Clinical Profile Of Patients With Acyanotic Congenital Heart Disease In Pediatric Age Group In Rural India

Smita Mundada\textsuperscript{1} Jagdish Kathwate\textsuperscript{1} Mukund Bajaj\textsuperscript{2} Sadhana Raut\textsuperscript{1}

\textsuperscript{1}Assistant professor, Dept of pediatrics, Govt. Medical college, Aurangabad, India.
\textsuperscript{2}Consulting cardiologist, Assistant professor, Dept of Cardiology, M.G.M hospital, Aurangabad, India.

Abstract: Introduction: Prevalence of CHD according to various studies is 8-10 per 1000 live births worldwide. According to status report on CHD, 10\% of the present infant mortality in countries may be accounted for by CHD. Pediatric cardiac care in developing countries is still in infancy. Methods: The present study was conducted on a total of 100 patients with acyanotic congenital heart diseases. A detailed clinical history and examination was carried out of every patient. Investigations like ECG and X-ray were done. The final diagnosis was confirmed by 2D-Echo and colour Doppler. Results and Conclusions: VSD was the most common type of acyanotic CHD, seen in 46\% of cases. This was followed by ASD (24\%), and then PDA (19\%). Significant proportions of patients (37\%) were diagnosed incidentally. Recurrent respiratory tract infections was the most common symptom (40\%). Weight of 68\% cases was below 10\% percentile while height was below 10\% percentile in 25\% of cases. The average age of presentation was late as compared to other studies. Rate of complications and growth retardation were seen in higher proportion of patients as compared to other studies. The study thus emphasis on greater attention in the field of paediatric cardiology in developing countries.

Key Words: ASD, Children, PDA, VSD.

I. Introduction

Congenital heart diseases (CHD) refer to structural and functional heart diseases, which are present at birth (CON, "together"; genitus, "born")\textsuperscript{1}. There are several reports suggesting the declining prevalence of rheumatic heart disease in our country. On the other hand, prevalence of CHD has remained same worldwide. Various studies have quoted it to range from 8-10 per 1000 live births\textsuperscript{2}. According to status report on CHD in India, 10\% of the present infant mortality may be accounted for by CHD\textsuperscript{3}. Pediatric cardiac care in India is still in infancy, there are very few specialized pediatric cardiology centers and are concentrated in certain regions of India. Training programmes exclusively dedicated to pediatric cardiology and pediatric cardiac surgery need to be established in centers with good standards of pediatric cardiac care. With the advent of various diagnostic tests like electrocardiography, roentgenography, phonocardiography, 2D-echocardiography, colour Doppler echocardiography, cardiac catheterization, angiography and magnetic resonance imaging, accurate diagnosis of any congenital malformation of the heart is possible\textsuperscript{4}. There are diagnostic tools available today by which an accurate diagnosis of CHD can be made even before birth. However these investigatory facilities are not available freely, the cost of investigation and risks involved in invasive techniques restrict their use repeatedly to assess the progress of the disease and decide the need to intervene surgically. Hence these newer methods of diagnosis should be used to confirm or rule out the diagnosis previously made with clinical observations. They should be used to supplement and not to substitute social information, which only a careful history and physical examination can supply. The present study is of acyanotic congenital heart diseases in pediatric age group. A clinical diagnosis was made after taking a detailed history, careful clinical examination, an electrocardiogram, a plain film of the chest, clinical acumen was evaluated in the form of provisional diagnosis and the final diagnosis reached with 2D-echocardiography and colour doppler.

Aims And Objectives

1. To study the relative incidence (frequency) of various types of acyanotic congenital heart diseases.
2. To correlate various variables like age and sex distribution, age of presentation, extracardiac malformations, complications, ECG and X-Ray abnormalities in acyanotic congenital heart diseases.

II. Material And Methods

This study included 100 cases of Acyanotic Congenital heart disease. It was conducted over a period of 24 months from Oct. 2011 to Sept. 2013 and included patients admitted to the pediatric ward and patients who attended the outpatient department. A proforma was prepared. Detailed case history was taken in every case. A special reference to the age of onset of symptoms, course of the illness, symptoms referable to cardiovascular system or complication of congenital heart disease was made. Past history of recurrent lower respiratory tract infections was the most common symptom (40\%). Weight of 68\% cases was below 10\% percentile in 25\% of cases. The average age of presentation was late as compared to other studies. Rate of complications and growth retardation were seen in higher proportion of patients as compared to other studies. The study thus emphasis on greater attention in the field of paediatric cardiology in developing countries.

www.iosrjournals.org
infections, chest pain, congestive cardiac failure, etc was noted. History of consanguineous marriage and a family history suggestive of heart disease were enquired. Antenatal history of drugs, infection, and birth order was noted. The motor and mental milestones, anthropometric measurements and effort tolerance were recorded. A thorough physical examination was carried out. Any associated anomalies were looked and noted for. The cardiovascular examination was done as per the proforma. The murmurs were graded into 6 grades according to Levine's classification. All the patients were then subjected to electrocardiograph and radiologic studies. As relevant to the cases various views of chest X-Ray were taken to delineate chamber enlargement, heart size, perfusion of lungs, position of liver and stomach. After the above procedures were complete, a provisional diagnosis reached after clinical examination. Other investigations like haemoglobin, leucocyte count, ESR, blood-culture etc, were carried as relevant to the individual case. The diagnosis reached after clinical examination, electrocardiography and X-Ray was revised and/or confirmed by 2-D echocardiography and colour Doppler which gives detailed structural abnormalities and an accurate diagnosis.

III. Results And Discussion

In present study, VSD ranked first in incidence among all acyanotic CHD. 46(46%) patients were diagnosed to have VSD. This was similar to the incidence quoted by Jain K. K. et al[5] i.e. 45.4%. Sharma et al[6] also founded incidence of VSD to be 56% in his study. However, incidence of VSD found by L. Kasturi et al[7] was 27%; that observed by Rashmi Kapoor and Shipra Gupta[8] was 21.3%. Mukul Misra et al[9] also observed the incidence of VSD to be 28.3%.

Age

As the patients studied were less than 12 years ages; 12(28%) of the patient were below 1 year of age; 10(22%) were between 1 - 3 years of age. 11 (25%) were between 4 - 6 years of age and 13 (27%) were between 7-12 years of age. Rashmi Kapoor et al[8] as in her study observed that 52% of patients were below the age of 3 years. Rene A. Arcilla Agusston et al[10] had similar observations.

Sex

36 patients with VSD were males and 10 females. Thus male to female ratio in our study in patients with VSD was 3.6:1. Ratio of male to female in the series of Wood et al[11] was 1.7:1 and that in Seizer et al[12] was 2.7:1. Jain K.K. et al and Bidewai DS et al[5] also showed male dominance in cases of VSD. However V. M. Vashishta[13] in his study observed female predominance in patients with VSD.

Symptoms

32 patients of VSD (70%) were symptomatic for various duration where as 14(30%) were asymptomatic. Devashish et al[14] in his study, observed that 22(78.57%) were symptomatic while 6 (21.43%) were asymptomatic. Earliest to manifest with symptoms was of 23 days. Majority of patients below the age of 1 year were asymptomatic. Beas et al[15] also noted the same. 11 patients had onset of symptoms before 1 year of age, 9 patients between 1 - 3 years of age, 6 between 4 - 6 years age and 6 between 7-12 years of age. Similar observations have been made by Nadas et al[16] and Hoffman et al[17]. History of repeated lower respiratory tract infection was the main symptom in 21 patients, 14 patients complaint of breathlessness. 10 patients were brought by parents with problem of feeding difficulty and 3 patients had failure to thrive. Wood et al[18]; Campbell et al[19] and Agusston et al[10] also observed similar symptoms in their study.

Type Of Vsd

20 (43%) of patients had perimembranous VSD making it most common type of VSD. 12(27%) patients were diagnosed with doubly committed VSD, 4(8.5%) had inlet VSD while 10(21.5%) patients had muscular VSD. The incidence of subtypes of VSD were comparable to that observed by Joseph Perloff et al at 20 as per as muscular VSD and inlet VSD are concerned. However Perloff mentions that approximately 80% of ventricular septal defects are perimembranous and only 5 to 7% have doubly committed VSD. But he also quotes that approximate incidence of doubly committed VSD is 27-30% in south-east Asia. The relative lower incidence (43%) of perimembranous VSD in the present study can be attributed to much higher incidence (27%) of doubly committed VSD.

Pulmonary Hypertension

The criteria for labelling the patient with diagnosis of pulmonary hypertension was clinically palpable P2 and confirmed by 2D-Echo as pulmonary artery systolic pressure calculated on the basis of TR jet. 43.9(9%) patients had pulmonary hypertension; none of them had eisenmenger's syndrome. 5 out of 9 patients had VSD.
Growth And Development
34 (74%) of patients with VSD had weight <10th percentile. 9 (19%) patients had height <10th percentile for their age and sex. Adams et al, Linde et al21 also observed significant growth disturbance in patients of VSD. All 5 patients with pulmonary hypertension had weight of less than 10th percentile. 4 out of 5(80%) had height of <10th percentile. Thus it was seen that development of pulmonary hypertension has huge bearing on growth and development of child. Similar observations were made by Birgul Varan et al22. One patient with sub-aortic VSD had moderate grade of Aortic regurgitation (AR). The prevalence of AR with VSD depends largely on the relative prevalence of doubly committed subarterial defects. Subarterial defects are common in Asians with an incidence of AR of approximately 30%. AR is mainly due to sagging or herniation of right coronary cusp23. One patient had left ventricle to right atrial shunt (Gerbodies defect). Though described first by Thurman in 1838, the publication of Gerbode and associates in 1959 stimulated renewed interest in the malformation23. One patient had polydactyl in upper limbs. One patient had features of trisomy 13-cleft palate, micro-ophthalmia, deafness and polydactyl24.

Ecg
In 21(45%) patients ECG were normal. Wood et al 11, Adams and Char et al also observed similar findings. 24(53%) patients had left axis deviation; while 22(48%) patients had LVH. Only 3(8%) patients had Biventricular hypertrophy. Riggs et al [25] also observed the same. ECG is a useful mirror of the physiologic derangement but not of the anatomic location of isolated VSD. The ECG is influenced by the presence and degree of volume overload of the left side of the heart and pressure overload of the right [20].

X Ray
19(41%) patients had normal cardiac contour. 19(41%) patients had LV type of cardiomegaly while 8(18%) patients had biventricular type of cardiomegaly. Increased lung perfusion was seen in 26(59%) patients. Keith et al[26], Campbell et al [19], Deives et al also observed similar findings. X-ray were mainly normal in patients with small restrictive type of VSD. BVH was mainly common in non-restrictive VSD particularly in infants.

Complication
2 patients presented in CCF. These two patients were infants with large non-restrictive VSD. 2 patients were admitted with pneumonia. Increased left to right shunt predisposes to the decreased local immunity of lung parenchyma.

Patent Ductus Arteriosus (Pda)
Incidence
In present study, there were total twenty four (24%) patients who were diagnosed as having PDA. This incidence was comparable with the incidence of 28% found by Sharma et al 6. However in most of other studies, incidence of PDA observed was much less. L Kasturi 7 noted it to be 11%; Mukul Mishra[9] observed it to be 9% and Rashmi Kapoor et al [8] noted it to be 14.6%.

Age
10 patients were below 1 year of age and 9 were between 1-3 years of age group; indicating 19 out of 24(80%) were diagnosed below 3 years of age. Rashmi Kapoor et al [8] also observed that 37 out of 41 patients with PDA were below the age of 3 years. This indicates decreasing number of patients with PDA with increase in age. This may indicate early diagnosis of PDA or spontaneous closure at early age.

Sex
17(70%) out of 24 patients were females.

Symptoms
Majority of the patients 20(83.33%) out of 24 patients were symptomatic. Devashish et al[27] in his study observed that all 8 patients with PDA were symptomatic. Symptoms of 4 patients started before age of 1 month. 17 out of 20 symptomatic patients had onset of symptoms before the age of 3 years. This finding of our study are similar to that of MoundAbottet al [28]; Steinberg et al [29] and Anderson et al[4]. 12 patients presented with recurrent lower respiratory tract infections. 4 complaint of Breathlessness. 8 patients were brought by parent with feeding difficulties and fatigability. This was in contrast to other studies by Steriberg et al[29] and M. Anderson et al [30] where majority of patients of PDA were symptomatic with dyspnoea.
Pulmonary Hypertension

3 (12.5%) patients with PDA had significant pulmonary hypertension. The incidence was comparable to incidence of 17.4% as found by R. J. Shepherd[31].

Growth And Development

17(73%) patients had weight of less than 10th percentile of their age and sex matched group. 8(33%) had height of less than 10th percentile. Adams et al[32], Linde et al[21] also demonstrated similar findings. All 3 (100%) patients with pulmonary hypertension had their weight and height below 10th percentile. Birgit Varan[22] in his report has stated that though inadequate calorie intake, mal-absorption and increased energy requirement may all contribute to growth retardation in CHD, it is the development of pulmonary hypertension that markedly affects the growth of a child with CHD.

Associated Congenital Anomalies

One patient had feature of Rubella syndrome. The baby was low birth weight and had failure to thrive in addition to PDA, bilateral Cataract and deafness. Interestingly mother did not give history of fever and rash in antenatal period. Cosh J. A. and Erigle M. A. et al[33] had described patients with maternal Rubella syndrome in their series. One patient had features of cerebral palsy due to hypoxic encephalopathy. There was definitive history of delayed cry at the time of delivery.

Ecg

10(41.75%) patient had normal ECG. Many publications reported normal ECG in uncomplicated PDA. Dozel et al observed normal ECG in 33% patients. Cruze et al[34], and Perloff et al[20] also observed the same. 14(48.45%) patients had left axis deviation. 12(50%) patients had LVH while 7(28.5%) patients had LA enlargement. 2(8.33%) patients had BVH. Keight et at [28] found BVH in 5% patients with PDA.

X Ray

10 patients had normal cardiac contour. 16(66%) patients had increased lung perfusion. Burwell et al found increased lung vascularity in 9 out of 9 patients in his series while Steinberg 98 and Perloff[20] found it in 50% of cases. 12 patients had Left Ventricular type of cardiomegaly, while 2 patients had Biventricular hypertrophy. Biventricular Hypertrophy correlated with development of pulmonary hypertension.

Complications

2 infants presented in CCF. 1 patient presented with left lower zone pneumonia. One patient was admitted with suspected infective endocarditis. Perloff [20] had described the complications of PDA on the same ground.

In genral VSD was the most common type of acyanotic CHD. [Table no.1] seen in 46% of cases. This was followed by PDA (24%) and then ASD (19%). PS was seen in 5% cases and HOCM. AS and coarctation or aorta in 2% each. ASD, though a much common condition was third in incidence. This may be justified by the fact that majority of patients with ASD are asymptomatic in first and second decade of life and so rarely seek medical attention. The same hold true for bicuspid aortic valve causing aortic stenosis/ regurgitation. There is male preponderance regarding the frequency (61%) of occurrence of acyanotic CHD. The entire patients were below 12 years of age. PDA was the most common form of acyanotic CHD in neonates. In contrast to other forms of CHD, PDA was common in males (70%). Significant proportions of patients (37%) were asymptomatic and were diagnosed incidentally. Recurrent respiratory tract infections was the most common symptom (40%) followed by feeding difficulties (24%) and breathlessness on exertion (21%). Very few patients were symptomatic (9%) in first month of life. Weight of 68% cases was below 10th percentile while height was below 10th percentile in 25% of cases. [Table no.2] Growth retardation was more profoundly affected in patients and pulmonary hypertension. [Table no.3] The incidence of pulmonary hypertension was 9%. All patients (100%) with pulmonary hypertension were small for weight (<10 percentile); 8 (89%) of these were with height less than 10th percentile. Thus, there is high incidence of malnutrition in patients with acyanotic CHD, especially with pulmonary hypertension (P<0.05). In 48 (46%) cases, mother’s age at pregnancy was between 21-25 years. In one case of ASD, age of mother was more than 35 years. This child had features of Down’s syndrome. Majority of patients (50%) were first by birth order while 36(36%) patients were second born. The increased incidence of CHD in 1st and IInd born child may be reflection of small family size. There was high incidence (9%) of consanguineous marriage seen in parents of cases with acyanotic CHD. Family history of congenital heart disease, though depicted very frequently in literature was found in only one case of HOCM. Aortic regurgitation was seen in one patient of subaortic VSD. One patient of VSD had Gerbodies defect. Polydactyl was seen in one patient. One patient with VSD had trisomy 13, hypoxic encephalopathy and features of rubella syndrome was seen in one patient each with PDA. One patient with ASD had Downs syndrome, while the other one had severe mitral regurgitation. One patient with pulmonary stenosis had
Noonan’s syndrome. Thus associated congenital anomalies are quite common in acyanotic CHD. 94% cases were accurately diagnosed clinically before being finally diagnosed by 2D-Echo. This proves the utility of clinical examination even in the age of development of newer modalities for early diagnosis of acyanotic CHD. The closest differential diagnosis of VSDis mitral regurgitation, while that of AS is mitral stenosis.20(43%) of cases of VSD were of perimembranous type. 12(27%) cases were of doubly committed VSD. Though rare world wide, it is seen more frequently in Asian population.21 patients with VSD had normal ECG. 24 (53%) had left axis deviation, while 22 (48%) patients had Left ventricular hypertrophy, ECG is a useful mirror of the physiologic derangements but not of the anatomic location of isolated VSD. 10 cases of PDA had normal ECG. RBBB, the hallmark of ASD was seen in 16(83.7%) cases. Right ventricular hypertrophy was the most consistent features in ECG in PS. Precordial bulge, which indicates structural heart diseases in early life, was seen in only 11 cases. Pulse was high volume in majority of cases of VSD and PDA. JVP was raised in 5 patients with CCF. Most of patients had pallor. Peripheral signs in general were not diagnostic. Apex beat in VSD and PDA was forceful ill sustained outside mid clavicular line. Apex beat in cases of AS, HOCM and coarctation of aorta was forceful, well sustained type. Patients with ASD and pulmonary stenosis had Right ventricular type of apex. Continuous machinery murmur was the diagnostic clinical feature of PDA. All patients with pulmonary hypertension had P2 Wide, split second heard sound was the hall mark of ASD. 36 patients (36%) had normal X-ray chest. Increased lung perfusion was seen in 52 (52%) cases. All 5 cases of PS had pulmonary oligemia. Biventricular cardiomegaly was seen in 10(10%) cases. X-ray was almost diagnostic in one case of coarctation of aorta with bilateral rib notching at inferior margins. Complications that were seen include pneumonia, CCF and suspected infective endocarditis. There were no deaths reported. Zero mortality in one case of coarctation of aorta with bilateral rib notching at inferior margins.

Table No.1: Distribution of Acyanotic Congenital Heart Diseases

<table>
<thead>
<tr>
<th>Heart Disease</th>
<th>No. of Cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD</td>
<td>46</td>
<td>46%</td>
</tr>
<tr>
<td>PDA</td>
<td>24</td>
<td>24%</td>
</tr>
<tr>
<td>ASD</td>
<td>19</td>
<td>19%</td>
</tr>
<tr>
<td>AS / AS+AR</td>
<td>2</td>
<td>2%</td>
</tr>
<tr>
<td>PS</td>
<td>5</td>
<td>2%</td>
</tr>
<tr>
<td>HOCM</td>
<td>2</td>
<td>2%</td>
</tr>
<tr>
<td>CoA</td>
<td>2</td>
<td>2%</td>
</tr>
<tr>
<td>Total</td>
<td>100</td>
<td>100%</td>
</tr>
</tbody>
</table>

Table No.2: Distribution According to Age and Sex.

<table>
<thead>
<tr>
<th>Cardiac Lesion</th>
<th>&lt;1mon</th>
<th>1m-1yr</th>
<th>1-3yrs</th>
<th>4-6yrs</th>
<th>7-12yrs</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>M</td>
<td>F</td>
<td>M</td>
<td>F</td>
<td>M</td>
<td>F</td>
<td>M</td>
</tr>
<tr>
<td>VSD</td>
<td>3</td>
<td>0</td>
<td>7</td>
<td>2</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>PDA</td>
<td>0</td>
<td>4</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>6</td>
</tr>
<tr>
<td>ASD</td>
<td>1</td>
<td>1</td>
<td>3</td>
<td>3</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>PS</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>AS</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>HOCM</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>CoA</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>4</td>
<td>5</td>
<td>13</td>
<td>8</td>
<td>13</td>
<td>10</td>
</tr>
</tbody>
</table>

Table No.3: Height & Weight In Percentile For Their Age & Sex Of Cases

<table>
<thead>
<tr>
<th>Cardiac Lesion</th>
<th>Height (in Centile)</th>
<th>Weight (in Centile)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>&lt;10&lt;sup&gt;th&lt;/sup&gt;</td>
<td>&gt;10&lt;sup&gt;th&lt;/sup&gt;</td>
</tr>
<tr>
<td></td>
<td>&lt;10&lt;sup&gt;th&lt;/sup&gt;</td>
<td>&gt;10&lt;sup&gt;th&lt;/sup&gt;</td>
</tr>
<tr>
<td>VSD</td>
<td>9</td>
<td>37</td>
</tr>
<tr>
<td>PDA</td>
<td>8</td>
<td>16</td>
</tr>
<tr>
<td>ASD</td>
<td>6</td>
<td>13</td>
</tr>
<tr>
<td>PS</td>
<td>1</td>
<td>4</td>
</tr>
</tbody>
</table>
Clinical Profile Of Patients With Aycanotic Congenital Heart Disease In Pediatric Age Group In India

<table>
<thead>
<tr>
<th>HOCM</th>
<th>2</th>
<th>2</th>
</tr>
</thead>
<tbody>
<tr>
<td>AS</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>CoA</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>25</td>
<td>75</td>
</tr>
</tbody>
</table>

VSD= Ventricular Septal Defect; PDA= Patent Ductus Arteriosus; ASD= Atrial Septal Defect; AS= Aortic Stenosis; Coa= Coarctation Of Aorta; PS= Pulmonary Stenosis; HOCM= Hypertropic Obstructive Cardiomyopathy.

While only 25% of patients had height less than 10 percentile, 68% of patients had weight of less than 10 percentile. 9 out of 46 (19%) patients with VSD had height of less than 10 percentile. 34 out of 46 (73%) patients with VSD had weight of less than 10 percentile. Number of patients with PDA with height and weight of less than 10 percentile were 8 (33%) and 17 (70%) respectively. Number of patients with ASD whose height and weight were less than 10 percentile were 6 (31.4%) and 12 (63%) respectively.

IV. Conclusions

CHD constitutes up to 25% of all congenital malformations in the neonatal period. Rheumatic fever and RHD have downward trend in prevalence, so congenital heart diseases are likely to become important contributors to morbidity and mortality among pediatric population. With increasing use of 2D-Echo, diagnostic rate has been enhanced leading to increasing prevalence of CHD. Pediatric cardiac care in India is still in infancy. Government is doing their best through programmes like NRHM (National Rural Health Mission). It is up to us to increase the awareness of the problem amongst the pediatricians through CME, seminars, symposia etc. Training programmes exclusively dedicated to pediatric cardiology and pediatric cardiac surgery need to be established in centres with good standards of pediatric cardiac care.

References

[26]. Rudolph A.M.: The changes in circulation at birth, their importance in C.H.D.
Clinical Profile Of Patients With Acyanotic Congenital Heart Disease In Pediatric Age Group In…