

Incidence of Heart Disease in Children at the National Institute of Cardiovascular Diseases

Pages with reference to book, From 300 To 305

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Abstract

Of 2824 children seen at the National Institute of Cardiovascular Diseases (N.I.C.V.D.), 2020 (71.6%) had congenital heart diseases. Incidence of symptomatic and cyanotic congenital cardiac lesions was higher than expected and asymptomatic, acyanotic lesions like semilunar valve stenosis, small V.S.D. and coarctation of aorta was less than expected and is mainly due to referral pattern of community physicians who are biased toward referral of symptomatic patients (JPMA 34: 300, 1984).

Introduction

There is paucity of reliable data concerning the prevalence of heart disease amongst children in Pakistan. A few reports of selected school children from Karachi¹ and Islamabad² show a prevalence rate of 1.8 -- 1.5 per 1,000 children for heart disease. The incidence of congenital and Rheumatic heart disease was in equal proportion in both studies. These studies are too selective as far as geographic, ethnic and socioeconomic factors are concerned. Another study by Ilyas et al. is better representative of geographic and socioeconomic variables which affect the prevalence data.³ This study was mainly concerned with Rheumatic heart disease and shows a prevalence rate of 7 and 11/1000 in urban and rural areas respectively. Unfortunately this study does not mention the prevalence rate of congenital heart disease nor is the information available regarding ages, socioeconomic factors and details of housing condition.

Study models comprising School children as indicator of the prevalence of heart disease amongst children in Pakistan is probably inappropriate since majority of children in lower socio-economic groups do not attend school. Further more school drop out rate is high since literacy rate for Pakistan is at most twenty five percent.

In our present Medical set up careful collection of hospital based data can serve a very useful role. Hospital based studies by Rahimtoola et al. show that 5.6 percent of Medical admissions at children's hospital JPMC were due to heart disease.^{4,5} The incidence of Rheumatic heart diseases (RHD) was significantly higher (3.5 percent) compared to congenital heart disease (2.5 percent).^{4,5} This data showing Rheumatic carditis as the main form of heart disease amongst children admitted to the hospital is consistent with experience in the third World^{6,7,8} and is also in keeping with the observations made in the developed countries during earlier period of their socio-economic progress.^{9,10}

We present our data of new patients attending the pediatric outpatient clinic at the N.I.C.V.D. With a view to determine the spectrum of heart disease in children and to determine the incidence of individual lesions

The pediatric Cardiology service at the N.I.C.V.D. was established in October, 1980. Only children with suspected heart disease are referred to the department of pediatric Cardiology.

Material and Method

The period of study was from January, 1980 to October, 1983. All new patients were registered and

given a registration card. The patients brought this card on subsequent visits so that the data files could be retrieved. A clinic sheet was made at each visit and filed along with reports of tests such as E.t.G., X-Ray chest and Echocardiographic examination. The initial examination included height and weight measurements and clinical examination of the cardiovascular system including blood pressure measurements. A 12 lead electrocardiogram and X-Ray chest were taken in each patient. The patients were then selected for specialised non invasive test such as echocardiography and invasive test such as cardiac catheterization. Patients seen at the Pediatric Cardiology clinics were 12 years of age or less. The patients were largely referred by physicians, clinical cardiologists and Pediatricians, hospital based or in general practice, predominantly from Sind and Frontier Provinces. Some patients were referred from Baluchistan (Quetta) and Punjab. In 949 patients (46% of the total number), the clinical diagnosis was confirmed by cardiac Catheterization and angiography. The clinical diagnostic categories were modified in accordance with the Echocardiographic or Cardiac Catheterization diagnosis.

Results

Two thousand eight hundred and twenty four new patients age 12 years and less were seen. Congenital Heart Disease was present in 2020 comprising 72 percent of patients. Acute or recurrent Rheumatic Heart Disease was present in 265 (9.4%) and no significant heart disease including functional murmurs was present in 472 (17%). Non congenital or systemic diseases affecting the cardiovascular system was seen in 67 (2.4%) included in the miscellaneous group (Table I).

Table I

**New Patients seen in Pediatric Cardiac Clinic
(1980 – 1983).**

	No.	(Incidence %)
Congenital Heart Disease	2020	71.6
Rheumatic Heart Disease	265	9.4
No Significant Heart Disease	472	16.7
Miscellaneous	67	2.4
	2824	100

Rheumatic fever without significant carditis was noted in a small number 15/265 (6%) of patients. Acute Rheumatic fever with mitral insufficiency alone was noted in 24/265 (9%) The total number of patients with Acute Rheumatic fever was 39/265 (15%). Mitral insufficiency was the most common lesion in patients; Rheumatic fever with acute and recurrent or chronic rheumatic carditis occurring in 130/265 (50%) of patients. Mitral Valve Stenosis was diagnosed and confirmed by echocardiography in 15/265 (9.6%). When one includes mitral stenosis in combination with mitral insufficiency the

incidence of Mitral Valve Stenosis increased to 56/265 (13.6%). The least common lesion seen was isolated; Aortic Insufficiency occurring in 7/265 patients (2.7%) (Table II).

Table II
RHD; Pediatric Cardiology Clinic, New Patients
(1980–1983).

Lesion	No.	%Incidence
Mitral Insufficiency (M.I.)	130	50.0
M.I. + M.S.	31	11.9
Mitral Stenosis (M.S.)	25	9.6
ARF With M.I.	24	9.2
M.I. + M.S.	23	8.8
ARF	15	5.8
Aortic Insufficiency (A.I.)	7	2.7
M.S., A.I., M.I.	5	1.9
M.S., A.I.	5	1.9
	265	100

Abbreviations

R.H.D. = Rheumatic Heart Disease; M.I. = Mitral Insufficiency; M.S. = Mitral Stenosis; ARF = Acute Rheumatic Fever; A.I. = Aortic Insufficiency.

Congenital Heart Disease was present in 2020 patients. The most common lesion was ventricular septal

defect occurring in 805/2020 (40%) of patients. The next common lesion was tetralogy of fallot (490/2020) occurring in 24% of patients. Malformation such as atrial septal defect, pulmonic valve stenosis, patent ductus arteriosus, occurred in approximately equal frequency (7.2-8%). Aortic Valve Stenosis was present in 76 cases, an incidence of 3.8%. The incidence of transposition of the great arteries was 3.1% (63/2020)(Table III).

Table III

**Congenital Heart Disease.
New Patients seen at Pediatric Out Patients
(1980 – 1983).**

	No.	C.H.D.	Incidence %
V.S.D.	805		39.0
T.O.F.	490		24.3
A.S.D.	166		8.2
P.V.S.	163		8.1
P.D.A.	146		7.2
A.V.S.	76		3.8
T.G.A.	63		3.1
Miscellaneous	111		5.5
	2020		100

Abbreviations

V.S.D. = Ventricular Septal Defect; T.O.F. = Tetralogy of Fallot; A.S.D. = Atrial Septal Defect; P.V.S. = Pulmonary Valve Stenosis; P.D.A. = Patent Ductus Arteriosus; A.V.S. = Aortic Valve Stenosis; T.G.A. = Transpositions of Great Arteries.

Miscellaneous group included III Congenital Heart Disease patients with complex lesion. The overall incidence of cyanotic lesions in the miscellaneous group was 55% (66/111) and cyanotic lesions was 84% (45/111). Cyanotic lesions were more common, 66/111 patients. Dextrocardia with V.S.D. and P.S., single ventricle and tricuspid atresia dominate this group of patients. Total anomalous pulmonary venous return Pseudotruncus, Eisenmenger Syndrome, double outlet right ventricle and Truncus arteriosus communis had an incidence between 0.39 to 0.05% amongst. The total congenital heart disease patients (Table IV).

Table IV
Miscellaneous C.H.D. Group, New Patients Pediatric
Cardiac Clinic (1980 – 1983).

Cyanotic lesions	No.	Incidence % C.H.D.
Dextrocardi With P.S. + V.S.D.	18	0.89
Single Ventricle + P.S. + V.S.D. (Univentricular Heart)	14	0.69
Tricuspid Atresia	13	0.64
TAPVR	8	0.39
Pseudotruncus	6	0.30
Eisenmenger Syndrome	3	0.15
DORV	3	0.15
Truncus Type I	1	0.05
	66	

Abbreviations

C.H.D. = Congenital Heart Disease; P.S. = Pulmonic Stenosis; V.S.D. = Ventricular Septal Defect; TAPVR= Total anomalous pulmonary Venous return; DORV = Double outlet right ventricle.

Acyanotic lesions were 45 of 111 patients and A.V. Canal defect was the most common lesion followed by coarctation of aorta (Table V).

Table V
New Patients Pediatric Cardiac Clinic Miscellaneous Group
(1980 – 1983).

Acyanotic Lesions	No.	Incidence % C.H.D.
A.V. Canal	13	0.64
CoAo	10	0.50
Ebsteins Anomaly	6	0.30
E.F.E.	6	0.30
Others	10	0.89
	45	

Abbreviations

A.V. Canal = Atrio Ventricular Canal Defect.

CoAo = Coarctation of Aorta

E.F.E. = Endocardial Fibroelastosis

Arteriosus and Aortic Valve Stenosis in descending order of frequency (Table VI. Cyanotic lesions were present in 619/2020 (31%) Congenital Heart patients.

Table VI
Congenital Heart Disease (Acyanotic).

	No.	Percentage
V.S.D.	805	57.5
A.S.D.	166	11.8
P.V.S.	163	11.6
P.D.A.	146	10.4
A.V.S.	76	5.4
Others	45	3.2
+		
	1401	100

Abbreviations

V.S.D.	=	Ventricular Septal Defect
A.S.D.	=	Atrial Septal Defect
P.V.S.	=	Pulmonary Valve Stenosis
P.D.A.	=	Patent Ductus Arteriosus
A.V.S.	=	Aortic Valve Stenosis

Tetralogy of Fallot was the single most common cyanotic lesions followed by Transposition of the great arteries (Table VII).

Table VII
Congenital Heart Disease (Cyanotic).

	No.	Percentage
T.O.F.	490	79.2
T.G.A.	63	10.2
Others	66	10.7
	619	100

Abbreviations

T.O.F. = Tetralogy of Fallot

T.G.A. = Transpositions of great arteries

Four hundred and seventy two patients (16.7%) had functional murmur. The diagnosis of these was made on clinical grounds suggested by a musical murmurs of grade 3/6 or less in intensity best heard at left sternal border and base of the heart. The murmur varied with respiration and posture. Echocardiogram and Chest X-Rays were within normal limits in these patients (Table I). Sixty seven patients had heart disease which was classified as non congenital or as a consequence to a generalised systemic disease. Prolapse Mitral Valve which is said to be very common was diagnosed in only Eleven patients (Table VIII).

Table VIII
Non Congenital Heart Disease Miscellaneous Group.

	No.
Myocarditis	17
Prolapse Mitral Valve	11
Dysrhythmia	11
Cardiomyopathies	7
Systemic Hypertension	6
E.F.E.	6
Anaemia	6
Pericardial Effusion (Tuberculous)	2
Pulmonary Arterial Hypertension (Idiopathic)	1
	67

Abbreviations

E.F.E. = Endocardial Fibroelastosis

Myocarditis comprised the largest number of patients in this group. Excluded from myocarditis group were the patients with classical picture of Endocardial fibro elastosis and hypertrophic cardiomyopathy. All patients had echocardiography for final diagnosis (Table VIII). Systemic hypertension due to various renal lesions was seen in six patients. One of those had renal artery stenosis which was surgically repaired with B.P. returning to normal after surgical repair.

Discussion

The frequency distribution of cardiac lesions in children presenting at the pediatric Cardiac Clinic at National Institute of Cardiovascular Diseases, Karachi, is presented. This was hospital based data and does not reflect the prevalence in general population or true incidence of heart disease in children in general or pediatric hospital. Epidemiologic studies reflecting the incidence of heart disease in children in Pakistani population are not available. The few reports which are available suffer from bias of design because the data base was limited to selected school or communities and did not present all the socioeconomic or ethnic groups from various regions of Pakistan.^{1,3}

Analysis of our data shows that the congenital heart disease was the most common heart disease observed and that symptomatic lesions dominate the series. For instance the incidence of tetralogy of Fallot (T.O.F.) was much higher than reported. The incidence of T.O.F. in series from Boston, U.S.A. and Toronto Canada was between 6-10% compared to 24% in our series (Table IX).

Table IX
Congenital Heart Disease At N.I.C.V.D. Pediatric Out Patient (1980 – 1983).

	Nicvd %	Cmhc* %	HSC** %	M. Abbot*** %
V.S.D.	39.9	11.4	20.0	6.2
T.O.F.	24.3	6.0	10.2	11.5
A.S.D.	8.2	4.7	5.4	8.3
P.V.S.	8.1	7.5	8.5	3.5
P.D.A.	7.2	15.5	12.1	10.5
A.V.S.	3.8	5.5	12.1	2.3
T.G.A.	3.1	4.7	5.4	4.5
Miscellaneous				
Dextrocardia	0.9	1.42	1.9	2.9
Tric. Atresia	0.6	1.0	1.9	
Single Ventricle	0.69	0.47	1.1	
A.V. Canal	0.69	2.7		
CoAo	0.49	8.1	5.6	8.5
	2020	6647*	10624**	1,000***

*Chmc = Children's Hospital Medical Centre, Boston

**Hsc = Hospital for sick children, Toronto

***MA = Maud Abbotts 1,000 autopsied cases of congenital heart disease.

The patients with Tetralogy of Fallot generally had severe limitation of activities and severe cyanosis and a good proportion had cyanotic spells. The incidence of ventricular septal defect was also much higher than reported although ventricular septal is an acyanotic lesions. However, in 10-15 percent of infants it can produce congestive cardiac failure, failure of growth and repeated respiratory infections. In this study large ventricular septal defect with evidence of congestive cardiac failure was commonly seen in older children. This spectrum of symptomatic large defects in older children is uncommon in reported experience. Small or moderately small V.S.D. does not produce symptoms. It seems likely that asymptomatic patients with ventricular septal defect are either not diagnosed or are managed by the physicians without referral to the N.I.C.V.D.

Patients are referred from up-country and symptoms usually prompt these referrals, therefore asymptomatic lesions are less evidence. Coarctation of aorta is a condition which can be easily detected by palpation of peripheral pulses. In our study the incidence was 0.49% compared to 6-8% in reported experience.¹¹ This suggests that since coarctation of aorta, which does not usually produce symptoms in older children, is not being detected by the referring physicians. A recent study of Co Ao in adults illustrate this point.¹² Patients with Coarctation of aorta included in this series were symptomatic due either to congestive cardiac failure, bacterial Endocarditis or associated lesions. It is reported that Co Ao and Aortic Valve Stenosis rarely occur in the third world¹³. Our data shows that it is certainly not true for Aortic Stenosis, Valvular, subvalvular or supra valvular types.

We have compared our data with large series from Boston U.S.A. and Toronto, Canada which are amongst the few large centres in North America. These data also suffer from referral patterns of those communities. It may be noted that detection of asymptomatic murmur or coarctation of aorta is much more likely in these communities since babies and children are examined at regular intervals throughout life.

The hospital based data is generally not adequate to assess incidence or prevalence in a community. However referral pattern can be studied by comparing data. The spectrum of lesions in our series is similar to the Boston and Toronto series but, symptomatic lesions dominate.

This suggests that asymptomatic lesions are not referred or are not being detected by the general physician. Our data can be used for the evaluation of priorities as to which lesions need greater concentration for hospital management. The two main lesions in our data were a large or moderately large symptomatic ventricular septal defect and tetralogy of fallot. Consequently much of our efforts have been devoted to these lesions. Transposition of the great vessels was the third common lesion and we are also devoting attention to its management. Our surgical team is getting introduced to newer lesions in order of priorities which have been assessed from this data.

Rheumatic fever is a major problem causing heart disease in Pakistan. NO definitive or unbiased data is available as to its prevalence in Pakistan. Our own data shows that severe carditis causing mitral valve regurgitation is common in those referred to N.I.C.V.D. The incidence of Acute Rheumatic Fever was small and it is because we are not the primary hospital where patients with joint pains and fever first come to. The data is much too selective and referral based to suggest the true incidence of Rheumatic fever. Most patients we see are severely symptomatic with moderate to gross cardiac enlargement in whom referring physician has tried the usual decongestant and digoxin therapy.

One interesting feature in our data is the 9.6% incidence of pure Mitral Stenosis in under 12 years age group. The occurrence of Mitral stenosis in children is peculiar to the developing countries. How frequent it really is has not been clarified; our data and reports from Pakistan and India suggest that its incidence may be significant in pediatric patients in this subcontinent^{3,8}.

Occurrence of mitral stenosis in children may be due to failure of secondary prophylaxis with penicillin so that recurrent episodes of Rheumatic fever cause fibrosis of mitral valve at a much younger age than reported in developing countries. It may be that these children have had recurrent episodes of rheumatic

fever without a diagnosis of rheumatic heart disease ever been made.

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