

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.

11. COMPLEX OCULAR SURFACE RECONSTRUCTION. KERATOPROSTHESIS.

Complex ocular surface reconstruction may be due to a variety of causes:

- Autoimmune diseases: pemphigus, pemphigoid, Stevens-Johnson syndrome, Lyell's syndrome, etc.
- Traumatic: chemical burns, accidents, etc.
- Congenital: ablepharon, first branchial arch syndrome, etc.
- Neurological: facial paralysis, trigeminal anaesthesia, etc.
- Infectious: trachoma, virosis, etc.

These conditions may harm the cornea severely hindering vision.

Patients with aniridia needing a limbal transplantation are also included.

Diagnostic procedures include elemental practices in a hospital ophthalmology department (biomicroscopy with slit lamp, tonometry, refractometry, keratometry, pachimetry, corneal topography) as well as more uncommon and complex procedures (confocal microscopy of the ocular surface, impression cytology, eye moisture measurement, eyelid dynamometry).

Therapeutic procedures include diverse reconstruction techniques affecting the cornea and its limbus (penetrating and lamellar keratoplasties, corneal epithelium limbal stem cells transplantation), the conjunctiva (conjunctival transpositions, amniotic membrane transplantation, caliciform cells implants) and the eyelids (various partial or extensive defects reconstructions; entropion, ectropion and symblepharon reconstructions; lagophthalmos). Sometimes it is required to turn to keratoprosthesis implantation.

Keratoprosthesis is an exceptional measure for severe bilateral keratopathy, for which cornea transplant has bad prognosis (graft survival rate is less than 50%). Therefore, it is only indicated for a single eye or, in bilateral cases, for surgery in the eye with better prognosis and previously guaranteeing retinal function to make possible for the patient to see. It is mainly *indicated* for leucomatous corneas with deep vascularisation, in which the corneal tissue has been substituted for fibrous tissue: chemical burns, advanced trachoma, advanced xerophthalmia, sclerocornea,

neuroparalytic keratitis, Stevens-Johnson, Lyell, pemphigus, total leucoma with considerable limbus anomalies, repeated or expected keratoplasty failure.

Basically, there are two types of keratoprosthesis, *synthetic* (artificial corneas), currently made of materials with higher biocompatibility and of better tolerance, with less complications during the postoperative period than the older models; and *osteo-odonto-keratoprosthesis*, which use a tooth from the patient as support of the artificial cornea, probably offering more long-lasting results. In both cases the main problem is the risk of rejection, being still very high with both types.

A. Rationale for the proposal^{1,2,3,4,5,6}

► Disease epidemiological data (incidence and prevalence).	Severe ocular surface alterations incidence is estimated at 400 cases/year ¹ . Cornea transplant prevalence is 60/millions people/year and the incidence is 40-50/million people/year ^{2,3} .
► Data on the use of complex ocular surface reconstruction technique.	There is no data available on limbal or amniotic membrane transplantations. In relation to keratoprosthesis, it is reserved for those cases in which cornea transplant surgery and combination of limbal with amniotic membrane and cornea transplantations have failed. <i>1 keratoprosthesis procedure is estimated out of 500-1000 cornea transplants.</i>

B. Guidelines to be followed by Centres, Services and Units in order to be designated as Reference Centres, Services and Units for complex ocular surface reconstruction.

► Experience of the Reference Centres, Services and Units: - Activity: <ul style="list-style-type: none"> • Number of complex ocular surface reconstructions (minimum and optimal) that should be performed to ensure an adequate care. 	- Cornea, limbal and amniotic membrane transplant and keratoprosthesis: Minimum 50. Optimal ≥ 100 .
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<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 programs^a.
<p>► Specific resources of the Reference Centres, Services and Units:</p> <ul style="list-style-type: none"> - Human resources required for the adequate implementation of complex ocular surface reconstruction. <p>Professional experience^b:</p> <ul style="list-style-type: none"> - Specific equipment required for the adequate implementation of complex ocular surface reconstruction. <p>► Resources from other units and services besides those belonging to the Reference Centres, Services and Units required for the adequate execution of complex ocular surface reconstruction.</p>	<ul style="list-style-type: none"> - 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 ocular plastic surgery and eyelid reconstruction techniques. - Ophthalmologists with experience in ocular surface, specially keratoplasty, amniotic membrane transplantations and glaucoma. - Ophthalmologists with experience in dacryology. - Nursing staff with experience in the care of patients with eye pathologies. - The service must have health authorization from the corresponding region for cornea, limbal and amniotic membrane transplants. - Ocular electrophysiological tests. - Collaboration with dentists for osteo-odonto-keratoprosthesis cases. - Pathological anatomy (tumoural pathologies assessment, ocular surface impression cytology assessment, etc.) - Electromyography for study of palpebral muscle function. - Clinical analysis (tear osmolality, PCR, lactoferrine assessment, lysozyme, etc) - Radiology.

<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 complex ocular surface reconstruction which at least must include: <ul style="list-style-type: none"> - Medical record number. - Date of birth. - Sex. - Admission date and discharge date. - Diagnosis procedures performed to the patient (ICD-9-CM). - Main diagnosis (ICD-9-CM). <ul style="list-style-type: none"> • Clinical situation at time of diagnosis (ROP stage) - Number and type of therapeutic procedures provided to the patient (ICD-9-CM): <ul style="list-style-type: none"> • Surgical procedures. • Keratoprosthesis type. • Other therapeutic procedures. - Date of the procedures. - Treatment results: <ul style="list-style-type: none"> • Pre- and post-operative final visual acuity. • Condition of the ocular surface and the preocular tear film. • Ocular and palpebral motility. • Survival time of the grafts/keratoprosthesis. • Aesthetic improvement and patient satisfaction. - Complications. - Patient's progress. - 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.

^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 Latkany R: Ocular Surface 2006;4:44-53.

² Dohlman CH. Keratoprosthesis. In: Krachmer JH, Mannis MJ, Holland EJ eds: Cornea. Surgery of the cornea and conjunctiva, vol III, St Louis: Mosby-Year Book; 1997; 1855-63.

³ Barron BA. Prosthokeratoplasty. In: Kaufman HE, Barron BA, McDonald, Walkman SR eds. The cornea. New Cork: Churchill Livingstone; 1988; 787-803

⁴ Shimazaki J, et al. Amniotic membrane transplantation for ocular surface reconstruction in patients with chemical and thermal Ophthalmology. 2000 Mar; 107(3): 411-2.

⁵ Rodríguez Martín J, *et al.* Transplante de limbo en patología de la superficie ocular. Arch Soc canar Oftalmol, 2003: 14.

⁶ López-García JS, *et al.* Tratamiento de la insuficiencia limbal grave mediante cirugía combinada de trasplante de limbo y trasplante de membrana amniótica. Arch Soc Esp Oftalmol. 2005, 80: 405-412.

⁵ Mateos E. Cirugía Plástica Ocular. Madrid; Ed. SEO. 2005.