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Epidemiological analysis of the congenital heart disease in adults

Abstract

Introduction: The congenital heart disease in adults is proving to be difficult to diagnose because of its complexity. Various studies show that its prevalence has been increasing in recent years in developing countries. However, in our country, we do not have a record of the systematized prevalence from group of cardiac abnormalities.

Objective: To analyze the frequency of presentation of congenital heart diseases in adults and that pathological entities are the most frequent in patients from the cardiology department of the General Hospital of Mexico.

Material and methods: retrospective study, cross-sectional, observational and descriptive, in a period of 24 months, from January 2013 to December 2014, which included 352 adult patients with a diagnosis of congenital heart diseases. We included patients with 18 years of age or older, which they have not been operated on before. The following variables were studied: sex, age, type of congenital heart disease syndromes associated and related complications.

Results: Our study included 352 patients, 205 (58.23%) female and 147 male (41.77%), mean age 31.68±14.4 years. Congenital heart diseases with the highest prevalence were atrial septal defect (27.84%), communication interventricular (15.05%), pulmonary stenosis (6.81%), patent ductus arteriosus (6.53%) and aorta bicuspid (5.96%). The associated complications were the most frequent pulmonary hypertension (15.58%) and the paradoxical embolism (1.98%).

Conclusions: The prevalence of congenital heart diseases in our population turned out to be similar to what has been reported at the international level.

Keywords: Congenital Heart Disease in adults, Septal defects, Cardiovascular abnormalities

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Abbreviations: CHD, Congenital Heart disease; CHDA, Congenital heart disease in adults; ASD, Atrial septal defect; CTGA, corrected transposition of the great arteries; VSD, Ventricular septal defect; PS, Pulmonary stenosis; EA, Ebstein´s anomaly; PDA, Persistence of ductus arteriosus; PFO, Patent foramen ovale; DAL, Double aortic lesion

Introduction

The congenital heart disease (CHD) are structural abnormalities of the heart, produced by cardiac embryogenesis errors, which may manifest in fetal, neonatal or pediatric age. Represent the most common congenital malformations, it is estimated that approximately 6 of every 100 newborns have a moderate or severe CHD;[1] Some patients may present with moderate defects that do not require surgery, and some others may go unnoticed and be discovered until adulthood, being these, the current population of adults with congenital heart disease, constitute a heterogeneous group with multiple alterations well known and various associations. The CHD that are detected after the pediatric age, are increasingly prevalent in developed countries.[2,3]

The initial manifestations of congenital heart disease in adults (CHDA) represent a range of symptoms, ranging from asymptomatic patients until such serious manifestations that compromise the quality of life; The symptoms of these disorders are not always proportional to the pathophysiologic conditions of these entities. The majority of the physiopathological alterations are not diagnosed early, and occasionally are complex or unusual associations that represent a challenge to both diagnostic and therapeutic.

The CHD are not unique to the pediatric population, although many of them are diagnosed and treated early. The medical and surgical advances have allowed the detection and early approach of most children with CHD, many of whom survive to adulthood, however, a large part of these patients require medical follow-up and re surgeries throughout his life. On the other hand, we have to patients who were not diagnosed in the pediatric stage, and that reach adulthood with symptoms variable, for which reason the diagnosis may be delayed so important if you do not have a high level of suspicion.

There are still no concrete data about the size of the population with CHDA. At the 32nd Bethesda Conference in 2000,[4] it was estimated that there were about 2.800 adults with CHD per million inhabitants, which is equivalent to 800 000 adults with CHD in the United States in which more than half were studying with a moderate complexity or severe. Recently in another systematic review that included patients from USA and United Kingdom of 10 publications about the prevalence of the CHDA, which reported that there were 3000 people per million inhabitants with CHDA.[5] In Mexico, as well as in other countries, there are no known official figures, however, according to the study of Alva et al. estimated that there are 300 thousand adults, treated and non-treated with CHD, with a likely annual increase of 15 thousand new cases.[6]

Some defects are diagnosed for the first time in adulthood, such

as for example the atrial septal defect (ASD), aortic coarctation, Ebstein's malformation and the corrected transposition of the great arteries (CTGA).[7,8] In a study carried out in the Super Specialty Hospital, National Medical Center "La Raza", echocardiographic studies were reviewed 9.833, of which 1.071 (10%) corresponded to CHDA, being the most frequent in this population, atrial septal defects (39%), bicuspid aorta (19%), ventricular septal defect (14%), persistent ductus arteriosus (10%), and aortic coarctation (7%)[9]. The spectrum of congenital heart disease can be classified in valvular lesions, malformation with short circuit, cyanogen heart disease and others (Table 1).

Table 1 Classification of Congenital Heart Disease in Adults

Valvular Lesions		**Malformations with short circuit**	**Cyanogenic Cardiopathies**	**Other**
Aortic Valve	Tricuspid Valve	Short Circuits		
Bivalve aorta		Interatrial comunication	Transposition of the great vessels	Common trunk
Aortic stenosis	Ebstein´s Anomaly	Intervetricular communication		
Valvular		Persistence of ductus arteriosus	Corrected transposition of the great vessels	Situs inversus
Sub-valvular	Tricuspid atresy	Atrioventricular canal		
Supra-valvular		Aorto pulmonary window	Tetralogy of Fallot	Hypertrophic cardiomyopathy
Aortic insufficiency			Unique ventricle	Dilated cardiomyopathy
Mitral Valve	Pulmonary Valve	Aneurysms		Cor tiratriatum
Mitral stenosis	Pulmonary stenosis	Aneurysm of the interatrial septum	Double way out of the right ventricle	
	Pulmonary artery	Broken valsalva sinus aneurysm		Aortic coarctation
Mitral valve in parachute	Stenosis of pulmonary branches	Coronary arteries aneurysm		
	Abnormal pulmonary venous drainage			Patent foramen ovale

Of the classification described above, it should be noted that, in some of them, patients do not reach adulthood, either because of their inability to adapt cardiopulmonary resuscitation or complications that these pathologies, examples of these are the tricuspid atresia, pulmonary atresia, stenosis of the pulmonary branches and transposition of the great vessels.

The clinical evaluation in patients with CHDA should be carried out in a rigorous and comprehensive. The analysis of the patient's clinical history is important for assessing past and current symptomatology, search for intercurrent episodes and the therapy. The physical examination is of the utmost importance that must include a thorough evaluation of the auscultatory findings, changes in blood pressure in the development of heart failure.[10]

The development in recent years of the echocardiography for the assessment of patients with CHD, its use in surgical areas cardiovascular and hemodynamic interventionism, has offered a significant improvement in the diagnosis and management of this group of patients.[11] The echocardiography in its different modalities, results in the diagnostic method of choice in patients with CHD, because it allows you to perform an anatomical and functional assessment of these pathologies, in addition to being a non-invasive, reproducible, and low-cost, despite the fact that the volumes are overestimated in comparison with the Bland-Altman analysis of 25 patients, showed a better clinical correlation with ventricular function using magnetic resonance that through the 3rd dimensional echocardiography.[12]

The transthoracic echocardiography in the evaluation of adult patients with CHDA, presents with frequency limitation in sonic window, which can be affected by the acoustic penetration, lung diseases and thoracic structural alterations; these conditions make it necessary to the implementation of the variant transesophageal echocardiography, which may be required in the evaluation and diagnosis in patients with suspected CHDA.[13]

The aim of the present study was to analyze the frequency of presentation of congenital heart disease in adults and that pathological

entities are the most frequent in patients in the Cardiology Service of the General Hospital of Mexico.

Materials and methods

Retrospective study, cross-sectional, observational and descriptive study that was developed in a period of 23 months, from January 2013 to November 2014. Clinical records were reviewed in the Cardiology Service of the General Hospital of Mexico of women and men, with a history of congenital heart diseases, whose diagnosis has been carried out at the age of 18 years of age or older. We analyzed the frequency of congenital, which entities are the most prevalent and complications most frequently enrolled in this group of patients. The following variables were studied: sex, age, type of congenital syndromes associated and complications of these diseases.

Quantitative variables were expressed as mean and standard deviation (SD), the qualitative variables are expressed in the form of ratios and percentages. We used the SPSS package version 22.0.0.0 for statistical analysis.

Results and discussion

The analysis of this study comprises from January 2013 to November 2014, 45.068 were reviewed clinical records of patients in the Cardiology Service of the General Hospital of Mexico, finding within these 352 cases that were diagnosed with congenital heart disease in the adult is not treated, which represents 0.78% of the total of the files reviewed (Figure 1).

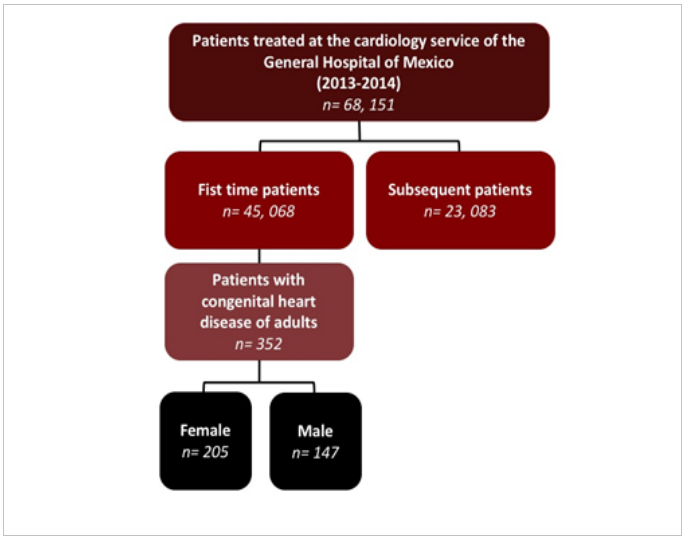

Figure 1 Diagram of patient selection and exclusion processes

The average age of diagnosis was 31.68 years (SD 14,44) for the total sample. With regard to the year 2013, the mean age was 31.23 years (SD 13,70), being the average age of the feminine gender of 10.917 years (SD 13.76), and the male gender of 16,916 years (SD 31.79). With regard to the year 2014, the mean age was 32.14 years (SD 15,17), being the average age of the feminine gender of 32,2 years (SD 14.16) and of the male gender of 37.9 years (SD 16,91) (Figure 2).

Inside of CHDA in our series, it was found that the most frequent was the atrial septal defect (ASD) 98 patients (27.76%), secondly the ventricular septal defect (VSD) in 53 patients (15.01%), thirdly the pulmonary stenosis (PE) in 24 patients (6.79%), followed by the persistence of ductus arteriosus (PDA) in 23 patients (6.51%), bicuspid aorta (BA) in 21 patients (5.94%), Ebstein's Anomaly (EA) in 17 patients (4.81%), aortic coarctation in 17 patients (4.81%), patent foramen ovale (PFO) in 15 patients (4.24%) and the double aortic lesion (DAL) (aortic stenosis + aortic insufficiency) in 10 patients (2.83%) and other diseases with less frequency (11.07%) and prolapse of the mitral valve, aortic stenosis, atrioventricular canal, dilated cardiomyopathy, corrected transposition of the great vessels, arteriovenous fistulas, pulmonary atresia, hypertrophic cardiomyopathy, common trunk, tetralogy of Fallot, interatrial septal aneurysm, septal aneurysm, double outlet right ventricle, and situs inversus (Figure 3)

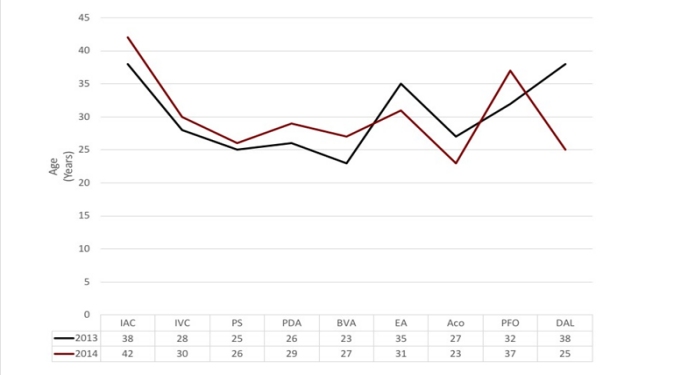

Figure 2 Mean age of diagnosis of congenital heart disease in adults.
IAC, Interauricular comunication; IVC, Interventricular comunication; PS, Pulmonary stenosis; PDA, Persistent ductus arteriosus; BVA, Bivalve aort; EA, Ebstein´s anomaly; Aortic coarctation; PFO, Patent foramen ovale; DAL, Doauble aortic lesion.

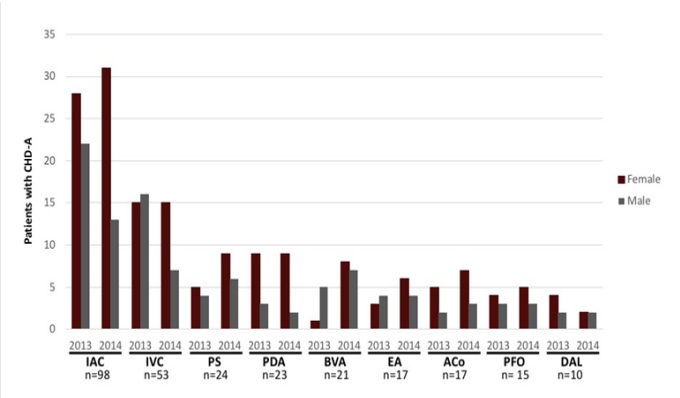

Figure 3 Frequency of CHD-A by year and sex.
IAC, Interauricular comunication; IVC, Interventricular comunication; PS, Pulmonary stenosis; PDA, Persistent ductus arteriosus; BVA, Bivalve aort; EA, Ebstein´s anomaly; Aortic coarctation; PFO, Patent foramen ovale; DAL, Doauble aortic lesion.

The association of congenital heart diseases with others was presented in 40 patients (11.04%) than in order of frequency is in the first place the ventricular septal defect and patent ductus arteriosus in 5 patients (1.41%) and others associations of congenital heart disease less frequently (<1%).

The presence of congenital heart disease associated with syndromes was present in 70 patients (19.83%), being the most frequent syndrome present in 27 patients (7.64%), followed by the

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Marfan Syndrome in 17 patients (4.81%), Turner syndrome in 9 patients (2.54%), Ehler-Danlos syndrome in 5 patients (1.41%) and Duchenne muscular dystrophy in 5 patients (1.41 %).

The complications associated with congenital heart disease was present in 67 patients (19.03%), being the most frequent pulmonary arterial hypertension (15.58%) and paradoxical embolism manifested as cerebral vascular event in 7 patients (1.98%).

The frequency of congenital heart disease in adults within the General Hospital of Mexico, does not vary with respect to what is reported in the international literature (Table 2), this is probably due to the high level of suspicion that has doctors on this set of pathologies, it is of great utility at the time of the comprehensive study of these patients, which leads to the increase in the incidence of these as they are intentionally starts to search for the presence of them. The frequency of each variety of congenital heart disease was similar compared with the frequency in the pediatric age, being for both groups the septal defects the most common such as atrial septal defect, ventricular septal defect, and the third is the persistence of the ductus arteriosus.[14,15]

Table 2 Frequency in different hospitals of congenital heart diseases in adults.

	Royal Hospital*	Mayo Clinic**	NICICH***	NMC "La Raza"****	General Hospital of Mexico
	n= 61 (%)	n= 391(%)	n= 651 (%)	n= 1071 (%)	n= 352 (%)
IAC	39	33	14	40	27
IVC	15	<1	18	14	15
PS	8	-	9	6	7
BVA	-	32	-	19	6
AS	5	-	12	4	<1
PDA	4	<1	8	10	6
ACo	3	2	4	7	5
EA	1	3	9	4	5
TF	1	3	9	3	<1
PA	-	3	3	<1	<1
TA	-	<1	3	<1	-
TGV	<1	-	3	1	<1
DWORV	-	1	-	<1	<1

IAC, Interatrial comunication; IVC, Interventricular comunication; PS, Pulmonary stenosis; BVA, Bivalve aort; AS, Aortic stenosis; PDA, Persistence of ductus arteriosus; ACo, Aortic coarctation; EA, Ebstenin´s anomaly; TF, Tetralogy of Fallot; PA, Pulmonary atresy; TA, Tricuspid atresy; TGV, Trasposition of the great vessels; DWORV, Double way out of the right ventricle.

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At the same time notes the predominance of females over males with a ratio 2:1 which is consistent with what is reported in the international reports.[1,5]

With regard to the age is observed a higher frequency among the age groups of 20 to 40 years, taking an age range of presentation that ranges from 18 years to 92 years of age, where you can show mild injuries and easily diagnosed, and occasionally rare and complex associations that represent a challenge to both diagnostic and therapeutic.

Within our group of study found patients with congenital rare and complex such as: double outlet right ventricle, the interatrial septum fistula, common trunk and pulmonary atresia, all of them diagnosed in adulthood and with a cardiopulmonary compromise important, however the average age of these patients over 30 years old and with several treatment options.

The associations between certain syndromes and congenital heart disease was relatively similar to that reported in the literature, showing a predominance with respect to the Down syndrome with septal defects, Marfan syndrome and valvular alterations and/or aortic and Turner syndrome and aortic coarctation.

Within this same study analyzed associations between congenital in 11.07% of the total sample, with a high frequency between the associations such as: IVC and PDA, IAC and IVC and PS, IVC and PS. It is also valued the complications associated with congenital heart disease, were found to be moderately frequent, reporting in the 19.03%, being the most relevant pulmonary arterial hypertension

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(15.58%), paradoxical embolism manifested as cerebral vascular event (1.98%) and pulmonary stenosis (0.84%).

Conclusion

Congenital heart disease in adults represent a set of increasingly frequent pathologies with a range of symptoms variable, which represents a diagnostic and therapeutic challenge. The increase in the incidence of these pathologies is mainly due to the fact that current technology allows us to evaluate with best quality and accuracy the cardiovascular status, this coupled with the high level of suspicion that the doctors to fully evaluate patients, which leads to intentionally look for these pathologies.

The increase in the incidence of these pathologies, leads to establish a multidisciplinary management for comprehensive and effective management of these patients. Adults with CEC present a high risk of long-term complications, so it is of the utmost importance the early diagnosis and the establishment of a therapeutic plan, which will provide a better quality of life and delay the onset of complications described above.

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