PAEDIATRIC CORNEAL SURGERY
Challenges abound, but advancing techniques and appropriate care make good outcomes possible
by Cheryl Guttman Krader in Milan

While advances in partial thickness procedures have revolutionised the field of corneal transplantation in adults, changes in paediatric corneal transplant surgery continue to occur at a slower pace. Currently, penetrating keratoplasty (PKP) remains the technique of choice for all graft procedures in children, and it will likely remain the dominant surgery, at least in younger children, considering the underlying indications for paediatric corneal transplantation, according to Gerald W Zaidman MD.

However, just as developments in technique enabled increased adoption of PKP in children, deep anterior lamellar keratoplasty (DALK) is being performed increasingly in paediatric eyes with appropriate indications, and it is likely that with future advances, there will be greater use of endothelial keratoplasty [ie, Descemet’s stripping (automated) endothelial keratoplasty (DSEAK)] in paediatric patients with endothelial dysfunction, he said.

Speaking during a joint symposium of the 2nd World Congress of Paediatric Ophthalmology and Strabismus and the 3rd EuCornea Congress, Dr Zaidman discussed corneal transplantation in children, focusing on PKP and DSEAK. Outcomes he reported from his personal experience and other published series showed that results for graft survival and visual function can be very good.

"While there was a general feeling for 60 to 70 years after the first human corneal graft procedure that corneal transplantation should not be done at all in children, the situation changed in the 1980s thanks to the work of some well-known corneal specialists who categorised paediatric corneal disease and described transplantation techniques for this population. Since then, outcomes have further improved thanks to the development of better methods," said Dr Zaidman, director of ophthalmology, Westchester Medical Centre, and professor of ophthalmology, New York Medical College, Valhalla, NY.

Differences in diagnoses The reason why PKP is the predominant type of corneal transplant surgery in children relates to the distribution of underlying diagnoses. Whereas data from North America show that among adults, endothelial disease is the leading indication for a corneal graft procedure followed by keratoconus, paediatric corneal transplantation is for a congenital corneal opacity in about two-thirds of cases and for endothelial disease in just about 20 per cent.

"Considering all children with congenital, acquired and trauma-related corneal opacities, the only patients who are candidates for endothelial keratoplasty are those with a congenital hereditary endothelial dystrophy (CHED), congenital glaucoma and corneal oedema, and failed grafts."

Endothelial keratoplasty pros and concerns DSAEK has several advantages compared with PKP as the partial thickness procedure is a closed globe operation, affords a stronger wound, minimises induced astigmatism and results in faster visual rehabilitation. However, a steep learning curve has limited its adoption even in the adult population, and the surgery is more technically challenging in children because of their shallower anterior chamber and the difficulty of visualising Descemet’s membrane, said Dr Zaidman.

In addition, limited available data indicate that re-bubbling is needed even more often in children than in adults (in about one-third of paediatric cases), and it must be done in the OR, he added.

Reviewing the literature on DSEAK in children, Dr Zaidman noted there have been only 33 reported cases of surgery for eyes with CHED. Mean age of the patients was eight years, with very few infants operated on, and the reporting surgeons all described the significant intraoperative challenges and frequent need for re-bubbling.

"However, as in adults, graft clarity was excellent and visual rehabilitation was faster than with PKP. Presumably the rates of endothelial cell loss and rejection are the same as in adults, but there are limited data available on those outcomes," Dr Zaidman said.

In 2007, Dr Zaidman published his experience with PKP in a series of 30 eyes (24 children) with Peter’s anomaly Type I [Am J Ophthalmol 2007;144:104-8]. Regrafting was needed in two eyes, and after a mean follow-up of 79 months, 90 per cent of eyes had a clear graft. In verbal children, visual acuity was 20/100 or better in 54 per cent of eyes, of which more than half had 20/50 or better visual acuity. No eyes were lost, and none had no light perception vision. Most of the eyes with visual acuity of 20/200 and worse had glaucoma. However, Dr Zaidman observed that they still had ambulatory vision, which is the goal of corneal transplant surgery in young children.

"The aim of the procedure is to provide vision that will allow the child to perform normal routine tasks, such as feeding and dressing one’s self, and to interact with the world. It is not to provide legal driving vision," said Dr Zaidman.

He also cited recently published results from the Australian graft registry that included 765 patients under 20 years of age [Ophthalmology 2011;118:492-7]. In the sub-group of children younger than five years old, in whom Peter’s anomaly/congenital defects was the leading underlying diagnosis, 56 per cent had a clear graft. The clear graft rate was 62 per cent among children ages five to 12, of whom almost two-thirds were operated on for keratoconus, and 90 per cent in those aged 13 to 19, who mostly had keratoconus.

“Graft clarity after PKP in children varies depending on patient age and underlying diagnosis. The results are better when the problem is more a pure corneal disease and there is less involvement in the anterior chamber,” Dr Zaidman said.

Despite its challenges, the surgery itself may not be the limiting step in achieving a good outcome after paediatric corneal transplantation, noted Dr Zaidman. Family cooperation plays a critical role as the postoperative course involves a marathon of exams, including multiple exams under anaesthesia, and over the long-term, vision outcome depends on treatment of amblyopia and glaucoma.