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SHORT COMMUNICATION

Penetrating keratoplasty in a newborn: Case report and analysis of current surgical trends in Italy

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PURPOSE. *The aims of this study were to describe bilateral penetrating keratoplasty (PK) in a newborn and to analyze the data of PKs performed in Italy during the 5-year period 1999-2003 in children under 4 years of age.*
METHODS. *A male newborn had PK at age 3 months and 5 months for near-blindness secondary to severe congenital corneal clouding in both eyes. The infant's explanted corneas were subjected to histochemical and ultrastructural analyses. Data regarding the number of PKs performed in Italy on 0-4-year-olds were obtained from the Web site of the Italian Ministry of Health.*
RESULTS. *The postoperative courses were uncomplicated, and 42 months of follow-up data show bilateral graft transparency and substantial improvement in visual acuity despite high-grade myopia and nystagmus. At the ultrastructural level, the main alterations involved the endothelial cells and Descemet membrane. A total of 45 PKs were performed in Italy on patients 0-4 years old from 1999 through 2003; only nine involved babies under 1 year of age.*
CONCLUSIONS. *In babies with congenital corneal opacities, early PK can reduce severe amblyopia. However, the risk of intra- and postoperative complications in PK is high. Based on the 42-month follow-up, the anatomic and functional results achieved in the current patient are satisfactory despite the presence of nystagmus and postoperative high-grade myopia. This study shows that PKs are rarely performed, in Italy, in children aged 0-4 years, and very few are done during the first year of life. (Eur J Ophthalmol 2008; 18: 290-3)*

KEY WORDS. *Pediatric keratoplasty, Congenital corneal clouding, Amblyopia*

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INTRODUCTION

Penetrating keratoplasty (PK) for congenital corneal opacity is rarely performed during the first months of life because it is associated with technical difficulties and a much higher rate of complications than those observed in adults (1-4). The use of PK in young infants is associated with risks related to the reduced scleral rigidity that characterizes this age group; a greater potential for inflammatory reactions in the anterior segment, which can accelerate graft rejection; the possibility of intraoperative complications stemming from the positive vitreous pressure in the developing eye; and/or the risk of poor postoperative visual acuity due to amblyopia and major refractive errors.

This report describes the results of bilateral corneal transplantation procedures performed in 2003 on a 3-month-old infant with intense corneal clouding, based on 42 months of follow-up. It also includes an analysis of the keratoplasties performed in Italy during the period 1999–2003 on children 4 years and younger.

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Case report

A 30-day-old male infant was referred to our department for near-total blindness and coarse, irregular nystagmoid eye movements. The baby had no signs of systemic disease. He was the first-born child of nonconsanguineous parents, and the family history was unremarkable.

The physical examination revealed no ocular anomalies other than marked clouding of both corneas. The diameters of the corneas, the irises, and the lenses were normal. No gross fundus anomalies were noted although visualization was markedly reduced by the corneal opacities. No abnormalities were noted on ocular ultrasonography. The axial lengths of the right and left eyes were 19.31 mm and 19.18 mm, respectively. The intraocular pressure (IOP) was 18 mmHg. Pachymetry revealed corneal thicknesses of approximately 950 mm in both eyes. Metabolic and laboratory tests were negative.

PKs were performed when the patient was 3 months old (right eye) and 5 months old (left eye). In both cases, we used a corneal flap with a diameter of 7.75 mm. The corneal grafts, which came from a 3-year-old donor (right eye) and a 17-year-old donor (left eye), were prepared with a Hessburg Barron vacuum trephine with a diameter of 7.5 mm. They were secured with double running sutures (10-0 nylon), which were removed after 6 months. Postoperative treatment included dexamethasone corticosteroid, and netilmicin drops four times a day for the first month and fluorometholone 0.1% drops twice a day for the next 45 days.

The infant's explanted corneas (and a specimen of one of the corneal grafts) were subjected to histochemical and ultrastructural analyses.

A few days after the operation, the infant began to show interest in and follow objects and luminous targets. On postoperative day 40, the implanted cornea was transparent, and the axial length of the right eye was 21.69 mm (versus 19.20 mm for the left eye). The IOP was 14 mmHg on the right and 18 mmHg on the left.

When the baby was 5 months old, the left eye was operated on, and the postoperative course was again uneventful.

When he was 9 months old, the nystagmus had diminished considerably, and the infant began to show clear interest in his surroundings. Examination under general anesthesia revealed that both of the grafts were transparent. The axial lengths had increased to 23.05 mm (right eye) and 22.04 mm (left eye). The IOP was 12 mmHg in both eyes. Corneal thickness was 550 mm in the right eye and 520 mm in the left. Refractive values obtained with the Nikon Retinomax K-plus 2 were –7.00 –5.50 x 50° for the right eye and –6.00 –3.00 x 125° for the left.

At 12 months of age, biometry revealed additional increases in the anteroposterior axis of both eyes (24.89 mm in the right eye, 23.55 mm in the left).

The most recent examination (when the child was 45 months old) revealed that both corneal transplants were transparent. Irregular, horizontal nystagmus was still present. Illiterate E visual acuity was 0.1 in the right eye (refraction: –10 –4 x 30°) and 0.15 in the left eye (refraction: –8 –2.50 x 100°), and the infant is wearing glasses with this optical correction.

The explanted tissues were characterized by very weak alcianophilia. At the ultrastructural level, the main alterations involved the endothelial cells and Descemet membrane. The endothelium was thickened and presented a severely altered cytotype and interdigitations between adjacent cells. Descemet membrane was thin (2.4 mm) and appeared completely amorphous. That of the donor cornea was almost twice as thick (4.2 mm) and contained figures typical of type VIII collagen in the form of pseudohexagonal polymers. In the explanted corneas, the keratocytes were arranged in uninterrupted rows with interdigitations between one cell and the next. The stromal fibrils lay parallel to one another and were normal in caliber (27 nm), but the interfibrillar spacing was only 15 nm (less than half that of the donor cornea: 35 nm). The findings were consistent with a diagnosis of combined dystrophy of the endothelial cells with secondary repercussions on Descemet membrane and the keratocytes with reduced space between the stromal fibrils.

Data regarding the number of PKs performed in Italy on 0–4-year-olds were obtained from the Web site of the Italian Ministry of Health (www.ministerosalute.it Accessed July 28, 2007), which systematically collects information on all hospital admissions in Italy. The information is based on hospital discharge abstracts and includes ICD-9-CM codes (International Classification of Diseases, ninth revision–Clinical Modification, 1997) for the discharge diagnoses and for all procedures performed during the hospitaliza-

tion. Access is currently limited to hospital admissions that occurred from January 1, 1999, through December 31, 2003. For each year of this period, we performed a search for procedure code 11.69 (corneal transplantation) among the records for all patients <1 year of age and those aged 1–4 years.
A total of 45 PKs were performed in Italy on patients 0–4 years old from 1999 through 2003. Nine involved children under 1 year of age, and this group includes the two operations described in the present report (both performed in 2003). The remaining 36 were performed on children aged 1–4 years: five in 1999, eight in 2000, nine in 2001, five in 2002, and nine in 2003.

DISCUSSION

In babies with congenital corneal opacities, early penetrating keratoplasty can reduce the risk of severe amblyopia. The earlier surgery is performed, the better the changes of visual recovery (1). Owing to technical advances, this procedure can now be performed successfully at an earlier age, but the risk of intra- and postoperative complications is high. Problems that arise during surgery are mainly related to the reduced rigidity of the sclera. As a result, the eye is more likely to collapse, and there is also a higher risk of prolapse of the intraocular contents when the cornea is removed (1, 5). Postoperative complications are linked to graft survival and refractive errors, which can be quite high after surgery of this type. There is also a real possibility of persistently severe amblyopia. The rate of success associated with PK in pediatric patients is highly variable depending on the patients and congenital pathology considered. In most cases, the child has Peters anomaly, sclerocornea, or congenital hereditary endothelial dystrophy (CHED). The chances of success seem to be fairly high in patients with CHED since this disease affects only the cornea (4, 6).
In the infant described in the present report, PKs were performed at the ages of 3 months and 5 months. The baby had presented with intense bilateral corneal clouding, which was caused by congenital changes in the corneal endothelium and Descemet membrane. Surgery produced an immediate improvement in the baby's visual capacity. At the age of 45 months, the corneal implants were still transparent. Visual acuities were 0.1 in the right eye and 0.15 on the left. In both eyes, nystagmus was associated with myopia (–10 in the right eye, –8 in the left). This substantial myopic shift can be attributed in part to genetic factors but in all probability it is also related to discrepancies in bulb growth before and after surgery. The corneal grafts came from donors aged 3 and 17 years, and these tissues were thus much more rigid than the native corneas.
Our analysis of data furnished by the Ministry of Health (5) shows that, in Italy, PKs are rarely performed in children aged 0–4 years, and very few are done during the first few months of life. During the 5-year period from 1999 through 2003, 36 keratoplasties were performed on children aged 1–4, but only nine were done on patients <1 year old.
In conclusion, based on our 42-month follow-up, the anatomic and functional results achieved with early PK in our patient are satisfactory, especially if we consider the severe sensory deprivation that was present before surgery. Although these findings are encouraging, further follow-up is needed to determine how long the corneal grafts will remain transparent.

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REFERENCES

1. Comer RM, Daya SM, O'Keefe M. Penetrating keratoplasty in infants. J AAPOS 2001; 5: 285-90.
2. McKellan K, Lai T, Grigg J, Billson F. Penetrating keratoplasty in children: visual and graft outcome. Br J Ophthalmol 2003; 87: 1212-4.
3. Patel HY, Ormonde S, Brookes NH, Moffatt LS, McGhee CNJ. The indications and outcome of paediatric corneal transplantation in New Zealand. Br J Ophthalmol 2005; 89: 404-8.
4. Al-Ghamdi A, Al-Rajhi A, Wagoner MD. Primary pediatric keratoplasty: indications, graft survival, and visual outcome. J AAPOS 2007; 11: 41-7.
5. Reidy JJ. Penetrating keratoplasty in infancy and early childhood. Curr Opin Ophthalmol 2001; 12: 258-61.
6. Schaumberg DA, Moyes AL, Gomes JAP, Reza Dana M. Corneal transplantation in young children with congenital hereditary endothelial dystrophy. Am J Ophthalmol 1999; 127: 3783-8.