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Review Article

Adults with Congenital Heart Disease in the Americas - Where we are today and where we are heading: A General View of the Inter-American Adult Congenital Heart Disease Council

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ABSTRACT

Congenital heart disease (CHD) in America has a global prevalence of 8-13 cases per 1,000 live births, with close to 45% being of medium and high complexity. Mortality continues to be high in Latin America and the Caribbean, with wide disparities in care, compared to North American countries. Fifty years ago, only 15% of children with CHD survived to age 18. Today, survival is greater than 90% in most countries worldwide. Currently, there are 2.2 million adults with congenital heart disease in the United States, with more than 1.8 million in South America. According to the Adult Congenital Heart Association, fewer than 10% of adults with congenital heart disease are estimated to be under specialized care, and more than 200 regional centers would be needed in the United States to meet the needs of this rapidly growing population. However, there are only 35 accredited programs in a total of 23 states. The scenario for Latin America and the Caribbean is bleaker and more distant, needing at least 249 programs, but lacking a formal census of centers and having few cardiologists who are experts in adult congenital heart disease. At the same time, there are insufficient paediatric cardiovascular centers. Thus, these countries have a double problem (children and adults with congenital heart disease, at the same time). In 2018, the Inter-American Society of Cardiology´s Inter-American Congenital Heart Disease Council was founded, which is working on various inter-American projects seeking to improve health care for adults with congenital heart disease.

Introduction

Congenital heart disease (CHD) is the most frequent congenital structural anomaly, worldwide. Prevalence at birth for every 1,000 live births (LBs) varies from country to country in the continent and is directly related to the birth rate and infant mortality rate. In general, we can say it is 8-13 cases for every 1,000 LBs. Previous research in 2013 by Hoffman J, *et al.* showed that 136,000 children with CHD are born every year in the Americas (65,000 in North America and 71,000 in South America) [1]. In North America, there are an average of 181 cases per day; in other words, there are 1.8 new CHD cases every 15 minutes. In South America, in turn, 197 cases are born each day, or 2.05 new cases every 15 minutes. Of all these cases, almost 45% have medium and high complexity, and at least 25% need surgical intervention within the first year of life, due to the severity of the defect [2]. Unfortunately, in Latin American and the Caribbean countries, especially low-income countries (LICs), only 8% have surgery in time. This leaves a large number of children without coverage and without timely access to cardiovascular healthcare systems, increasing early childhood mortality (in the first year of life). Every year, 15 million children die or fall severely ill due to CHD in some part of the world. Seen another way, on average, every two seconds, a child dies because of CHD. More than 90% of deaths occur in low and middle-income countries (LMICs) [3-5].

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Global Burden of Congenital Heart Disease in the Americas

Data on basic health indicators in the Americas reported in 2018 by the World Health Organization´s (WHO) Regional Office for the Americas and the Pan American Health Organization (PAHO) show that mortality in children under five years old due to non-communicable diseases is three times higher (18.6 vs 6.8) in LMICs compared to high-income countries (HICs) [6]. A large part of this mortality is due to CHDs, especially in children under one year old. The most recent study by the Global Burden of Disease (GBD 2017) showed that, during 2017, CHD was the underlying cause of an estimated 261,247 deaths globally (95% uncertainty interval [UI] 216,567 - 308,159). In the Americas, mortality per 100,000 children under the age of one year was 84·1 to 37·8 in HICs in North America vs 159·3 to 85·6 for southern Latin America, and 218·6 to 193·4 for the Caribbean, from 1990 to 2017, respectively. Of all CHD deaths in 2017, 180,624 (69%) occurred in infants under one year old [7].

The birth rate in Latin America and the Caribbean is estimated to be at least twice the rate of North America. In 2019, the mean population (millions of inhabitants) was 425.2 in South America, 219.2 in Central America and the Caribbean, 329.2 in the United States (US) and 37.4 in Canada [8]. A large number of births with CHD would be expected for Latin America and the Caribbean, as a result of the birth rate. Epidemiologically, the six most common lesions (atrial septal defect, pulmonary valve stenosis, aortic valve stenosis, coarctation of the aorta, ventricular septal defect and patent ductus arteriosus) comprise 67% of CHDs at birth. While not all of these lesions are amenable to interventional correction, many subsets are, and may contribute to as many as 30% of all detected CHD [9]. These CHDs are considered to have a good prognosis when they are diagnosed and treated in time, and, of all the variety of congenital heart defects, when they are not associated with other defects (brain or respiratory tract malformations or genetic syndromes), they may even be considered to be cured.

The problem for Latin American and Caribbean nations is that diagnosis continues to be delayed, and the chance of surgical repair is remote for many children. As a result, congenital lesions with a good prognosis end up claiming the lives of many children before their first birthday, increasing pediatric losses due to simple defects, which does not happen in developed countries. In Latin America, many children with CHD have associated comorbidities such as malnutrition, parasitic diseases and superimposed infections. Consequently, causes of death such as pneumonia, superinfected bronchiolitis, myocarditis and sudden death, among others, often mask a previously undiagnosed CHD. It is also common to find high complexity CHDs (conal-truncal CHDs, heterotaxy syndromes) together with other major malformations or complex genetic syndromes (trisomy 18, DiGeorge syndrome and Down syndrome, among others) which increase the clinical expression and severity.

Research has been done on the fact that a variety of sociodemographic factors affect the incidence and recurrence of several CHDs. A recent study characterized the risk factors of fathers and mothers of children with CHD. A total of 500 families in a region in Colombia answered a questionnaire to identify their sociodemographic factors and preconception risk factors. Families with as many as three children with CHD were found. Parents with two or more children with CHD were associated with a middle socioeconomic status, low level of education (p= 0.013) and rural residence (p=0.041). Preconception risk factors for CHDs were related to exposure to fertilizers (p=0.024), exposure to fuels (p=0.025), the use of antihypertensive medications (p=0.037), alcohol consumption (p=0.042) and the use of cocaine (p=0.039) [10].

Where We are Today

Fifty years ago, only 15% of children with CHD survived and reached the age of 18 [11]. Today, survival is more than 90% in most countries worldwide. The projected number of ACHDs for 2020 is 2.2 million for the US and more than 1.8 million for South America [12]. The HICs in America (Canada and the US) have significantly decreased child mortality in those under five years old. Communicable diseases (gastrointestinal and respiratory infections, rheumatic fever, tuberculosis and parasitosis, among others) are no longer a problem, contrary to the situation in the rest of the countries in Latin America and the Caribbean where they are still prevalent. Tropical diseases such as dengue fever, malaria, yellow fever and, more recently, zika, cause thousands of illnesses and disabilities every year, and are a real public health problem in these areas. Rheumatic fever alone affects 15.6 million people worldwide with more than 98% of cases occurring in LMICs [13].

Health policies for Latin America and the Caribbean have concentrated efforts and resources on the fight against these diseases, and at the same time, non-communicable diseases like CHDs continue to cause high mortality in children under one year old. Therefore, CHD care in childhood is also a priority. Currently, ever-greater efforts are being made to improve early detection of CHDs and provide access to prompt surgical treatment. However, there are still many gaps in care and access to specialized cardiovascular services. Undoubtedly, the support of non-governmental organizations (NGOs) has managed to improve the results. A 2013 survey of members of the World Society for Pediatric and Congenital Heart Surgery and PediHeart on the scope of NGOs which provide pediatric cardiovascular care in LMICs showed that the highest percentage of outreach efforts were concentrated in South and Central America (42%), followed by Africa (18%), Europe (17%), Asia (17%), and the Asia-Western Pacific region (6%) [14].

The current American Heart Association and American College of Cardiology guidelines recommend that adults with moderate and severe CHD be seen every 12 to 24 months by a cardiologist with specific CHD expertise at a regional CHD center [15]. Therefore, with the current estimate of 2.2 million ACHDs living in the US, It is conceivable that >200 ACHD regional centers will be required to meet the needs of this rapidly growing population. Just in the US, every year, approximately 20,000 CHD patients enter adulthood, and the number of adults with CHD has already surpassed the number of children. The rapid increase in complex CHDs and the distance separating many communities from the larger, developed cities in each country (where the specialized care centers function) are problems for this population´s follow up, even in developed countries. According to the Adult Congenital Heart Association (ACHA), it is estimated that less than 10% of ACHDs in the US are under specialized care. As of this paper´s writing, the ACHA ACHD Accreditation Program had accredited 35 programs in a total of 23 states [16]. At the same time, there are approximately 308 board certified ACHD cardiologists; however, the patient: ACHD cardiologist ratio is very low (4,220: 0.1).

For Latin America and the Caribbean, the real problem is that pediatric cardiovascular centers are still lacking, and specialized ACHD centers

are even further away from complete development. The differences in mortality between HICs and LMICs are almost 7 times more frequent (3% vs 20%, respectively) [17]. However, could be even greater if we consider that in many adults the deaths reported by unknown cause have a hidden CHD. Due to poor access to timely diagnosis and the vast majority of studies only report patient data in tertiary centers.

The Congenital Heart Disease Committee of the European Association for Cardio-Thoracic Surgery (EACTS) published the following recommendations for pediatric cardiovascular center functioning in Latin America [18]:

i. A minimum of 250 procedures per year;
ii. More than 100 procedures on newborns and children under the age of one;
iii. A minimum of three surgeries per week and 126 surgeries per year for each surgeon;
iv. Sites with a lower-case volume (≤ 250 patients operated on per year) may be considered to be functional hospitals if the results are similar to those of centers which handle higher volumes.

According to the EACTS recommendations, there should be one cardiovascular surgical center for every four million inhabitants. The reality is that in most Latin American countries, the best-case scenario is three to five at the most. Mexico alone, with a current population of 135.5 million people, would need at least 33 centers. Colombia, with a population of 49.4 million people would need at least 12 centers. Analyzing the Latin American and Caribbean situation, where reducing child mortality continues to be a priority, the scenario is further complicated by the simultaneous presence of CHD survivors who are already adults and also need specialized care. Thus, there are two large problems at the same time (children vs ACHDs), for which efforts need to be directed along two simultaneous lines of action. However, the economic costs and the maintenance of specialized hospitals require strong health policies in order for this to be achieved.

In Latin America, there is still no official number of ACHD specialized centers. Extrapolating the current ACHD population in Latin America and the Caribbean, at least 279 centers would be needed to meet the needs of this population. According to the study by Kempny A, *et al*., there are only 0.4 ACHD centers per 10 million people in South America [19], and likewise very few ACHD cardiologists with formal training. It is generally accepted that 10%, or at the most, 15% of CHDs are diagnosed in adulthood. This is accepted for American HICs, with most of these being low complexity congenital defects. For developing countries, the scenario is different, with more cases of adults not having been diagnosed early and not having been treated adequately, and the diagnosis of complex defects is not uncommon. Thus, the clinical presentation and natural evolution will be more severe. A recent study in Mexico showed that adults represented almost 30% of newly diagnosed cases, and another 30% were undertreated (diagnosed and treated in childhood, but, for various reasons, did not continue or finish treatment for their CHD) [20].

To conclude the current scenario, it is understandable that Latin American and Caribbean countries need to prepare for an avalanche of ACHDs with more serious and severe clinical forms, greater complications and disabilities. And, at the same time, they will need to continue to improve pediatric CHD care. Healthcare systems are practically obligated to improve the care conditions of both children and adult CHD patients.

Where We are Headed

The disparities in access to CHD care for the pediatric population will continue to be a constant struggle in Latin America and the Caribbean. This is precisely what was proposed by the United Nations as one of the Sustainable Development Goals for 2030, specifically Goal 3, which aims to end preventable deaths in newborns and children under five years of age. All countries aim to reduce neonatal mortality to at most 12 per 1,000 live births and under-5 mortality to at most 25 per 1,000 live births [21]. The less fortunate countries in America will need to concentrate their efforts to achieve this. As was mentioned previously, there are two concurrent problems today, since the growing ACHD population is already present in the hospitals of many countries who are still unprepared for this avalanche of patients.

These ACHD patients will undoubtedly be a big problem for these countries´ healthcare systems, not just because of the cost of their care, but also because this population is still made up of many young people who should be in optimal condition to participate in a productive economic system. A large part of the effort to improve CHD care has aroused the interest of various CHD specialists; thus, over the last few years, The World Database for Pediatric and Congenital Heart Surgery: A collaboration with the Registro Nacional de Cirugía Cardiaca Pediátrica (RENACCAPE) was created. This database seeks to improve the general care of children with CHD [22]. The United States is preparing and working hard to improve results and coverage for the ACHD population. They have recently studied the projected growth of the adult congenital heart to 2050, using combined National Vital Statistics System data and National Health Interview Survey data to determine the prevalence of CHD. At the same time, they are developing and creating ACHD programs throughout the country [23].

The Canadian system has been a worldwide pioneer and they have very good care coverage. They face some problems common to developed countries, such as ACHD losses to follow up, although to a lesser degree. In Latin America and the Caribbean we are working hard to construct a joint organization to strengthen and unify the Spanish-speaking and non-Spanish speaking American nations. Thus, in mid-2018, the Adult Congenital Heart Disease Council of the Inter-American Society of Cardiology (ACHDC-IASC) was founded, composed of several specialists in congenital heart disease from various countries in the continent [24]. Its goal is to integrate all the cardiologists in charge of ACHD management in the Central American, Caribbean and South American countries; there are delegates from each country who are in direct contact with the council leaders.
Its objectives are to:

i. Work together on ACHD registry projects;
ii. Further educational activities and cooperation among the various countries;
iii. Seek integration and cooperation with other global ACHD care societies like the International Society for Adult Congenital Heart Disease (ISACHD), and the Working Group on Adult Congenital Heart Disease of the European Society of Cardiology (EuroGUCH);
iv. Foster research in Latin America and the Caribbean;

v. Maintain active participation of the representative members from each country;
vi. Develop the directory of ACHD specialists in Latin America and the Caribbean;
vii. Implement the development of Latin American ACHD guidelines, among others.

During 2019, at least two inter-American research projects on ACHD were begun. Furthermore, several South American countries: Argentina, Chile, Brazil and Colombia, have begun to cooperate on a worldwide ACHD research project led by ISACHD. We expect that, with this contribution, in a few years we will be able to have a more exact understanding of the reality of the ACHD population in this area of the continent. Likewise, with the inter-American project results, we hope to have a more precise diagnosis of the actual situation of the ACHD population living in this region.

Conclusion

Child mortality due to congenital heart disease in children under the age of one continues to be a problem for Latin American and Caribbean countries, and, at the same time, the number of adults with congenital heart disease demands specialized care. Despite all the efforts of American countries, there is still a wide disparity in the care and management of children and adults with congenital heart disease. Even the US has a deficit of specialized centers; however, with better financial resources, it is preparing and projecting care for the ACHD population up to 2050. For Latin America and the Caribbean, the formation of the Adult Congenital Heart Disease Council of the Inter-American Society of Cardiology is a hope for integrating Spanish-speaking and non-Spanish speaking countries in a variety of inter-American projects to improve ACHD care.

Conflicts of Interest

None.

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