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.

4. CONGENITAL ANOMALIES OF THE EYE DEVELOPMENT (globe and eye lids defects)[1,2,3,4,5]

Congenital anomalies of the ocular development include many, rare, and varied conditions that may be classified as follows:

1 Defects of the globe: anophthalmos, microphthalmos.
2 Defects of the lids: ptosis, epicanthal folds, entropion, ectropion, trichiasis, cryptophthalmos, ankyloblepharon, colobomas, etc.
3 Defects of the ocular surface: dermoid cysts, dermolipoma, sclerocornea, dystrophies.
4 Anterior chamber dysgenesis.
5 Defects of the iris: aniridia, coloboma, polycoria, corectopia, etc.
6 Persistent foetal vasculature syndrome: hyperplastic primary vitreous.

Defects of the ocular surface and defects of the iris were included under the file "Complex ocular surface reconstructions", anterior chamber dysgenesis was included under "Congenital glaucoma and childhood glaucoma", and the persistent foetal vasculature syndrome under "Advanced retinopathy of prematurity". Therefore, this piece will deal with congenital defects of the globe and eye lids.

A. *Rationale for the proposal*

► Epidemiological data on congenital anomalies of the globe and eye lids (incidence and prevalence).	In Spain, it is estimated an incidence of 21.34 anophthalmos and microphthalmos for every 100,000 births, approximately 84 cases every year. In terms of congenital eye lids defects, 240 cases a year are estimated[1,2,3].

B. ***Guidelines to be followed by Centres, Services and Units in order to be designated as Reference Centres, Services and Units treating congenital anomalies of the globe and eye lids.***

► Experience of the Reference Centres, Services and Units: - Activity: • Number of procedures that should be performed in a year to ensure an adequate care. - Other data: research on the subject, postgraduate teaching, continuing training, etc.	 - >50 eyelid surgeries (ptosis, epicanthal folds, entropion, ectropion, trichiasis, cryptophthalmos, ankyloblepharon, colobomas, palpebral reconstruction, etc.) in children per year (an average of 50 procedures per year in 5 years). - >30 enucleations and orbital cavity reconstructions. - Accredited postgraduate teaching. - Participation in research projects and publications in the field[a]. - Continuing training programs[a].
► Specific resources of the Reference Centres, Services and Units: - Human resources required for the adequate care of congenital anomalies of the globe and eye lids. Professional experience[b]:	 - 2 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 eyelid surgery (an average of 50 procedures per year in 5 years) and in enucleations and orbital cavity reconstructions (an average of 30 procedures per year in 5 years). - Nursing staff with experience of at least 5 years in the care of paediatric patients and in ocular pathology.

- Specific equipment required for the adequate care of congenital anomalies of the globe and eye lids. ► Resources from other units and services besides those belonging to the Reference Centres, Services and Units required for the adequate care of congenital anomalies of the globe and eye lids.	- Equipment for routine ophthalmologic examination. - CT scan and MRI for orbit exploration in anophthalmos. - Anaesthetic services with experience in paediatric patients[b]. - Intensive care unit with experience in paediatric patients[b]. - Paediatric services. - Radiology. - ENT services. - Maxillofacial services. - Neurology services. - Neurosurgical services. - Plastic surgery services.
► Clinical results indicators of the Reference Centres, Services and Units [c]:	**The indicators will be agreed with the Units that will be designated.**
► 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)	- Filling up the complete MBDS of hospital discharge. - The unit must have a *registry of patients* with congenital anomalies of the globe and eye lids which at least must include: - 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). • Type and characteristics of the congenital defect of the globe and eye lids. - Number and type of therapeutic procedures provided to the patient (ICD-9-CM): • Surgical procedures performed associated to congenital defects of the globe and eye lids (ptosis, epicanthal folds, entropion, ectropion, trichiasis, cryptophthalmos, ankyloblepharon, colobomas, palpebral reconstruction, enucleations, orbital cavity

	reconstructions, etc.) • Other therapeutic procedures. - Results of the care procedures: • External prosthesis adaptation at the end of the treatment. • Congenital defect corrected a year after. • Patient satisfaction. - Intraoperative and postoperative complications (ICD-9-CM). - 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.*

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