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

32. COMPREHENSIVE CARE FOR THE NEWBORN WITH CONGENITAL HEART DISEASE AND FOR THE CHILD WITH CONGENITAL HEART DISEASE (ARISTOTLE ≥3)

Care of patients with congenital heart disease has improved in the last years to become a continuum covering from the foetal period to adulthood.

It may be classified according to the different periods of the patient's life and, in fact, this is followed by daily clinical practice; however, these patients optimal care benefits from the interaction and the close and continuous feedback among the various people involved in the different care stages. Therefore, an ideal reference service must be able to provide care at the best level and with the best quality rate throughout all the range of ages in those processes of high complexity. Frequently, these complex processes require continuing and continuous care only possible in services with this focus.

Procedures during the neonatal period bring together a great variety of very different pathologies, most of them with a very limited number of cases. These pathologies are also very variable in terms of complexity and results[1]. Classifying its complexity in terms of the European Aristotle score allows including the most complex pathologies in reference services or units[2]. Those in the higher levels of complexity (Aristotle 3-5) represent around 15-20% of all heart surgeries in children performed in Spain and include, among others, pulmonary atresia (when using extracorporeal circulation during surgery), complete atrioventricular canal defect (CAVC), tetralogy of Fallot, Fontan procedure, heart transplantation, total anomalous pulmonary venous drainage, transposition of the great arteries (TGA), truncus arteriosus, interruption of the aortic arch (IAA) and hypoplastic left heart syndrome.

Care of these rare pathologies of complex treatment, with results depending on the experience of the surgical teams and in the activity volume, recommends the proposal for reference centre, services and units.

Transportation of these patients is also an essential part of the comprehensive care since its conditions decisively affect the result of the surgical procedure and the prognosis of these patients.

A. Rationale for the proposal

► Epidemiological data on congenital heart disease (incidence and prevalence).	Every year in Spain there are around 400,000 children born. Eight out of every 1,000 are born with a heart condition; however, the average number of patients requiring a surgical procedure is 51 per million population (around 2,000 in the whole country per year). This number in Europe corresponds to 62 per million. From these, between 15 and 20% reach the level 3 or higher in the Aristotle risk score, which implies a number of 300-400 children/year with complex heart disease undergoing surgical procedure.
► Data on the use of diagnosis and therapeutic procedures.	In Spain a total of 2,000 surgical procedures and 1,000 therapeutic catheterization are performed in a year in the 17 centres with medical-surgical activity, performing 60% of the procedures with extracorporeal circulation and 40% with closed-heart surgery. Approximately 15-20% of the patients have been treated for complex heart disease.

B. Guidelines to be followed by Centres, Services and Units in order to be designated as Reference Centres, Services and Units treating newborns and children with congenital heart disease (Aristotle ≥3)

► Experience of the Reference Centres, Services and Units: - Activity: • Number of procedures that should be performed in a year to ensure an adequate care of newborns and children with congenital heart disease.	- 150 procedures in congenital heart disease (surgical procedures and percutaneous coronary interventions) per year in patients under 18 years old, at least 20% performed in newborns.

• Number of procedures similar to those specific to the designation requested that should be performed in a year to ensure an adequate care of newborns and children with congenital heart disease. - Other data: research on the subject, postgraduate teaching, continuing training, publications, etc.	- Hospital activity accredited in complementary techniques: ◆ Foetal echocardiography: 300 per year. ◆ Intraoperative, transesophageal, or transepicardial ultrasound in children: 75 per year. - Accredited postgraduate teaching: unit participation in the internship and residency programme in cardiology and cardiovascular surgery of the centre. - Participation in research projects and publications in the field[a]. - 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 decisions and coordinate treatments. - Availability of a surgical research programme authorized by the centre's board of directors, with special focus on circulatory care. The required training in more complex and less frequent techniques relies on this type of care. The possibility of having simulators for continuous education and training brings added value.
► Specific resources of the Reference Centres, Services and Units: - Human resources required for the adequate care of newborns and children with congenital heart disease.	- Surgical team available 24 hours a day, every day of the year, ready to perform surgery in less than 2 hours. - Interventionist cardiology team, available 24 hours a day, every day of the year, ready to perform surgery in less than 2 hours. - 3 surgeons from the hospital staff, full-time or part-time dedicated to heart surgery in newborn and children. - 5 cardiologists or paediatricians, full-time or part-time dedicated to congenital heart disease in the foetus and child, at least one of them engaged in interventionist cardiology.

	- 2 perfusionists, full-time or part-time dedicated to heart surgery in newborns and children. - Nursing, surgical, and hemodynamic staff. - 1 administrative assistant.
- Basic education of the team members [b].	- Cardiologists or paediatricians with experience and dedication in the last 3 years in paediatric patients with congenital heart disease. - Surgeons with 5 years experience in congenital heart disease surgery, at least one of them must have performed a total of 200 surgeries in the last 5 years. - Interventionist cardiology team with experience from having performed a minimum of 130 interventionist surgical procedures in congenital heart disease, in the last 2 years. - Perfusionists with experience in treating paediatric patients with congenital heart diseases, ECMO (extracorporeal membrane oxygenation) and circulatory care. - Nursing, surgical, and hemodynamic staff with experience in paediatric patients with congenital heart disease.
- Specific equipment required for the adequate care of newborns and children with congenital heart disease.	- High end hemodynamic room, biplane recommended, including flat detector and with the possibility to perform hybrid procedures. - Intraoperative transthoracic, transepicardial, and transesophageal ultrasound scanner available for all age groups. - High end ultrasound equipment with 3D reconstruction scanners. -Telemetry in hospitalization ward. - Circulatory care programme including ECMO and univentricular and biventricular care systems. - Complete cardiology equipment: Ergometry, oxygen consumption, pulse oximetry, Holter for arrhythmias and arterial pressure, defibrillators. - Habitual equipment for heart surgery: extracorporeal circulation; hypothermia equipment; gas, ion and pH control; complete monitoring.

- Resources from other units and services besides those belonging to the Reference Centres, Services and Units required for the adequate care of newborns and children with congenital heart disease [b].	- Electrophysiology unit with experience in paediatric patients with congenital heart disease. - Intensive care services/ unit with experience in paediatric patients with congenital heart disease, heart surgery post operative care, and training in ECMO. - Neonatal services/unit with experience in paediatric patients with congenital heart disease and heart surgery post operative care. - Anaesthesia services/unit with experience in paediatric surgery, paediatric heart surgery and treating paediatric and newborn patients. - Gynaecology and Obstetrics services/unit with the option to admit the mother before the birth of the child with a diagnosed congenital heart disease. - Foetal medicine programme, accredited by the Board of Directors of the Centre, including foetal interventionism and programming of measures to undertake before, during and after delivery, including the option to admit the mother before the birth of the child when he/she has a congenital heart disease. - Paediatrics services/unit with experience in patients with congenital heart disease. - Paediatric surgery services/unit. - Diagnostic imaging services/unit, performing a minimum of 50 CT scan and MR in children per year and able to perform complex cardiovascular analysis. - Rehabilitation services/unit with at least one physiotherapist with experience in paediatric patients with congenital heart disease. - Pathological anatomy services/unit, with experience in congenital heart disease. - Genetics services/unit. - Immunology services/unit. - Social workers services/unit. -Programme authorized by the centre's board of directors for transportation and accommodation of high risk patients, in conjunction with paediatrics services, A&E services, and transportation units. - Active heart transplantation programme and authorized according to the Royal Decree 2070/1999, December 30th, establishing the general basis for clinical harvesting and use of human organs and the territorial coordination in donation and

	transplantation of organs and tissues, allowing for response and continuity to the needs arising from the treatment of patients with complex congenital heart disease (complications or poor progress). - Facilities for surgical research, allowing long-term adequate staff training and qualifications in rare procedures, without morbidity costs for the patients. - It is advisable to have an integrated bidirectional information system with the patients' referral hospitals, allowing development of telemedicine programmes for overseen distance-care of the patients.
► 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. - Participation of the Unit in the European Registry "Congenital Database" [6] and in the Registry of the Spanish Society for Thoracic and Cardiovascular Surgery[4] is advisable. - The unit must have a *registry of patients* with congenital heart disease 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 congenital heart disease.

	- Other diagnosis (ICD-9-CM). - Diagnostic procedures provided to the patient (ICD-9-CM): Type of procedure and date when it was provided. - Therapeutic procedures provided to the patient (ICD-9-CM): Type of procedure and date when it was provided. ◆ Therapeutic catheterization. ◆ Surgical procedures performed to the patient. - Complications (ICD-9-CM). - Patient monitoring: New surgeries, interventionist catheterization, doctor's visits, etc. - 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 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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