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.

3. CONGENITAL GLAUCOMA AND CHILDHOOD GLAUCOMA[1,2,3,4,5,6,7,8]

The most commonly used *classification* for congenital glaucoma is the Shaffer-Weis classification, which categorizes it from an etiological perspective:

A/ Primary congenital glaucoma due to abnormal development of the angle structures: It appears clinically isolated, as an entity in itself and not related to other syndromes or ocular defects, except for the alterations of the trabecular meshwork causing ocular hypertension.
B/ Glaucoma associated with other ocular and/or systemic anomalies: Certain syndromes affecting more than one organ such as aniridia, Axenfeld-Rieger anomaly, Peters anomaly, Lowe syndrome, Sturge-Weber syndrome, neurofibromatosis, etc.
C/ Secondary glaucoma following other ocular conditions such as trauma, infections, tumours, etc.: It occurs together with other malformations or ocular defects, being noteworthy:

- Glaucoma complicating the evolution of vitreo-retinal defects, vitreo-retinal dysplasia, hyperplastic primary vitreous, retinopathy of prematurity.
- Glaucoma as final stage of certain ocular pathologies such as long-standing retinal detachment, certain ocular traumas, Coast disease, ocular tumours, etc.
- Glaucoma following uveitis, especially after anterior and intermediate types.
- Glaucoma following a cataract surgery.
- Glaucoma due to dislocation of the crystalline during childhood (Marfan syndrome, Weii-Marchesani, homocystinuria).

Primary congenital glaucoma is a condition characterized by high ocular pressure and anomalies in the embryonic development of the iridocorneal angle structures, not associated with other ocular or systemic anomalies. This elevated intraocular pressure during the first months/years of life causes a thinning of the sclera, less rigid at that age, and turns into severe and irreversible changes of the ocular structures (optical nerve and globe)

The *diagnosis*[3,7] is suspected by clinical exam and confirmed through a series of tests that require anaesthetic sedation: Measurement of corneal diameter; calculation of intraocular pressure (IOP); exam of the anterior segment and gonioscopy (by using Goldmann, Koeppe, Ritch and Worst

lenses, the most widely used since they allow visualizing the angle as it will be seen during surgery); ophthalmoscopy to examine the papillae and evaluate the effects on the optical nerve. Determining the axis length of the globe is important for diagnosing and monitoring the evolution of the patient with glaucoma.

These patients will require during their first years of life (until three or four years of age) multiple examinations under anaesthetic sedation, for diagnostic purposes as well as for monitoring.

Treatment[9,10,11,18,19,21] is surgical; medical treatment is consigned to preparation for the surgical procedure or to cases where surgical procedures to control the progress of the glaucomatous damage fail.
The surgical techniques[5] used depend on the degree of development of the glaucomatous damage. Thus, if the angular structures are visible, goniotomy is preferred, providing good results. If the angle is not visible at the time of surgery, then trabeculectomy is used.
Failure of these techniques forces the use of other procedures, similar to those used for adults, with higher long term risks for children, such as simple trabeculectomy, trabeculectomy using antimitotics (its use in children is controversial), insertion of drainage devices, and the cyclodestruction of the ciliary body using laser.
Diagnosis and early referral to a reference centre would allow providing the surgical techniques with the best results (goniotomy instead of tabeculectomy).

A. *Rationale for the proposal*

► Epidemiological data on congenital and childhood glaucoma (incidence and prevalence).	The *incidence* of primary congenital glaucoma, considering only group A, fluctuates from 1/10,000 to 1/15,000 newborns, according to the populations considered. If groups A, B and C are taken, the figure rises to 1/2,000 newborns. The approximate annual incidence, for group A only, is 30-45 cases in Spain out of 454,951 births (2004 rates from the Spanish National Statistics Institute). Considering all the groups (A, B, and C) the incidence rises to 250 cases per year. The incidence expected in each region would be as follows (2004 birth rates from the Spanish National Statistics Institute is used):

	<table><tr><th>Region</th><th>Number of births</th><th>Number of cases group A</th><th>Number of cases group A, B, and C</th></tr><tr><td>Andalusia</td><td>89,516</td><td>7</td><td>44</td></tr><tr><td>Aragon</td><td>11,484</td><td>1</td><td>6</td></tr><tr><td>Asturias</td><td>7,324</td><td>< 1</td><td>3</td></tr><tr><td>Balearic Islands</td><td>10,717</td><td>1</td><td>6</td></tr><tr><td>Canary Islands</td><td>19,073</td><td>2</td><td>10</td></tr><tr><td>Cantabria</td><td>4,961</td><td>< 1</td><td>2</td></tr><tr><td>Castile-La Mancha</td><td>17,900</td><td>2</td><td>9</td></tr><tr><td>Castile and Leon</td><td>18,923</td><td>1</td><td>9</td></tr><tr><td>Catalonia</td><td>77,176</td><td>6</td><td>38</td></tr><tr><td>Valencian Community</td><td>48,990</td><td>4</td><td>25</td></tr><tr><td>Extremadura</td><td>9,897</td><td>1</td><td>5</td></tr><tr><td>Galicia</td><td>20,692</td><td>2</td><td>10</td></tr><tr><td>La Rioja</td><td>2,919</td><td>< 1</td><td>1</td></tr><tr><td>Madrid</td><td>69,727</td><td>6</td><td>35</td></tr><tr><td>Murcia</td><td>16,709</td><td>2</td><td>8</td></tr><tr><td>Navarre</td><td>6,342</td><td>< 1</td><td>3</td></tr><tr><td>Basque Country</td><td>19,756</td><td>2</td><td>10</td></tr><tr><td>Ceuta</td><td>1,282</td><td>< 1</td><td>< 1</td></tr><tr><td>Melilla</td><td>1,189</td><td>< 1</td><td>< 1</td></tr></table> In a Spanish research analyzing more than a million births in order to evaluate congenital ocular malformations[12,13,14,15], the *prevalence* of congenital glaucoma was 2.85/100,000 newborns. Glaucoma is bilateral in 75-80% of the cases and affects more to males (63%)[7,8,9,10].
► Data on the use of diagnosis and therapeutic procedures.	Hospital Clínico San Carlos in Madrid[21] has the highest statistics on diagnosis and treatment of congenital glaucoma in Spain; 573 cases have been examined between 1969 and 1999.

B. ***Guidelines to be followed by Centres, Services and Units in order to be designated as Reference Centres, Services and Units treating congenital and childhood glaucoma.***

► Experience of the Reference Centres, Services and Units: - Activity: • Number of procedures that should be performed in a year to ensure an adequate care.	10 congenital glaucoma surgical procedures in a year and 10 procedures related to other complex types of secondary glaucoma (traumatic, aphakic, etc.) appearing during paediatric age are required to maintain a level of knowledge and experience needed in the medical and surgical treatment as well as for postoperative management and patient monitoring.
- Other data: research on the subject, postgraduate teaching, continuing training, etc.	- 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 and childhood glaucoma.	- Ophthalmology unit with, at least, 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.
Professional experience[b]**.**	- Ophthalmologists with experience of at least 5 years in congenital glaucoma surgery and in the care of paediatric patients. - Unit staff with experience of at least 5 years in the care of paediatric patients, ocular pathologies and vision rehabilitation (with knowledge of the visual characteristics of the amblyopic eye in order to determine visual acuity, and of the basic guidelines for eye patching in order to avoid deprivation amblyopia in the healthy eye).

- Equipment required for the adequate treatment of congenital and childhood glaucoma.	- Specific equipment: - Portable slit lamp. - Gonioscopy lenses for examination and surgery. - Goniotomy knives. - At least one of the following techniques for evaluating and assessing the development of glaucoma damage: digital photography, OCT, GDX or HRT. - Ophthalmology services basic equipment: Surgical microscope with coaxial light, tonometers, portable biometer to measure the axial length of the globe, binocular ophthalmoscope, manual and computer campimeters.
► Resources from other units and services besides those belonging to the Reference Centres, Services and Units required for the adequate care of congenital and childhood glaucoma.	- Anaesthesia services with experience in paediatric patients[b]. - Intensive care unit with experience in paediatric patients[b]. - Paediatric Services.
► Procedure and 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 and childhood glaucoma which at least must include: - Medical record number. - Date of birth. - Sex. - Admission date and discharge date. - Patient's age at the time of admission in the centre. - Patient's age at the time of diagnosis.

	- Diagnosis procedures performed to the patient (ICD-9-CM). - Main diagnosis (ICD-9-CM) • Etiology of glaucoma. • Clinical situation at the time of diagnosis: Mainly the condition of corneal transparency. - Number and type of theraputic procedures provided to the patient (ICD-9-CM): • Surgical techniques performed: Goniotomy, trabeculotomy, trabeculectomy and others. • Other therapeutic procedures. - Techniques results: • Control of intraocular pressure using tonometry. • Control of globe growth using biometry. - Number and type of repeat surgeries: For goniotomy more than one may be needed, for trabeculotomy and trabeculectomy only one, and when these procedures fail a cyclodestruction with laser or a drainage device is then performed. - Intraoperative and postoperative complications (ICD-9-CM): • Anterior chamber haemorrhage. • Corneal leukoma. • Cataract. - Final condition after visual rehabilitation care: Visual acuity at the end of treatment to recover from amblyopia. - Follow-ups. - 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] Collins ET. Congenital defects of the iris and glaucoma. Trans Ophthalmol Soc UK 1893; 13: 114.
[2] Cross FR. Congenital hidrophthalmos. Trans Ophthalmol Soc UK 1896; 16: 340
[3] García Sanchez J, Zato M, Fernández – Vila PC. Glaucoma congénito Madrid, 1982; 16 –17.
[4] Rojas, B, Ramírez, AI, De-Hoz, R, et al. Cambios estructurales del ángulo de la cámara anterior en el glaucoma congénito: Comparación con el desarrollo normal. Arch Soc Esp Oftalmol 2006 Feb.
[5] Becker B. Shaffer R. Diagnosis and theraphy of the glaucomas. 2nd Ed. St. Louis; 1961.
[6] Hoskins HD Jr, Shaffer RN, Hetherington J. Anatomical classification of developmental glaucomas. Arch Ophthalmol 1984; 102:1.331-1.334.
[7] Wong PC, Dickens CJ, Hoskins HD. Duane´s Clinical Ophthalmology. Vol 3. Philadelphia: J.B. Lippincott Company, 1995;1-18.
[8] Shields MB. Glaucomas del desarrollo con anomalías asociadas. In: Panamericana Ed. Glaucomas Shields 2nd. Ed. Buenos Aires: Ester Levi A, 1987: 208-222.
[9] Fonseca A, Abelairas J, Rodríguez Sánchez JM, Peralta J. Actualización en cirugía oftálmica pediátrica. Madrid, 2000.
[10] Cioffi GA.Goniotomy and trabeculotomy Glaucoma surgery 2nd Ed. American Academy of Ophthalmology San Francisco 1998; 196-203.
[11] Azuara-Blanco A, Wilson RP, Spaeth GL, Schmidt CM, Augsburg C. Filtration procedures supplemented with mytomicin C in the management of childhood glaucoma. Br J Ophthalmol1999 Feb; 83(2): 151-156.
[12] 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.
[13] 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.

[14] Bermejo Sánchez E, Ayala Garces A, Félix Rodríguez V, Martín Bermejo M, Blanco García M, Egues Jimeno J, Huertas Camacho H, Jiménez Munoz-Delgado N, Paisan Grisolia L, Martínez-Frías ML. [Anophthalmia/micro-ophthalmia in syndromes: epidemiology study of newborns in Spain] An Esp Pediatr 1996 Sep; 45(3):269-75.
[15] Bermejo Sánchez E, Ayala Garces A, Felix Rodríguez V, Martín Bermejo M, Blanco García M, Egues Jimeno J, Huertas Camacho H, Jiménez Munoz-Delgado N, Paisan Grisolia L, Martínez-Frías ML [Sindrome de Anophthalmia/micro-ophthalmia: estudio de epidemiología de recién nacidos en España. Am J Med Genet A 2006 Jun 1; 140 (11): 1148-55.
[16] Vogt G,Horvath-Puho E, Czeizel AE. A population-based case-control study of isolated primary congenital glaucoma. Am J Med Genet A 2006 Jun 1; 140 (11):1148-55.
[17] Trivedi RH, Wilson ME Jr, Golub RL. Incidence and risk factors for glaucoma after pediatric cataract surgery with and without intraocular lens implantation. J AAPOS 2006 Apr; 10(2):117-23.
[18] Beck AD. Diagnosis and management of pediatric glaucoma Ophthalmol Clin North Am 2001 Sep; 14(3):501-12.
[19] Meyer G, Schwenn O, Grehn F. [Trabeculotomy in congenital glaucoma: comparison to goniotomy]. Ophthalmologe 2000 Sep; 97(9):623-8.
[20] Walton DS, Katsavounidou G. Newborn primary congenital glaucoma: 2005 update. J Pediatr Ophthalmol Strabismus 2005 Nov-Dec; 42(6):333-41; quiz 365-6. Comment in: J Pediatr Ophthalmol Strabismus 2005 Nov-Dec; 42(6):332.
[21] García Sánchez J. Estudio retrospectivo de glaucoma congénito de 1969 a 1999.