Congenital Heart Disease in Children Visiting JPMC.

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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 disease is the more common of the two. Indeed Keith(3) cites a 20:1 ratio for congenital methods the disease.

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

In 1783 Hunter described a case who had occurs of paroxysmal dyspnoea which occurs of paroxysmal disease.

In 1866 Peacock published his book on the last disease.

In 1888 Etienne Fallot reported a series of clarified the syndrome bearing his

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 Department, Unit-1 JPMC. Karachi. drugs by the mother, infections, toxaemia of pregnancy, hydramnios and other conditions of ALDAL AUOTMINAL ANALA AU etiological significance.

Clinical examination of the case was done by the same paediatricians and all complicated cases were also seen by a cardiologist. 100 0801

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, tend on the high visites when

Age, presenting symptoms, clinical signs, investigations and follow-up in the ward were recorded in all cases sixyle , vioteth lo sixylana

studies and electrocardiogram. In some cases off Five cases had cardiac catheterization; three with cyanotic heart disease and two of the potentially cyanotic group. There is scarce information on the incidence

and 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.

experience and clinical acumen of the physician

who examines the case.

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 1 Congenital Heart Disease. Paediatrics

July, 1964 to June, 1971. Total Admission.

7480

Total Number of Congenital Heart

and Disease Cases. International 190-2.5%

Acvanotic and Potentially Cyanotic b Cyanotic. tion 62nd angiocardiograms), and th18 bectacular a (% 30.16) surgical treatment fo (% 40.66) n of

In the past Rheumatic heart disease was

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. A hund A nesnet still

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.

I Congenital Heart D)isease.	Acvan	ersian
Potentially Cyanotic.	2-4	2-1	dta

rtment of Paediatrics, JPMC, Karachi.

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	-				
Pulmona	ry Stenos	5 2	(MC	17(2	C10 500501
54	1	3	4	711	210.52%
(Controllati	ion —	1	4.	1	0.52%
	-	-	-	1	20,52°.0°.
Fibroelas	stosis	********	-	2	1.05%
THE PARTY OF THE P		1	-1-	-	1.05/20
WSD	1	2	2	75	39.46%
mr.	1	0	(M))∂	7 3
				23	12.10%(M)
ASD					1 1
				16	7.90%
Defin P	rimum De	fakt	- Ann	-	2
	rimuin De	rect	2	5	2.63%
Amom. Pu	ılm. Venor	is Retur	n	2	1.0504
	0	10	20	4	47.05%
Elisermen	ger's Synd	rome 1)	3.68)	14 (08 24.7 36667 878
Lindiagno	sed	The state of the s		2	%20.1 toms, Deptt.
- LURIS	PINK, Na	trics, J	Pacdia	10	toms. Deptt.
Timal:	Retar				
Total .	Retur	7. 7	13	1	68.94%
	Growth	(11)	2971 3	TERRATA	Дуг риоса М
- 4.1					~

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

There were six cases of Down's syndrome; and ostium primum defect and two had VSD.

case-each of transposition of the great and Fallot's tetraolgy were also noted in with Down's syndrome though, Bergassociates (1) have reported that transformed the great vessels is not found in the great vessels in the great vessels is not found in the great vessels in the great vessels is not found in the great vessels in the great vessels in the great vessels is not found in the great vessels in the grea

Table V shows that over half the cases with came with respiratory infection and 15 with due to cardiac failure. Cases of ASD

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,

	Noniciation
Fallot's Tetralogy	28 14.73%
Transposition G.V.	mu4ing 2010%
Tricuspid Atresia	sizotzalgodd 7 5 y q 2,63%
Truncus Arteriosus	Fisenmenger's % 20,1 Tei2alogy
Single Ventricle 2	V.D 3 in 1.88%
Ebstein's Anomaly	Tricuspid Arresia %20, I Venticle
Undiagnosed	Temous Arteriosus
Total: ₂₉ 18	59 31.06%
	The state of the s

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

0-6 Mnth. ————————————————————————————————————	6 Mnth 1 Year ————————————————————————————————————	Years	2-4 Years 7(2M) 4	4-6 Years	8-12 Years	Total 1 1 75
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6 2		4	7(2M) 4	5 3		
6 2		4	7(2M) 4	5	izono4	
6 2		4	4	3	menting C	
(M)	5					23
(M)		4	4	1		16
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	3	24 05	2	2		4
5	7	3	6(1M)	6	1	28
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3	í	1				5
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0.2	2	Accounts			-	324
a nis mo		à t	7 1	11.7	-	2
10	5	2		34313	o ammi	17
60	47	30	26	21	6	190
.50)	(24.73)	(15.78)	(13.68)	(11.05)	(3.71)	(100.0)
	1 - 5 2 3 2 - 10	5 7 2 1(M) 3 1 2 1 - 2 10 5	1 — 1 - 3 2 1(M) 1 3 1 1 2 1 — 2 - 2 — 10 5 2	(M) 1 2(1M) 1 1 - 1 - 1 - 2 1 - 2 5 7 3 6(1M) 2 1(M) 1 - 3 1 1 - 2 2 1 1 10 5 2 - 1 60 47 30 26	2 5 4 4 1 1	2 5 4 4 1 1 1 1 1 1 1 1 1 1 1 1 1

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

Diagnoses	Cyanosis	Failure to Thrive	Dyspnoea	Murmur	Res. Inf.	Retar ded Growth	Total
Coarctation	t of Paedistr	cpartmen	d	1.	VIEL VIEW CO	ero al de de	Milks
P.S.	·		*331	SOUND POUR	ni age. in	er tivet vedits	Dan 1851
V.S.D.		8	15	oldus ni z	45	incignating	75
P.D.A.	mmain	allocia Ten	4	2	17		23
A.S.D.		9	15/170	nen's kynder	CI 15	via diene e	16
Ostium Primum	_I.D m	ottia 4 anta	T	The street	3	10000000000	
Fibroelastosis		-	2	DAG DAG DE	FE 3 2323D TEI	mura minis	5
Anom. P.V.R.		A biquoin	T 1 1 1 1 2 1	ani to n	0111200211111	le_dose-s	200
Eisenmenger's			211 00	ton mile on	2	ra'i . U s'il br	4
Fallot's Tetralogy	17	2	Bern T	4	orbers s'm	5	28
1000 5					(HEA	MIPLEGIA)
Transposition G.V.	2	1111	2 1	I MAN MOUTE	nave_repu	1) santiacest	2114
Tricuspid Atresia	3	_	1/11 101	is not tour	sloden a	organis lo	Bout on
Single Ventricle	1 6/100	1 1 1	A lasse	ai s -T eff je	vi -li nist	tudi -i ire di	3
Truncus Arteriosus	_	1	1		T	-	2
Ebstein's Anomaly	b	sao ca sibni	5	_	20 01103 21	3	2
Undiagnosed	8	2	5	of the cases	2 2	awada V ul	17
Total:	31	29	34	14	73	91111 01	190

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

Diagnoses	Clubbing	Enlarged	MUI	RMUR		Loud	PD 1
v of 20% (39 care)	Fingers	Heart	Sys	Dias	CCF	P2	Total
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DA most oda be	bably lessen	45	75	8	25	26	75
SD.	-	14	23	23	14	15	23
Primum	-	6	16	3 10	se, 21 died	13	16
elastosis		3	5	2	3	3	5
P.V.R.	ETRIB LA T to	dique 2 .ao	garate , D.	isca co - E.C	al Harth	Compani	HIV 210
enger's	_	2	2	_	1	2	2
Tetralogy	20	4	4	-	2	4	4
position G.V.	20	1	28	lumin o/	3	33	28
Atresia	2	2	2	-	2	2	4
Ventricle	1	1	4	-	1		5
Arteriosus	1	1	- 3		1	1	Hollar 3 to
s Anomaly		2	2		1	_	2
magnosed		10	0112	29	1		2
THE PROPERTY OF	26			2	10	5	17
	26	101	170	38	65	71	190

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

Diagnoses (1)	Enlarged Heart	Absence of Pulm. Conus	Pulmo nary Plet hora	Pulmo- nary Oligaemia	Other Changes	Total
Interctation	1				(1) Rib Notching	
Stenosis	1	-			(1) Rio radicining	dochd
[3D.	35		(2) 15		here to be the mand	1
DA	16	HV	A 814 A	triati Lied en	(10) D1	75
			12 27 X 12 1/3	Cherry Winds	(10) Pulmonary	23
SD.	7	Parkers .	7		Conus+	
			25	-	(4) Pulmonary	16
Primum Primum	4		2 4		Conus+ Column	
	Bull Discou		- 4	die i	(3) Pulmonary	5
incelastosis	2				Conus+	
P.V.R.	2 2	THE WAY	2	-	(a) a designation	2
menger's	4		2	-	(2) R.A.+(Fig. of Eight)	2 2
3	7		4		(4) Pulmonary	4
Tetralogy	8	30			Conus+	
position G.V.	4	28		28	(1) Dextrocardia	28
pid Atresia	5	-	3	1	(3) Narrow Pedicle	4
Ventricle		5	-	5	_	5
Arteriosus	2	-	01 1	(2) 2	(2) Globular Heart	3
's Anomaly	2 2 2	2	_	2	(2) Broad Pedicle	2
agnosed		_		100 <u>100 100 100 100 100 100 100 100 100</u>	(2) R.A.+	1
	10	3	6	6	(2) P.C.+	17
	103	38	56	20		190

Pakistan Heart Journal Dr. Razia J. Rahimtoola, Dr. Ismat Mojid, Dr. Hamid Shafqat Vol. 13 No. 2 and Dr. Asma Fauzia Qureishi

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

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

Table VIII Congenital Heart I		2	2		Anom. P.V.R.
Diagnoses — &	Normal	R.V.H.	L.V.H.	Bîventri Cular	FalatoT Tetra redtO Transpositiot esgnädO Trieuspid Atresa
Coarctation		- 3	1 1	1 -	Single Ventricle-
P.S. ²	-	1 1	1_	1_	Truncus Arteriosus
v.s.p. 5	29	0112	0123	_12	Ebstein's Anomaly Undragnosed (I)
65 71 190 A.G.	88 2	170	101	26	Heart Block
Paediatrics, JPMC, Kardia,A	Deptt. of	Rayl Chest.		l Heart D	
				Inc	comp. BBB(Rt).
Other Changes mumira Totilao	Pulmo-	(5)	Absence of Pulm.	Enlarged Heart	Diagnoses (1)
	d Oligaemia	eft Axis & R	C_{on} HV2	116411	Paroxysmal
(1) Rib Notching sisotsaled				1	Tachycardia.
Anom. P.V.R.		(2)		1	Pulmonary Stenosis
(10) Pulmonary 23		RAH & RV	H _	35	V.S.S. —
Conus+ crammagia	processo	4			_ 4
Fallot's Tetralogy (4) Conus+	3	25	_	7	Cl.2.A
Transposition G.V. Pulmor (3)		A 2		A 2	Osti4m Primum-
Conus+ aisentA biquoriT			5		- , , 5 ,
(2) R.A.+(Fig. of Light) slgniS	_	c	_ 1	2	Fibroelastosis (2)
(4) Pulmonary 4	*****	4	-	4	Bizarre enger errazia
Truncus Arteriosus - zumo	- 0.6	 /		2	$ \frac{2}{1}$
(1) Dextrocardia (3) Narrow PylamonA s'nisted3	87	£ -	87	8 - 4	Fallot's Tetralogy Transposition (9)V.
2	5		5	5	Freuspid AtrHAN
(2) Globular HeartsesongaibnU	2 (2)	1 10	-2	2 3	Sing 1 Ventric (1)
(2) Broad Pedicle 2 (2) R.A.+	2		7	2	Ebstein's Allomary ruk
(2) P C + 17	9 20	9 70	3	0124	Indiagnosed
Total	39	70	50	ε01 ε01	— 190 Fotal:

Heart Journal & Congenital Heart Disease in Chidren Visiting JPMC TWO 13 No. 2 and Dr. Asma Fauxia Qureishi

Table IX Congenital Heart Disease. Results. Deptt. of Paediatrics, JPMC, Karachi. Table X Congenital Heart Disease. Comparative Percentage. Depit. of Paediatric

Diagnoses		I1	npro		Poo Resu		LAN	WA	Expired	Total	Referred to Heart
TPMC -	FPMC	bbot\$'s	A	cith's	Ä.	Vadas		lune	I	n size	Clinic
Karation notation	Kuruchi	Series		Series	1	Series		ood's	W	1	MAGNICE 1
Heart R.	Pacd	0001)		(380)	1	(3786)			2.—	Î	1
Cl. (200) . D.Z.	(190)	PM).		STIS .	_	Kai	5	748)		75	34
ASD.		-	19				2		2	23	9
Orium Primum Dei	Forefile ()	07.6	14	00 -	-		1		1	16	6
Fibroelastosis	ecic.0	3.50	4	7.00		11.97		2.00		Scnosis	Pulmonary
P.V.R.1	0.52	8.50	1	6.00	_	4.99	1	9.00	2	2 2	— (Darctatio
Tabot's Tetralogy Tabsposition G.V.	1.05		12	1.00	11		2		2 3	4	2 I 21 roclast
Trouspid Atresia Ventricle	39.46	6.20		22.00	1	19.97	2	9.30	2 2	5	1 VIS.D.
Anteriosus Anteriosus Anomaly	12.10	10.50		17.00		12.31	1 1	15.00	2	3 2	P.D.A.
Undagnosed. 01	7.90	3.30	4	7.00	_	10.04	2	21.50	11	2 17	.CI.22A
	2.63	(6	119 2.63	2.00	14 7.36		19 10.(0)	38 (20.0)	190 (100.0)	(38.90)
Against Medic	al Advice.	4.00		2.00				77			Anom. P

Scussion . 3.00

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

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

treation							
Diagnosis	Paul Wood's Series	Nadas Series	Keith's Series (380)	Abbots's Series (1000	JPMC Karachi Paed	JPMC Karachi Heart	Fin- Land (777-
75 94 34 24 23 9	(748)	(3786)	(300)	PM).	(190)	Cl. (200)	(111- PM)
Pulmonary Stenosis	12.00	11.97	7.00	3.50	0.52	15.50	A.S.D. O cio na
Coarctation	9.00	4.99	6.00	8.50	0.52	1.00	9.5
Fibroelastosis	8 _ 8		1.00	\$1 -	1.05	Yearning V	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	(0) = (20.0)	3.94	2.00	(62,6	2.63		6.5
Anom. P.V.R.	_	_	2.00	4.00	1.05	gains () ledig	A fl ed 8
Eisenmenger's	3.00	Trble	3.00	_	2.10	1.00	Discus
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	nio) recies A	(He <u>sti</u> Cli Wood's an	1.00	2.10	1.05	1.00	9.5
Single Ventricle	er hamd, is h s. It is pos	die 🚅 no insa todio	2.00	2.70	1.58	is study of c	dT —
Ebstein's Anomaly	1.00	living in respiratory	1.00	immon detec ne others, Fe	1.05	hat VSD is od, and that	shown t
Undiagnosed (Miscellaneous)	3.70	11.30	io di eniral	37.40	8.94	y, P <u>DA</u> and il number	8.4

The incidence of ASD is highest in Paul series while the incidence of transposifrom of the great vessels is high in the series from Firmilan-A

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

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

wledgement

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-: 0: cular Discases (Pakistan), Karachi

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