ORIGINAL ARTICLE

Profile of Congenital Heart Disease and Correlation to Risk Adjustment for Surgery; An Echocardiographic Study

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ABSTRACT

Objective: To determine the pattern and profile of Congenital Heart Diseases (CHD) in paediatric patients (age 1 day to 18 years) presenting to a paediatric tertiary referral centre and its correlation to risk adjustment for surgery for congenital heart disease.

Study Design: Descriptive case series.

Place and Duration of Study: Paediatric Cardiology Department, Armed Forces Institute of Cardiology/National Institute of Heart Diseases, Rawalpindi (AFIC/NIHD).

Patients and Methods: Over a period of 6 months, 1149 cases underwent 2-D echocardiography. It was a non-probability purposive sampling.

Results: This study showed 25% of all referrals had normal hearts. A male preponderance (38%) was observed from 1 year to 5 years age group. Nineteen percent of the cases were categorized as cyanotic CHD with the remaining as acyanotic variety. Tetralogy of Fallot (TOF) represented 10%, Ventricular Septal Defects (VSD) 24%, followed by Patent Ductus Arteriosus (PDA) and Atrial Septal Defect (ASD), which comprised 6.6% and 6.5% respectively. VSD was the most common association in patients with more complex CHD (10%) followed by PDA in 3% and ASD in 1.2% of the cases. Most of the cases were category 2 in the RACHS-1 scoring system.

Conclusion: VSD and TOF formed the major groups of cases profiled. Most of the cases recommended for surgery for congenital heart disease belonged to the risk category 2 (28.1%) followed by the risk category 1 (12.7%) of the RACHS-1 scoring system.

Key words: *Congenital heart disease. Acyanotic congenital heart lesions. Cyanotic congenital heart lesions. Ventricular septal defect. Tetralogy of fallot.*

INTRODUCTION

The internationally reported incidence of congenital heart disease is 8/1000 live births.[1-5] In Pakistan, as indeed in any developing country, there is no accurate data to predict prevalence of Congenital Heart Disease (CHD) at birth, since most births occur in villages and in remote areas and at homes or in ill-equipped basic health units.[6-12] This study was conducted on more or less similar lines to the previous local studies, aiming to sample the profile of congenital heart disease of cases reporting to Armed Forces Institute of Cardiology/ National Institute of Heart Diseases (AFIC/NIHD), Pakistan and correlate the surgical risk involved in correcting these ailments. This could help us prioritize learning management skills for these ailing patients, especially surgery.

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PATIENTS AND METHODS

A total number of 1149 undiagnosed patients from "outpatient department" and "emergency reception" of AFIC/NIHD, Rawalpindi were included in the study. Diagnosis of the cases was based on the 2D echocardiographic examination. Disposal and follow-up including those reporting for repeat echocardiography were not included in the study. Factors that could affect the quality of the study e.g. the level of patient comfort (crying, hyperactive, uncomfortable and apprehensive, severely ill or cyanotic requiring immediate management) were addressed. A hierarchic system of classification was adopted. A brief history and a brief clinical examination of the cardiovascular system along with any relevant examination were made for individual patients. Pulse oximetry saturation data was recorded. Acquired childhood diseases like rheumatic fever and its sequela and viral carditis leading to dilated cardiomyopathy were entered for completeness sake.

The echocardiography was carried out by experienced paediatric echocardiographers. Toshiba echocardiography machine model Aplio-VX UJUR-700-A and saturation monitor PM-5000 Shenzen Mindray Bio-Medical Electronics Co. Ltd., was used for this purpose. For sedation, oral chloral hydrate 50 mg/kg body weight and oral midazolam 0.15 mg/kg was used.

The study design was non-probability convenience sampling. Demographic variables (age and sex), saturation data and findings of echocardiographic examination were recorded and transferred to SPSS 11. A descriptive statistical analysis was undertaken. The data was reviewed for age at presentation, gender and type of heart lesion. Types of lesions were reviewed for its size, severity and other aspects. Patients with structurally and functionally normal hearts were grouped separately. CHD was further subdivided into cyanotic and acyanotic heart diseases. Children with cardiomyopathy were placed in the group of myocardial disease. The study was primarily descriptive and included no comparison; therefore, no test of significance was applied. Categories were then grouped according to the RACHS-1 (risk adjustment for congenital heart surgery) scoring system[29] and results analyzed.

RESULTS

This study showed a male predominance of cases with congenital heart diseases. Of the total 1149 cases, 733 cases (64%) were males with a male-female ratio of 1.85:1, (Table I). Five percent of the cases reporting for echocardiography were less than 1 month of age. Sixteen percent of the cases ranged 1 month to 1 year of age. Thirty eight percent of the cases were in the 1 year to 5 years age group. Twenty two percent of the cases were between 5 years to 12 years of age. Four percent of the cases were between the ages of 12 years to 18 years. In the remaining 14% of the cases age could not be recorded.

Of the cases, 19% were categorized as cyanotic congenital hearts and the remaining as acyanotic variety. Tetralogy of fallot which included variety with pulmonary atresia was found to represent 10% of the cases with male-female ratio of 3.2:1. This was followed by tricuspid atresia representing 2.7% and transposed vessel relationship was found in 3% of all cases from simple TGAs to more complex congenital heart anatomies with single ventricles.

Among the acyanotic hearts, 269 (24%) of the cases had ventricular septal defects with restrictive variety comprising of 16% of the cases. Perimembranous area was the commonest location identified (19%). The frequency age distribution of cases with restrictive VSD suggested that maximum cases were seen in the $8^{1/2}$ years age group. Out of a total of 76 large VSDs with unrestricted flow, 50% were older than 1 year of age with significant pulmonary hypertension. VSD was also found to be the most common association in patients with more complex congenital heart diseases (10%) followed by PDA in 3% of the cases and ASD in 1.2% of the cases. PDA comprised 6.6 % of the total cases with equal male and female distribution and secundum atrial septal defects represented 6.5 % of the total cases with male-female ratio of 1.5:1.

Table I: Sex, and percentage distribution of cases

	Male	Female	Percent
Normal	189	91	25
Cyanotic heart diseases			
TOF with pulmonary stenosis	77	24	9
DORV, TOF	4		0.3
DORV, TGA, PS	3	2	0.5
DORV, PHT		1	0.1
Tricuspid atresia, pulmonary stenosis	10	9	1.7
Tricuspid atresia, VSD, unrestricted flow	5	5	1.0
Total anomalous pulmonary venous return (TAPVR) supracardiac	3	2	0.4
TAPVR mixed	1		0.1
Partial anomalous pulmonary venous return (PAPVR)	1		0.1
TGA simple	4	2	0.5
TGA VSD	2		0.2
TGA, VSD, PS	4	1	0.4
TGA, PS	1		0.1
TGA, complete AVSD	1		0.1
Pulmonary atresia, TOF	6	7	1.1
Pulmonary atresia, dTGA,	2		0.2
Isolated ventricular inversion	1		0.1
Double inlet left ventricle (DILV)	2	1	0.7
Single ventricle DTGA unrestricted flow	2		0.2
Cortriatriatum	2	1	0.3
Multiple congenital anomalies	6	6	1.0
Truncus (all varieties)	7	4	1.1
Acyanotic variety			
ASD	36	23	5.1
Partial AVSD	5	2	0.6
Complete AVSD	13	6	1.9
ASD VSD	3	2	0.6
ASD, PS	3	2	0.4
ASD PDA	2	1	0.3
VSD restrictive	120	62	15.9
VSD unrestrictive	57	26	7.5
VSD pulmonary stenosis	4	7	0.3
VSD, PDA	1		0.1
VSD, ASD, PDA			0.1
Moderate PDA	11	14	2.2
Large PDA	4	2	0.5
Small PDA	22	21	3.7
PDA and coarctation		2	0.2
Coarctation of aorta	3	2	0.5
Corrected TGA	7	2	0.8
Corrected TGA, VSD	1	1	0.2
AP window	2		0.2
Valvular AS	20	7	2.4
Congenital bicuspid aortic valve	1		0.1
Subaortic membrane	1		0.1
Isolated PS	11	13	2.1
Branch pulmonary artery origin stenosis	1	1	0.3
Supramitral membrane	1		0.1
Congenital MS		1	0.1
MLP	4	1	0.6
Parachute mitral			0.1
Ebstein's anomaly	2		0.17

Continued.....

(Table I, continued) **Miscellaneous**

	Male	Female	Percent
Spongiform cardiomyopathy	1		.1
Hypertrophic cardiomyopathy	1	3	.3
Rheumatic valvular heart disease	18	8	2.3
DCM	14	1	1.3
Myocarditis	7	7	1.2
Pericardial effusion (non-surgical)	1	-	0.1
Total cases	733	395	1128 + 21 missing gender entry

Ventricular septal defect (VSD), atrial septal defect (ASD), patent ductus arteriosus (PDA), tetrology of fallot (TOF), tricuspid atresia/stenosis/ (TA) Epstein anomaly, mitral atresia, mitral regurgitation (MR), mitral leaflet prolapse (MLP), mitral stenosis (MS), pulmonary atresia (PA), partial and total anomalous pulmonary venous drainage (PAPVR and TAPVR), transposition of great vessels (TGA), corrected transposition of great vessels (CTGA), truncus arteriosus (TA), co-arctation of aorta (co-A), pulmonary stenosis (PS), aortic stenosis (AS), aortic regurgitation (AR), double outlet ventricle right or left (DORV, DOLV), hypoplastic left heart syndrome (HLHS), vascular rings and slings, aorto-plumonary window (AP window) dilated cardiomyopathy (DCM).

Categorization of cases recommended for surgery, 52.1%, according to the consensus-based method of risk adjustment for in-hospital mortality among children younger than 18 years for congenital heart disease (designated RACHS-1)[14] suggested that 28.1% of the cases belonged to the risk category 2 followed by 12.7% to the risk category 1 of the RACHS-1 scoring system. Risk group 3 comprised 9.1% and risk group 4 made-up 2.2% of the cases.

DISCUSSION

This study does not represent the incidence or prevalence of CHD in the total population of this country. Majority of cases were males (64%), which may be due to the predominance of CHD in males[1-5] or it could be due to more male reporting due to cultural eccentricity.[13] Twenty-one percent of the cases referred were less than one year of age as compared to approximately 40% of the cases in the one year to 5 years age group, correlating well with similar data in previous studies.[7] This can be explained by the fact that in Pakistan, most births occur without supervision of a paediatrician. Expenses involved in travelling, and income loss due to lost work days is a deterrent that often results in further delaying the condition to beyond treatment. In addition, delay may also result from inaccurate diagnosis. In addition, condition such as ASDs, might not be detected until they appear in adult life. Many families seek advice from quacks and “Peers” because of common beliefs and myths.[13]

In this study, VSD was present in 24% of the cases. Approximately 20% of patients in CHD registries have VSD as a solitary lesion.[15,30] Unlike this study, Hoffman found VSD was slightly more common in females.[16] Our data suggested that half of the cases who had VSDs with unrestricted pulmonary flow belonged to the 5-10 years age group. This very particular group of children require diagnosis and surgery before pulmonary hypertension starts to develop, preferably before 6 months of age.[17,19,29,32] Similar situation holds true for large PDAs, which need to be diagnosed well within 1 year age group. In this study, PDA and ASD had almost equal distribution, 6.6% and 6.5% respectively. International work also suggests a similar findings that secundum ASDs represent 6-10% of all cardiac anomalies and are more frequent in females than males.[1,18]

TOF (all varieties) with a male dominance was found to be the most common cyanotic heart disease accounting for 10% of all cases as compared to the figures of 3.5% to 9% in various international studies. In Baltimore-Washington Infant Study (BWIS), TOF accounted for 6.8% of all forms of CHD, and was the most common form of cyanotic CHD.[15] In this study, TOF with pulmonary stenosis accounted for 85% of the cases compared to 79.7% in the BWIS 1981-1989.

Six cases (0.5%) of simple transposition were included in this study. Unfortunately all these cases were more than 1 month old further proving the claim that early referrals and diagnosis for congenital hearts can make a difference.[13] Tricuspid atresia was found in 2.7% of all cases as compared to 3% of patients reported in the New England Regional Infant Cardiac Program.[1]

Aortic stenosis was found in 2.4% of the cases and had strong male dominance comparing well with international literature with figures between 3-8%.[1-5] In this study, aortic stenosis (sub-valvular, valvular and supravalvular) was primarily valvular and congenital in origin.[20,21] Isolated pulmonary stenosis present in 2.1% of all cases in this study had slight female dominance compared to 8-10% cited in international literature with equal male-female ratio.[22]

As VSD and TOF formed the major chunk of cases profiled at AFIC/NIHD, most of them belonged to the risk category 2 followed by the ASD and PDA, risk category-1 of the RACHS-1 scoring system. The in-hospital mortality for the Paediatric Cardiac Care Consortium data were 0.4% in category 1, 3.8% in 2, 8.5% in 3, 19.4% in 4, and 47.7% in 6. There were less cases in category 5 to estimate mortality rates.[14,31] Since majority of the cases belonged to the category 2 of the scoring system, this study showed that we need to develop and prioritize learning management skills for this very subset of patients.

CONCLUSION

VSD and TOF followed by PDA and ASD formed the major groups of cases profiled. Most of the cases recommended for surgery for congenital heart disease belonged to the risk category 2 (28.1%) followed by the risk category 1 (12.7%) of the RACHS-1 scoring system.

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