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Original Research Article

Spectrum of Acyanotic Congenital Heart Disease in Rajasthan: A Hospital Based Study

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ABSTRACT

Objective: To assess the spectrum of acyanotic congenital heart disease in pediatric age group (0-18 years) in South Rajasthan.

Methods: This is a single centre observational study, conducted at a tertiary care hospital of south Rajasthan. Pediatric patients referred to department of cardiology with suspected clinical diagnosis of congenital heart disease and on echocardiographic evaluation found to be having acyanotic congenital heart disease were included in the study. After inclusion, the patients were classified according to the cardiac defects and age at presentation.

Results: Over the study period of one year, a total of 537 patients were diagnosed as having congenital heart disease-398 (74.2%) of which were acyanotic. More than one defect was present in 44% patients and the frequency of multiple defects decreased with age. Overall ASD, VSD and PDA were equally distributed (~40% each). ASD and PDA were predominant lesions in neonates while VSD was the most common lesion in post neonatal life. Less common defects reported were valvular pulmonary stenosis, coarctation of aorta, persistent left superior vena cava, AV canal defect, branch pulmonary stenosis, aortic stenosis, lutembacher syndrome, subaortic membrane, atrial septal aneurysm, congenital mitral stenosis and aortopulmonary window. Four cases had dextrocardia.

Conclusion: The study presents the overall spectrum of various congenital acyanotic heart diseases in Indian children. This study highlights the proportional burden of individual acyanotic CHD in varied age groups and suggests large scale studies to better characterize the epidemiology of these easily missed and potentially treatable congenital diseases

Keywords: Congenital Heart Disease; Acyanotic Congenital Heart Disease.

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INTRODUCTION

Congenital heart disease (CHD) is the commonest of all birth defects and is the most common type of heart disease among children.[1] CHD is defined as a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance.[2] The prevalence of CHD in India ranges between 3.9 – 26.4 per 1000 live births, in hospital based studies.[3-5] This heavy burden emphasizes the importance of this group of heart diseases. Amongst CHD majority are acyanotic CHDs which account for about two third of CHD burden.[2,4-5] With improvement of pediatric cardiac care, their survival to adulthood has increased leading to increasing number of surviving adult patients with uncorrected congenital heart diseases. Majority of acyanotic CHD are simple and potentially correctable. Over the last few years, rapid improvement in percutaneous interventional therapies for acyanotic CHD has occured and majority of these can now be dealt without exposing the patients to an open surgical procedure in early life. However clinicians either miss the diagnosis because of lack of suspicion or based upon their misconceptions about natural history falsely assure parents regarding large defects also. This leads to delayed diagnosis or improper follow-ups and may be detrimental. Few retrospective studies from various parts of India have addressed the overall childhood spectrum (0-18 yrs) of CHD.[6-8] A study from Bangalore evaluated the spectrum of cyanotic CHD.[9] Studies with a relative spectrum of congenital acyanotic heart disease in overall children of 0-18 years are largely missing. No study from Rajasthan has yet addressed the issue. This prospective observational study was undertaken to evaluate the frequency of various forms of congenital acyanotic heart disease diagnosed by 2D echocardiography in children. The study has demonstrated the relative proportion of cases with acyanotic CHD evaluated in a tertiary care centre of South Rajasthan.

MATERIALS AND METHODS

This study is a single centre prospective observational study, conducted at Department of Cardiology of a tertiary care hospital of south Rajasthan from July 2015 to June 2016. The objective of the study was to evaluate spectrum of congenital acyanotic heart disease in pediatric age group (0-18 yrs) patients. All the children of 0-18 yrs age group referred from department of pediatrics with clinical suspicion of heart disease, neonates referred for screening of congenital heart disease or patients attending cardiology outpatient department with suspected heart disease were subjected to clinical examination followed by comprehensive transthoracic echocardiography using standardized equipment (Vivid 7 from General Electric Company). For younger children pediatric probe 7S (3.5 – 8.0 MHz) and for older children M4S probe (2-5 MHz) was used. Those who were found to have acyanotic congenital heart disease were included in the study. Excluded patients were those with normal echocardiography (e.g. neonates with normal screening echo or patients with cardiac failure and respiratory distress due to noncardiac causes like anemia), with acquired heart diseases (e.g. Rheumatic heart disease, myocarditis or pericardial effusion) or those with congenital cyanotic heart diseases (included in another ongoing registry of cyanotic heart disease at the centre). The patients of previously established diagnosis of acyanotic CHD who came for follow up were also excluded to avoid duplication. After final inclusion in the study, patients were classified according to the cardiac defects and age at presentation. Children were arbitrarily divided into five groups, namely early neonatal (<7 days), late neonatal (7d-1 month), infancy (1-12 months), preschool age (>1–5 years) and school age/adolescence (6–18 years).

Table 1: Age and Gender distribution of Study population

	Males	Females
< 1 week	34	40
1wk-1month	51	47
>1month- 1yr	56	45
>1-5 yrs	33	25
>5-18 yrs	29	39
Total	203(51%)	195(49%)

Table 2: Age and gender wise distribution of common acyanotic heart disease

	Age Wise Distribution					Gender Wise Distribution		Total	% of acyanotic CHD (n=398)	% of overall CHD (n=537)
	<1wk	1wk- 1 m	>1m-1yr	>1-5yr	>5-18yr	Males	Females			
Single lesion	28	41	63	42	50	118	106	224	56.3%	41.7%
Multiple lesion	46	57	38	16	17	85	89	174	43.7%	32.4%
ASD	42	56	28	9	23	73	85	158	39.7%	29.4%
VSD	20	24	51	27	29	79	72	151	37.9%	28.1%
PDA	44	57	32	14	9	76	80	156	39.2%	29.1%
PFO	18	23	14	3	0	30	28	58	14.6%	10.8%
ASD+PDA	22	23	6	4	3	27	31	58	14.6%	10.8%
ASD+VSD	15	13	9	3	2	17	25	42	10.6%	7.8%
VSD+PDA	7	8	6	1	0	8	14	22	5.5%	4.1%
ASD+VSD+PDA	5	5	2	0	0	3	9	12	3.0%	2.2%

Table 3: Contribution of less common congenital acyanotic heart diseases in overall spectrum

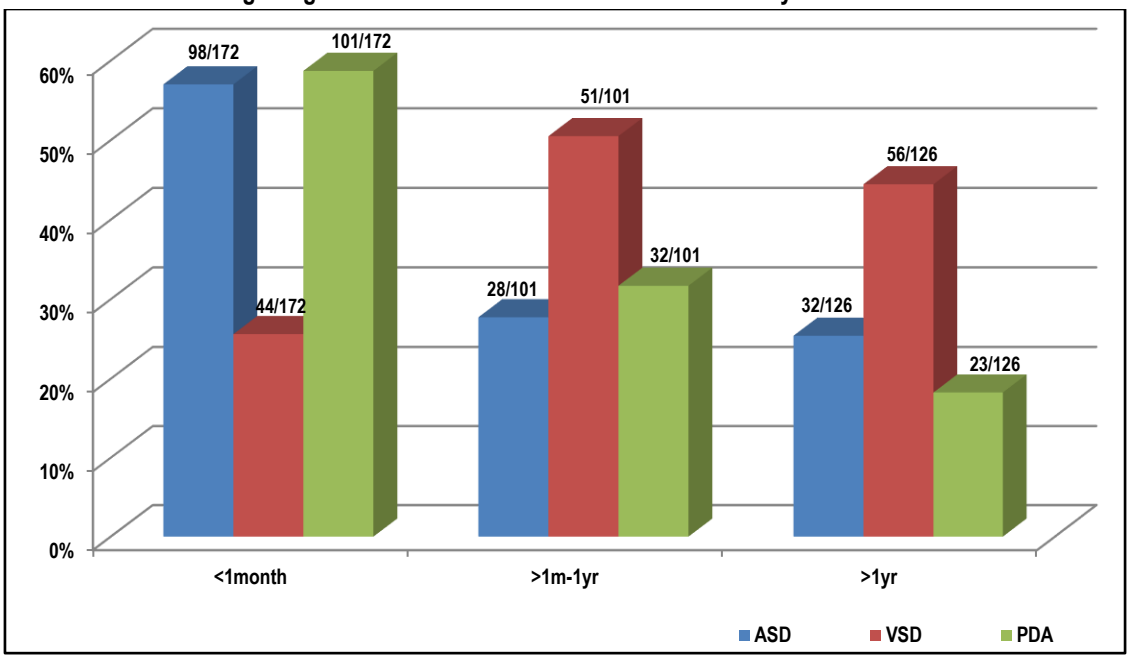
Disease	Number of patients (%)
Valvular Pulmonary Stenosis	15 (3.8%)
Coarctation of Aorta (CoA)	12 (3.0%)
Persistent Left superior Vena Cava	8 (2.0%)
AV canal defect	7 (1.8%)
Branch Pulmonary artery Stenosis	6 (1.5%)
Aortic Stenosis	6 (1.5%)
Lutembacher syndrome	4 (1.0%)
Sub aortic membrane	2 (0.5%)
Atrial septal aneurysm	2 (0.5%)
Congenital mitral stenosis	1 (0.3%)
Aorto-pulmonary Window	1 (0.3%)
Dextrocardia	4 (1.0%)
VSD+ AR	4 (1.0%)
BiAoV + CoA	3 (0.8%)

RESULTS

During the study period a total of 1105 children of age 0-18 years were evaluated. Of these 568 were either normal or had acquired heart disease and therefore excluded from the study. Remaining 537 were having CHD. Among these 139 (25.8%) were having cyanotic CHD and excluded. Rest 398 patients constitute our study group i.e. acyanotic CHD (74.2% of overall CHD) of which 203 (51%) were males and 195 (49%) were females. Age and gender distribution of study population is depicted in Table 1. Isolated single heart defect was present in 224 (56%) while combination of multiple defects was seen in 174 (44%) cases. Majority of acyanotic defects were Atrial septal defect (ASD), ventricular septal defect (VSD) and Patent ductus arteriosus (PDA); all were nearly equally distributed. About 40% of all congenital acyanotic heart disease (~30% of overall CHD) patients had either of these. Among combined lesions, the combination of ASD and PDA was the most common (58 cases i.e. 14.6%). Combined lesions were mostly distributed in younger study subsets (mostly infants) with remarkably decreased number seen in later age group. (Table2). Among patients with VSDs (n=151); 96 (63.6%) were distributed in perimembranous location. Others were muscular (17.2%), subaortic (8.6%) or inlet (5.3%) VSDs. Multiple VSDs were present in 12 (7.9%) of these patients.

Apart from these major isolated or combined defects, various minor lesions were also encountered. This involved 15 cases of valvular pulmonary stenosis, 12 cases of coarctation of aorta (3 were associated with bicuspid aortic valve) and 8 cases of persistent left superior vena cava (LSVC). Three of LSVC were associated with Ostium secundum ASD (1 of which had additional branch pulmonary stenosis), 1 case associated with sinus venosus ASD, 1 case with bicuspid aortic valve and AS, 2 cases with combined OS ASD and VSD (one of which had additional coarctation) and 1 with complete AVSD & PDA. There were 7 cases of AV canal defect (4 of which were complete AVSD), 6 cases of branch pulmonary stenosis, 6 cases of aortic stenosis (3 of which had bicuspid aortic valve), 4 cases of lutembacher syndrome, 2 each of subaortic membrane and atrial septal aneurysm, 1 each of congenital MS (mitral arcade) and aortopulmonary window. There were 4 cases of AR, all were associated with large VSD. Four cases had dextrocardia. (Table 3) After excluding the neonatal period (during which PVR may be physiologically high), 24 patients (10.6%) had increased PA pressures (>40 mm Hg). Three patients (0.7%) had infective endocarditis (all three had VSD), 4 (1%) had LV dysfunction and 1 had RV dysfunction.

Fig 1: Age wise distribution of three most common acyanotic CHD



DISCUSSION

The prevalence of CHD in India ranges between 3.9 – 26.4 per 1000 live births.[3-5] In our study we found that among all patients diagnosed to have CHD (N= 537), 398 were having acyanotic CHD. This accounts for 74.2 % of overall burden of CHD. Prior large hospital and community based studies favour our finding.[2,8,10-11] In our study ASD, VSD and PDA contribute equally (~40% each) to the overall burden. Though there are no studies regarding spectrum of acyanotic CHD per se, most of prior large scale studies evaluating prevalence of CHD have reported VSD to be the most common lesion.[11-13] This can be explained by differences in relative population age. In our study 273 out of 398 patients (68.6%) were infants of < 1 yr age. This included 172 (43% of overall) neonates of <1 month. This is attributed to increasing number of hospital deliveries and increasing awareness and prompt referral on suspicion of congenital heart defects. If we analyse the neonatal population of various studies we find that ASD and PDA outnumbered VSD in these age groups.[7-8] On careful analysis of age distribution as illustrated in Figure 1, we find that PDA was the most common lesion in neonatal age group. The spectrum of children older than 1 month in our study shows the predominance of VSD as most common defect which is in accordance with most other studies. Studies based on echocardiography[14-16] have shown that in the term infant the ductus arteriosus is almost always closed by four to seven days after birth. Therefore, studies done a few days after birth will record a larger number of subjects with a PDA than studies done after three weeks.[17] Frequency of multiple defects also decreases

with age (Table 2) This is explained by spontaneous closure of one or more defect(s). CHD comprises one of the major diseases in the pediatric age group with majority being acyanotic CHD. However there are very few Indian studies stating the whole spectrum of these diseases and none from Rajasthan, hence this effort in this direction.

LIMITATIONS

This study is a single centre hospital based study. This cannot depict the true incidence or prevalence of the disease in the region, which requires large scale community based epidemiological studies. Secondly being hospital based, this also suffers from referral bias. Only the part of iceberg that is caught because of symptoms or auscultation by an astute clinician is picked up and asymptomatic patients are inevitably missed. Thirdly as we know that a significant number of these defects spontaneously close with age, exact natural history cannot be truly estimated in a single contact observational study. Although a rough estimate can be obtained by age-wise relative distribution of these defects, true estimation will require longer duration studies with meticulous follow up.

CONCLUSIONS

Although this study does not provide any novel data, this study is relevant in presenting spectrum of various congenital acyanotic heart diseases in Indian children. This study highlights the proportional burden of individual acyanotic CHD in varied age groups and suggests large scale studies to better characterize the epidemiology of these easily missed and potentially treatable congenital diseases.

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