33. COMPRENHENSIVE CARE FOR THE ADULT WITH CONGENITAL HEART DISEASE (CHD)

This medical-surgical unit must refer to the group of techniques, procedures and experience required to provide an optimal response to adult patients with congenital heart disease (CHD).

In short, the fundamental reasons justifying the creation of Units for Congenital Heart Disease in Adults are:
1. High prevalence: 250 cases per 100,000 population (115,395 patients in the National Health System).
2. High increase on patients with CHD growing up into adulthood in the next decades, demanding careful consideration of new care needs.
3. Complexity: Physiology of certain CHD previously repaired or palliate may be difficult to comprehend by the cardiologist without special training in paediatric cardiology. On the other hand, part of the medical problems in adults exceeds the point of view of paediatric cardiologists. Clinical assessment of these patients is complex, as well as the surgical technique, requiring surgeons with broad experience in these types of pathologies.
4. Placement of these patients in the hospitalization ward, doctor offices or examination rooms is also complicated. A paediatric environment is not adequate for the continuous care of adults growing up; however most of the cardiology services are not adapted yet to handle this demand. The traditional gap between paediatrics and adult medicine has to be broken since there is a need for work teams where cardiologists and cardiac surgeons with experience in children and adults collaborate[1].

Surgery of CHD is directed to a growing group of pathologies since there is a higher number of survivors of heart surgery during childhood requiring surgical correction of the heart disease long-term effects, acquired heart disease of a heart with a congenital condition (arrhythmia, valvulopathy, myocardiopathy, etc.), there are also congenital conditions with delayed manifestation (interauricular communication, anomalous drainage, mitral pathology, etc) requiring surgical correction. It is a heterogeneous group requiring treatment in a specialized unit since the techniques needed exceed the usual knowledge and techniques of a heart surgery unit for adults. Pulmonary failure and right ventricular dilation in operated tetralogy of Fallot, in particular, as well as Fontan procedure failure are increasing. Procedures are diverse: valve prosthesis or conduits, arrhythmia ablation, residual defects closure, etc. Most of them require extracorporeal circulation and in some cases ventricular assist and/or heart transplantation.

Surgery for congenital heart disease in adults may be[2]:
- Curative, if it corrects the defect without leaving long-term effects or residual defects.
- Reconstructive or corrective, when it reconstructs or repairs the structural defects leaving long-term effects or unavoidable residual damages.
- Palliative, alleviating the symptoms but leaving the basic structural defect without repair.
- Transplanting, replacing the affected organ: heart, lungs, or both.
- Mixed techniques: interventionist catheterization and surgery.

Under these conditions, patients with congenital heart disease may require a specialized unit when:
- Requiring emergency admission due to cardiovascular condition.
- Having to perform cardiac catheterization or electrophysiological study.
- Requiring a surgical cardiovascular procedure or through therapeutic catheterization.
- Requiring non-cardiac surgery when having a complex congenital heart disease.
- Requiring monitoring in high risk pregnancy units.

A. Rationale for the proposal

► Epidemiological data on congenital heart disease in adults (incidence and prevalence).	Congenital heart diseases are the most frequent defects at birth: 8-10 out of 1,000 children are born with a heart or major blood vessels defect and at least 6 out of 1,000 have a moderate or severe heart disease. Until not long ago, only 50% reach adulthood. According to the British Cardiac Society Working Party[3], 85% of children operated on congenital heart disease reach adulthood due to the development of heart surgery and percutaneous techniques. 110,000 is estimated to be the current number of adults with congenital heart disease, 250 cases per 100,000 population. In Spain, around 1,700 cases per year undergo surgery, meaning that 1,300 children reach adulthood. Out of these, a minority will require repeat surgeries; however, many will require specialized medical monitoring, electrophysiological studies, interventionism and hemodynamics. Nowadays, population of adults with Congenital Heart Disease includes two main groups: a) Heart disease with natural survival.

	b) Heart disease with some type of therapeutic procedure (surgery or percutaneous procedure) performed during childhood.
► Data on the use of techniques, technologies, or diagnosis and therapeutic procedures.	Those patients with more complex CHD (20-25%) may require monitoring in a specialized unit. 25-40% may be followed-up by their usual cardiologist, but they may require in the future access to specialized care. The rest of the patients (40%) with simple or repaired conditions would require little or no specialized monitoring.

B. Guidelines to be followed by Centres, Services and Units in order to be designated as Reference Centres, Services and Units treating congenital heart disease in adults

► 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 congenital heart disease in adults.	- 40 procedures in congenital heart disease (surgical procedures and percutaneous coronary interventions) per year in adults. (Correction of residual defects, closure of the intraventricular and interauricular communication, valve prosthesis, conduits, etc.)
• Number of procedures similar to those specific to the designation requested that should be performed in a year to ensure an adequate care of congenital heart disease in adults.	- The cardiovascular surgery services must perform 450 heart surgeries per year. - Participation, authorized by the centre's board of directors, in an active programme of congenital heart disease in children.
- Other data: research on the subject, postgraduate teaching, continuing training, publications, etc.	- 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 congenital heart disease in adults. - Basic education of the team members [b].	The unit must be located in an adult medical environment, coordinated with a unit of congenital heart disease in children. - 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. - 2 cardiologists, at least one specialized in congenital heart disease in adults. - 2 cardiac surgeons, full-time or part-time dedicated to congenital heart disease in adults. - 2 perfusionists working full-time or part-time - Nursing, surgical, and hemodynamic staff. - 1 administrative assistant. - Cardiologists with: ◆ 2 year experience in paediatric cardiology patients in a centre with an active programme in percutaneous surgery and interventionism. ◆ 2 years experience in treating adult patients with congenital heart disease. - Cardiac surgeons with 5 year experience in heart surgery and congenital heart disease, with minimum 2 years experience in treating adult patients with congenital heart disease. - Interventionist cardiology team with experience from having performed a minimum of 130 interventionist surgical procedures in congenital heart disease.

	- Perfusionists with experience in congenital heart diseases, ECMO and circulatory care. - Nursing, surgical, and hemodynamics staff with experience in patients with congenital heart disease.
- Specific equipment required for the adequate care of congenital heart disease in adults.	- 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. - Hospitalization: ◆ Hospitalization unit, within the general cardiology hospitalization ward or as a unit dedicated to CHD in adults. ◆ Telemetry in hospitalization ward. - Specific care for congenital heart disease in adults. - High end hemodynamic room, biplane recommended, including flat detector and with the possibility to perform hybrid procedures, intravascular and intracardiac ultrasound scanner (IVUS) available, with at least one specialist in hemodynamics with experience in percutaneous interventionism in congenital heart disease. - Intraoperative transthoracic, transepicardial, and transesophageal ultrasound scanner available for all age groups. - High end ultrasound equipment with 3D reconstruction scanners. - Circulatory care programme including ECMO and univentricular and biventricular care systems.
► Resources from other units and services besides those belonging to the Reference Centres, Services and Units required for the adequate care of congenital heart disease in adults [b].	- Electrophysiology unit with experience in patients with congenital heart disease, with at least one electrophysiologist with experience in treating arrhythmias in congenital heart disease, pacemaker ablation and implantation, and defibrillators. - Intensive care services/ unit with experience in patients with congenital heart disease, heart surgery post operative care, and training in ECMO. - Anaesthesia services/unit with experience in congenital heart disease in all age

	groups, especially in adults. - Gynaecology and Obstetrics services/unit, with experience in contraception, high risk pregnancy, foetal echocardiography. - Diagnostic imaging services/unit, able to perform cardiac imaging studies, with at least MR and CT scan; as well as a radiologist and cardiologist with experience in congenital heart disease who must perform and interpret the cardiac imaging studies. - Rehabilitation services/unit with at least one physiotherapist with experience in patients with congenital heart disease. - Genetics services/unit. - Pathological anatomy services/unit, with experience in congenital heart disease. - Immunology services/unit. - Internal medicine services/unit. - Pulmonology services/unit. - Neurology services/unit. - Endocrinology services/unit. - General surgery services/unit. - Nephrology 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 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

	patient's referral hospital, 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[7] 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). - 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. - 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.*

Bibliography:

1. Jakob Klcovansky, Lars Søndergaard, Morten Helvind, and Henrik Ørbæk Andersen Cardiac surgery in grown-up congenital heart patients. Will the surgical workload increase? (Institutional report - Congenital) Interact CardioVasc Thorac Surg 2008;7:84-89.
2. Daebritz SH. Update in adult congenital cardiac surgery. Pediatr Cardiol. 2007 Mar-Apr;28(2):96-104. Epub 2007 May 4.
3. Grown-up congenital heart (GUCH) disease: current needs and provision of service for adolescents and adults with congenital heart disease in the UK. Report of the British Cardiac Society Working Party. Heart. 2002 Sep;88 Suppl 1:i1-14.
4. Karamlou T, Diggs BS, Person T, Ungerleider RM, Welke KF. National practice patterns for management of adult congenital heart disease: operation by pediatric heart surgeons decreases in-hospital death. Circulation. 2008 Dec 2;118(23):2321-2.
5. Management of Grown Up Congenital Heart Disease The Task Force on the Management of Grown Up Congenital Heart Disease of the European Society of Cardiology European Heart Journal (2003) 24, 1035–1084.
6. Registro de la Sociedad Española de Cirugía Torácica y Cardiovascular (Registry of the Spanish Society for Thoracic and Cardiovascular Surgery). 2007 www.sectcv.es.
7. EACTS European Congenital Database. www.eacts.org.