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SPECTRUM OF CONGENITAL HEART DISEASES (CHD) IN OLDER CHILDREN AND ADULT POPULATION

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All the authors contributed significantly to the research that resulted in the submitted manuscript.

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ABSTRACT

Objective: To determine the spectrum of congenital heart diseases in older children and adult population.

Methodology: This was a retrospective cross-sectional study. Data retrieved from computerized database of Echocardiography section of Cardiology department Lady Reading Hospital Peshawar from Feb: 2004 to Oct: 2011.Newly diagnosed CHD patients' age more than 15 years who have not undergone previous intervention (Surgical or Transcatheter) were included in the study. Patients were subdivided into 3 groups on the basis of age. Group A include age 16-35years, group B 36-50years and group C more than 50 years. Frequencies of various types of CHD were determined using SPSS version 16.

Results: A total of 1413 patients were included in the study. Female patients were 742 (52.5%).Mean age in years was 27.91 ±11.94 (range 16-80). Number of patients in different groups were as follow: Group A= 80.2%(1133), Group B=15.2%(215) and Group C= 4.6%(65). The relative no of patients with different diseases were ASD 30.6%(432), VSD 18.9%(267),PDA 18.5%(261), P.S 12.7%(180), Mixed lesions 7.7%(109), TOF 7.14%(101), TGA 1.5%(21), Co arctation of aorta1.2%(17), Ebstien`s anomaly 0.9%(13), A-V canal defect 0.6%(9), Truncus Arteriosus 0.2%(3).ASD was the most frequent CHD in all age groups. There was no patient with Coarctation of Aorta in group C.

Conclusion: CHD is a frequent diagnosis in population above 15 years of age. Attempts should be made to provide early diagnosis so that defects can be corrected in infancy or early childhood to avoid permanent damage.

Key Words: Atrial Septal Defect, Ventricular Septal Defect, Pulmonic Stenosis, Co-arctation of Aorta, Persistant Ductus Arterios.

INTRODUCTION

Congenital heart defects are common group of birth defects, occurring in approximately0.8-1% of live births.[1-4] Historically, most patients with CHD died in early childhood. The improvements in diagnostic, interventional and critical care skills have resulted in survival of approximately 90% of these children to adulthood.[5] Now, for the first time in history, estimates suggest that more adults than children are living with congenital heart disease, and this population is estimated to be growing at 5% per year.[6] Furthermore, whereas in the past it was thought that congenital heart defects seen in adults represented the mild and simple portion of the spectrum, it now seems that the number of adults with conditions of moderate or severe complexity outweighs that of children. This puts a very different face on the requirements for research and education needed to maintain optimal health and prevent secondary disability in this population. The limited data available suggest that only a fraction of adult patients with congenital heart disease are followed up by appropriately trained physicians in a setting in which required services are available.

In September 2004, the National Heart, Lung, and Blood Institute (NHLBI) and the Office of Rare Diseases, both components of the National Institutes of Health, Department of Health and Human Services, jointly convened a Working Group on research in adult congenital heart disease (ACHD). The Working Group started with the premise that a systematic approach to the understanding and management of ACHD would best meet the needs of patients. Components of such an approach include identifying and educating patients, providing diagnostic and treatment services, identifying risk factors for adverse outcomes, and expanding the pool of appropriately trained individuals to care for patients with ACHD.[7]

Due to non-structured health infra-structure in our country, Pakistan, large numbers of children with CHDs are deprived of definitive cardiac surgeries, resulting in increasing number of adults with uncorrected heart defects. This group is commonly known as grown up patient with CHD (GUCH) and carries numerous inherent complications including cardiac failure, infective endocarditis, Arrhythmias, pulmonary hypertension (HTN) and cerebral events.[4,8] Maternal mortality is alarmingly high in GUCH women.[8]

This retrospective study was undertaken with aim of determining spectrum of congenital heart diseases (CHD) in older children and adult population.

METHODOLOGY

This study was conducted at Cardiology department, Postgraduate Medical Institute, Lady Reading Hospital, Peshawar. This department has the facilities for echocardiography and computerized data base is available. 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 andconfirmation of diagnosis and management.

From February 2004 to October 2011, computerized data of all patients with CHD was reviewed. This study included all newly diagnosed CHD patients' age more than 15 years who have not undergone previous intervention (Surgical or Transcatheter) were included in the study. The diagnosis was based on Trans-thoracic echocardiography (2-D and Doppler) performed by experienced echocardiographer and reviewed by the consultant cardiologist. All echo-cardiographic 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 ofaccounting system.

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 Feb: 2004 to Oct: 2011., covering more than seven years and six months. Patients were subdivided into 3 groups on the basis of age. Group A include patients of age 16-35years, group B 36-50 years and group C more than 50 years. Frequency of various types of CHD was determined using SPSS version 16. Data was presented as tables and graphs.

RESULTS

In this study, 1413 adult patients, diagnosed to have congenital heart diseases been included. Out of 1413 patients, 742 (52.5%) were female and 671 (47.5%) were males. Mean age was 27.91 years (range 16-80)SD 11.94 (Table 1). Number of patients in different groups were as follow: Group A=80.2% (1133), Group B= 15.2% (215) and Group C=4.6% (65).

The relative no of patients with common congenital heart diseases were ASD 30.6% (432), VSD 18.9% (267), PDA 18.5% (261), P.S 12.7% (180), Mixed lesions (≥2 lesions) 7.7% (109) and TOF 7.14% (101) {Figure 1}. Frequency of less common congenital heart diseases were, TGA 1.5%(21) and Coarctation of aorta 1.2% (17), Ebstien`s anomaly 0.9% (13), A-V canal defect 0.6% (9), Truncus Arteriosus 0.2% (3) { Figure 1}.

ASD was the most common anomaly in all age groups and its was more frequent in female population (Table 2 & 3). In male patients the most common anomaly was VSD (Table 2).

There was no patient with Coarctation of Aorta and Truncusarteriosus in group C. ASD, PDA and PS were more frequent in female patients while VSD, TOF and TGA were more in male patients.

Table1: Patient Characteristics and Demographic Data

Overall (n=1413)	
Age years±SD	27.91±11.94
Female	52.5% (742)
Group A (16-35 yrs)	80.2% (1133)
Group B (36-50 yrs)	15.2% (215)
Group C (>50 yrs)	4.6% (65)

Figure 1: Frequency of common CHD in adults

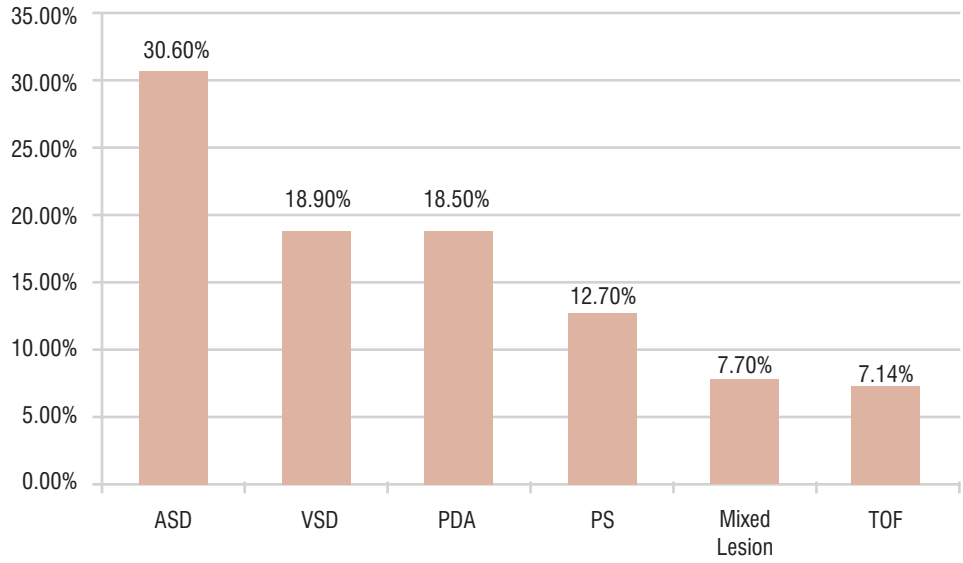

Figure 2: Frequency of uncommon CHD in adults

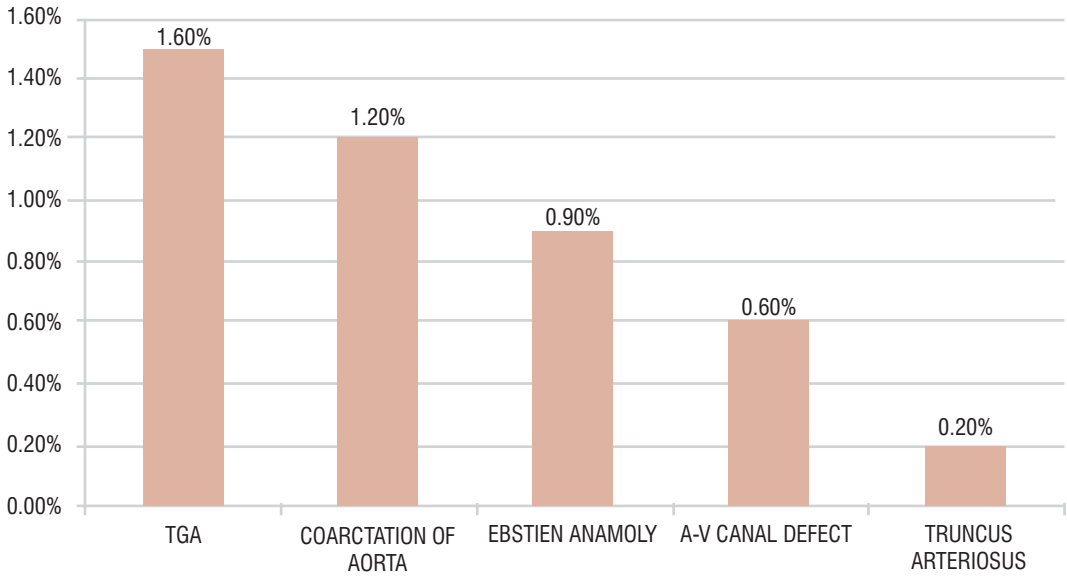

Table 2: Gender Distribution of Adult CHD

OVERALL (n=1413)			
Disease	**Female**	**Male**	**Total**
ASD	281	151	432
VSD	110	157	267
PDA	142	119	261
PS	95	85	180
Mixed Lesions	52	58	110
TOF	38	63	101
TGA	9	11	20
Coarctation of Aorta	8	9	17
Ebstien Anamoly	4	9	13
A-V Canal Defect	3	6	9
Truncus Arteriosus	0	3	3
Total	742	671	1413

Table 3: Age-wise distribution of Adult CHD

OVERALL (n=1413)				
DISEASE	**GROUP A(16-35)**	**GROUP B(36-50)**	**GROUP C(>50)**	**TOTAL**
ASD	293	99	40	432
VSD	227	33	7	267
PDA	228	32	1	261
PS	159	20	1	180
MIXED LESIONS	82	17	11	110
TOF	93	6	2	101
TGA	16	3	1	20
COARCTATION	15	2	0	17
EBSTIEN ANAMOLY	10	2	1	13
A-V CANAL DEFECT	7	1	1	9
TOTAL	1133	215	65	1413

DISCUSSION

Congenital cardiac abnormalities are very important causes of morbidity and mortality inpaediatric age group patients.[9] The initial presentation of congenital heart disease in adult life is now uncommon. The specialty of adult congenital heart disease is largely concerned with long term follow up of patients with previously diagnosed lesions, many of whom have undergone a palliative or theoretically corrective procedure, and who are best managed by an appropriate specialist in a tertiary centre.[10]

The last 50 years have witnessed dramatic changes for the once threatened and limited life of the baby born with congenital heart disease. The advances in echocardiography, anesthesia, intensive care, and particularly cardiac surgery have facilitated the survival of babies born with even the most complex cardiac anomalies. Fifty years ago, only 25% of these infants would survive beyond the first year of life, but now more than 95% will

survive to adulthood. This triumph of survival, which has evolved over the last few decades, has led to a "new population"of adults with congenital heart disease. This population size is estimated to be approximately 800, 000 in the U.S.[11]

They are seldom "cured" by surgery and continue to have cardiac problems. Much time, money, and effort has been devoted to secure their survival, and unfortunately, very little thought has been given to providing for their long-term care. These survivors are extraordinarily courageous and usually, determined to work, contribute to society, and be as normal as possible. In adulthood, they often receive no care or suboptimal care, perhaps the worst of any cardiovascular subspecialty. The cardiology community serves them poorly, and, as we look to the future, we must make provision for lifelong care by trained physicians with expertise in their complex problems.[12]

In our study the most common adult congenital heart defect was ASD (30.6%). Other studies have also reported ASD as the most common adult congenital heart disease.[10,13] ASDs are common, accounting for one third of all congenital defects diagnosed in adulthood. These are detected regularly at an adult echocardiography service as an incidental finding.[10]

In our study ASD was the most frequent lesion in female patient. Other studies have also reported ASD as the most common anomaly in female patients.[14,15]

VSD was the second most common anomaly noted in our study. Previous studies have also reported VSD as the second most common CHD in adult populations.[10,13] while in paediatric age group VSD is the most common CHD[16] but many of these close during the initial few months of age.

In our study PDA was found in 18.5% patients , this is higher than previous reports of 7-10%,[15] in adult patients whileWihajaman et.all: in his study reported PDA in 12.8% cases in paediatric age group.[16] P.S was found in 12.7% patients, Mixed lesions 7.7% (109).

Tetrology of Fallot was the commonestcyanotic heart disease accounting for 7.14% cases in our study. In a review article M. Elizabeth Brickner et all: also stated that in adults, the most common causes of cyanotic congenital heart disease are tetralogy of Fallot and Eisenmenger's syndrome.[17] Although there is no local data available about the frequency of adult congenital heart diseases, studies in paediatric age group from Pakistan alsofavor TOF as the leading cyanotic congenital heart disease.[16,18] In our study majority of patients with TOF were males. 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.

In our study frequency of less common congenital heart diseases were TGA 1.5% and Coarctation of aorta 1.2% , Ebstien`s anomaly 0.9% , A-V canal defect 0.6% , Truncus Arteriosus 0.2%. There is no local data available on the frequency of these disorders in adult population.

CONCLUSION

CHD is a frequent diagnosis in population above 15 years of age. ASD and VSD are the major acyanotic cardiac lesions and TOF is the major cyanotic congenital heart disease. Attempts should be made to provide early diagnosis so that defects can be corrected in infancy or early childhood to avoid permanent damage. A large population based study is required to estimate the actual magnitude and prevalence of adult congenital heart diseases in Pakistan.

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