CONGENITAL HEART DISEASE IN SCHOOL CHILDREN AND ADULTS IN SINGAPORE *

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The knowledge of the incidence of a disease is the cornerstone of any study in morbidity, and the awareness must modify the approach in medicine. Such knowledge or awareness is essential for a correct appreciation of the extent of a disease locally, and the geographical variation of pattern may help in the elucidation of the aetiology.

To be effective, incidence must be reliable, and this demands reliable statistical information, and rigid and accurate diagnostic criteria. The increasing popularity of Western medicine in the postwar years makes available sufficient information and material. Accurate diagnoses would imply not only familiarity with current advances, and acquisition of new technique and equipment but also the requisite personnel, and a strict assessment of individual errors.

Material for this study is drawn from the following sources: -

1. Cases seen in the cardiac clinic of Medical Unit II.
2. Cases seen in the consultative cardiac clinic in the Singapore Anti-Tuberculosis Association.
3. Referred from School Health Clinics.
4. Referred from doctors in charge of police and officials.
5. Referred from general practitioners.

The period of registering was from 1954 to 1957, and the period of observation extended to 1962. The ages extended from five onwards. The closure of the registry was because the Paediatrics Units began to accept cases up to 10 years old from the end of 1956, and the period of observation was to enable that every case should have been followed up for at least 4 years.

Methods of study included the usual history taking and clinical examination, electrocardiograph, fluoroscopy and, in addition, angiocardiogram in selected cases. Catheterisation was not attempted because of the lack of facilities during that period.

The classification adopted was aimed to be simple, comprehensive and also of value in the selection of cases for treatment. Congenital heart disease was defined as an anomaly of the heart either in function or structure or both at the time of birth, and because of the practical difficulties in distinguishing between congenital and acquired diseases, and the absence of any useful medical or surgical aid at the time of study, conduction and rhythmic disturbances were excluded from the series. The classification was similar to that proposed by Bing (1958) and Marple (1953) as follows: -

1. Those with increased pulmonary circulation — atrial septal defect, patent ductus arteriosus, Eisenmenger's syndrome, ventricular septal defect, aorto-pulmonary fistula.
2. Those with decreased pulmonary circulation for example, Fallot's tetralogy, pulmonary stenosis, tricuspid atresia, absence of pulmonary arteries.
3. Septal anomaly: -

   Atrial septal defect, ventricular septal defect, cor biloculaire, cor triloculaire.
4. Others: -
   a. Vessel anomaly of the aortic arch, aorta and pulmonary arteries, for example, Coarctation, dextroposition, transposition.
   b. Valvular anomaly, for example, valvular stenosis, and subvalvular stenosis, infundubular stenosis and bicuspid valves.
   c. Vessel anomaly of the coronary systems and large veins, for example, abnormal superior vena cava.
   d. Myocardial and pericardial anomaly.

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TABLE SHOWING AGE, INCIDENCE AND RACIAL BREAKDOWN.

AGE	CHINESE		MALAY		INDIAN		OTHERS		TOTAL	
	M.	F.	M.	F.	M.	F.	M.	F.	M.	F.
0 – 5 **	0	2	0	0	0	1	0	0	0	3
6 – 10	20	17	3	0	3	1	0	0	26	13
11 – 15	15	24	3	1	2	0	0	0	20	25
16 – 20	11	3	3	0	1	0	0	0	15	3
21 – 25	7	4	3	1	2	1	1	0	13	6
26 – 30	3	5	1	0	0	0	0	0	4	5
31 – 35	2	0	1	0	1	0	0	0	4	0
36 & above	4	10	0	1	2	1	0	0	6	12
Total	62	70	14	3	11	4	1	0	88	77

**As the majority of the cases under four are looked after by the Paediatric Unit, the absence of cases in this age group must mean that most of the cases go to the Paediatricians.

TABLE.

SHOWING THE DIFFERENT GROUPS AND THE ASSOCIATED FINDINGS & COMPLICATIONS

Type of Anomaly	*Number of Cases*	*%*	** *Muir* (2)	*Abbott* (15)	*Gibson*
Atrial Septal Defect	66	40.0	16.5	3.3	10.0
Patent Ductus Arteriosus	22	13.3	—	10.5	4.5
Ventricular Septal Defect	24	14.5	18.1	6.2	8.5
Fallot's Tetralogy	20	12.2	7.4	8.5	20.5
Eisenmenger Complex	3	1.8	2.3	—	2.0
Pulmonary Stenosis	14	8.3	1.7	4.0	15.5
Lutenbacher Syndrome	1	0.6	—	—	—
Coarctation of Aorta	2	1.2	3.3	3.5	4.0
Dextrocardia	4	2.4	—	—	—
Situs Inversus	5	3.0	—	—	—
Tricuspid Atresia	1	0.6	—	1.9	—
Undiagnosed — Probably 1 patent foramen ovale, 1 aorto-pulmonary fistula and 1 aortic stenosis	3	1.3			
Total	165				

**Necropsy incidence must differ from case incidence in that poor prognosis cases will predominate in the former, and comparison of the two may give a guide to prognosis.

Incidence may be on figures drawn from necropsy, hospital attendances and regional surveys. Each of these has its own shortcomings. The degree of acceptance of post-mortems differs locally. The major racial groups are Malay, Chinese and Indian. The Indians on the whole are reasonably receptive, the Chinese less so and the Malays totally resistant. In fact in 1957, the Minister of Health passed a ruling that to approach a Malay family for consent for necropsy would be a breach of discipline! This sort of ruling would be injurious to the Malay community because their disease would be least understood, and hence they would stand to benefit least from modern medicine. It would also mean that necropsy incidence for Malays would be totally worthless. Necropsy for Chinese infants, however, was generally obtainable, and hence necropsy figures for Chinese would show a strong bias for babies.

Hospital incidence figures would depend on the utilisation rate of the communities, and it is noted that, taking Chinese as the norm, Indians tend to use the hospital services twice as much, whereas the Malays halved or less. There is also a female reluctance to come to the hospital with M: F ratio of 2: 1 of hospital incidence other than the paediatric group. Regional surveys on theoretical grounds would be most reflective of the population incidence, but local surveys in the form of miniature chest X-ray survey accepted only cases after the age of 14 — an age which excluded a large population of congenital heart disease. Bearing these in mind, the following figures were of interest: -

GENERAL HOSPITAL ANNUAL RETURNS.

Year	No. of Admns.	No. of Congenital Heart	Percentage
1954	26,693	145	0.6%
1955	27,415	168	0.6%
1956	31,445	205	0.6%
1957	33,507	214	0.6%

Hanam and Chew (1961) reported an incidence of 0.3% for a regional chest X-ray survey, and Shanmugaratnam (1950) and Muir (1960) an incidence of 2.8% and 2.1% for necropsy respectively. Jacobius and Moore however reported an incidence of 8.1% in necropsy.

In the present series of 165 cases, the breakdown would suggest that 53.9% of cases were below the age of 15. This would suggest that chest X-ray survey would miss out at least half of the cases. Taking into consideration the above factors, the population incidence would appear to be in the region of 0.7% to 0.8%. Most of the cases, to be expected, were Chinese, but the number of Malay cases appeared high considering the extent of their use of hospital facilities. It could be that the incidence is higher in the Malays (Table).

INDIVIDUAL GROUPS

1. Atrial Septal Defects

In this series of 165 cases, there are 66 cases of A.S.D. equivalent to 40% of the series. Muir found in local necropsies an incidence rate of 11.9 and 16.5% respectively in 2 different series. This large disparity between the living and dead probably reflects the comparatively good prognosis of A.S.D. as compared to other congenital defects as after a time especially after adolescence, the surviving cases will have a high proportion of A.S.D. Hanam and Chew (1961) computed in their series an incidence of 51.7% and since their series has no cases under 14 years of age, the higher figure would further support the speculation that A.S.D. has a better survival prognosis. Reference to Table shows a M: F ratio of 5: 3 which, on consideration of factors mentioned in the preceding paragraph, would confirm the reported female bias of the disease. Right bundle branch block was found in only 40% of the E.C.G. (Table).

TABLE — ELECTROCARDIOGRAPH

Right ventricular hypertrophy	23
Right bundle branch block	19
Normal	6
Total	48

This finding is in agreement with that of Braunwald et al (1955) but differs from that of Barker et al (1950) who stated that more than 90% would have a RBBB pattern.

The age incidence showed a sharp drop after the age of 35, and this may indicate that

locally at least, the long term outlook of A.S.D. is not good, for the sharp fall must suggest that the cases die off in the latter thirties, unless one wishes to consider the extreme improbability of spontaneous closure at a later period of life. Such has been reported, but the happening is rare.

TABLE — AGE AND SEX BREAKDOWN

Age	Male	Female	Total
6– 5	0	0	0
6–10	8	5	13
11–15	3	6	9
16–20	7	3	10
21–25	6	0	6
26–30	2	2	4
31-35	2	0	2
36–40	0	0	0
41–45	1	1	2
46–50	0	0	0
51 +	1	1	2
Total	30	18	48

If this contention is correct, ASD population died off early, and the sharp drop would suggest that the decade at risk is between 31 to 40 with the tendency beginning in the 31 to 35 group. Wood (1950) reported that cases might live to the fifth or sixth decade. The usual expectation of life is accepted as about 35 years of age, and would appear to be indirectly confirmed by the present series.

2. Patent Ductus Arteriosus

There are 18 cases with an M: F ratio of 4: 14 showing a very marked bias in the female. Again, there is a sudden fall in incidence from the age group 26–30 and, as argued previously, this would indicate the local prognosis of PDA is a grim one — with expected survival up to 26 only. Shapiro and Keys (1963) reported an expectation life of 35 to 40 years. Although spontaneous closure in later life has been reported, such events are rare, and the probability is that they begin to die in the late second decade.

This evidence must make the cause for the early surgical intervention very strong indeed, particularly when the operation is relatively safe and the cure usually complete (See Table).

TABLE — AGE AND SEX BREAKDOWN

Age	Male	Female	Total
4– 5	0	0	0
6–10	2	4	6
11–15	0	4	4
16–20	1	2	3
21–25	1	2	3
26–30	0	1	1
31–35	0	0	0
36–40	0	0	0
41–45	0	1	1
46–50	0	0	0
51 +	0	0	0
Total	4	14	18

It would appear that the usually experienced well-being of this condition is deceptive, and the true prognosis is grave without surgery.

3. Ventricular Septal Defect

There are 19 cases altogether with a sex ratio of M: F = 8: 11, showing a female bias (See Table).

TABLE — AGE AND SEX BREAKDOWN

Age	Male	Female	Total
6–10	0	1	1
11–15	3	6	9
16–20	0	0	0
21–25	0	0	0
26–30	0	1	1
31–35	0	0	0
36–40	0	1	1
41–45	1	0	1
46–50	0	0	0
51 +	0	1	1
Total	4	10	14

As no attempt is made to differentiate between VSD of different sizes and positions, the analysis in age distribution would be of less importance in considering the survival, for Roger's disease is well known to have little haemodynamic disturbances and a very good outlook as far as prognosis is concerned, whereas the high VSD is more inclined to the production of pulmonary hypertension with a more gloomy consequence. Mixture of two

such groups would invalidate any attempt to deduce the life expectation from the age distribution.

It is reported that VSD are much more liable to subacute bacterial endocarditis. Gelfman and Levine (1942) reported 23% of all ages of Roger's disease had subacute bacterial endocarditis and 51% of them had the complication within a period of 2 years.

In the present series, no case had developed subacute bacterial endocarditis, and it may be that this is due to the improved state of health because of the regular surveillance afforded by the necessity of the follow up.

4. Fallot's Tetralogy

There are 15 cases in the series with a M: F ratio of 11: 4 which is insignificant from the point of sexual bias as the normal ratio of patients seeking treatment in the General Hospital has consistently been 2: 1 throughout the years. Of these cases, the majority occurs in the age group of 5 to 20 years and no surviving cases after 25 years of age was seen in this series (See Table).

TABLE — AGE AND SEX BREAKDOWN

Type	Male	Female	Total
4– 5	0	0	0
6–10	3*	2	5
11–15	5	1	6
16–20	3	0	3
21–30	0	1	1
31 +	No case		
Total	11	4	15

* (one case died and was found to be truncus).

Although Fallot's tetralogy living to a ripe old age has been reported in which one lived to the age of 60 and the others to 62, it would appear that in general, the outlook is poor towards the third decade. Postmortem figures also suggested a high incidence of death during the first year of life, and again at a later age in the twenties (Muir 1960). This finding is of considerable importance as there are surgical measures to relieve the cardiac stress of a Fallot's tetralogy, and if there is a risk of death in the early age groups, the case can be made for recommending surgery in selected cases early in life — probably in the first year of life.

There is some ground for believing that one of the group of Fallot's tetralogy is dependent on the patency of the ductus arteriosus to survive, because of the severity of the pulmonary stenosis, and the early closure of the ductus in most cases in the first month produces a fatal result (Gwee 1958). In other words, there may be two periods of maximal death in Fallot's tetralogy. This would seem to be true in the series of Muir (1960) locally and has been also been reported elsewhere (Rowe et al 1955).

5. Pulmonary Stenosis

There are 8 cases in this series with a M: F ratio of 3: 5 which appears to be a significant sexual bias. The distribution is sporadic in the age groups suggesting that survival to a good age is possible. No case, however, has occurred after 35, but in view of the small total number, no definite conclusion should be drawn (See Table).

TABLE — AGE AND SEX BREAKDOWN

Age Group	Male	Female	Total
4– 5	0	0	0
6–10	2	1	3
11–15	0	1	1
16–20	0	1	1
21–25	0	0	0
26–30	0	1	1
31–35	0	0	0
36–40	1	0	1
41–45	0	1	1
46–50	0	0	0
51 +	0	0	0
Total	3	5	8

It is perhaps worth noting that a systolic murmur with thrill in the pulmonary area, though highly suggestive of a diagnosis of pulmonary stenosis, is by no means diagnostic. Two cases were seen during the collection of this series with systolic murmur and thrill in the pulmonary area. One had a soft chest wall, and a depressed upper chest. The murmur was apparently due to the local com-

pressive effect of the costal cartilage as the murmur and thrill were markedly reduced when the patients sat up and took in a deep breath, and the signs increased when the upper left costal cartilages were compressed manually. One of these cases died, and at postmortem, there was no local disease in the pulmonary valve, and the pulmonary artery was normal. The other, after a year or so, when she grew up, lost both her thrill and her murmur (Gwee 1962).

6. Coarctation of the Aorta

Only 2 cases were found. The selection of cases above 4 years of age must have excluded many cases as suggested by Muir's series where coarctation occurred in 4.6% and 8.3%.

7. Dextrocardia

5 cases were found and were complete situs inversus. M: F ratio is 2: 3 suggesting a female bias (See Table).

AGE AND SEX BREAKDOWN

Age group	Male	Female	Total
4– 5	1	0	1
6–10	1	1	2
11–15*	0	0	0
16–50*	0	0	0
51 +	0	2	2
Total	2	3	5

* The absence of cases in these age groups is not well understood, as the prognosis being good, there does not seem any reason why there should be no cases. One probable explanation is that the majority of the cases came from referrals, and as the defect is symptomless, the patients do not come up until they are old when other complaints bring them along.

8. Miscellaneous

A case of cyanotic heart disease was sent up as a case of Fallot's tetralogy and was found to be one of tricuspid atresia. He survived to the age of 20. A number of left-sided superior vena cava was confirmed at angiocardiogram examinations, and since they are symptomless, and require no treatment, they are omitted from the series, especially when a significant proportion of the cases in the series refused angiocardiogram.

SUMMARY

A study of congenital heart disease in school children and adults in Singapore was made, and the population incidence was worked out to be in the region of 0.7 to 0.8%.

It was contended that the distribution in age group would indicate a life expectancy trend, and the results were analysed accordingly. Apart from situs inversus, all the listed forms of congenital heart disease have serious limitation to survival.

REFERENCES

Braunwald, E., Sapir, S.O., Donoso, E., Grishman, A. (1955): A study of the electrocardiogram and vectorcardiogram in Congenital Heart Disease. Amer. Heart Jour., 50: 323.

Barker, J. M., Magidson, O. and Wood, P. (1950): Atrial Septal Defect. Brit. Heart Jour., 12: 277.

Bing, J. R. (1958): "Catheterisation of the heart" Advances in Int. Medicine. The Year Book Publishers Inc., U.S.A.

Gelfman, R., Levine, S.A. (1942): The Incidence of Acute and Subacute Bacterial Endocarditis in Congenital Heart Disease. Jour. Med. Sc., 204: 846.

Gwee, A.L. and Nadarajah, I. (1960): A Survey of Tetanus in Singapore. S.M.J., 1: 4, 164.

Gwee, A.L. (1958): Congenital Heart Disease with special reference to Fallot's tetralogy. — "Refresher Course in Cardiology". Annals of Academy, Singapore, Vol. 1.

Gwee, A.L. (1962): Unpublished material.

Hanam, E. and Chew, A. (1961): Heart Disease from a case-finding Tuberculosis Survey in Singapore, 1958. S.M.J., 2: 1, 3.

Jacobius, H.L. and Moore, R.A. Incidence of Congenital Anomalies in Autopsies at New York Hospital. Jour. Tech. Meth. XVIII, 133.

Marple, C.D. (1953): Clinical Cardiology. 1st Ed. Baltimore, Maryland.

Muir, C.S. (1960): Incidence of Congenital Heart Disease in Singapore. Brit. Heart Jour., 22: 2, 243.

Rowe, R.D., Vlad, P. and Keith, I.D. (1955): Experiences with 180 cases of tetralogy of Fallot in infants and children. J. Canad. Med. Assoc., 73: 1, 23.

Shapiro, M.J. and Keys, A. (1943): The Prognosis of untreated Patent Ductus Arteriosus and the Results of Surgical Intervention. Amer. J. Med. Sci., 206: 174.

Wood, P. (1950): Congenital Heart Disease: A review of its clinical aspects in the light of experience gained by means of modern technique B.M.J., 2: 639, 693.