analesdepediatría

www.analesdepediatria.org

EDITORIAL

The development of Spanish paediatric cardiology and its impact on congenital heart disease management☆

El desarrollo de la cardiología pediátrica española y su impacto en el manejo de las cardiopatías congénitas

D.C. Albert Brotons

Unidad de Cardiología Pediátrica, Hospital Universitari Vall d'Hebron, Barcelona, Spain

Paediatric cardiology began in Spain with the opening of the Paediatric Cardiology Units at the public children's hospitals La Paz in Madrid (1965) and Vall d'Hebron in Barcelona (1967). During those early years a very important part in its development was played by Dr. Manuel Quero, a distinguished figure with an international reputation.[1] From the 1990s, paediatric cardiac surgery developed at a national level to the point it has reached today, with numerous tertiary cardiological units appearing in Spain, where every kind of procedure is performed, with results similar to international standards.

The treatment of congenital heart disease has progressed spectacularly in the last few years. Thanks to advances in both diagnostic and therapeutic techniques, 85% of children born with heart conditions that were fatal a few years ago now survive to adulthood.[2]

Congenital heart disease is characterised by its great variability of symptoms, from both an anatomical and a physiopathological point of view, making it difficult to arrive at an exact diagnosis, with a variety of treatments available. It is therefore essential that it should be managed by specialists to achieve good therapeutic outcomes and a good quality of life. Increased survival of children with congenital heart disease has led to development, expansion and organisational changes in the structure of units dedicated to paediatric cardiology, with the gradual appearance of adult congenital heart disease units. Multidisciplinary units have now been created, in which professionals from different specialities, such as obstetricians, paediatricians, cardiologists, radiologists, surgeons, anaesthetists, intensive care specialists, psychologists, nurses, etc., with specific knowledge of congenital heart disease, work together in an interconnected way.[3,4]

The first congenital heart disease operation was performed in Boston in 1938 and consisted of closing a patent ductus arteriosus using lateral thoracotomy. This can be said to be the start of the development of cardiology in congenital heart disease, for which there had previously been no curative treatment. The advent of the membrane oxygenator in 1958 marked a radical change in surgery, making it possible to ''stop'' the heart and keep the body oxygenated by extracorporeal circulation (ECC) while the operation was performed. Ongoing technological progress led to the introduction of neonatal heart surgery in the 1980s, in which the figure of Dr. Aldo Castañeda stands out as one of the pioneer surgeons. These advances in surgery, together with clinical anatomical studies and the development of new technologies such as electrocardiography, radiology, cardiac catheterisation, etc. shaped the course of this speciality.

Surgical procedures for congenital heart disease can be divided into closed-heart surgery (without the need for ECC, usually by lateral thoracotomy) and open-heart surgery (with ECC). Another way of referring to types of surgery is to classify them into anatomical (correction of the defect by correcting the underlying anatomical problem) and physiological (surgical corrections that make the heart work

☆ Please cite this article as: Albert Brotons DC. El desarrollo de la cardiología pediátrica española y su impacto en el manejo de las cardiopatías congénitas. An Pediatr (Barc). 2015;83:295–296.

E-mail address: dimpnacalila@gmail.com

properly, even though the underlying anatomical problem has not been corrected). In Spain, they are usually named after the surgeon who first performed the procedure. If the patient has two well-functioning ventricles of similar size and a vessel of adequate size, biventricular or corrective surgery is performed, and if there is only one ventricle we use (Fontan-type) single-ventricle procedures, where the blood from the venae cavae is diverted directly to the pulmonary arteries. Both surgeries require that the pulmonary artery branches be anatomically correct. In some kinds of heart disease only palliative actions can be taken (the patient's heart, despite the interventions, will never be able to function like a normal heart) and they are operated by inserting prostheses in the pulmonary position which have to be replaced as the patient grows, inevitably involving successive operations with the risk that they may fail and a heart transplant may be required as a last resort.

Interventionist catheterisation is currently solving problems of simple and isolated heart conditions and resolving the complications that arise during followup, leading to a reduction in the number of operations. There are specific devices for closing septal defects as well as interatrial defects and some interventricular ones, patent ducti arteriosi and fistulas, and narrowed vessels can be widened, as in coarctation of the aorta or valvuloplasties for valve stenosis (even in foetal surgery), and more recently insertion of endovascular devices (stents) to widen vessels and valves in pulmonary and aortic position by catheterisation.[3]

As regards to recognition of this speciality, paediatric cardiology was established in the United States as a Section of the American Academy of Pediatrics in 1961 and is currently recognised in several European countries, including the United Kingdom, Germany, Sweden and Portugal. In Spain, the process leading to its recognition was begun in the 1970s by Dr. Quero and it is currently in the process of being approved as a ''specific training area''. A growing awareness that it constituted a ''different and recognised speciality'' led to the creation of the Paediatric Cardiology Scientific Sections within the Spanish Association of Paediatrics (AEP) and the Spanish Cardiology Society (SEC) in the late 1960s, operating under the general regulations of those societies. The Spanish Paediatric Cardiology and Congenital Heart Disease Society (SECPCC) was founded in 2001; it now has 317 members, including nearly all the paediatric[5] cardiologists and a high percentage of the heart surgeons specialising in congenital heart disease in Spain, as well as other specialists working in this field. In a survey of Spanish paediatric cardiology units carried out by SECPCC in 2014 to find out the current situation, we obtained data from 80 hospitals in 13 autonomous communities, recording the existence of 45 units with full-time paediatric cardiologists and 17 hospitals with surgical activity in congenital heart disease, 39 surgeons specialising in congenital heart disease and vascular care, and the performance of nearly 2000 operations on congenital heart disease during that year.

To sum up, the field of congenital heart disease has progressed from the anatomical study of heart conditions and their embryological origin to the development of imaging techniques for diagnosing them, and to therapeutic advances, focusing on developing percutaneous procedures or minimally invasive surgical treatment, and we are currently at the stage of developing genetic studies. Advances in prenatal diagnosis have reduced the incidence of the most severe heart conditions and will certainly have an impact on the incidence and overall prevalence of congenital heart disease in the next few years. Nevertheless, we must offer the children that are born all the benefits of technological progress. As of now, children suffering from congenital heart disease still have a life expectancy well below that of the general population, and we must therefore continue to improve the management of heart disease so as to increase their survival and their quality of life.

References

1. Tynan MJ, Becker AE, MaCarthey FJ, Quero Jimenez M, Shinebourne EA, Anderson RH. Nomenclature and classification of congenital heart disease. Br Heart J. 1979;41:544–53.
2. Subirana MT, Oliver JM, Sáez JM, Zunzunegui JL. Cardiología Pediátrica y Cardiopatías congénitas: del feto al adulto. Rev Esp Cardiol. 2012;65:50–8.
3. Sable C, Foster E, Uzak K, Bjornsen K, Canobbio MM, Conolly HM, et al., American Heart Association Congenital Heart Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, Council on Clinical Cardiology, and Council on Peripheral Vascular Disease. Best practices in managing transition to adulthood for adolescents with congenital heart disease: the transition process and medical and psychosocial issues. A scientific statement from the American Heart Association. Circulation. 2011;123:1454–85.
4. Warnes CA, Williams RG, Bashore TM, Child JS, Conolly HM, Dearani JA, et al., American College of Cardiology, American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease), American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, Society of Thoracic Surgeons. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease). Developed in collaboration with the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol. 2008;52:e143–263.
5. Santos de Soto J. Registro español sobre organización, recursos y actividades en Cardiología pediátrica. An Pediatr (Barc). 2004;61:51–61.