FREQUENCY OF CONGENITAL HEART DISEASES IN PATIENTS UNDER THE AGE OF TWELVE YEARS AT LADY READING HOSPITAL PESHAWAR

Wahaj Aman, Akhtar Sherin, Mohammad Hafizullah

Department of Cardiology,
Postgraduate Medical Institute, Lady Reading Hospital, Peshawar

ABSTRACT

Objective: To find out the frequency of various congenital heart diseases (CHD) in patients less than 12 years of age at Lady Reading hospital, Peshawar.

Material and Methods: This retrospective study was conducted at Department of Cardiology, Postgraduate Medical Institute, Lady Reading Hospital, Peshawar. Patients referred for echocardiography were reviewed. Trans-thoracic two dimensional echocardiography and Doppler studies were done on all cases. The results were analyzed from the data retrieved from the computerized data base of Cardiology department from July 1998 to December 2005 covering almost seven years and six months. Records of patient aged twelve and below and labeled as congenital heart disease were analyzed on SPSS windows version 10.

Results: Out of 3072 patients with CHD, 83.7% were acyanotic heart diseases. Ventricular septal defect was present in 1248 patients (40.6%), atrial septal defects (ASD) were 493 (16%) comprising of ASD secundum in 397 (12.9%) and ASD primum in 96 (3.1%) patients. PDA was present in 394 patients (12.8%). A total of 473 patients (15.4%) had the tetrology of Fallot and 236 patients (7.7%) had pulmonic stenosis. Complex congenital heart diseases were present in 198 (6.4%) patients and coarctation of aorta was found in 0.9%.

Conclusion: Majority of the congenital cardiac anomalies in patients less than 12 years of age are acyanotic. VSD and ASD are the major acyanotic and TOF is the major cyanotic congenital heart disease.

Key Words: Congenital Heart Disease, Echocardiography, VSD, ASD, TOF, PDA, Pulmonic Stenosis, Complex Cardiac Anomalies

INTRODUCTION

Congenital heart disease, i.e. a 'structural abnormality of the heart or intra thoracic great vessels that is actually or potentially of functional significance[1] has an incidence of 8-10 cases per 1000 live births.[2,3]. As it is not possible to detect all lesions in childhood, the true figure may be much higher than this. Prevalence rate of CHD for whites is significantly higher than for blacks or Mexican-Americans [4]. In Sudan, a prevalence rate of congenital heart disease in children aged 5-15, has been reported as 2.0 per 1000 [5], and CHD accounts for 3.9% of the total hospital admissions for cardiovascular disease in Sudan [6]. The prevalence rate reported from other African countries is 3 per 1000 from Uganda[7] and 3.6 per 1000 from Nigeria[8].

In developing countries it is not an easy task to perform a birth prevalence study of CHD[9]. Majority of births still take place at home and routine neonatal screening is not common due to financial constraints and other cultural and religious issues. A hospital based study from Karachi estimated that congenital heart disease in the neonatal period was 4 per 1000 live births[10].

Due to the advances of heart surgery, interventional cardiology and medical treatment the earlier high mortality rate of congenital heart disease has declined from 80 % to 20 %. Nowadays more than 80 % of children with congenital heart disease reach adulthood [11,12]. However without surgery, only 10 to 15 per cent of patients with CHD survive to adolescence and adult life. Worldwide CHD accounts for 2 % of heart diseases that present in adulthood [13]. Some

defects are lethal during infancy or childhood and many more are trivial or correct spontaneously, e.g. small septal defects. Severe and moderately severe congenital heart disease usually necessitates expert cardiology care. Majority of patients with CHD are picked up in early infancy due to cyanosis in cyanotic heart diseases or in childhood when loud murmur is picked up by the attending physicians. Ventricular septal defect is the most common amongst the acyanotic heart diseases and Tetrology of Fallot is the commonest cyanotic heart disease[14,15].

This study was conducted to find out the frequency of various congenital heart diseases (CHD) in patients less than 12 years of age referred for echocardiography.

MATERIAL AND METHODS

This study was conducted at Cardiology department, Postgraduate Medical Institute, Lady Reading Hospital, Peshawar. This department has the facilities for echocardiography and Doppler since 1984. Computerized data base is available since 1998 and is available online. Being the only tertiary referral centre of NWFP with facilities for invasive and interventional procedures patients are referred from all over the province for opinion and confirmation of diagnosis.

This study was conducted on patients diagnosed as congenital heart disease based on echocardiographic examination under the age of twelve years, from July 1998 to December 2005. All patients referred for echocardiography by in hospital doctors, and general practitioners from all over the province were reviewed. Trans-thoracic echocardiography and Doppler studies were performed by experienced echocardiographers and reviewed by the consultant for the confirmation of the diagnosis. All echocardiographic data was saved in purpose built software running under Multi User Unix OS, and FoxPro data base, since 1991 and updated in 1998 with a capability of accounting system. This is unique in Cardiology and has software features like patients information system, accounting system, management information system, inventory control system. Statistical analysis is available on-line. The results were analyzed from the data retrieved from the computerized data base of Cardiology department of Postgraduate Medical Institute, Lady Reading Hospital, Peshawar from July 1998 to December 2005, covering almost seven years and six months. Records of patient aged twelve and below labeled as congenital heart disease were further analyzed on SPSS windows version 10 for descriptive analysis and nonpaired t test was used to compare the means. Threshold of .05 was used to determine the significance level.

The patient's age, gender and address was noted. The electrocardiograph and chest radiograph of the patients was evaluated for clinical correlation. Echocardiography was done on Toshiba color Doppler ultrasound (SS-A 270 A). Standard M-mode and 2 D examination was followed by Doppler examination. The diagnosis was reviewed on spot by second physician for opinion, confirmation and clinical correlation. The following end points were noted: The number of cases with congenital heart disease and type of congenital heart disease. Male and females of any age were considered. Cases with features of rheumatic heart disease and other acquired heart disease including mitral valve prolapse were excluded from this study. Clearly defined and unambiguous cases with congenital heart disease were included for example Ventricular Septal Defect VSD), Atrial Septal Defect (ASD), Atrial Septal Defect primum (ASDP), Patent Ductus Arteriosus (PDA), Pulmonary Stenosis (PS) and Tetrology of Fallot (TOF).

RESULTS

In this study, 3072 patients under the age of twelve years, diagnosed to have congenital heart diseases referred to Cardiology department, Postgraduate Medical Institute, Lady Reading Hospital from 1998 to 2005 have been included. Out of 3072 patients, 1869 (60.8%) were males and 1203 (39.2%) were females.

Our data reveals that acyanotic heart diseases make the major bulk claiming about 80 % of the share. Among the congenital heart disease VSD claims the biggest segment totaling 40.6% of the total. ASD is 16 % of which one in five have ASD primum. Of cyanotic heart diseases the commonest lesion was Fallot's tetrology constituting 15.4% of the total population of patients with congenital heart diseases. Another 6.4% accounted for complex congenital heart diseases presenting as cyanosed babies.

FREQUENCY OF CONGENITAL HEART DISEASES

Disease	Number (n=3072)	%age
Ventricular septal defect	1248	40.6
Atrial septal defect Secundum	397	12.9
Atrial septal defect primum	96	3.1
Patent ductus arteriosus	394	12.8
Tetrology of Fallot	473	15.4
Pulmonary stenosis	236	7.7
Complex	198	6.4
Coarctation of Aorta	30	0.9

Table 1

VSD: Out of 3072 patients, 1248 patients (40.6 %) had VSD. Out of 1248 patients, 780 (62.5%) were males and 468 (36.8%) were females. Mean age for males was 4. 40 ± 4.36 (CI 4.09 4.7). Mean age for females was 4. 25 ± 4.4 (CI 3.85 4.65). The number of patients presenting under the age of I year were 566 (45%), and 869 (69.3%) of patients were below the age of 5 years.

ASD Secundum: The total number of patients diagnosed as atrial septal defect was 493 (16.04%), out of which 397 (80.5%) had ASD secundum. Out of these there were 288 (72.5%) patients under the age of 5 and 190 (48%) were brought in before their first birthday. Males dominated making the bulk: 246 (62%) as against 151(38%) being females. In female patients the mean age was 3.77 ± 3.55 and (CI 3.20 to 4.35). In male patients, the mean age was 3.65 ± 3.39 and CI 3.23 to 4.08.

ASD Primum : There were 96 patients found with the diagnosis of atrial septal defect primum forming one fifth of total population labeled as ASD. Out of these 81(84.3%) were under the age of 5 and 68(70.8%) were below the age of one. There were 58 (60.4%) males with a mean age of 2.40 ± 1.0 and CI 1.58 to 3.21. Thirty eight were females with a mean age of 2.87 ± 2.25 and CI 1.72 to 4.02. ASD primum presented more in males and at earlier age as against ASD secundum and more patients presented with in one year due to the seriousness of defect.

PDA: 394 patients (12.8%) were found having PDA. 292 (74.11%) of them were under the age of 5 and 176 (44.67%) were under the age of 1. Females 207 (52.5%) were more than males 187 (47.46%). The female patients had a mean age of 3.80 ± 3.47 and the CI 3.32 to 4.27. The mean age of males was 3.49 ± 3.36 and CI 3.01 to 3.98. Sex distribution was nearly same and presentation was a little earlier in males.

TOF: There were a total of 473 patients (15.4%) suffering from the Tetrology of Fallot. Out of these 337 (71.2%) were under 5 and 197(41.6%) under 1. There were surprisingly only 165 (34.88%) females with 3.73 ± 3.47 mean age and the CI 3.20 to 4.27. There were 308 (65.11%) patients who were males. They had a slightly higher mean age of 3.89 ± 3.40 and CI 3.51 to 4.27. Males dominated the presentation but females presented a little earlier.

PS: there were 236 patients (7.7%) found with pulmonic stenosis. 141 (59.75%) patients were below the age of 5 and 73 (30.93%) below the age of 1. Female patients were 93 (39.40%) and the males were 143 (60.59%). The mean ages were higher than usual being 4.78 ± 3.78 for females and 5.13 ± 3.97for males. And the CI were from 4.01 to 5.56 for females and 4.47 to 5.78 for males.

Complex congenital heart diseases: There were 198 (6.4%) grouped as complex congenital heart diseases patients. Below the age of 5 were 154 (77.77%) and below the age of 1 there were 107 (54.04%). The females were approximately half of the males the numbers being 66 (33.34%) females

SEX AND AGE DISTRIBUTION

DISEASE	Males		Female		Mean age	
	Number	%age	Number	%age	Males	Females
Ventricular septal defect n=1248	780	62.5	468	36.8	4.4 ± 4.36	4.25 ± 4.4
Atrial septal defect Secundum n=397	246	62	151	38	3.65 ± 3.39	3.77 ± 3.55
Atrial septal defect primum n=96	58	60.4	38	39.6	2.40 ± 1.0	2.87 ± 2.25
Persistent atrial duct n=394	187	47.5	207	52.5	3.49 ± 3.36	3.80 ± 3.47
Tetrology of Fallot n=473	308	65.1	165	34.9	3.89 ± 3.4	3.73 ± 3.47
Pulmonary stenosis n= 236	143	60.6	93	39.4	5.13 ± 2.97	4.78 ± 2.65
Complex n= 198	132	66. 7	66	33.3	3.32 ± 3.29	3.03 ± 2.97
Coarctation of Aorta n=30	15	50	15	50	2.2 ± 2.17	2.0 ± 0.7

Table 2

COMPARISON OF FREQUENCY OF VARIOUS CONGENITAL HEART DISEASES IN PAKISTAN

	Sadiq M[20]	Rahim F[19]	*Ahmad R et al*[21]	Present study
No of Patients	6620	150	398	3072
Ventricular Septal Defect	1343 (32.1%)	69 (46%)	180 (42.2%)	1248 (40.6)
Atrial Septal Defect	552 (13.2)	4 (2.6 %)	56 (14.08%)	493 (16%)
Patent Ducts Arterious	536 (12.8%)	4 (2.6 %)	39 (9.79%)	394 (12.8%)
Pulmonary stenosis	336 (8.03%)	10 (6.6 %)	28 (7%)	236 (7.7%)
Fallot's Tetrology	372 (16.1%)	38 (25.3 %)	38 (9.54%)	473 (15.4%)

Table 3

and 132 (66.67%) males. The mean female age was 3.03 ± 2.97 and CI 2.30 to 3.76. Males had a mean age of 3.32 ± 3.29 and CI 2.73 to 3.90.

Coarctation of Aorta: There were 30 patients (0.9%) with coarctation of aorta. Half of them were males with mean age of 2.2 ± 2.17 years and half were females with mean age of 2.0 ± 0.7 years.

DISCUSSION

Congenital cardiac abnormalities are very important causes of morbidity and mortality in paediatric age group patients. Aetiology of congenital heart disease is usually unknown, however it is often associated with maternal perinatal rubella infection, maternal alcoholic use, maternal drug treatment and radiation and various genetic and chromosomal abnormalities[16].

The magnitude of the CHD cannot be accurately estimated as majority of studies are hospital based and there are very few population based studies available to know the epidemiology of CHD[17,18]. Congenital heart defects need careful attention and management as it effects the younger generation. The main ailments that effect the younger generation are VSD, ASD, PDA, Fallot's Tetralogy and Pulmonic Stenosis. The last two decades have witnessed great progress in the diagnosis and management of cardiac ailments in children. Echocardiography has revolutionized the management and has offered us a new noninvasive tool in our hands which has offered us new insights into the understanding the myths of all congenital heart diseases. The ease of the technique and being noninvasive hence being able to be repeated frequently, has provided us useful information into the progress of various cardiac lesions. This has also changed the role of cardiac catheterization laboratory, where the emphasis has changed from diagnosis to therapeutic interventions. With the advances in interventional cardiology many lesions can be dilated or closed in the cardiac catheterization laboratory. Earlier definitive diagnosis has opened new doors to definitive treatment like interventional procedures and cardiac surgical procedures which has changed the outlook of otherwise seriously ill patients.

The data on local demography is at the best very sketchy and hospital based. There are no country based statistics reflecting the trend of different cardiac disorders. Ours is the biggest series consisting of children less than twelve years of age from both sexes and based on definitive diagnosis made by echocardiography. Being the pioneer in the field of cardiology and echocardiography in the province, most of the patients labeled as congenital heart disease are referred for a primary or second opinion from all over the province. Our centre being the only referral centre for the province hence reflects the trend of the province.

In our study around eighty percent of patients were acyanotic while others were cyanotic. This figure correlates with the international and national data which report about two thirds being acyanotic[17-20]. Males outnumbered females in our results, also in accordance with other studies from this area[19,21,22].

We had maximum number of patients diagnosed as VSD (40.6%). This finding is in accordance with other studies from Pakistan[14, 15, 19-22] (Table 3). In all these studies, VSD was the commonest acyanotic heart disease, with variable frequency. In our study VSD was more common in males and the mean

age was 4.4 ± 4.36 years for males and 4.25± 4.4 years for females. Mean age of presentation was 4.8 years in another study[20]. Almost half of the patients presented under the age I year and more than thirds had been diagnosed before their fifth birthday. Unlike cyanotic heart diseases where symptomatic patients are brought in infancy, these cases are diagnosed a bit later as majority of these cases are picked up by loud murmurs by the attending physicians.

The second most common CHD in our study was ASD (16%) majority being of Secundum type (12.9%). Another larger study from Pakistan had similar results where ASD was present in 13.2% of cases, with ASD Secundum as more common than ASD primum[20]. Other studies from Pakistan also showed ASD as the second common CHD[21,23]. However Rahim et al[19] reported aortic stenosis and Burki et al[15] reported TOF as the second leading CHD. ASD Secundum is the most commonly diagnosed CHD in patients presenting in adult age group[24].

Persistent atrial duct was present in 12.8% cases in our study. Other studies have reported 12.8%[20] and 9.79%[21] cases of persistent atrial duct. Rahim et al[19] reported 2.6 % and Burki et al[15] reported 8.77% cases of persistent atrial duct. Majority of children with PDA were either infants or older children and number of preterms referred for echocardiography was small. This again is likely to underestimate the true prevalence. The facilities for neonatal ventilation in our setup hardly exist and majority of preterm babies needing ventilation are not likely to survive.

Tetrology of Fallot was the commonest cyanotic heart disease accounting for 15.8% cases in our study. TOF is the most common cyanotic heart disease[11]. Local studies from Pakistan also favoring TOF as the leading cyanotic congenital heart disease[14,19,20]. Rahim et al[19] reported TOF in 25.3% cases of CHD. In our study majority of patients with TOF were males. Patients with TOF can be asymptomatic unless right heart failure or arrythmias occur in due course of time. In adults majority of the patients are usually postoperative. However in our population un-operative TOF can be seen due to shortage of facilities for cardiac surgery.

Other relatively less common congenital cardiac anomalies in our study were PS and complex congenital heart defects in 7.7% and 6.4% of cases respectively. PS has been reported in 7-8 % of cases in various local studies[19-21]. Complex congenital cardiac lesions have been reported in 5.2% cases by Sadiq M et al[20]. Coarctation of aorta was very rare (0.9%) in our study, endorsing the findings of Sadiq et al (1.34%).

In developed world, surgery and catheter based intervention have provided a very high probability of survival in children with congenital heart disease[27]. Management of children with CHD by interdisciplinary team, including the GP, the cardiologist, the psychotherapist and the physiotherapist can help the child to have a normal life style[28]. However such a management cannot be provided to our patients in general. Efforts should be taken to diagnose CHD at the earliest possible age and more surgical interventions be undertaken to improve the survival of patients with various congenital cardiac anomalies.

CONCLUSION

Congenital heart diseases are still common in our set up and majority of the congenital cardiac anomalies in patients less than 12 years of age are acyanotic. VSD and ASD are the major acyanotic cardiac lesions and TOF is the major cyanotic congenital heart disease. In order to estimate the actual magnitude and prevalence of congenital heart diseases in Pakistan, a large population based study is urgently required.

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Address for Correspondence:
Prof Muhammad Hafizullah
Head Department of Cardiology,
Postgraduate Medical Institute,
Govt. Lady Reading Hospital Peshawar