

## Adults with Congenital Heart Disease, and the New Patients in Cardiology

For some time now, many cardiologists for adults report that they are consulted by patients with a congenital heart disease of various degrees of severity, and are facing a real problem when making a decision against a complication, managing sequelae, or correcting a residual lesion.

Undoubtedly, the problem colleagues have to solve is indeed complex, for several reasons. First of all, congenital heart defects include a wide variety of lesions, each of which covers a range of subtypes, usually associated with two or more abnormalities, and their severity rating is a true Kafkaesque maze which still does not satisfy the leader experts in this topic. Second, over time, cardiologists for adults are not that acquainted with the pathophysiology of congenital heart diseases learned in college and during residency, so it is difficult for them to manage decompensations. Third, being acquainted with the different surgical techniques applied at different stages of the development of medicine in this field requires additional information, which is not always available. Finally, the extraordinary progress of interventional catheterization requires a renewed awareness of its availability and its application in each case.

Today, the increased number of adults with congenital heart diseases—due to their better survival—will require major tertiary healthcare services and academic programs to train new specialists and retrain cardiologists in general. (1) Paradoxically, there is a new scenario in which cardiologists for adults and for children meet again.

Until 1950s, medicine for adults and medicine for children were convened in the same conference every time a congenital heart defect was dealt with, and there was a main and essential character: the pathologist. The reasons were obvious. At those times, less than 20% of the patients born with these defects reached adulthood, many of them had severe defects, and only those with mild and complex defects exceptionally survived. (2)

In 1954, with the advent of extracorporeal circulation, advances in cardiovascular surgery offered a remarkable survival to patients with ventricular septal defect, tetralogy of Fallot, transposition of the great vessels, and univentricular heart. Later on, the development of interventional catheterization further improved this survival, having a great impact in the 1980s: children born in that decade could reach adulthood in 85% of the cases. (3)

This success in survival, due to advances in medicine in recent decades, has produced a new population of adults with congenital heart disease. These new patients have special characteristics, which are important to

know and investigate.

To describe the state of the art of this subject, Dr. Carola A. Warnes, from Mayo Clinic, in the United States, wrote in 2005 an article with a suggestive heading: *“The adult with congenital heart disease. Born to be bad?”*. (4) In this context, some patients have minor defects, so they will never need surgery. In other cases, the defect may not be detected until adulthood. However, most of them have undergone surgery before, and in many cases they are considered “cured”.

The perception of “cure” has been encouraged by the surgical description of “total correction”, which is applied in many repair surgeries for complex congenital defects. In fact, there is not always a cure for congenital heart diseases, perhaps with the exception of ligation and successful section of the ductus arteriosus. All the other repaired lesions have a potential capacity to develop residual lesions and sequelae. The perception of “cure” has serious consequences. Patients do not seek to understand the nature of their abnormality, do not realize the need for continued medical advice, and forget to take prophylactic antibiotics. As a result, sequelae and residual lesions are detected only when patients have symptoms.

These patients are different from those with an acquired heart disease. For a patient with congenital heart disease who usually has a complication like arrhythmia, cardiologists unfamiliar with these conditions often focus their decisions on electrophysiological aspects, unaware of the underlying hemodynamic problems associated with the onset of arrhythmia. In the same way, it is difficult for many colleagues to detect subtle changes in exercise capacity until they are significant for those patients, who have to live with heart problems for life. In general, dyspnea and exercise limitations are associated with residual valvular lesions and/or ventricular dysfunction, which are often severe and irreversible.

To address these new challenges of adults with congenital heart disease, a group of experts gathered in the 32<sup>nd</sup> Bethesda Conference (2001) for the purpose of finding answers to five questions: 1) number of adult patients with this condition, 2) special needs of these patients, 3) workforce that will be required, 4) recommendations for efficient offers and successful results in a health system, and 5) necessary steps to ensure the access of patients to proper health care. (5)

At that conference, it was estimated that for the year 2000, the number of adults with congenital heart disease in the United States would reach 787,800, out of which 368,800 (46.8%) would have simple problems,

302,500 (38.4%) of moderate severity, and 117,000 (14.8%) would be of high complexity.

The estimation model of this population for the year 2000 was based on the following data: a) birth rates from 1940 to 1989, b) incidence of congenital heart disease applied (6.2‰ live births), and c) survival of the disease depending on the period of birth (1940-1959, 1960-1979, 1980-1989) and on its severity (simple, moderate, and complex). (6)

Applying this same model in our country, it is estimated that there would be about 115,460 adult patients with congenital heart diseases, who would be distributed as 54,035 simple, 44,337 moderate, and 17,088 complex. According to some experts, it would be more realistic to estimate 70% of those data, in which case the total number would be 80,822.

In any case, these figures are important as burden of disease. If we consider only the presence of significant arrhythmias, one of the most important sequelae in this population, prevalence reaches 10% in samples from younger patients, and so there would be at least 8,082 patients in Argentina who will require specialized care in the field of arrhythmology. (7) The health care system must be prepared to face different residual lesions, sequelae and complications such as arrhythmia, persistent shunt, valve diseases, myocardial dysfunction, vascular lesions (stenosis, expansions, etc.), problems with prostheses, infections, thromboembolic events, pulmonary hypertension, bleeding, alterations of organs and systems, in addition to acquired cardiovascular diseases that may be associated in these patients.

Pregnancy and delivery in women with congenital heart disease are a special chapter of cardiology to cover all the risks for the mother and child. Advances in the pathophysiologic knowledge of this group of patients has resulted in improved care, reducing non-punishable abortion to extreme cases. Also, comprehensive care of mother and child is complemented by monitoring fetal development, and recurrence of cardiac defects which, as we know, is high in this population.

The Argentine Society of Cardiology is working actively for the formation of this new specialty, encouraging the development of centers with programs for the care of adults with congenital heart disease. We do not know exactly how many centers are being developed. Based on international experience, it is assumed that our country needs about 20 centers, that is, one center per two million inhabitants.

A total of 55 centers in the United States, 15 in Canada and 70 in Europe are reported in the First World. In Europe, only 7.1% of the estimated 1.800.000 adults with congenital heart disease are treated in specialized centers, which is an alarmingly low percentage. (8, 9)

A final message. In our country, 64% of the adult patients with congenital heart disease die of pulmonary hypertension, heart failure, and sudden death at an average age of 30 years, according to a recent report. (10) It would be a laudable goal that the coming

local reports informed us that the age at which these patients die is close to that achieved in the First World: 48.8 years (11), and as a result of the work performed by Argentinean cardiologists. At least we have a goal. As the philosopher would say: Argentines, let's get down to it.

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