CONGENITAL HEART DISEASE AMONGST MALAYSIAN CHILDREN.

RICHARD O. JOHNSON, BEE HONG JOHNSON, ANDREW W. GRIEVE,

INTRODUCTION

CONGENTIAL HEART disease provides a major contribution to the work load (inpatient and ambulatory) of the paediatric, medical and surgical services of the University Hospital. To date, University Hospital has been the major referral centre for heart disease in Malaysia, being the only hospital with "open heart" surgical capability.

The purpose of this brief report is to indicate the incidence of congenital heart defects in Malaysian children as experienced by the paediatric section of the University Hospital Cardiac Service.

SUBJECTS

During the 2½ years from November 1975 to April 1978 1,148 children under the age of 12 years, were referred for cardiac evaluation. 1,037 were diagnosed as having congenital heart disease. Of the remaining 111, 47 had rheumatic heart disease and 64 were considered not to have heart disease at all. Infants with heart disease (especially sick neonates) were referred infrequently, and most cases of childhood rheumatic heart disease in University Hospital, and presumably elsewhere in Malaysia, were managed without referral to the cardiac unit. It is believed therefore that both of these groups are significantly under-represented.

The Department of Paediatrics, University of Malaya, Kuala Lumpur, Malaysia.

RICHARD O. JOHNSON, M.B., B.S. (N.S.W.), F.R.A.C.P., B.Sc Med. (N.S.W.)

BEE HONG JOHNSON, M.B., B.S. (N.S.W.)

ANDREW W. GRIEVE, M.B., B.S. (Lond.), M.R.C.S., L.R.C.P., M.R.C.P. (U.K.)

Address for reprints: Dr. R.O. Johnson, 23 Rowan Street, Orange, NSW, 2800, Australia.

METHODS

All patients were assessed at least once by one or more of the authors, and in many instances by members of the adult cardiac service also. In every case clinical diagnosis was aided by chest radiographs and electrocardiograph. Other non-invasive investigations (haemoglobin determination, arterial blood-gas studies, phonocardiography, and echocardiography) were employed as diagnostic aids where appropriate.

Clinical diagnosis alone was available for three quarters of the children with congenital heart disease. Confirmation of diagnosis was possible for only 24.8% of cases. In this latter group, confirmation was by cardiac catheterization (with cardiac angiography when indicated) in 43%, by cardiac catheterization and subsequent surgery in 26.5%, by surgery alone in 26.5% (mostly patent ductus arterious), by echocardiography alone in 1% (hypertrophic obstructive cardiomyopathy, 1 case, and mitral valve prolapse, 2 cases) and by autopsy in only 3%.

INCIDENCE OF DIFFERENT CONGENITAL HEART DEFECTS

We found simple Ventricular Septal Defect (VSD), Patent Ductus Arteriosus (PDA), Tetralogy of Fallot, secundum Atrial Septal Defect (ASD), Pulmonary Stenosis (PS) with intact interventricular septum, Endocardial Cushion Defect and Transposition of the Great Arteries (TGA), in that order, to be the 7 most common defects. These defects also ranked within the first 10 in other series (Loh, 1969; Nadas and Fyler, 1972; Keith *et al.*, 1967).

Simple VSD, accounting for 40.4% of our cases, was more common than usually reported (31.4% by Loh (1969), 19.4% by Nadas and Fyler (1972)). This may be due to overdiagnosis of simple VSD and to under-representation of other defects (especially those which present in early infancy) rather than to an actual higher incidence of simple VSD.

Table I

Incidence of congenital heart defects by diagnosis

Rank Order of Lesion	Diagnosis	Number of Cases	Percent of Total	Number of cases with Confirmed Diagnosis
1.	Ventricular Septal Defect (VSD) Simple	419	40.4	22
2.	Patent Ductus Arteriosus (PDA)	156	15.0	102
3.	Tetralogy of Fallot	148	14.3	35
4.	Atrial Septal Defect (ASD) Secundum	53	5.1	17
5.	Pulmonary Stenosis (PS) with intact interventricular septum	52	5.0	14
6.	Endocardial Cushion Defect	22	2.1	5
7.	Transposition of the Great Arteries (TGA)	17	1.6	7
8.	Dextrocardias	16	1.5	
9.	Pulmonary Atresia with VSD	15	1.5	5
10.	Myocardial Diseases	12	1.2	7
11.	Arrythmias	12	1.2	
12.	VSD with PDA	9	0.9	6
13.	Tricuspid Atresia	9	0.9	2
14.	VSD with ASD	8	0.8	7
15.	Coarctation of Aorta	8	0.8	3
16.	Aortic Stenosis — congenital	8	0.8	3
17.	VSD with PS	7	0.7	2
18.	Total Anomalous Pulmonary Venous Drainage	7	0.7	3
19.	Mitral Incompetence (MI) — non-Rheumatic	7	0.7	2
20.	VSD with Aortic Incompetence	4	0.4	3
21.	Ebstein's Anomaly	4	0.4	2
22.	Pulmonary Hypertension — idiopathic	4	0.4	1
23.	VSD with coarctation of aorta	3	0.3	2
24.	VSD with MI	3	0.3	0
25.	Corrected TGA	3	0.3	0
26.	DOuble outlet right ventricle	2	0.2	2
27.	Coronary arterio-venous fistula	2	0.2	2
28.	Sinus of Valsalva fistula	2	0.2	1
29.	Truncus Arteriosus	2	0.2	0
30.	Aortic Incompetence — non-Rheumatic	2	0.2	0
31.	Hypoplastic Left Heart	1	0.1	0
	Complex Cyanotic — type undertermined	18	1.7	0
	VSD with other defects	2	0.2	2
	TOTAL	1,037	100.3%	257

VSD with PS (and left to right shunt) was very much less common in our experience (0.7%) than reported by Loh (1969) (3.2%) or Nadas and Fyler (1972) (3.1%). Tetralogy of Fallot accounted in our experience for a larger proportion of congenital heart disease (14.3%) than found by Loh (1969) (9.4%), and was the commonest cyanotic lesion. TGA was diagnosed in only 1.6% of our cases compared with 4.7 — 7.6% reported by others. This is probably an under-representation as most cases of TGA present in early infancy and many die before referral for cardiac evaluation. Also some of the "Complex Cyanotic — type undertermined" group of patients may have transposition.

We found low incidence of Coarctation of Aorta (0.8%) and congenital Aortic Stenosis (AS) (0.8%), both of which lesions can be diagnosed clinically with a fair degree of confidence. Low incidence of these lesions was also reported by Loh (1969) in Singapore and is in sharp contrast to Western experience, where coarctation and AS are common. Nadas and Fyler (1972) ranked

Coarctation of Aorta third (8.1%) and Aortic Stenosis sixth (5.5%).

Hypoplastic Left Heart was infrequently diagnosed in Singapore (6 cases, 0.5%) (Loh, 1969) and only once in this report, despite its quite common occurrence in Western centres. It is highly likely that the true incidence of this lesion is much higher than we found, as most cases of Hypoplastic Left Heart, a uniformly lethal defect, die within the first week of life. It may be speculated, however, that Hypoplastic Left Heart, like other left sided obstructive lesions, has a truly lower incidence in Malaysia and Singapore than in Western countries.

The spectrum of congenital heart defects encountered at the University Hospital, Kuala Lumpur, is similar to that described in other cardiac centres. We must emphasise that in 75% of our cases, the diagnosis was made clinically and this may partly account for the difference in the incidence of specific cardiac defects compared to those reported in Western countries. Documentation of the true incidence and distribution of the various congenital cardiac lesion in this country will have to wait until full investigative cardiac services are available for every child suspected of harbouring a heart defect.

ACKNOWLEDGEMENTS

Sincere thanks to our colleagues in the adult cardiology service and the cardiothoracic surgical service for their invaluable advice and expertise, so generously given; to the staff of the Cardio-respiratory and Cardiac Catheter Laboratories; and to Miss Florence Aw Yong for typing the manuscript.

REFERENCES

Keith, J.D., Rowe, R.D. and Vlad, P. (1967). "Heart Disease in Infancy and Childhood". 2nd Edition, Mac-Millan Company, New York.

Loh T.F. (1969). Congenital Heart Disease in Singapore — A Review of 5 years (1964-1968). *J. Singapore Paediatric Society*. 11(2), 89 — 100.

Nadas, A.S., and Fyler, D.C. (1972). "Pediatric Cardiology" 3rd Edition, W.B. Saunders Company, Philadelphia.