

PATTERN OF CONGENITAL HEART DISEASE IN THE SOUTHWESTERN REGION OF SAUDI ARABIA

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Background: The aim of this study was to assess the frequency of various forms of congenital heart disease (CHD) among affected children in the Southwestern region of Saudi Arabia.

Patients and Methods: During the study period (July 1994 to June 1996), 608 children were referred to Asir Central Hospital as having CHD. All the children were evaluated by a pediatric cardiologist and had electrocardiogram and echocardiogram.

Results: Of the 608 patients, only 335, comprising 162 males and 173 females, had CHD. The male to female ratio was 0.9:1. The frequency of various forms of CHD was as follows: ventricular septal defect (VSD) 32.5%; patent ductus arteriosus 15.8%; atrial septal defect 10.4%; pulmonary stenosis 10.1%; atrioventricular septal defect and mitral valve prolapse, 3.6% each; aortic coarctation/interruption 3.3%; obstructive aortic valve lesions 2.7%; tetralogy of Fallot 4.5%; common ventricle 2.7%; pulmonary atresia (PA) with VSD 1.8%; D-transposition of the great arteries 1.5%; Ebstein anomaly 1.5%; and isolated PA 1.2%. Other lesions were extremely rare.

Conclusion: The distribution in this study is similar to that reported in previous studies from other parts of the world, except for the lower incidence of obstructive aortic valve lesions. *Ann Saudi Med 1998;18(5):393-395.*

Key words: Congenital heart disease.

Congenital heart disease (CHD) is one of the common forms of major congenital malformation in the Southwestern region of Saudi Arabia, and therefore, knowledge of the relative frequencies of different lesions is important to physicians in the process of training, management and the planning of health care systems. There have been a few reports^{1,2} addressing the pattern of CHD in some regions of Saudi Arabia.

Materials and Methods

This study was conducted in Asir Central Hospital (ACH), which is the main tertiary hospital for the Southwestern region of the Kingdom of Saudi Arabia. The study period extended from July 1994 to June 1996, and during this period ACH was the only hospital in the region with appropriate facilities to assess children suspected to have CHD. All children referred to the Pediatric Cardiology clinic were examined by the author and had electrocardiograms. A two-dimensional and Doppler echocardiogram was done and interpreted by the author, regardless of the clinical impression. Only children with CHD were reviewed. The relative frequencies of specific forms of CHD in the group were determined.

CHD is defined here as a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance, and therefore, a persistent left superior vena cava, inferior vena cava, azygous continuity and right-sided aortic arch are excluded. Arrhythmias unassociated with structural abnormalities are also not included, because there is often uncertainty about their congenital origin. A patent ductus arteriosus in premature babies and in term babies younger than 15 days of age is not considered a form of CHD.

Results

During the study period, 608 patients were evaluated and only 335 patients were diagnosed as having CHD. There were 162 males and 173 females, and male:female ratio was 0.9:1. Two hundred and eighty patients had acyanotic CHD, while 55 patients had cyanotic CHD. The frequencies of specific forms of CHD are shown in Table 1 and comparison is made with the frequencies reported in previous studies (Table 2).

Ventricular septal defect (VSD) was the most frequent form of CHD. It was found in 109 patients (32.5%), 49 male patients and 60 female patients, with male to female ratio of 0.8:1. It was found in isolation in 83 of the patients (76%). The following lesions were found in association: atrial septal defect, 12 patients (11%); patent ductus arteriosus, six patients (5.5%); aortic coarctation or interruption, five patients (4.6%), and pulmonary stenosis, three patients (2.8%). The mean age at diagnosis was 12.2 months. Seventy-three patients (67%)

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TABLE 1. *Distribution of types of congenital heart disease.*

Lesion	# of patients (%)	Male:female	Mean age at presentation (mon)
VSD	109 (32.5)	0.8:1	12.2
PDA	53 (15.8)	0.8:1	11.4
ASD	35 (10.4)	0.9:1	28
PS	34 (10.1)	1.1:1	48
AVSD	12 (3.6)	0.3:1	9.5
MVP	12 (3.6)	0.2:1	112
Aortic coarctation/ interruption	11 (3.3)	10:1	54
AS and/or bicuspid valve	9 (2.7)	8:1	66
Peripheral PS	5 (1.5)	1.5:1	7.7
TOF	15 (4.5)	1.1:1	6
Common ventricle	9 (2.7)	8:1	7.5
PA with VSD	6 (1.8)	0.2:1	8.4
D-TGA	5 (1.5)	4:1	0.2
Ebstein anomaly	5 (1.5)	Females	11
PA (isolated)	4 (1.2)	0.3:1	0.3
DORV	3 (0.9)	2:1	12
TAPVC	2 (0.6)	Males	1
L-TGA	2 (0.6)	Males	12
Truncus arteriosus	2 (0.6)	Females	1.6
Tricuspid atresia	1 (0.3)	Females	2.5
Hypoplastic left heart	1 (0.3)	Males	0.1
Total	335 (100)	0.9:1	

VSD=ventricular septal defect; PDA=patent ductus arteriosus; ASD=atrial septal defect; PS=pulmonary stenosis; AVSD=atrioventricular septal defect; MVP=mitral valve prolapse; AS=valvar aortic stenosis; TOF=tetralogy of Fallot; PA=valvar pulmonary atresia; TGA=transposition of the great arteries; DORV=double outlet right ventricle; TAPVC=total anomalous pulmonary venous connection.

were diagnosed at or before six months of age, while 36 patients (33%) were diagnosed at a later age.

Patent ductus arteriosus (PDA) was the second most frequent form of CHD, occurring in 53 patients (15.8%), 23 males and 30 females, with male to female ratio of 0.8:1. VSD occurred in six of them (11.3%), and associated valvar pulmonary stenosis occurred in one. The mean age at diagnosis was 11.4 months. Thirty-two patients (60%) were diagnosed at or before 6 months of age, while 21 patients (40%) were diagnosed at a later age.

Fifty-five patients (16.4%) had cyanotic CHD. Tetralogy of Fallot (TOF) was the most common form of cyanotic CHD, occurring in 15 patients (4.5% of the whole group). The mean age at diagnosis of TOF was six months, and all but one patient were diagnosed before the age of one. Eleven patients (73.3%) of those with TOF had cyanosis by the time the diagnosis was established, while in four patients the cyanosis had not yet developed on presentation. The latter is explained by the mild pulmonary stenosis. Pulmonary atresia with VSD occurred in six patients (1.8% of the whole group).

Common ventricle defects occurred in nine patients (2.7%)—this includes severe unbalanced forms of

atrioventricular septal defect (AV canal), and excludes tricuspid atresia, pulmonary atresia with hypoplastic right ventricle, and hypoplastic left heart syndrome. D-transposition of the great arteries (D-TGA) occurred in five patients (1.5%), three of whom had small VSDs, while one had coarctation of the aorta; all presented in the first week of life. Ebstein malformation of the tricuspid valve occurred in five patients (1.5%), one of whom had mitral stenosis as well. Pulmonary atresia with intact ventricular septum occurred in four patients (1.2%), while double-outlet right ventricle occurred in three patients (0.9%). L-transposition of the great arteries, total anomalous pulmonary venous connection and truncus arteriosus were very rare; each was encountered in two patients (0.6%). Classical tricuspid atresia and hypoplastic left heart syndrome were found only in one patient each (0.3%).

Discussion

This study does not address the incidence of CHD in the population but shows the relative frequency of various forms of CHD, and although it is hospital-based, it would be reasonable to believe that it represents the population in the Southwestern region for the following reasons. First, the hospital in which the study was conducted is a tertiary health care center and is the only hospital in the region during the study period with appropriate facilities to assess children suspected of having CHD. Second is the ease of access to the service by the whole population of the Southwestern region, and this is attested to by the high number of patients with innocent murmurs referred to the hospital. Erroneous diagnosis and overestimation of particular forms of CHD (e.g., VSD) are avoided by confirming the clinical diagnosis with the help of echocardiography, including Doppler assessment. On the other hand, underestimation is also avoided by subjecting all referred patients to echocardiography even when the provisional diagnosis suggested by the pediatric cardiologist is innocent murmur, and when cardiac signs are absent. If the patient had cardiac symptoms, echocardiography was performed for the purpose of the study.

The most common form of CHD is VSD (32.5%), which is consistent with the findings of previous studies from Saudi Arabia,^{1,2} and from other parts of the world.³⁻⁵ The second most common lesion in this study is PDA, which is also consistent with the findings of previous studies,⁴⁻⁶ however, the frequency of 15.8% is much higher than that reported in studies from Buraidah, Saudi Arabia (8%),¹ North America (5.5%),^{3,4} and Britain (5.5%-10.6%),⁷ but is similar to the frequencies reported from Australia⁵ (15.4%), and Cape Town (16.1%).⁸ A higher frequency of 20.9% was reported from Nigeria.⁶ The relatively high frequency of PDA in the present study may be explained by racial variations, or by the fact that echocardiographic assessment in this study (not used in most of the previous studies) was sensitive enough to diagnose very small PDAs that may not be suspected on clinical grounds. In this study, the most common congenital valve lesion to present in childhood is pulmonary valve stenosis (10.1%). Obstructive

TABLE 2. Percentages of various types of congenital heart diseases in different countries.

Lesion	Present study	California	Minnesota	Australia	Nigeria	Blackpool	Cape Town
VSD	32.5	31.3	34.6	28.2	38.8	28.1	21.8
PDA	15.8	5.5	10.6	15.4	20.9	6.5	16.1
ASD	10.4	6.1	7.3	13.4	–	8.3	17
PS	10.1	13.5	5	14.2	6.0	2.7	9.2
AVSD	3.6	3.7	4.5	–	9.0	7.4	–
MVP	3.6	–	–	–	–	–	–
Coarctation	3.3	5.5	5.6	5.1	–	5.6	6.1
AS and/or bicuspid valve	2.7	3.7	6.1	3.8	3.0	4.1	4.4
Peripheral PS	1.5	–	–	–	–	–	–
TOF	4.5	3.7	5	7.3	3.0	8.6	12.2
Common ventricle	2.7	0.6	0	–	–	1.5	0.7
PA with VSD	1.8	–	–	–	–	–	–
D-TGA	1.5	3.7	7.8	5.1	1.5	5.6	1.8
Ebstein anomaly	1.5	–	–	–	–	–	–
Hypoplastic rt. heart (PA + TA)	1.5	0.6	3.4	–	4.5	1.5	3.7
DORV	0.9	0.6	0	–	–	0	–
TAPVC	0.6	0.6	2.8	14.9	1.5	2.1	0.5
L-TGA	0.6	–	–	–	–	–	–
Truncus arteriosus	0.6	2.5	0	–	–	1.2	0.7
Hypoplastic lt. heart	0.3	0.6	4.5	–	1.5	3.3	–
Miscellaneous	–	17.8	2.8	14.9	10.4	13.6	6.5
Total	335	163	179	1325	67	338	1439

aortic valve lesions were uncommon, occurring in 2.7%. This frequency is similar to that reported from other regions of Saudi Arabia, 3% and 1.6%,^{1,2} and lower than that reported from other parts of the world.^{4,7,8} This may be explained by racial variation. Mitral valve prolapse is found in only 3.6%; this lesion was not included in the previous studies.

The age at which the diagnosis is made is very important for the outcome; for example, patients with large left to right shunt across VSD or PDA may develop pulmonary vascular disease if the diagnosis is made late. Fortunately, this complication rarely occurs before two years of age, but children with atrioventricular septal defect may develop it at an earlier age. Another example where age at diagnosis is important is D-TGA, as it influences the type of surgery and the outcome. The fact that the majority of our patients with VSDs and PDAs were diagnosed at or before six months of age and that all patients with D-TGA were diagnosed during the first week of life probably indicates that the age at which CHD is detected at the primary and secondary health care centers in our region is within acceptable limits.

This study gives only an overview of the pattern of CHD in

the Southwestern region of Saudi Arabia. An epidemiological study to look at the incidence and prevalence of CHD in Saudi Arabia is highly recommended, as this will help in planning the health care system.

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