Congenital Heart Disease in Children Visiting JPMC.

By
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Introduction

Interest in congenital heart disease has increased in recent year due to advent of better methods of diagnoses (such as cardiac catheterisation and angiocardiograms), and the spectacular advances in surgical treatment for correction of cardiac defects.

In the past Rheumatic heart disease was believed to be the most common form of cardiac disease in children. Recently, however, it has become obvious that in the West congenital heart disease is the more common of the two. Indeed Keith(3) cites a 20:1 ratio for congenital to rheumatic heart disease.

The first account of congenital heart disease was in 1672 when a case of cyanotic heart disease resembling tetralogy of Fallot was reported by Nils Stensen. A hundred years later, in 1777, Sandifort wrote an account of a child who suffered from the same condition.

In 1783 Hunter described a case who had attacks of paroxysmal dyspnoea which occurs in cyanotic heart disease.

In 1866 Peacock published his book on congenital heart disease.

In 1888 Etienne Fallot reported a series of cases which clarified the syndrome bearing his name.

The brilliant work of Helen Taussig from 1930 onwards led to the first heart operation—the Blalock—Taussig shunt in 1944. Later it was followed by other heart operations.

The major therapeutic problems in pediatric cardiology are confined to early infancy, since serious cardiac malformations and mortality are comparatively high in the first year of life.

According to Taussig, a large number of malformations can be delineated by careful analysis of history, physical examination, X-ray studies and electrocardiogram. In some cases additional studies are required, such as catheterisation and angiocardiograms.

There is scarce information on the incidence of congenital heart diseae in children alone especially in Pakistan. as most series published include adults and children. Autopsy series show more serious lesions while clinical diagnoses favour others, and differ according to the experience and clinical acumen of the physician who examines the case.

Material and Methods

190 cases of congenital heart disease were studied in Unit-I of the Children's Hospital, Jinnah Postgraduate Medical Centre, from July 1968 to June, 1974, a six year period. This is purely a clinical study and no autopsies were performed.

A detailed history was taken on admission, including prenatal history, such as intake of drugs by the mother, infections, toxaemia of pregnancy, hydramnios and other conditions of etiological significance.

Clinical examination of the case was done by the same paediatricians and all complicated cases were also seen by a cardiologist.

Standard 12 Lead Electro-cardiograms and chest X-Rays in three positions (P.A., Left lateral and right anterior oblique) were done on admission in all cases except those seriously ill. They were repeated in complicated cases as and when necessary.

Age, presenting symptoms, clinical signs, investigations and follow-up in the ward were recorded in all cases.

Five cases had cardiac catheterization; three with cyanotic heart disease and two of the potentially cyanotic group.

In the younger age group (Under one year) cyanosis may occur in other conditions such as pulmonary disorders and intracranial lesions; therefore only those cases with obvious clinical signs of heart disease were included in this study.

Results

During the period of this study the total admission in the ward were 7480, and cases of congenital heart disease were 190, i.e. of the 2.5% admissions (Table I); of these approximately 69% were acyanotic and potentially cyanotic, and 31% were of the cyanotic group. There was a preponderance of males over famales-109 males (57.3%) to 81 females (42.7%).

Table I Congenital Heart Disease. Paediatrics Department, Unit-1 JPMC. Karachi.

July, 1964 to June, 1971.	
Total Admission.	7480
Total Number of Congenital Heart Disease Cases.	190—2.5%
Acyanotic and Potentially Cyanotic	Cyanotic.
131 (63.94%)	59 (31.06%)

The incidence of consanguinous (first cousin) marriages was very high i.e. 71.2% compared to about 45% in the general population. A positive correlation has been reported between advanced maternal age and incidence of congenital heart disease, but this was not found in our study. A higher incidence has also been reported in premature infants, but this could not be confirmed as most mothers did not know the birth weights of these children.

In the acyanotic and potentially cyanotic group the three most common defects seen were ventricular septal defects 39.46%, patient ductus arterious-12.1% and atrial septal defect-7.9% (Table II).

Amongst the 59 cases of cyanotic congenital heart disease (Table III), 14.73% cases of Fallot's tetralogy were seen; 7.62% cases were undiagnosed, the majority (11 cases) of these having expired within 48 hours of admission.

Table II Congenital Heart Disease. Acyanotic and Potentially Cyanotic.
Department of Paediatrics, JPMC, Karachi.

Pulmonary Stenosis	1	0.52%
Coarctation	1	0.52%
Fibroelastosis	2	1.05%
VSD	75	39.46%
PDA	23	12.10%
ASD	16	7.90%
Ostium Primum Defect	5	2.63%
Anom. Pulm. Venous Return.	2	1.05%
Eisenmenger's Syndrome	4	2.66%
Undiagnosed	2	1.05%
Total:	131	68.94%

Almost 56% cases were under one year and 72% under two years of age. The incidence declines with increasing age as seen in Table IV.

There were six cases of Down's syndrome; two had ostium primum defect and two had VSD. One case-each of transposition of the great vessels and Fallot's tetraolgy were also noted in association with Down's syndrome though, Berg and his associates(1) have reported that transposition of the great vessels is not found in mongolism and that tetralogy of Fallot is seen less frequently than in controls.

Table V shows that over half the cases with VSD came with respiratory infection and 15 with dyspnoea due to cardiac failure. Cases of ASD and ostium primum defect presented with failure to thrive and respiratory infection. Of the 28 cases of Fallot's tetralogy 17 were brought for cyanosis and five with growth retardation; one case had associated hemiplegia.

Clinically (Table VI) the majority of cases presented with cardiomegaly, murmurs and signs of cardiac failure. Only three cases of Fallot's tetrology had cardiomegaly and cardiac failure. However, in the cyanotic group of the 31 cases other than Fallot's tetralogy, 16 came with CCF.

Roentgenologic findings (Table VII) were consistent with the diagnoses made, as were the findings on electrocardiography (Table VIII). One case of dextrocardia was seen amongst the 28 cases of Fallot's tetralogy. One case of VSD had complete heart block. Paroxysmal atrial tachycardia was found in one case with an ostium primum defect, and auricular fibrillation was seen in one case of Ebstein's anomaly.

Table III Congenital Heart Disease. Cyanotic.
Department of Paediatrics, JPMC, Karachi.

Fallot's Tetralogy	28	14.73%
Transposition G.V.	4	2.10%
Tricuspid Atresia	5	2.63%
Truncus Arteriosus	2	1.05%
Single Ventricle	3	1.88%
Ebstein's Anomaly	2	1.05%
Undiagnosed	15	7.62%
Total:	59	31.06%

Table IV Congenital Heart Disease-Age Incidence. Deptt. of Paediatrics, JPMC, Karachi.

Diagnosis	0-6 *Mnth.*	6 Mnth 1 *Year*	1-2 *Years*	2-4 *Years*	4-6 *Years*	8-12 *Years*	*Total*
Coarctation	—	—	—	—	1	—	1
P.S.	—	—	—	—	1	—	1
V.S.D.	28	19	12	7(2M)	5	4	75
P.D.A.	6	5	4	4	3	1	23
A.S.D.	2	5	4	4	1	—	16
Ostium Primum	1(M)	1	2(1M)	1	—	—	5
Fibroelastosis	1	—	1	—	—	—	2
Anom. P.V.R.	—	—	—	1	1	—	2
Eisenmenger's	—	—	—	2	2	—	4
Fallot's Tetralogy	5	7	3	6(1M)	6	1	28
Transposition G.V.	2	1(M)	1	—	—	—	4
Tricuspid Atresia	3	1	1	—	—	—	5
Single ventricle	2	1	—	—	—	—	3
Truncus Arteriosus	—	2	—	—	—	—	2
Ebstein's Anomaly	—	—	—	1	1	—	2
Undiagnosed	10	5	2	—	—	—	17
Total:	60	47	30	26	21	6	190
Percentage:	(31.50)	(24.73)	(15.78)	(13.68)	(11.05)	(3.71)	(100.0)

Table V Congenital Heart Disease: Symptoms. Deptt. of Paediatrics, JPMC, Karachi.

Diagnoses	*Cyanosis*	*Failure to Thrive*	*Dyspnoea*	*Murmur*	*Res. Inf.*	*Retarded Growth*	*Total*
Coarctation	—	—	—	1	—	—	1
P.S.	—	—	—	1	—	—	1
V.S.D.	—	8	15	5	45	2	75
P.D.A.	—	—	4	2	17	—	23
A.S.D.	—	9	—	1	5	1	16
Ostium Primum	—	4	—	—	3	—	5
Fibroelastosis	—	—	2	—	—	—	2
Anom. P.V.R.	—	1	1	—	—	—	2
Eisenmenger's	—	—	2	—	2	—	4
Fallot's Tetralogy	17	2	—	4	—	5 (HEMIPLEGIA)	28
Transposition G.V.	2	1	1	—	—	—	4
Tricuspid Atresia	3	—	1	—	1	—	5
Single Ventricle	1	1	1	—	—	—	3
Truncus Arteriosus	—	1	1	—	—	—	2
Ebstein's Anomaly	—	—	5	—	—	3	2
Undiagnosed	8	2	5	—	2	—	17
Total:	31	29	34	14	73	9	190

Table VI Congenital Heart Disease-Clinical Signs: Deptt. of Paediatrics, JPMC, Karachi.

Diagnoses	*Clubbing Fingers*	*Enlarged Heart*	*MURMUR Sys*	*Dias*	*CCF*	*Loud P2*	*Total*
Coarctation	—	—	1	—	—	—	1
P.S.	—	1	1	—	—	—	1
V.S.D.	—	45	75	8	25	26	75
P.D.A.	—	14	23	23	14	15	23
A.S.D.	—	6	16	3	—	13	16
Ostium Primum	—	3	5	2	3	3	5
Fibroelastosis	—	2	—	—	1	—	2
Anom. P.V.R.	—	2	2	—	1	2	2
Eisenmenger's	—	4	4	—	2	4	4
Fallot's Tetralogy	20	3	28	—	3	—	28
Transposition G.V.	2	4	2	—	2	2	4
Tricuspid Atresia	2	3	4	—	1	—	5
Single Ventricle	1	1	3	—	1	1	3
Truncus Arteriosus	1	1	1	—	1	—	2
Ebstein's Anomaly	—	2	2	—	1	—	2
Undiagnosed	—	10	12	2	10	5	17
Total:	26	101	170	38	65	71	190

Table VII Congenital Heart Disease. X-Ray Chest. Deptt. of Paediatrics, JPMC, Karachi.

Diagnoses	*Enlarged Heart*	*Absence of Pulm. Conus*	*Pulmonary Plethora*	*Pulmonary Oligaemia*	*Other Changes*	*Total*
Coarctation	1	—	—	—	(1) Rib Notching	1
Pulmonary Stenosis	1	—	—	—	—	1
V.S.D.	35	—	15	—	—	75
P.D.A.	16	—	14	—	(10) Pulmonary Conus+	23
A.S.D.	7	—	7	—	(4) Pulmonary Conus+	16
Ostium Primum	4	—	4	—	(3) Pulmonary Conus+	5
Fibroelastosis	2	—	—	—	—	2
Anom. P.V.R.	2	—	2	—	(2) R.A.+(Fig. of Eight)	2
Eisenmenger's	4	—	4	—	(4) Pulmonary Conus+	4
Fallot's Tetralogy	8	28	—	28	(1) Dextrocardia	28
Transposition G.V.	4	—	3	1	(3) Narrow Pedicle	4
Tricuspid Atresia	5	5	—	5	—	5
Single Ventricle	2	—	1	2	(2) Globular Heart	3
Truncus Arteriosus	2	2	—	2	(2) Broad Pedicle	2
Ebstein's Anomaly	2	—	—	—	(2) R.A.+	1
Undiagnosed	10	3	6	6	(2) P.C.+	17
Total:	103	38	56	20		190

Of the total number of cases studied, 119 improved (62%) and 71 cases (38.5%) were referred to the Heart Clinic for further investigations and possible surgical treatment (Table IX). These were chiefly cases of cyanotic heart disease and PDA and VSD, who had come with complications. Of these, 27 died, chiefly due to cardiac failure and respiratory infections. Of the 17 undiagnosed cases also, 11 expired. This gives a total mortality of 20% (38 cases). The cases that left against medical advise (10%) were mostly in a serious condition and this has probably lessened the mortality in this study.

Table VIII Congenital Heart Disease. E.C.G. Changes. Deptt. of Paediatrics, JPMC, Karachi.

Diagnoses	*Normal*	*R.V.H.*	*L.V.H.*	*Biventri Cular*	*Other Changes*	*Total*
Coarctation	—	—	1	—	—	1
P.S.	—	1	—	—	—	1
V.S.D.	29	10	23	12	(1) Heart Block	75
P.D.A.	2	—	16	5	—	23
A.S.D.	5	11	—	—	(8) Incomp. BBB(Rt).	16
Ostium Primum	—	(5) Left Axis & RVH.	—	—	(1) Paroxysmal Tachycardia.	5
Fibroelastosis	—	—	2	—	—	2
Anom. P.V.R.	—	(2) RAH & RVH	—	—	—	2
Eisenmenger's	—	4	—	—	—	4
Fallot's Tetralogy	3	25	—	—	—	28
Transposition G.V.	—	2	—	2	—	4
Tricuspid Atresia	—	—	5	—	—	5
Single Ventricle	—	—	1	—	(2) Bizarre	3
Truncus Arteriosus	—	—	—	2	—	2
Ebstein's Anomaly	—	—	—	—	(2) RAH	2
Undiagnoses	(2)	10	2	3	(1) Aur. Fibrill.	17
Total	39	70	50	24	—	190

Table IX Congenital Heart Disease. Results. Deptt. of Paediatrics, JPMC, Karachi.

Diagnoses	*Improved*	*Poor Results*	**LAMA*	*Expired*	*Total*	*Referred to Heart Clinic*
Coarctation	—	1	—	—	1	1
P.S.	—	1	—	—	1	1
V.S.D.	62	—	5	8	75	34
P.D.A.	19	—	2	2	23	9
A.S.D.	14	—	1	1	16	6
Ostium Primum Defect	4	—	—	1	5	—
Fibroelastosis	—	—	—	2	2	—
Anom. P.V.R.	1	—	1	—	2	1
Eisenmenger's	2	—	—	2	4	2
Fallot's Tetralogy	12	11	2	3	28	16
Transposition G.V.	1	—	1	2	4	1
Tricuspid Atresia	—	1	2	2	5	1
Single Ventricle	—	—	1	2	3	—
Truncus Arteriosus	—	—	1	1	2	—
Ebstein's Anomaly	—	—	1	1	2	—
Undiagnosed	4	—	2	11	17	2
	119 (62.63)	14 (7.36)	19 (10.0)	38 (20.0)	190 (100.0)	74 (38.90)

*Left Against Medical Advice.

Discussion

Exact diagnoses were not possible in all cases due to lack of facilities for detailed and sophisticated investigations. Taussig's(7) descriptions with X-ray and Electrocardiograms were followed in the clinical assessments of the cases. No autopsies were performed.

This study of congenital cardiac disease has shown that VSD is the most common defect encountered, and that amongst the others, Fallot's tetralogy, PDA and ASD make up 8-15% of the total number of patients with congenital cardiac defects.

Table X shows the incidence in our series as compared to studies carried out in the U.S.A., U.K. and Finland. There is also one study from Karachi which includes both children and adults. The incidence of pulmonary stenosis and coarctation of the norta was very low, 0.52% each, in our study. This is in contract to a 15.5% incidence of pulmonary stenosis in Dr. Shafaqat(6) (Heart Clinic) series which is similar to Paul Wood's and Nadas(4). Our incidence of VSD, on the other hand, is higher than in any of the other series. It is possible that cases of VSD living in poor socio-economic conditions get respiratory infections more frequently and are brought to the hospital at an early age. While Pulmonary stenosis do comparatively well in the paediatric age group.

Table X Congenital Heart Disease. Comparative Percentage. Deptt. of Paediatrics, JPMC, Karachi

Diagnosis	*Paul Wood's Series* (748)	*Nadas Series* (3786)	*Keith's Series* (380)	*Abbott's Series* (1000 PM).	*JPMC Karachi Paed* (190)	*JPMC Karachi Heart* Cl. (200)	*Fin-Land* (777-*PM*)
Pulmonary Stenosis	12.00	11.97	7.00	3.50	0.52	15.50	—
Coarctation	9.00	4.99	6.00	8.50	0.52	1.00	9.5
Fibroelastosis	—	—	1.00	—	1.05	—	5.8
V.S.D.	9.30	19.97	22.00	6.20	39.46	21.00	13.3
P.D.A.	15.00	12.31	17.00	10.50	12.10	12.00	6.05
A.S.D.	21.50	10.04	7.00	3.30	7.90	16.00	9.1
Ostium Primum Defect	—	3.94	2.00	—	2.63	—	6.5
Anom. P.V.R.	—	—	2.00	4.00	1.05	—	—
Eisenmenger's	3.00	—	3.00	—	2.10	1.00	—
Fallot's Tetralogy	11.10	14.55	11.00	11.50	14.73	16.50	5.9
Transposition G.V.	1.00	3.96	8.00	4.90	2.10	1.50	13.1
Tricuspid Atresia	1.50	1.24	3.00	1.60	2.63	1.50	2.8
Truncus Arteriosus	—	—	1.00	2.10	1.05	1.00	9.5
Single Ventricle	—	—	2.00	2.70	1.58	—	—
Ebstein's Anomaly	1.00	—	1.00	—	1.05	—	—
Undiagnosed (Miscellaneous)	3.70	11.30	—	37.40	8.94	—	8.4

The incidence of ASD is highest in Paul Woods' series while the incidence of transposition of the great vessels is high in the series from Finland.

The incidence of truncus arteriosus has been reported to be high in Asia but comparison shows shows that our incidence is similar to those in studies from the west. Perhaps more cases can be diagnosed after cardiac catheterisation and angiograms are more freely available.

As no true population studies of the prevalence or incidence of various congenital heart diseases in Pakistan are available, this study attempts to give the breakdown of various congenital heart diseases as seen in one large pediatric center. While such a study cannot accurately define incidence or prevalence as in a true population sampling but will still reflect the true picture to some extent and provide a working picture in the absence of definitive works.

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References

1. Berg, J.M. Crome, L. and France, N.E. Congenital Cardiac Malformations in Mongolism. Brit. Heart J., 22:331-1960.
2. Keith, J.D., Congenital Heart Disease in Parsons, L., Modern Trends in Paediatrics. London Butherworth & Co., 1951.
3. Keith, J.D., Rowe' R.D. and Vlad, P. Heart Disease in Infancy and Childhood. 2nd Edition Newyork. MacMillan Co., 1967.
4. Nadas, A.S., Paediatric Cardiology, 3rd Edition. Philadelphia. W.B. Saunders Co., 1972.
5. Potter, Edith L., Pathology of Fetus and Newborn. Chicago Year Book Publishers. 1952.
6 Shafaqat, S.H. Professor of Cardiology, J.P.M.C., Karachi. Personal Communication.
7. Taussig, Helen B., Congenital Malformations of the Heart, Vol. 1, 2nd Edition, Harvard University Press, 1960.