

Guidelines to be followed by centres, services and units in order to be designated as Reference Centres, Services and Units of the National Health System as agreed by the Interterritorial Board

23. PENETRATING KERATOPLASTY IN CHILDREN

Keratoplasty refers to corneal transplantation or corneal graft, in which the damaged corneal tissue is replaced by a donor tissue. Full thickness corneal transplantation is referred to as penetrating keratoplasty in children.

Based on data from published research in countries similar to Spain^{5, 6,7,9}, it can be stated that keratoplasty in children is due to various types of pathologies, all of them severe, of low incidence and with poor visual prognosis.

Paediatric keratoplasty causes that should be cared for in a Spanish National Health Service reference unit are below:

- Congenital^{4,5,6}: Anomalies in eye development (the most frequent are Peters anomaly, polymorphous corneal dystrophy, and sclerocornea). Corneal anomalies resulting from another ocular congenital pathology that may require corneal transplantation during their evolution (congenital glaucoma) are also included within this group⁸.
- Trauma acquired: following trauma affecting the cornea⁸.

A statistic study on congenital ocular defects carried out by the Spanish team Bermejo y Martínez Frias^{1, 2} with a sample of more than a million consecutive births found out an incidence of corneal conditions of 3.1 out of every 100,000 births.

Paediatric corneal transplantation has a higher number of complications and failures, associated pathologies, and higher monitoring problems than for adults, resulting in the need to be performed in specialized services^{3,9,10,11} with support units: paediatricians, anaesthetists and ophthalmologists with experience in paediatric patients.

A. Rationale for the proposal

▶ Data on the use of penetrating keratoplasty in children.	50 <i>transplantations</i> are performed in <i>children per year</i> (population under 14 years of age). (According to data from the Spanish National Transplant Organization, there are around 2,500
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	<p>corneal transplantation per year and according to bibliography from developed countries approximately 2-3% of the keratoplasty performed in a year are in children^{10,11}.)</p> <p>There are few data on keratoplasty results¹⁰ in children due to different reasons:</p> <ol style="list-style-type: none"> 1. Paediatric cases, due to a more complex evolution and poorer results which hide and alter results data in adults, are usually excluded from research published on this topic³. 2. Low incidence of these processes leads to no research published in Spain on results in children. 3. Statistics from other countries are not comparable to our environment. (For example, there are various publications in India, a country with high incidence of corneal problems due to weather and sanitary conditions, which are not comparable to our country.)⁸
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B. Guidelines to be followed by Centres, Services and Units in order to be designated as Reference Centres, Services and Units performing penetrating keratoplasty in children

<p>► Experience of the Reference Centres, Services and Units:</p> <p>- Activity:</p> <ul style="list-style-type: none"> • Number of penetrating keratoplasty in children (minimum and optimal) that should be performed in a year to ensure an adequate care. • Number of procedures (minimum and optimal) that should be performed in a year of techniques, technologies and procedures similar to those specific to the 	<p>- 5 to 10 penetrating keratoplasty in children.</p> <p>- Minimum of 10 surgical procedures of anterior segment in children (congenital glaucoma, congenital cataract, dermoid cyst...)</p>
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<p>designation requested.</p> <ul style="list-style-type: none"> - Other data: research on the subject, postgraduate teaching, continuing training, etc. 	<ul style="list-style-type: none"> - Accredited postgraduate teaching. - Participation in research projects and publications in the field^a. - Continuing training programme.
<p>► Specific resources of the Reference Centres, Services and Units:</p> <ul style="list-style-type: none"> - Human resources required for adequate carrying out of penetrating keratoplasty in children. <p>Professional experience^b:</p> <ul style="list-style-type: none"> - Specific equipment required for adequate carrying out of penetrating keratoplasty in children. 	<ul style="list-style-type: none"> - Two ophthalmologists. - 24 hour continuous ophthalmic care, given the need for postoperative monitoring and the possibility of complications during the first hours. - Nursing staff, surgical auxiliaries and technicians. - Ophthalmologists with experience in: <ul style="list-style-type: none"> • Corneal microsurgery (experience of five years, having performed a minimum of 25-30 corneal transplantation in children.) • Postsurgical problems and monitoring of the paediatric transplant patient in order to be prepared for secondary complications to this surgery (wound dehiscence, secondary glaucoma, endophthalmitis, graft rejection...) • Monitoring of frequent anomalies associated to corneal alterations leading to transplantation (experience in anterior pole surgery, congenital and paediatric cataract, paediatric glaucoma, and autoimmune therapies.) - Unit with experience in visual rehabilitation for paediatric patients, for follow-up and adequate treatment of amblyopia, necessary so that anatomical success accompanies functional results. - Nursing staff with experience in the care of ophthalmologic paediatric patients. - State-of-the-art surgical microscope. - Corneal transplantation equipment with trephines of different sizes, since small size grafts may be needed due to the children eye characteristics.

<p>► Resources from other units and services besides those belonging to the Reference Centres, Services and Units required for adequate carrying out of penetrating keratoplasty in children.</p>	<ul style="list-style-type: none"> - Blepharostats for paediatric patients. - Corneal topographer, corneal pachymeter. - Hand held refractor keratometer - Endothelial microscope. - Portable slit lamp. - Fixed slit lamp. - Tonometer for ocular pressure assessment. - Ocular ultrasound. - Paediatric ophthalmology office basic equipment with refraction unit. - Anaesthesia services with experience in paediatric patients^b. - Intensive care unit with experience in paediatric patients^b. - Paediatric Services. - Radiology services. - Pathological Anatomy.
<p>► Procedure and clinical results indicators of the Reference Centres, Services and Units ^c:</p>	<p>The indicators will be agreed with the Units that will be designated.</p>
<p>► Existence of an adequate IT system (Type of data that the IT system must include to allow identification of the activity and evaluation of the quality of the services provided)</p>	<ul style="list-style-type: none"> - Filling up the complete MBDS of hospital discharge. - The unit must have a <i>registry of patients</i> who have undergone penetrating keratoplasty, which at least must include the following information: <ul style="list-style-type: none"> - Medical record number. - Date of birth. - Sex. - Date of admission and date of discharge. - Family history (mother, siblings, multiple births...) - Diagnosis procedures performed to the patient (ICD-9-CM). - Main diagnosis (ICD-9-CM). <ul style="list-style-type: none"> • Clinical situation at the time of diagnosis (corneal condition).

	<ul style="list-style-type: none"> - Number and type of therapeutic procedures provided to the patient (ICD-9-CM): <ul style="list-style-type: none"> • Penetrating keratoplasty. • Other therapeutic procedures. - Treatment results. <ul style="list-style-type: none"> • Corneal graft a year after. - Complications. <ul style="list-style-type: none"> • Graft opacification. • Wound dehiscences. • Graft rejection, acute or chronic. • Endophthalmitis • Glaucoma. • Cataract. - Patient's progress. <p>- The unit must have the required data which should be sent to the Spanish National Health Service Reference Centres, Services and Units Appointment Commission Secretariat for yearly reference unit monitoring.</p>
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^a *Criteria to be assessed by the Appointment Commission.*

^b *Experience will be accredited by certification from the hospital manager.*

^c *Clinical results standards, agreed to by the experts group, will be assessed, initially by the Appointment Commission, while in the qualification process, as more information from the Reference Centres, Services and Units is being obtained. Once qualified by the Appointment Commission, the Quality Agency will authorize its compliance, as for the rest of guidelines.*

Bibliography:

¹ Bermejo E, Martínez-Frías ML. Congenital eye malformations: clinical-epidemiological analysis of 1,124,654 consecutive births in Spain. *Am J Med Genet.* 1998 Feb 17; 75(5):497-504

² Bermejo E, Martínez-Frías ML. Malformaciones Congénitas de los ojos: análisis clínico-epidemiológico de 1.124.654 nacimientos consecutivos en España. *An Esp Pediatr.* 1996 Sep; 45(3):269-75.

- ³ Thompson RW Jr, Price MO, Bowers PJ, Price FW Jr. Long-term graft survival after penetrating keratoplasty. *Ophthalmology* 2003 Jul;110 (7):1396-402.
- ⁴ Yang LL, Lambert SR, Lynn MJ, Stulting RD. Long-term results of corneal graft survival in infants and children with peters anomaly. *Ophthalmology* 1999 Apr;106(4):833-48.
- ⁵ Yang LL, Lambert SR. Peters' anomaly. A synopsis of surgical management and visual outcome. *Ophthalmol Clin North Am* 2001 Sep;14(3):467-77.
- ⁶ Watts P, Michaeli-Cohen A, Abdolell M, Rootman D. Resultados de queratoplastia lamellar para dermoides límbicos en niños. *J AAPOS* 2002 Aug; 6 (4):209-15
- ⁷ Dana MR, Moyes AL, Gomes JA, Rosheim KM, Schaumberg DA, Laibson PR, Holland EJ, Sugar A, Sugar J. The indications for and outcome in pediatric keratoplasty. A multicenter study. *Ophthalmology* 1995 Aug; 102 (8):1129-38.
- ⁸ Aasuri MK, Garg P, Gokhle N, Gupta S. Penetrating keratoplasty in children. *Cornea* 2000 Mar; 19 (2):140-4.
- ⁹ Comer RM, Daya SM, O'Keefe M. Penetrating keratoplasty in children. *J AAPOS* 2001 Oct; 5 (5): 285-90.
- ¹⁰ Fonseca A, Abelairas J, Rodríguez- Sánchez JM, Peralta J. Actualización en cirugía oftálmica pediátrica Madrid 2006.
- ¹¹ Patel HY, Ormonde S, Brookes NH, Moffatt LS, McGhee CN. The indications and outcome of paediatric corneal transplantation in New Zealand: 1991-2003. *Br J Ophthalmol* 2005 Apr; 89 (4):404-8.