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

27. PAEDIATRIC ORTHOPEDICS: ORTHOPAEDIC TREATMENT OF NEUROMUSCULAR DISORDERS (CEREBRAL PALSY, MYELOMENINGOCELE), CONGENITAL MALFORMATION (CONGENITAL SHORT FEMUR, TIBIAL/FIBULAR AGENESIS), BONE DYSPLASIAS (OSTEOGENESIS IMPERFECTA, ARTHROGRYPOSIS) AND MAJOR LIMB LENGHTHENING.

Paediatric orthopaedics involves a group of medical, surgical and rehabilitation treatments focused on correction of the musculoskeletal system's deformities and/or dysfunctions of the child and adolescent caused by a group of complex pathologies that may be classified in:

- **Congenital malformations**: including disorders such as proximal femoral focal deficiency or tibial/fibular agenesis, causing important defects, with shortening of the limb and joint alteration.
- **Bone dysplasias**: including alterations such as osteogenesis imperfecta, arthrogryposis, or achondroplasia, each of them with specific circumstances such as bone fragility, joint stiffness or dwarfism.
- **Neuromuscular disorders**: including **deformities secondary** to cerebral palsy or myelomeningocele, treatable through orthopaedic surgery.

The current tendency in caring for these pathologies involves multidisciplinary treatment where specialists in paediatric orthopaedic surgery collaborate with specialists in paediatric neurology, rehabilitation, neurosurgery and paediatric urology. The objective of the treatment is social integration of the child with the most physical ability and less psychic damage possible. After adequate clinical assessment of the patient it is fundamental to reach a diagnosis as precise as possible in order to understand the vital and functional prognosis of the patient.

There is not an objective evaluation system of results for these patients. For more of a decade, in neighbouring countries, Gait Analysis systems are being developed trying to register, in an objective way, the improvement of patients' gait by using video, electromyography and oxygen consumption. There are still few units in our country where this type of research may be performed in order to be clinically applicable.

Congenital malformations are very diverse, being detected in the newborn or in the first months of life. They may affect either upper and lower limbs or the spine. The degree of involvement is also diverse, from minor conditions which may be corrected with early actions to major deformities requiring complex surgical treatment. In any case, diagnosis in the first days of life is important in order to take action from the beginning, as in the congenital hip dislocation and club foot, whose condition worsens if the treatment is delayed. There are more striking malformations, such as proximal femoral focal deficiency or tibial/fibular agenesis, whose treatment requires, in a first surgical step, performing

joint reconstruction so that in a second step limb lengthening through external bone lengtheners is performed, a demanding technique with many possible complications.

Limb lengthening is a technique involving callus distraction after osteotomy by using a fixation system, a mixed method (external fixator and intramedullary nail) or an exclusive intramedullary distraction technique. After a few days waiting period, during which the haematoma originating from the osteotomy becomes organized, lengthening starts with a rate of 1mm/day, until the length difference is compensated or the desired length is reached. After this stage, the lengthened callus must consolidate until achieving the mechanical resistance desired and allowing supporting weight loads without deformation or fracture of the regenerate bone. Finally, when the bone has the desired characteristics, the external fixator is removed, placing temporal immobilization or protection during the following 2-4 months.
The external fixator, circular or monolateral, must provide enough stability during the lengthening phase and have available the required modularity in order to perform the appropriate adjustments when needed. The circular and monolateral systems are used indiscriminately at the tibial level; however, in other locations monolateral systems are used for better comfort of the patient.
The bone lengthening technique must be generally associated to concomitant surgery of soft tissue, such as tenotomy, in order to avoid joint retraction, along with a physiotherapy programme during the lengthening stage and afterwards.
Complications during the treatment period are numerous and diverse, with variable incidence and severity. Clinical monitoring of the patient must be very close in order to detect as soon as possible alterations that may be occurring so as to treat them and avoid long-term effects weakening the results.

Bone dysplasias are other important group of conditions during skeletal growth also affecting in a major way the life of these children. Conditions such as osteogenesis imperfecta have a complex treatment, requiring major commitment given the many fractures and progressive deformity that it involves. There are surgical techniques to perform realignment of deformed bones, such as the Fassier-Duval method, with telescopic intramedullary nailing that protects the bone as it grows. At the same time, the bisphosphonates treatment has a predominant role, usually performed through paediatric endocrinology. In arthrogryposis there is limiting joint stiffness which may only be mitigated by corrective surgeries and a long rehabilitation treatment. These are dependent but intelligent children who also need orthopaedic and social help during their whole life. Achondroplasia and other types of dwarfism require multiple lengthening surgeries in specialized centres.

In **myelomeningocele**, after surgical closure of the defect during the first hours and assessment whether there is need or not for valve bypass, the patient is clinically assessed to determine the degree of involvement, which decides the functional prognosis for each case. **From the orthopaedic surgery point of view** and given that it is a type of flaccid paralysis affecting lower extremities and spine, there must be special monitoring of the deformities affecting feet, knees, hips and spine specially in patients able to move. The most frequent surgical techniques performed on these patients are:

- Foot: surgery on club foot associated to the deformity; surgical treatment of talipes calcaneus and talipes equinusvalgus through tendon transfer; arthroereisis of the subastragalar joint according to the techniques of Grice, Judet-Cavalier or current variations. Surgery of bone deformities at ankle level by epiphysiodesis or osteotomy.
- Knee: Flexion deformity: Soft tissue surgery at the popliteal fossa height combining tendon transfers or extension osteotomies.
- Hip: Cases of dislocation or subluxation require relocation of proximal femur and/or acetabulum, with or without combining tendon transfer of the psoas muscle.
- Spine: Surgical treatment of dorsolumbar kyphoscoliosis.

In the case of **cerebral palsy**, after rehabilitation care during the first years of life and according to the success achieved, the purpose **from the orthopaedic perspective** is to perform the required surgeries in the musculoskeletal system before age 9-10, achieving at the same time the most appropriate social and school integration of the child, in order to avoid annual surgeries that disturb the child psycho-social relationship. The cerebral palsy type most frequently treated is the spastic type; monoplegia, diplegia or quadriplegia types indistinctively. In the latest two, the degree of acetabular coverage in hip must be watched since it will decisively influence the functional prognosis of the patient's walking and pain. Different surgical techniques are based in muscular unbalances, performing soft tissue surgery: tenotomies, tendon lengthening with or without being combined to osteotomies.
In patients with spastic quadriplegia, spine alignment and pelvic obliquity deserve special attention, generally associated to hip dislocation and subluxation.
Surgery in upper limbs is less frequently indicated, being performed in elbow or wrist flexion or thumb-in-palm, performing lengthening and/or tendon transfers or wrist arthrodesis.

A. ***Rationale for the proposal***

► Epidemiological data (incidence and prevalence).	Generally these are rare conditions; incidence varies from 1/50,000 newborns for osteogenesis imperfecta, 0.7/10,000 for arthrogryposis, 1/25,000 for achondroplasia to 0.9/10,000 for short congenital femur. More frequent are cerebral palsy, 2-2.5/1,000 newborns, although with variable degrees and complex cases being less frequents (more than 80% show musculoskeletal system deformities requiring correction), and myelomeningocele, with one case every 1,000 births per year, 50% of them showing musculoskeletal system deformities requiring correction. In both pathologies, musculoskeletal system deformities requiring referral to a reference unit of the National Health System would be the most severe, decreasing the frequency of these cases to 2.8/10,000 newborns.

	According to the Spanish National Statistics Institute, in 2006 there were 482,957 births in Spain, and following the previous data on incidence it could be estimated that in 2006 in Spain there were: - 9.7 newborns with osteogenesis imperfecta. - 33.8 newborns with arthrogryposis. - 19.3 newborns with achondroplasia. - 43.5 newborns with short congenital femur. - 1,207 newborns with cerebral palsy and 483 newborns with myelomeningocele (cases with musculoskeletal system deformities requiring referral to a reference unit of the National Health System would be the most severe: 135 newborns). Total without including cerebral palsy and myelomeningocele: 106 cases. Total including musculoskeletal system deformities secondary to cerebral palsy and myelomeningocele that would require to be transferred to a reference unit of the National Health System: 241 cases.

B. Guidelines to be followed by Centres, Services and Units in order to be designated as Reference Centres, Services and Units providing paediatric orthopaedics

► Experience of the Reference Centres, Services and Units: - Activity: • Number of paediatric orthopaedic procedures that should be performed in a year to ensure an adequate care.	- 30 complex procedures per year. Types: lengthening for short congenital femur or achondroplasia, hip neurological dislocation, multiple osteotomies and femur realignment in osteogenesis imperfecta with telescopic nail.
• Number of procedures that should be performed in a year of techniques, technologies and procedures similar to those specific to the designation requested.	- Between 100 and 150 simple procedures per year, related to similar pathologies with less complexity, such as: Achilles lengthening, ischiotibial lengthening, adductor tenotomy.

- Other data: research on the subject, postgraduate teaching, continuing training, etc.	- Accredited postgraduate teaching: unit participation in the internship and residency programme of the centre. - Participation in research projects and publications in the field[a]. - Continuing training programme standardized, and authorized by the centre board of directors. - Multidisciplinary clinical sessions (surgeons, neurologists, urologists, rehabilitation therapists…), at least once a month, in order to coordinate treatments.
► Specific resources of the Reference Centres, Services and Units:	
- Human resources required for the adequate performing of paediatric orthopaedics.	The Unit must have: - Surgeon coordinating the Unit. - At least, 2 surgeons specialized in orthopaedic and trauma surgery with specific dedication to paediatric orthopaedics. - Nursing staff, surgical auxiliaries and technicians. - Non medical staff.
- Basic education of the team members[b].	- At least 2 of the surgeons specialized in orthopaedic and trauma surgery with 5 years of experience in paediatric orthopaedics. - Nursing staff, surgical auxiliaries and technicians with experience in paediatric patients and care of these types of pathologies.
- Equipment required for the adequate performing of paediatric orthopaedics.	- Operating theatre ready for surgery on paediatric patients and orthopaedic surgery: orthopaedic surgery instruments, osteotomy and osteosynthesis instruments (drills, plates, intramedullary nails, external fixator and bone lengtheners), image magnifier.
► Resources from other units or services besides those belonging to the Reference Centres, Services and Units required for the adequate performing of paediatric orthopaedics[b].	- Paediatric services/unit with 3 years experience in treating this type of pathologies. - Neurology services/unit with 3 years experience in treating this type of pathologies and paediatric patients. - Neurosurgery services/unit with 3 years experience in treating this type of pathologies and paediatric patients.

	- Neurophysiology services/unit with 3 years experience in treating this type of pathologies and paediatric patients. - Urology services/unit with 3 years experience in treating this type of pathologies and paediatric patients. - Rehabilitation services/unit including physiotherapists with 3 years experience in orthopaedic treatment of congenital malformations, bone dysplasias and neuromuscular conditions and treating paediatric patients. - Endocrinology services/unit with 3 years experience in bone dysplasia and treating paediatric patients. - Intensive care services/unit with experience in treating paediatric patients. - Anaesthesia services/unit with experience in treating paediatric patients. - Radiodiagnosis services/unit with 3 years experience in treating this type of pathologies and paediatric patients. - Angiology and vascular surgery services/unit with experience in treating paediatric patients. - Plastic and reconstructive surgery services/unit with experience in treating paediatric patients. - Psychiatric services/unit with 3 years experience in treating this type of pathologies and paediatric patients. -Clinical psychology services/unit with 3 years experience in treating this type of pathologies and paediatric patients. - Social workers services/unit.
► 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* treated in the paediatric orthopaedics unit which at least must include: - First and last name. - Medical record number. - Date of birth and sex. - Home address and telephone number.

	- Prenatal and perinatal information. - Date of admission, surgery and discharge. - Circumstances of the discharge (home, hospital transfer, voluntary, death, transfer to a healthcare centre, other.) - Diagnostic procedures provided to the patient (ICD-9-CM): - Main diagnosis (ICD-9-CM). • Injury site and characteristics. - Number and type of therapeutic procedures provided to the patient (ICD-9-CM): • Surgical technique. • Other therapeutic procedures. - Surgical procedure results: functional improvement after treatment, degree of independence achieved after surgery, as well as the orthesis required after treatment. - Complications: Bedsores of pressure ulcers, peripheral venous thrombosis, infections, post operative fractures, etc. - Monitoring: Follow-up in doctor's office and operating theatre. Changes in the patient's degree of dependence, repeat surgeries. - 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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