46. COMPLEX PAEDIATRIC NEUROSURGERY

Due to the fact that neurosurgical pathology of children is completely different in many aspects to the adult, Paediatric Neurosurgery appears first in the USA with Ingraham and Matson during the 1950s and later in Europe.

Complex paediatric neurosurgical pathology includes a group of special clinical conditions with low prevalence triggering the need for Reference Centres, Services and Units with the purpose to resolve them with efficiency. The current tendency to manage many of these pathologies is treating them in a multidisciplinary way in specific units where specialists in paediatric neurosurgery collaborate in a comprehensive way with other specialists. Under these circumstances are considered the following conditions:

- **Syndromic craniosynostosis:** This group of rare pathology includes more than 150 syndromes that are related to complex craniosynostosis causing severe cranial deformities limiting the normal encephalic development. The most common are: Crouzon, Apert, Saethre-Chotzen, and Pfeiffer syndromes, etc., evolving with premature closure of two or more cranial suture and other anomalies. Some of these syndromes frequently show intracranial hypertension, hydrocephalus, Arnold-Chiari and severe respiratory and deglutition disorders.

- **Complex tumours:** These are tumours which given their location, size and intrinsic tumour characteristics are difficult to remove in a satisfactory way. These fundamentally include those in brain basal ganglia, third ventricle, hypothalamic-pituitary region, brain stem, pineal region, cerebellopontine angle, cranial base and spinal cord. Also those large tumours in the lateral ventricles and in the functional cortical brain areas requiring cortical mapping, and some in the vertebral column which due to their extension, size and localization are complex and require treatment in collaboration with units of paediatric spine surgery.

- **Vascular pathology:** Within the brain, this group includes arterial aneurysms, arteriovenous malformations, vascular stenosis, venous malformations and vascular pathologies or tumours requiring brain revascularization. The spinal cord and close areas include arteriovenous fistulas, arteriovenous malformations and aneurysms. Treatment requires experts in vascular microsurgery and endovascular procedures.

- **Craniospinal defects:** Complex conditions in this group are rare and include: large encephalocele in the base or cranial convexity, extensive aplasia cutis congenita in the vertex area, hypertelorism with cranioencephalic anomalies, extensive dysplasia of the cranial base, Chiari malformation with compression from the odontoid, major stenosis of the craniovertebral region related to achondroplasia, mucopolysaccharidosis and neurofibromatosis, severe craniovertebral instability (in Down syndrome or other pathologies), difficult
rachischisis (large cervical, dorsal or lumbar myelomeningoceles), large intra- or extraspinal lipomas with medullary-radicular tethering, caudal regression syndrome, tethered cord related to vertebral or dural anomalies.

- **Other complex pathologies:** Those pathologies requiring collaboration of a more expert service in specific topics, such as some neurosurgical complications of premature babies with intraventricular haemorrhage and severe infections of the central nervous system.

**A. Rationale for the proposal**

<table>
<thead>
<tr>
<th>Epidemiological data (incidence and prevalence).</th>
<th>In Spain, the estimated incidence per year is:</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Cranial suture pathology: 1/3,000 newborns.</td>
<td></td>
</tr>
<tr>
<td>- Open lumbosacral dysraphia: 1/10,000 newborns.</td>
<td></td>
</tr>
<tr>
<td>- Close lumbosacral dysraphia: 1/5,000 newborns.</td>
<td></td>
</tr>
<tr>
<td>- Complex craniofacial syndromes: 1/500,000 newborns.</td>
<td></td>
</tr>
<tr>
<td>- Carniopagus: 1/1,000,000 newborns.</td>
<td></td>
</tr>
<tr>
<td>- Complex vascular pathology: 1/10,000 children &lt; 14 years old.</td>
<td></td>
</tr>
<tr>
<td>- Brain tumours: 2-3 cases/100,000 children &lt; 14 years old.</td>
<td></td>
</tr>
<tr>
<td>- Hydrocephalus during children development: 1/1,000 children &lt; 14 years old, of these 10% are complex hydrocephalus.</td>
<td></td>
</tr>
<tr>
<td>- Large premature babies with risk of intraventricular haemorrhage: 1/1,000 newborns.</td>
<td></td>
</tr>
<tr>
<td>50% &lt; 750 g</td>
<td></td>
</tr>
<tr>
<td>25% ≥ 750 &lt;1,000 g</td>
<td></td>
</tr>
<tr>
<td>12% ≥ 1,000 &lt; 1,200 g</td>
<td></td>
</tr>
</tbody>
</table>

**B. Guidelines to be followed by Centres, Services and Units in order to be designated as Reference Centres, Services and Units performing complex paediatric neurosurgery:**

- Experience of the Reference Centres, Services and Units:
- **Activity:**
  - Number of paediatric neurosurgical procedures that should be performed in a year to ensure an adequate care.

- **Other data:** research on the subject, postgraduate teaching, continuing training.

- **Activity:**
  - Number of procedures that should be performed in a year similar to those specific to the designation requested to ensure an adequate care in complex paediatric neurosurgery.

- **Minimum of 150 non-complex paediatric (≤14 years old) neurosurgical procedures in a year as average in the last 3 years, including, at least:**
  - 15 surgeries for craniosynostosis.
  - 15 surgeries for brain tumours.
  - 20 surgeries of cephalorachidian fluid pathologies (non-tumoural hydrocephalus, slit ventricle, benign intracranial hypertension, chronic valvular dysfunction)
  - 15 spine procedures (myelomeningoceles, Chiari, spinal cysts, tethered cord, extramedullary spinal tumours, spinal disc hernias, spine fixation, spasticity).

- **Minimum of 15 (optimal 30) complex paediatric (≤14 years old) neurosurgical procedures as average in the last 3 years.**

  Complex procedures: Syndromic craniosynostosis, complex tumours, complex vascular pathology (arterial aneurysms, arteriovenous malformations, venous malformations, brain revascularization and arteriovenous fistulas), craniospinal defects (large encephaloceles in the base or cranial convexity, extensive aplasia cutis congenital, hypertelorism with cranioencephalic anomalies, extensive dysplasia of the cranial base, Chiari malformation with compression from the odontoid, major stenosis of the craniocervical region related to achondroplasia, mucopolysaccharidosis and neurofibromatosis, severe craniocervical instability, difficult rachischisis, intra- or extraspinal lipomas with medullary-radicular tethering, caudal regression syndrome, tethered cord related to vertebral or dural anomalies), neurosurgical complications of premature babies with intraventricular haemorrhage and severe infections of the central nervous system.

- **Accredited postgraduate teaching:** Unit participation in the internship and residency programme of the Centre.
| publications, etc. | - Participation in research projects and publications in the field.
- Continuing training programme standardized and authorized by the centre’s board of directors.
- Clinical multidisciplinary sessions, at least once a month, in order to make clinical decisions and coordinate treatments. |
| Specific resources of the Reference Centres, Services and Units: | - Human resources required for adequately performing complex paediatric neurosurgery.
- Basic education of the team members. |
| [►] | - Specific equipment required for adequately performing complex paediatric neurosurgery. |
| Multidisciplinary care provided by: | - Neurosurgeons with 5 years experience, at least one of them, in paediatric pathology and performing a minimum of 125 neurosurgical procedures in a year in children ≤14 years old, as average in the last 3 years.
- Specialists with at least 5 year experience in paediatric neurosurgical pathology.
- Neurophysiologist with at least 5 year experience in paediatric neurosurgical pathology.
- Hospital and surgical nursing staff with experience in neurosurgical paediatric patients. |
| - 2 neurosurgeons, dedicating >75% of their work time to treating paediatric neurosurgical pathology.
- 2 specialists with experience in paediatric neurosurgical pathology.
- 1 neurophysiologist, available for intraoperative neurosurgical monitoring.
- Neurosurgical care available 24 hours, given the need for post operative monitoring. | - Hospital and surgical nursing staff.
- Hospitalization unit:
  - Availability of hospital beds where 24 hours monitoring (EEG/ICP/Other) as well as supervision (24 hours a day) of admitted patients by qualified staff is ensured.
- Hospitalization must be paediatric, in an environment adequate for the child needs, in exclusive paediatric areas.

<table>
<thead>
<tr>
<th>Surgical equipment:</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Specific standard and microsurgical instruments, including high speed drills and materials for cranial remodelling.</td>
</tr>
<tr>
<td>• Surgical microscope.</td>
</tr>
<tr>
<td>• Neuronomapper and/or stereotaxis system.</td>
</tr>
<tr>
<td>• Paediatric neuroendoscope.</td>
</tr>
<tr>
<td>• Ultrasonic aspirator.</td>
</tr>
<tr>
<td>• Bipolar coagulator, radiofrequency and laser sources available.</td>
</tr>
<tr>
<td>• Ultrasound scanner for intraoperative use to localize injuries.</td>
</tr>
<tr>
<td>• Portable angiography scanner available.</td>
</tr>
<tr>
<td>• Intracranial pressure (ICP) monitoring equipment with a system for graphic recording of the signal.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Neurophysiological equipment:</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Digital video-EEG equipment for continuous monitoring (with at least 64 channels). Electrocorticography</td>
</tr>
<tr>
<td>• Cortical mapping equipment with stimulation electrodes.</td>
</tr>
<tr>
<td>• Short and long latency evoked potentials equipment for intraoperative monitoring.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Neuropsychological equipment:</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Neuropsychological test battery for brain dysfunction assessment.</td>
</tr>
</tbody>
</table>

- Anaesthesia services/unit with experience in paediatric patients with neurological pathology.
- Intensive care services/unit with experience in paediatric patients with neurological pathology.
- Paediatrics services/unit.
- Paediatric surgery services/unit.
- Oncology services/unit with experience in paediatric patients with neurological
- Radiotherapy oncology services/unit with experience in paediatric patients with neurological pathology.
- Diagnostic imaging services/unit with neuroimaging techniques, with experience in paediatric patients with neurological pathology and having available:
  - CT scan.
  - Brain angiography.
  - High resolution MRI and functional mapping.
- Rehabilitation services/unit available, with a specific paediatric area and with experience in paediatric patients with neurological pathology.
- Pathological anatomy services/unit with experience in paediatric neuropathology.
- Maxillofacial surgery services/unit with experience in paediatric patients with neurological pathology.
- Cardiovascular surgery services/unit with experience in paediatric patients.
- Plastic surgery services/unit.
- Orthopaedic surgery and trauma services/unit with experience in paediatric patients.
- Ophthalmology services/unit with experience in paediatric patients.
- ENT services/unit with experience in paediatric patients.
- Clinical psychology services/unit available with experience in paediatric patients with neurological pathology.
- Nuclear medicine services/unit available with experience in paediatric patients.
- Interventional radiology services/unit available.

<table>
<thead>
<tr>
<th>Procedure and clinical results indicators of the Reference Centres, Services and Units</th>
</tr>
</thead>
<tbody>
<tr>
<td>The indicators will be agreed with the Units that will be designated.</td>
</tr>
</tbody>
</table>

- Filling up the complete MBDS of hospital discharge.
- The unit must have a registry of patients who have undergone paediatric neurosurgery which at least must include:
  - Medical record number.
- Date of birth.
- Sex.
- Patient’s habitual region of residence.
- Admission date and discharge date.
- Type of admission (Emergency, planned, other).
- Type of discharge (Home, hospital transfer, voluntary, death, transfer to a healthcare centre, other.)
- Service in charge of patient’s discharge.
- Main diagnosis (ICD-9-CM).
  - Type of injury.
- Other diagnosis (ICD-9-CM).
- Diagnostic procedures provided to the patient (ICD-9-CM).
- Therapeutic procedures provided to the patient (ICD-9-CM):
  - Number and type of surgical procedure provided to the patient related to the surgery.
  - Other therapeutic procedures: Type of procedure and date when it was provided.
- Complications (ICD-9-CM):
  - After effects of surgery.
- Patient’s monitoring.

- 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: