

A STUDY OF HEART DISEASE IN CHILDREN IN MALTA

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This paper is based on a study of 352 children referred to the Paediatric Cardiology Unit, St. Luke's Hospital, since its inception in January, 1966, until 16th September, 1968.

All children were examined by at least two physicians. The usual clinical diagnostic criteria and methods, including radiology and electrocardiography, were used in assessing the cases. In some the clinical diagnosis has been confirmed at operation or during catheterisation and angiocardiology carried out at St. Mary's Hospital, London.

This Unit receives the vast majority of heart cases in children in Malta and these cases may be taken as a fair representation of heart disease in Maltese children.

The purpose of this study is to compare the incidence of the various cardiac anomalies, congenital or acquired, in a small island like Malta with that of other countries, while a study has also been made of the presenting symptoms and other relevant clinical features.

The total number of children seen and their diagnosis is seen in Table 1. The largest

number of cases (159 or 45.2%) were congenital while the next most frequent condition was Rheumatic heart disease (74 or 21%). The remainder are made up of a miscellaneous group and included 44 children (12.5%) who are being followed up for so-called "innocent systolic murmurs", 35 infants (9.9%) who were found to have thymic hyperplasia and 24 (6.8%) with cardiomegaly, still under investigation.

The frequency of the different congenital anomalies studied at the Unit is shown in Table 2. A very high incidence of ventricular septal defects (44.7%) was found in our series. This agrees with the studies of Perry (1931) and Muir and Brown (1934)

TABLE 2
Percentage distribution of the different congenital anomalies

<i>Type of Defect</i>	<i>No. of Cases</i>	<i>Percentage</i>
Ventricular septal defect	71	44.7
Tetralogy of Fallot	22	13.8
Atrial septal defect	15	9.4
Pulmonary stenosis	12	7.5
Aortic stenosis	11	6.9
Patent ductus arteriosus	5	3.1
Coarctation	1	0.63
V.S.D. + P.S.	6	3.8
V.S.D. + A.S.D.	3	1.9
V.S.D. + P.D.A.	2	0.8
A.S.D. + P.S.	2	0.8
Fallot + P.D.A.	1	0.63
V.S.D. + A.I.	1	0.63
Dextrocardia	1	0.63
Eisenmenger	1	0.63
Persistent Truncus	1	0.63
Single Ventricle	1	0.63
Tricuspid Atresia	1	0.63
R-Sided Aortic Arch	1	0.63
Idiopathic Dilatation of P.A.	1	0.63
Total	159	

TABLE 1

Total number of infants and children referred to the paediatric cardiology unit during period January 1966 - September 1963 and their diagnostic classification

<i>Classification</i>	<i>No. of Cases</i>	<i>Percentage</i>
Congenital heart disease	159	45.2
Rheumatic heart disease	14	21.0
Cardiomegaly	24	6.8
"Innocent murmurs"	44	12.5
Paroxysmal tachycardia	5	1.4
Sinus tachycardia	7	2.0
Thymic hyperplasia	35	9.9
Normal hearts	4	1.1
Total	352	

TABLE 3
Percentage distribution of
different malformations in
infants and children
(Comparison of different series)

<i>Type of defect</i>	<i>Keith Et al. (1958)</i>	<i>Stuckey (1954)</i>	<i>Hay (1963)</i>	<i>Present series (1968)</i>
Ventricular septal defect	25	20	28	44.7
Patent Ductus Arteriosus	17	18	14	3.5
Atrial Septal Defect	7	11	15	9.4
Pulmonary Stenosis	7	8.6	17	6.9
Tetralogy of Fallot	11	15	4	13.8
Aortic Stenosis	4	—	12	6.9
Coarctation	6	3.7	2.5	0.63
Transposition	8	—	—	—
Tricuspid Atresia	3	—	0.3	0.63
Truncus	1	—	—	0.63
All others	11	—	7.2	11.5
No. of Cases	Not Given	426	291	159

who studied school children. but other series shown in Table 3 give much lower figures (20 - 28%). The high incidence of V.S.D. in our series might be due partly to the lower incidence of symptomless anomalies such as patent ductus arteriosus or coarctation who are still undiagnosed and have not yet come forward or who have presented themselves to physicians seeing older patients. Many

children with severe defects who have died in infancy may not have been included in our series. The incidence of atrial septal defect in the series dealing with children and adults is to be noted since this condition can also be symptomless until adult age.

The frequency of the presenting symptoms in the more common congenital heart anomalies is shown in Tables 4.01, 4.02, 4.03, 4.04, 4.05.

The commonest presenting symptoms were dyspnoea and cyanosis (18.2%) which is to be expected. Many children (26.4%) were discovered to have a murmur

TABLE 4.01
Presenting symptoms in
congenital heart disease

<i>No. of Cases</i>	<i>Presenting Symptom or Sign</i>	<i>Percentage</i>
44	Dyspnoea	27.6
42	Murmur	26.4
29	Cyanosis	18.2
22	Resp. Tract Infection	13.8
6	Fatigue	3.9
5	Acute heart failure	3.1
4	Failure to thrive	2.5
2	Chest pain	1.25
1	Cardiomegaly	0.6
1	Cong. Oesophageal Atresia	0.6
1	Cough & epistaxis	0.6
1	Routine for R.T.I.	0.6
1	Dizziness	0.6
159		

TABLE 4.02
Presenting symptoms in
ventricular septal defect

<i>No. of Cases</i>	<i>Presenting Symptom or Sign</i>	<i>Percentage</i>
27	Murmur on routine exam.	38.0
20	Dyspnoea	28.2
13	Recurrent U.R.T.I.	18.3
3	Failure to thrive	4.2
3	Fatigue	4.2
5	Acute heart failure	7.0

71 **Note** Eight of these patients were mongols, one was a cretin.

TABLE 4.03
Presenting symptoms in
atrial septal defect

<i>No. of Cases</i>	<i>Presenting Symptom or Sign</i>	<i>Percentage</i>
9	Dyspnoea	60.0
2	Murmur	13.3
2	U.R.T.I.	13.3
1	Fatigue	6.66
1	Failure to thrive	6.66
17	Note Two (2) pts. had associated cong. defects (cretinism hare-lip). Another had the W.P.W. syndrome.	

on routine examination. Another common mode of presentation was recurrent respiratory tract infection. Unusual but with very interesting presenting symptoms were the 2 children who had aortic valvular stenosis

TABLE 4.04
Presenting symptoms in
congenital aortic stenosis

<i>No. of Cases</i>	<i>Presenting Symptom or Sign</i>	<i>Percentage</i>
5	Murmur	45.4
3	Dyspnoea	27.1
2	Chest pain	18.1
1	Fatigue	9.09
11	Note One patient had gargoylism.	

TABLE 4.05
Presenting symptoms in
pulmonary stenosis

<i>No. of Cases</i>	<i>Presenting Symptom or Sign</i>	<i>Percentage</i>
6	Murmur	50.0
2	Dyspnoea	16.7
2	U.R.T.I.	16.7
1	Cardiomegaly	8.3
1	Fatigue	8.3
12	Note Two of these patients were sisters.	

who complained of chest pain, and the child with congenital heart block whose chief complaint was severe dizziness, a symptom usually associated with adult patients.

An interesting association is V.S.D. with other congenital anomalies, 12.6% of our V.S.D., of whom eight were mongols and the other a cretin, had an associated congenital lesion. This association has also been noted by Silvy (1934) and Evans (1950) who observed that the commonest congenital heart anomaly associated with mongolism was V.S.D.

Table 5.01 shows the different types of rheumatic valvular lesions that make up the 74 cases in this series, while tables 5.02 and 5.03 show the presenting symptoms in mitral insufficiency and mitral stenosis.

TABLE 5.01
Percentage distribution of
the different valvular lesions
in rheumatic heart disease

<i>Type of Valvular Lesion</i>	<i>No. of Cases</i>	<i>Percentage</i>
Mitral Incompetence	46	62.2
Mitral Stenosis	10	13.5
M.S. c M.I.	8	10.8
Aortic Stenosis	2	2.7
Aortic Incompetence	2	2.7
A.S. c A.I.	2	2.7
Mixed Mitral & Aortic	4	5.4
Total	74	

TABLE 5.02
Presenting symptoms in
mitral incompetence

<i>No. of Cases</i>	<i>Presenting Symptom or Sign</i>	<i>Percentage</i>
22	Acute rheumatic fever	47.8
5	U.R.T.I.	10.9
4	Joint pains	8.7
4	Dyspnoea	8.7
4	Acute heart failure	8.7
3	Tonsillitis	6.52
2	Murmur	4.4
2	Chorea	4.4
46		

TABLE 5.03
Presenting symptoms in
mitral stenosis

No. of Cases	Presenting Symptom or Sign	Percentage
7	Murmur	20.0
2	Dyspnoea	70.0
1	U.R.T.I.	10.0
10		

M.I. very often follows an acute episode of Rh. fever. 22 patients (47.8%) were first seen during an acute attack of Rh. fever. Other less common modes of presentation were joint pains, chorea, recurrent upper respiratory tract infection and acute tonsillitis. Dyspnoea is an uncommon mode of presentation in children (only 8.7%).

The mode of presentation of M.S. on the other hand is very different. As can be expected the number of cases with M.S. is much smaller than that of M.I. cases. However, the few cases seen were all severe cases, two of whom had tight M.S. at the ages of seven and nine, bearing in mind that the usual latent symptom-free period is usually in the region of 19 years (Faul Wood Series). Dyspnoea was by far the commonest presenting symptom (70%).

Of the 74 cases of rheumatic heart disease 50 gave a history of rheumatic fever, 16 had no past history, while 8 suffered from equivalents (sore throats, epistaxis, skin lesions).

Most of the cases classified as "cardiomegaly", "innocent murmurs" and "thymic hyperplasia" are still being followed up and will be the subject of a separate study. Of the five cases of supraventricular paroxysmal tachycardia, four had their attacks terminated by intramuscular deslanoside U.S.P. (Cedilanid).

TABLE 6
Presenting symptoms in cases
diagnosed as cardiomegaly —
under investigation

No. of Cases	Presenting Symptom or Sign	Percentage
7	Dyspnoea	29.2
5	U.R.T.I.	20.8
3	Cyanosis	12.5
2	Homozygous (major) thalassaemia	8.3
2	Tachycardia	8.3
2	Murmur	8.3
1	Hypercalcemia	4.2
1	Failure to thrive	4.2
1	Acute heart failure	4.2
24		

One case requiring an additional small dose of propranolol suffered two relapses of supraventricular paroxysmal tachycardia and his electrocardiogram showed the Wolf-Parkinson-White syndrome.

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