Anatomical success rate following penetrating keratoplasty in childhood including infants and neonates has increased significantly. The incidence of vision restoration in children following penetrating keratoplasty is still low. Visual prognosis has been reported to be better in younger children and is likely determined by the incidence and severity of amblyopia. In recent study of 65 grafts on 58 eyes of 52 children (mean age 10.6 years SD 4.3 years) 38% achieved BCVA 6/9 or better and 60% had BCVA 6/18 or better.20 Visual acuity has been reported to be significantly better for the acquired indications as compared to the congenital ones. Significant numbers of patients in this study had keratoconus as an indication for penetrating keratoplasty (Fig 9). Results from developing countries are less favourable. In a study from India nearly 1/3rd patients achieved > or = 20/400 vision and nearly 50% of these achieved > or = 20/50.5 Allograft rejection, infective keratitis and glaucoma were major causes of graft failure. The overall long term (10 years) probability of maintaining clear graft after initial penetrating keratoplasty for Peter□s anomaly is 35% +/- 0.6%.21 Eyes with severe disease, larger donor cornea, co-existing central nervous system abnormalities and anterior synechia were reported to have significantly poorer outcome

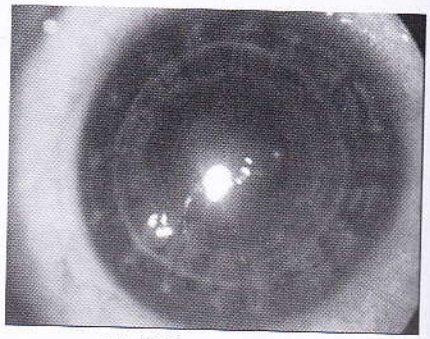


Fig 9.Penetrating keratoplasty for keratoconus.

than the eyes without these factors. Children with severe form of Peter□s anomaly may require multiple grafts to have functional vision. One of our patient with severe Peter□s anomaly had undergone four corneal transplants during 14 years and enjoyed good vision. He sadly died recently due to severe CNS disease (Fig 10).

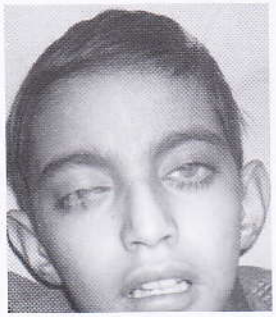


Fig 10.Regraft in a child with bilateral Peter□s anomaly (LE)

Repeat Corneal Graft

In case the child develops graft failure a repeat graft should be considered. In a recent study irreversible graft rejection has been reported as the commonest cause of graft failure.22 Patients need to be explained that subsequent corneal grafts have less chances of success. In one of the studies, of 27 repeat grafts undergoing second graft 19% were successful and of six graft undergoing third graft none succeeded. Repeat graft may be indicated to prevent dense and irreversible amblyopia. However in case the child is having unilateral corneal opacity, parents should be explained that even if the graft may becomes opaque, later on chances of improvement of vision by performing repeat graft will be there. In adults if the grafts fails due to the graft rejection, it is advisable to wait for 6 months before a repeat graft is performed. It is aimed to bring down the inflammation in the graft to minimum and to decrease the incidence of allograft rejection. At times it may be difficult to ascertain whether the graft failed due to allograft rejection or due to some other cause. It is better to treat it as allograft rejection. In younger children it may not be possible to wait for 6 months due to danger of development of amblyopia. In these cases the repeat graft may be performed at 3 months after the initial graft has failed. One can wait a little longer in case the child is six year old or more. Children suffering from perforated corneal ulcers need therapeutic penetrating keratoplasty and these grafts usually become opaque due to chronic inflammation. However, successful regraft can be performed for these patients at later date and both vision improvement and graft clarity can be obtained (Fig 11).

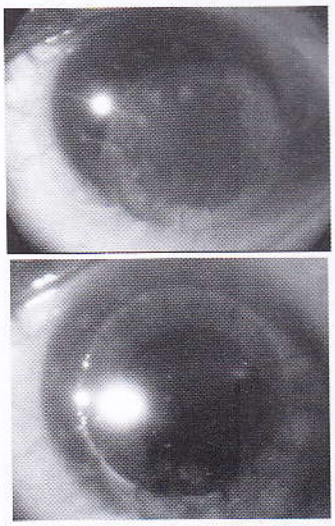


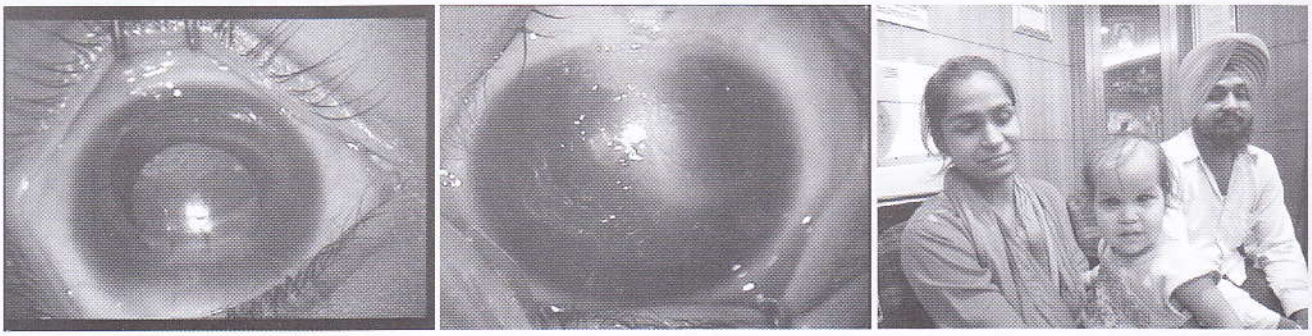
Fig 11.Regraft for opaque therapeutic graft
Before regraft
After regraft

Keratoprosthesis

Infants and children who are at high risk of graft rejection and subsequent graft failure may be benefited with keratoprosthesis or artificial corneal transplantation.23 Keratoprosthesis transplantation means placing an optical device in the host cornea. It is immunologically inert and has the advantage that graft rejection does not occur. Recently, a custom made Boston type 1 keratoprosthesis is available and can be designed to correct refractive errors including aphakia. AlphaCor, a synthetic cornea made up of hydrophilic polymer poly (2-hydroxyethyl methacrylate) is another keratoprosthesis used in high risk cases for corneal transplant surgery.24 The alphaCor is implanted in a corneal stromal lamellar pocket in a two stage procedure. In the first stage 3600 peritomy and debridement of corneal ephithelium is done. A superior 1800 limbal incision at 50% of depth is extended into the corneal stroma forming an intralamellar pocket is made. A central 3.5 mm posterior corneal trephination is performed. The device is placed with in the corneal pocket and paralimbal incision is closed. After 8 to 12 weeks anterior trephination (3mm) is done to expose the optic of the device. Keratoprosthesis helps the corneal surgeon to rehabilitate those corneal blinds having visual potential, but who are unlikely to be benefited by performing penetrating keratoplasty using human donor cornea.

Pediatric corneal transplant surgery is a team effort it involves combined effort of corneal surgeon

assisted by glaucoma specialist, pediatrician, anaesthetist, counselors and rehabilitation team(Fig 12 a,b,c). Rehabilitation of blinds due to corneal disease



Before PKP | After PKP | Child with bilateral PKP

Fig 12 Penetrating keratoplasty in a child with buphthalmos and opaque cornea

Children with bilateral corneal opacities either congenital, chemical burns, Stevens Johnson Syndrome may not be successfully visually rehabilitated even after performing repeated corneal grafts. The parents of these children should be counseled to get their children admitted to blind schools to provide educational and vocational training to these children. These children can lead independent life and contribute to the development of the society if proper facilities and opportunities are provided.

Summary & Conclusions

Advancement in micro-surgical techniques, quality eye banking and better anaesthesia facilities have made it possible to undertake corneal transplant in a neonate as soon as the diagnosis is made and corneal transplant surgery advised. Although surgery is technically demanding but it is possible to provide clear visual axis during critical period of visual development. Treatment and prevention of development of amblyopia in neonates is extremely important even after successful penetrating keratoplasty.

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