Clinical and echocardiographic patterns of congenital heart diseases in adults in Karbala Province, Iraq

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Objective To determine the patterns of congenital heart diseases in adults in Karbala province.
Methods A total of 85 patients aged more than 16 years were examined at Al-Hussain Medical City and Al-Hindiya General Hospital from June 2006 to December 2014. History, physical examination, electrocardiograms and transthoracic echocardiograms were done for them.
Results Seventy-eight of patients were less than 30 years old and two patients were more than 50 years old. Atrial septal defect was the most common primary diagnosis (28 patients) followed by pulmonary stenosis (22 patients) and ventricular septal defect (18 patients). Seven patients had patent ductus arteriosus and five patients had tetralogy of Fallot. Three patients had dextro-transposition of the great arteries and three had atrioventricular septal defect. There was one patient in each of the other categories (single ventricle, Ebstien anomaly, coarctation of aorta, subaortic ridge and bicuspid aortic valve).
Conclusion Atrial septal defect was the most common primary diagnosis. The percentage of patients with coarctation of aorta and aortic valve diseases was much less than in other studies. The ages of study patients were less than the patients in Western countries especially patients with tetralogy of Fallot, dextro-transposition of the great arteries, atrioventricular septal defect and single ventricle and coarctation of aorta.
Keywords congenital heart disease, echocardiographic, karbala

Introduction

Congenital heart disease (CHD) is a common birth defect. Advances in paediatric cardiology and cardiac surgery contributed to the survival of the majority of these patients to adulthood.[1,2] These successful efforts have changed the fate for CHD creating a large population of adolescent and adult patients.[3] One quarter of these patients had special educational and occupational demands.[4] Physical and emotional maturity is the primary prerequisite for the transfer of adolescent patients into adult care territory. The age at which this takes place may range from the mid-teens to the mid-twenties depending upon the patient. Preparing young patients for successful transfer to an adult healthcare provider at a subsequent time should commence by the age of 12 years.[5] Patients more than 16 years old are managed by adult cardiologists.[6]

Although evidence-based medicine is the base of contemporary cardiology practice, data about adolescent and adult survivors of CHD is sparse.[7] This paucity of data is more prominent in Iraq, especially Karbala province.

The aim of this study is to find the patterns of CHD in adults (aCHD) in Karbala province.

Methods and Materials

Research ethics and patient consent

Ethics Committee at Karbala Health Directorate provided approval for the study. All patients provided informed verbal consent.

Study population

A total of 85 patients examined at Al-Hussein Medical City and Al-Hindiya General Hospital from June 2006 to December 2014. These patients were referred to echocardiography units for suspicion or previous diagnosis of CHD. All patients were more than 16 years old. History and clinical examination were performed for each patient.

Echocardiographic Data

The study patients underwent transthoracic echocardiography using Philips EnVisor C machine (Philips medical Systems, USA).

Echocardiographic Definitions

Congenital Heart Disease (CHD)

CHD is defined as a structural abnormality of the heart or intrathoracic great vessels that is of functional significance.[8] If a case had more than one independent lesion, each one was counted separately.[9] Definitions of individual lesions were based on published studies and ACC/AHA guidelines for the management of adults with congenital and valvular heart disease.[10–15]

Statistical Analysis

Statistical analysis was done using Excel 2013 (Microsoft Corporation, USA), and compared using the chi-square (χ^2) test for statistical analysis of data. $P < 0.05$ is the level of significance.

Results

A total of 85 patients were examined (35 males and 50 females), female–male was ratio 1.4. Age range was between 16 and 80 years with a mean age of 29 (±12.5) years. Distribution of patients according to age groups is shown in Table 1. Atrial septal defect (ASD) was the most common lesion (28 patients, 30.8%), followed by pulmonary stenosis (PS) (22 patients, 24.2%), then ventricular septal defect (VSD) (18 patients, 19.7%). The number of patients with patent

ductus arteriosus (PDA) was seven (7.7%) and tetralogy of Fallot (TOF) five (5.5%). Patients with dextro-transposition of the great arteries (d-TGA) and atrioventricular (AV) canal defect were three (3.3%) for each category. There was one patient (1.1%) with each of other abnormalities: single ventricle, Ebstien anomaly, coarctation of aorta (COA), subaortic ridge and bicuspid aortic valve (BAV) (Table 2).

The percentage of ostium secundum was 87%, ostium primum 10% and sinus venosus 3% of ASD defects. Age distribution for patients with ASD, VSD, PS, PDA and TOF is shown in Fig. 1.

The percentage of VSD types was perimembranous 79%, muscular 8.5%, and inlet 12.5%. Gender distribution in patients with ASD, VSD and PS is shown in Table 3. ASD occurred in females in 68%, PS in 64% and VSD in 44%. In patients with PS, 45% had mild stenosis (Fig. 2).

Discussion

Age and Gender Distribution

Seventy-eight patients (92%) were less than 30 years of age, seven patients (8%) were more than 40 years, and two patients (2.2%) were more than 50 years of age. In the Mayo Clinic series, patients with more than 40 years of age constituted 50% while in the Toronto series, they constituted 30% of ACHD patients.[16] A Lebanese study done by Hannoush et al. found that 16% of patients with acyanotic heart disease and no patients with cyanotic heart disease were more than 50 years old.[17] In a European study done by Engelfriet et al., 21% of the ACHD patients aged more than 50 years.[18]

Reasons for these age differences may be related to differences in survival of patients, availability of surgery and/or in population age distribution between the countries. In USA, persons above 45 years old constitute 39% of the population,[19] whereas in Iraq, they constitute 12%.[20]

There is a significant predominance of females in the current study with 50 patients (59%) ($P < 0.05$). This is comparable to the European study (52%),[18] whereas in Lebanese study, there was a slight predominance of men (52%).[17]

Table 1. **Numbers (and percentages) of patients according to age groups**

Age at examination (years)	Number (%) of patients
≤ 20	42 (49.4)
21–30	24 (28.2)
31–40	12 (14.1)
41–50	5 (5.8)
51–60	1 (1.1)
61–70	–
> 70	1 (1.1)
Total	85 (100)

Table 2. **Numbers (and percentages) of ACHD lesions**

Lesion	Number (%) of defects
ASD	28 (30.8)
PS	22 (24.2)
VSD	18 (19.7)
PDA	7 (7.7)
T.O.F	5 (5.5)
D-TGA	3 (3.3)
A-V canal	3 (3.3)
Single ventricle	1 (1.1)
Ebstien anomaly	1 (1.1)
C O A	1 (1.1)
Subaortic ridge	1 (1.1)
BAV	1 (1.1)
Total	91 (100)

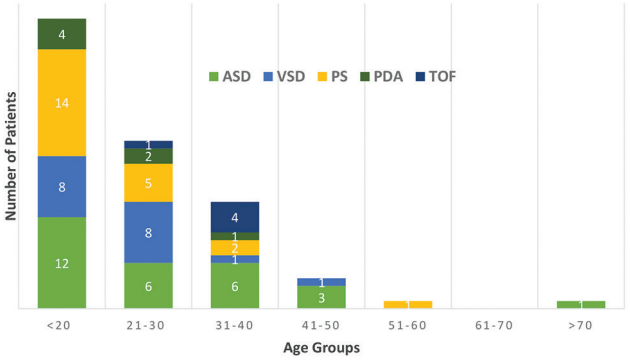

Fig. 1 **Age distribution by decade for patients with ASD, VSD, PS, PDA & TOF.**

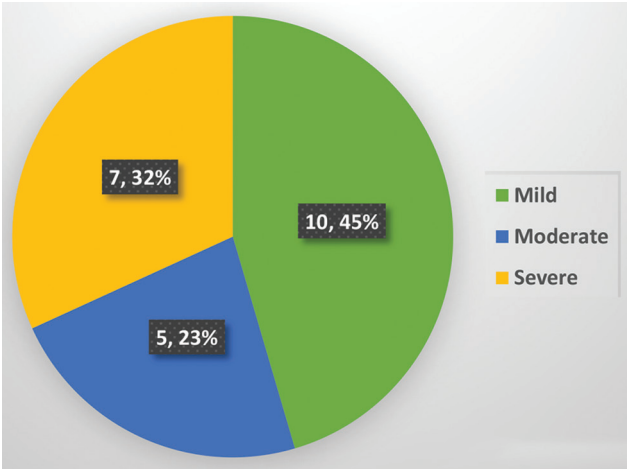

Fig. 2 **Severity of PS.**

Table 3. **Gender distribution in patients with ASD, VSD and PS (numbers and percentages)**

ACHD	Male (%)	Female (%)	Total %
ASD	9 (32)	19 (68)	28
VSD	10 (56)	8 (44)	18
PS	8 (36)	14 (64)	22

Congenital Defects

Atrial Septal Defect

Atrial septal defect was the commonest lesion accounting for 30.8% of the cases. This percentage was more than European (22%),[18] US (20.6%)[6,21] and Egyptian (20%)[22] and less than Lebanese (53%)[17] and Indian (44.5%)[23] studies.

The higher number of ASDs may be attributed to racial differences,[24] inclusion of small defects or patent foramen ovale with a tiny left-to-right shunt[6] or referral bias.[17]

ASD occurred in women two times as often as in men in our study, and this was similar to European and US studies.[18,25,26] Female-to-male ratio in the patients with ASD was 1.2 in Hannoush et al. study.[17]

Three ASD patients had mitral valve prolapse and three had PS. These are well-recognized associations.[15,27–29]

The majority of our patients (87%) had ostium secundum defects, which is more than the 75% mentioned in the literature.[30] The percentage of primum defects in our study was less than the literature (10% vs 15%) so as sinus venosus defects (3% vs 10%).[30] In Hannoush et al. study, 86% had secundum, 9% had primum, and 5% had sinus venosus ASD,[17] which is close to our results.

Four patients (15%) of ASD patients were more than 40 years of age. These constituted 57% of total patients who are more than 40 years of age.

The number of ASD patients presenting in adulthood might be the half because of the absence of complete diagnosis in childhood.[6] This is because patients with ASDs often have no symptoms until the third or fourth decades of life even with substantial left-to-right shunting.[30] One of our ASD patients was diagnosed in his eighth decade and was the oldest patient in the study. Patients with an unrepaired ASD may survive into the eighth or ninth decade of life.[31]

Pulmonary Stenosis

PS constituted 24.2% of cases in our study, which is more than the literature numbers ranging between 10 and 16.3%.[6,21,30] Hannoush et al. described pulmonary stenosis in 6% of ACHD.[17] Reasons for these differences may be racial.[24] Grech[12] and Botto et al.[24] reported increment in the rate of pulmonic stenosis. This increment may be attributed to "methodological divergences" due to different definitions of mild pulmonary stenosis.[12] It may be the result of improved diagnosis and reporting for the less severe defects due to increased availability of two dimensional and colour-Doppler echocardiography.[24] Forty-five percent of our patients had mild stenosis. Females constituted 64% of our patients with PS, which is close to Europe (58%).[32]

In 95% of patients in our study, the stenosis was valvular which is close to the literature.[30] More than 63% were less than 20 years old, and one patient (4.5%) was more than 40 yr. Without treatment, patients with severe stenosis may die from congestive heart failure or ventricular arrhythmias.[6] About 60% of these patients will require intervention by 10 years after diagnosis.[33]

Ventricular Septal Defect

It is the most common congenital heart defect at birth.[34] The prevalence is much less in adults because there is a high incidence of spontaneous closure of small VSDs.[35,36]

VSD in our study constituted 19.7% of ACHD cases, which is close to the numbers in the US.[6,21] Hannoush et al. described ventricular septal defects in 11% of ACHD[17] and in Europe it was15%.[21]

Male–female ratio in our study was nearly equal which is similar to US and European studies.[18,33]

Perimembranous type constituted 79% of VSD patients, which is close to the US literature.[30] The percentage of the muscular type was less than the literature (8.5% vs 20%), whereas that of the inlet type VSD was more than the literature (12.5% vs 5%).[30]

Untreated VSDs in adults are almost always small defects with little hemodynamic significance and low risk of pulmonary hypertension.[6]

Eighty-nine percent of our VSD patients were less than 30 years old. Because there is no reason to expect a high mortality rate in these subjects, the reduction in VSD numbers with age may represent late spontaneous closure rather than death.[6] Spontaneous closure rate of 80% up to the age of 70 years has been described.[36–42]

Patent Ductus Arteriosus

It constituted 7.7% of cases, which is less than the literature numbers, and it may reach double this percentage.[6,21,30] This variability may be due to methodological differences related to the population groups, age of consideration, and detection methods.[39]

One patient in our study (14%) was more than 30 years of age. The natural history studies imply that only 50% of patients with a PDA live for 20 years and only 10% of them live for 50 years.[6]

Tetralogy of Fallot

It constituted 5.5% of cases in our study which is close to the US literature numbers.[6,21] The percentage was 11% in Lebanon.[17]

None of our patients was beyond fourth decade. Untreated patients with tetralogy of Fallot die young, usually from hypoxaemia, brain abscess, stroke, or occasionally myocardial failure.[6] Only 25% of persons with TOF are alive after the age of 10 years, and thereafter, the mortality rate is 6.4% per year.[43,44] Most patients die before the end of their second decade.[45,46]

Aortic Valve

Bicuspid Aortic Valve

It accounts for 1.1% among our patients despite the reported incidence of 1% to 2% of the population. This probably because a BAV may develop significant obstruction or regurgitation after midlife, with a peak age range for surgical intervention between 60 and 80 years.[36] Botto et al. reported racial variations in prevalence of aortic valve disease.[2] In Hannoush et al. study, valvar aortic disease was seen in 6% of ACHD patients.[17]

SubAS constituted 1.1% among our patients. The prevalence of discrete SubAS among ACHD patients has been reported to be 6.5% in US studies.[47] Hannoush et al. described SubAS in 2% of ACHD patients.[17]

D-TGA

It constituted 3.3% of ACHD cases in our study, which is more than the percentage in the US (1.8%).[6,21] Hannoush et al. described l-TGA in 2% and didn't describe d-TGA.[17] Racial variations in prevalence of d-TGA were reported by Botto et al.[24] All our patients were less than 30 years old. Without treatment, 95% of patients die in the first year, mainly from hypoxaemia, and very few reach adult life.[48]

AV Canal Defect

It constituted 3.3% of ACHD cases in our study, which is close to the literature numbers.[6,21] All patients were less than 40 years old. Few survive past childhood if untreated, because of early death from congestive heart failure or pulmonary vascular disease.[6]

Single Ventricle

It constituted 1.1% of ACHD cases, which is much more than the percentage in the US (0.3%).[6,21] Hannoush et al. described single ventricle in 7% of ACHD cases.[17] These differences may be attributed to differences in risk factors such as paternal smoking and alcohol consumption.[49] Our patient was less than 30 years old and died after enrollment in study.

Coarctation of Aorta

It constituted 1.1% of ACHD cases in our study, which is much less than the percentage in the US (8.4%).[6,21] and Europe (13%).[18] Hannoush et al. described COA 2% of ACHD.[17] Racial variations in the prevalence of COA were reported.[24] Our patient was less than 20 years old.

Ebstein Anomaly

It constituted 1.1% among ACHD patients of in our study, which is close to US literature.[50] Hannoush et al. described Ebstein anomaly in 4% of ACHD.[17] Our patient was in 5th decade.

Conclusions

1. Karbala ACHD patients were younger than the patients in Western countries. No patients with Tetralogy of Fallot, d-TGA, AV canal defect, single ventricle and coarctation of aorta were beyond fourth decade of life.
2. There is a significant predominance of females.
3. Atrial septal defect was the most common primary diagnosis with a significant predominance of females.
4. Patients with ASD and VSD may live a normal life span.
5. The percentages of ostium secundum ASDs, inlet VSD, PS and d-TGA were more common than the literature. The percentages of muscular VSD, coarctation of the aorta and aortic valve diseases were less than the literature.

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Conflict of Interest

The authors declare that there is no conflict of interest. ■

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