

CONGENITAL HEART DISEASE IN SINGAPORE — PRESENT PROBLEMS AND FUTURE PERSPECTIVES

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SYNOPSIS

Congenital heart disease has become the top killer amongst children admitted to the University Department of Paediatrics, Singapore General Hospital in the last 5 years. Analysis of the last 1000 cases of congenital heart disease seen in the Department reveals that 95% of them are due to multifactorial inheritance. Prevention aims at elimination of the remaining 5% of the cases that are due to primary environmental causes like congenital rubella infection or to primary genetic causes like Down's anomaly, will have little effect on the incidence of congenital heart disease in Singapore. As it is now feasible to salvage many children with congenital heart disease who were previously inoperable, it is morally unjustified to see these children die and suffer. Singapore, with her intention to achieve medical excellence, cannot afford to neglect the field of paediatric cardiothoracic surgery. As congenital heart disease has outnumbered acquired heart disease in terms of open heart surgery by 4 : 1 in recent years in Singapore, priority in cardiac catheterization should be given to patients with congenital heart disease. With the existing infrastructures, further progress to excellence is feasible provided we are prepared to invest our time, energy and a little extra cost to upgrade our skills and to provide adequately trained personnel and equipments to tackle this pressing problem.

INTRODUCTION

In recent years, Singapore has improved considerably in economic growth and together with it, better housing and education, higher standard of medical practice and more efficient delivery of health care to the people. It is therefore not surprising that Singapore has done better, in terms of infant mortality rate, which is generally considered as a good index of overall health status of the community, than many of the advanced Western countries like United Kingdom, United States, Australia and New Zealand (1). However, with the control of infectious diseases (2) and malnutrition which were prevalent two decades ago, the medical problems which face the paediatricians in institutional practice in the eighties will be very different compared to those ten to twenty years ago. Congenital heart disease (CHD), for example, with its incidence around 6 to 8 per 1000 live births (3) has become the pressing problem to doctors caring for infants, children and adolescents in Singapore.

The purpose of this paper is to survey the importance of CHD as a cause of mortality in the past two decades in the University Department of Paediatrics, Singapore General Hospital, to analyse the aetiological cause of CHD and to examine the currently available medical services, both diagnostic and therapeutic, in order to propose preventive measures and to suggest plans for further improvement on the existing facilities.

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MATERIALS AND METHODS

The University Department of Paediatrics, Singapore General Hospital, is one of the two largest paediatric departments in Singapore. It has 120 beds with a 3-bed intensive care unit. The annual admission rate is around 5000. The age limit for admission is up to 12 years.

The Departmental Weekly Mortality Record from 1966 to 1980 inclusive was analysed to obtain the death rate per year and the top five causes of death in these three 5-year periods. The Cardiac Clinic Registration and Cardiac Catheterization Record Books provided information on aetiology of last 1000 cases of CHD seen in the Department from 1973 to 1981 as well as the number of patients registered and catheterized from 1974 to 1980.

The information on the distribution of cases of cardiac surgery from 1973 to 1978 from the Annual Report of the Department of Cardiothoracic Surgery, Tan Tock Seng Hospital, was analysed to assess the relative importance of CHD as compared to acquired heart disease in the field of cardiac surgery. Finally the percentage time allocated for paediatric cases at cardiac catheterization from 1971 to 1980 was also analysed from the Annual Report of the Cardiovascular Laboratory, Singapore General Hospital.

RESULTS

Table 1 shows the total departmental mortality expressed in case fatality rate (%) for children under 12 years of age admitted to the University Department of Paediatrics from 1966 to 1980. Over these 15 years, there is a significant drop in case fatality rate from 3.972% in 1966 to 0.858% in 1980.

Table 2 shows the absolute numbers and percent-

ages of the top five causes of death in hospitalised children in the three 5-year periods from 1966 to 1980. Figure 1 shows the relative trend of the major causes of mortality in graphic form. Note that CHD has always been an important killer since 1966.

Table 3 tabulates the aetiological causes of CHD in the last 1000 cases seen in the Department. For comparison, Western figures quoted by Nora (4) were also included.

Table 4 shows the total number of patients with CHD registered at the Cardiac Clinic as well as total number of such patients who were catheterized from 1974 to 1980. It must be stressed that not all patients with CHD admitted to the Department were registered at the Cardiac Clinic. Hence the cases presented are only minimal figures.

Table 5 tabulates the yearly number of cases who underwent cardiac surgery (both closed heart and open heart operations) in the Department of Cardiovascular Surgery, Tan Tock Seng Hospital from 1970 to 1978. In this 9-year period, the distribution of cases of cardiac surgery is tabulated according to aetiology of heart condition (congenital or acquired) and type of operation (closed or open heart surgery) in Table 6.

Figure 2 shows the yearly number of patients with CHD and with acquired heart disease who underwent open heart surgery in the Department of Cardiothoracic Surgery from 1970 to 1978. It is important to note that the total number of open heart surgery has gone up four folds over the years and CHD accounts for nearly 80% of all open heart operations in Singapore.

Table 7 shows the yearly number of cardiac catheterizations done by paediatric cardiologists in the 10-year period from 1971 to 1980 in the Cardiovascular Laboratory, Singapore General Hospital. Note that only one-third of the catheterization time was allotted to paediatric cases.

TABLE 1: TOTAL DEPARTMENTAL MORTALITY IN HOSPITALISED CHILDREN (0-12 YEARS) FROM 1966 TO 1980

YEAR	TOTAL ADMISSION	TOTAL DEATH	CASE FATALITY RATE (%)
1966	6873	273	3.972
1967	6586	244	3.705
1968	6308	216	3.424
1969	6524	179	2.744
1970	6356	169	2.659
1971	6380	158	2.477
1972	6200	144	2.323
1973	6637	178	2.682
1974	4876	144	2.953
1975	3820	86	2.251
1976	4787	85	1.776
1977	4318	73	1.691
1978	4375	59	1.349
1979	5484	70	1.277
1980	5124	44	0.858
TOTAL	84648	2122	2.507

TABLE 2: MAJOR CAUSES OF DEATH IN HOSPITALISED CHILDREN (0-12 YEARS)

YEAR	MAJOR CAUSES OF DEATH					TOTAL DEATH
	NO. 1	NO. 2	NO. 3	NO. 4	NO. 5	
1966-1970	BRONCHOPNEUMONIA 288 (27%)	CHD* 164 (15%)	GASTROENTERITIS 90 (8%)	MALIGNANCY 62 (6%)	KERNICTERUS 45 (4%)	1081 (100%)
1971-1975	BRONCHOPNEUMONIA 127 (18%)	CHD* 109 (15%)	GASTROENTERITIS 63 (9%)	MALIGNANCY 43 (6%)	SEPTICAEMIA 41 (6%)	710 (100%)
1976-1980	CHD* 72 (22%)	MALIGNANCY 56 (17%)	BRONCHOPNEUMONIA 53 (16%)	SEPTICAEMIA 16 (5%)	GASTROENTERITIS 7 (2%)	331 (100%)

*CHD = CONGENITAL HEART DISEASE

TABLE 3: AETIOLOGY OF CONGENITAL HEART DISEASE

	SINGAPORE FIGURES		WESTERN FIGURES
PRIMARYLY GENETIC FACTORS			
CHROMOSOMAL	24	(2.4%)	5%
SINGLE MUTANT GENE	9	(0.9%)	3%
PRIMARYLY ENVIRONMENTAL FACTORS			
RUBELLA	14	(1.4%)	1%
OTHERS	0		1%
GENETIC-ENVIRONMENTAL INTERACTION	953	(95.3%)	90%
	1000	(100%)	100%

TABLE 4: DEPARTMENTAL CARDIAC CLINIC AND CARDIAC CATHETERIZATION REGISTRY

YEAR	TOTAL NO. REGISTERED	TOTAL NO. CATHETERIZED
1974	192	30
1975	98	41
1976	57	52
1977	93	63
1978	88	77
1979	124	69
1980	95	40
TOTAL	747	372

DISCUSSION

From the analysis of the mortality record of the Department, it is gratifying to note that over the years from 1966 to 1980, there is a steady decline in case fatality rate, defined as the total number of death per 100 hospitalised children aged below 12 years (Table 1). This is partly due to better general health status of our children in recent years as reflected by the declining infant mortality rate (1) and partly due to improved standard of preventive as well as therapeutic and intensive paediatric care.

However, when the major causes of death in hospitalised children in the Department are examined

more closely (Table 2, Figure 1), it is revealing that CHD has become the number one killer in the last five years. Hence, if the standard of paediatric health care is to improve further, paediatricians should now spend more time and energy to tackle this pressing problem.

There are three immediate fundamental questions we have to answer before we deal with the problem of CHD in childhood:

- (1) Can CHD be prevented?
- (2) Is it feasible to salvage children with CHD?
- (3) Should we attempt to tackle this problem at all in terms of cost and manpower deployment?

TABLE 5: YEARLY NUMBER OF CARDIAC SURGERY IN TAN TOCK SENG HOSPITAL FROM 1970 TO 1978

YEAR	CLOSED HEART OPERATION			OPEN HEART OPERATION			ALL HEART OPERATION		
	CHD*	AHD†	TOTAL	CHD*	AHD†	TOTAL	CHD*	AHD†	TOTAL
1970	60	29	89	50	0	50	110	29	139
1971	69	38	107	84	8	92	153	46	199
1972	72	47	119	90	25	115	162	72	234
1973	67	33	100	98	41	139	165	74	239
1974	56	45	101	105	33	138	161	78	239
1975	66	70	136	127	36	163	193	106	299
1976	57	80	137	103	33	136	160	113	273
1977	42	84	126	126	33	159	168	117	285
1978	66	76	142	162	41	203	228	117	345
GRAND TOTAL	555	502	1057	945	250	1195	1500	752	2252

* CHD = CONGENITAL HEART DISEASE
 † AHD = ACQUIRED HEART DISEASE

TABLE 6: DISTRIBUTION OF CASES OF CARDIAC SURGERY FROM 1970-1978 (INCLUSIVE) IN TAN TOCK SENG HOSPITAL

TYPE OF CARDIAC OPERATION	TYPE OF CARDIAC DISEASE		
	CONGENITAL HEART DISEASE	ACQUIRED HEART DISEASE	ALL HEART DISEASE
CLOSED HEART OPERATION	555 (53%)	502 (47%)	1057 (47%)
OPEN HEART OPERATION	945 (79%)	250 (21%)	1195 (53%)
ALL HEART OPERATION	1500 (67%)	752 (33%)	2252 (100%)

TABLE 7: DISTRIBUTION OF CARDIAC CATHETERIZATION TIME

YEAR	TOTAL NO. OF CATHETERIZATION	NO. DONE BY PAEDIATRIC CARDIOLOGISTS	% TIME ALLOCATED TO PAEDIATRIC CASES
1971	115	49	43%
1972	221	70	32%
1973	263	107	41%
1974	204	77	38%
1975	362	110	30%
1976	335	123	37%
1977	305	142	47%
1978	401	195	49%
1979	401	152	38%
1980	427	126	30%
TOTAL	3034	1151	38%

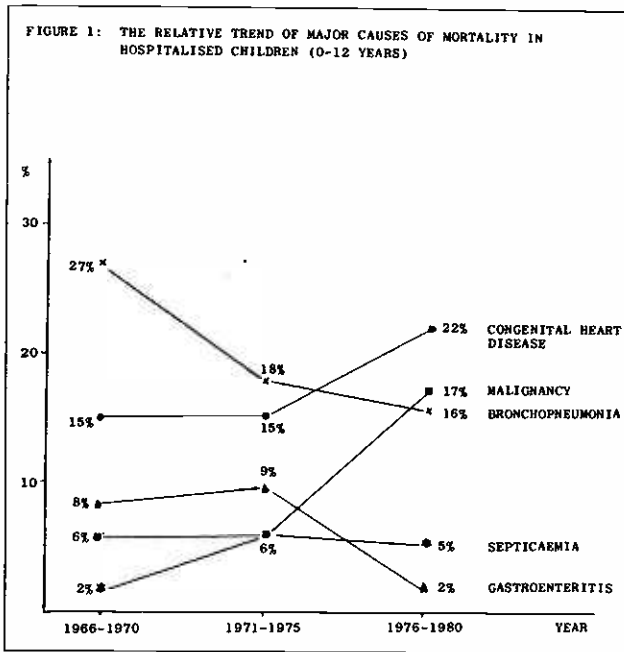


Figure 1: The relative trend of major causes of mortality in hospitalised children (0 - 12 years)

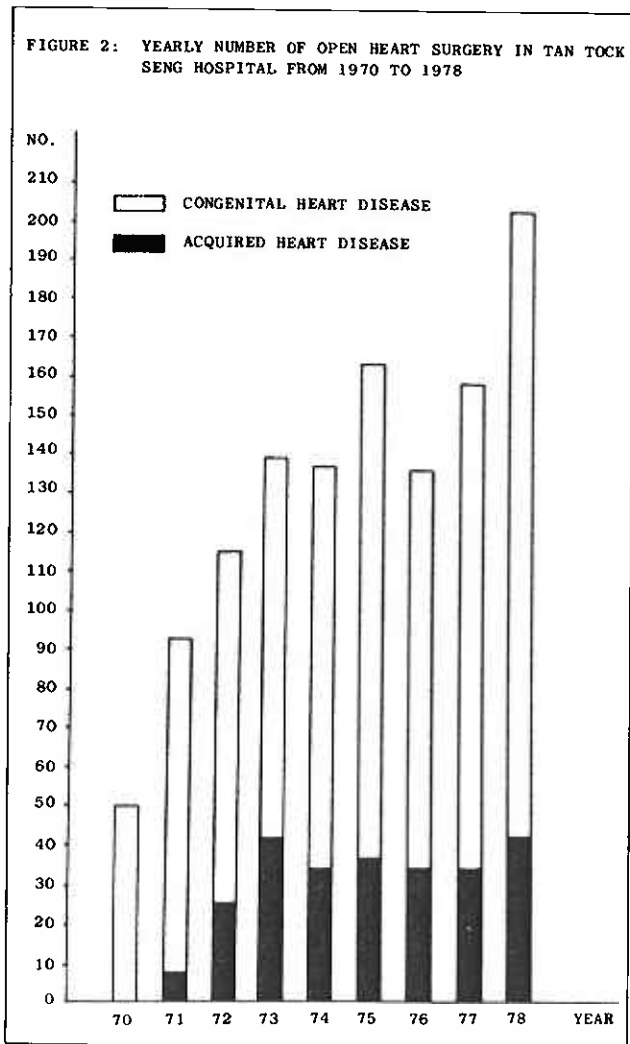


Figure 2: Yearly number of open heart surgery in Tan Tock Seng Hospital from 1970 to 1978

Prevention is better than cure. The key question is whether CHD can be prevented. In order to answer this question, the aetiologic causes of CHD must be studied. Table 3 shows that only 5% of the 1000 cases of CHD seen in the Department are due to preventable causes. Primary environmental factors like congenital rubella syndrome only accounts for 1.4% of the cases, while primary genetic factors like Down's anomaly and Mendelian Genetic disorders are responsible for only 0.9% in our patients. The aetiologic cause of CHD for the majority of cases, however, is multifactorial, due to a combination of underlying genetic propensity and some obscure environmental triggers, like irradiation, viral infections and drugs. This situation is not unique to Singapore but is also seen in the West (4) (Table 3). Although preventive measures for CHD have been emphasized (5, 6), it is certain that, even with total elimination of congenital rubella infection by immunisation and with aggressive genetic counselling to prevent recurrence of CHD caused by primary genetic factors, the incidence of CHD will remain largely unchanged. In short, CHD cannot be eliminated at this time by preventive measures.

Is it then feasible to salvage children with CHD? With a few exceptions like small ventricular septal defect and mild pulmonary stenosis, CHD is a serious disease. It is estimated that about 40% of children born with CHD die within the first year of life unless treated surgically (7). Newer techniques in anaesthesia like profound hypothermia and circulatory arrest (8, 9, 10, 11, 12) however, have ensured safety in open heart surgery even during infancy (13). Early cardiac operation is made possible partly also because of accurate pre-operative diagnosis by modern non-invasive as well as invasive investigations, like 2-D echocardiography (14) and cardiac catheterization using improved catheters designed for specific purposes (15). There is little doubt that operative correction for many different CHD's is feasible now even in young infants.

The next decision we have to make is whether we want to tackle this problem at all taking into account the cost and manpower. In this regard, we should first review the existing infrastructure in terms of diagnostic facilities and therapeutic service and identify their shortcomings. Since 1973, the Department has set up a registry for all CHD cases who are seen in the Department and also a registry for those cases who have been catheterized (Table 4). We see more than 100 new CHD patients per year, of whom about 50% are catheterized. However many deserving candidates are deprived of a chance for surgery because of lack of catheterization time. In the Department of Cardiothoracic Surgery, over the 9-year period from 1970 to 1978, there is a steady increase in the activity of cardiac surgery, both in the closed heart as well as open heart operations (Table 5). In the same period, CHD accounts for two-thirds of all cardiac surgery (Table 6). Of the open heart surgery, CHD outnumbers acquired heart disease by 4 : 1. Figure 2 shows that while the number of acquired heart disease for open heart surgery remains fairly constant over the years, the number of CHD for open heart surgery has increased five folds. This is not surprising as the

standard of living in Singapore has improved, rheumatic heart disease which has been the commonest chronic cardiac problem that requires surgery in adults before, has decreased significantly in number in recent years. This being so, cardiac surgeons therefore spend much of their time tackling CHD rather than acquired heart disease.

Unfortunately investigatory support for CHD is not given the same weightage in Singapore. Table 7 shows that in the 10-year period from 1971 to 1980, the average percentage time allocated to paediatric cases for cardiac catheterization is only 38%. This is disproportionately low compared to the fact that 75% of open heart surgery cases are CHD. Many of our patients die before cardiac catheterization can be carried out because of long waiting time. Optimal timing of investigation including catheterization has been stressed (16) and many deaths from CHD can actually be prevented if timely investigation and operation are carried out (17).

CHD is not only responsible for high mortality but also much morbidity in those children before they succumb to the disease. It is morally unjustified to see these children suffer and die when successful treatment is now available. Singapore, with her intention to become the centre of medical excellence in this part of the world, cannot afford to neglect the field of paediatric cardiothoracic surgery. With the existing infrastructures, further progress to excellence is within our reach provided we are prepared to invest our time, energy and a little extra cost to upgrade our skills, both diagnostic and therapeutic, and to provide adequately trained supportive staffs, so that we can effectively tackle this number one killer amongst hospitalised children.

Looking into the future, it is important to realise that the scenario of cardiac surgery in the management of CHD is changing. The role of surgery, being mainly palliative in the past (18), has become more definitive and aggressive now (19). Corrective open heart surgery is offered to young infants in many centres with good results (20, 21, 22, 23, 24, 25). In addition, many previously inoperable complex cardiac malformation can now be surgically managed because of the recently available conduits (26). As Singapore is striving to achieve excellence in the field of paediatric cardiovascular surgery, our approach should be positive (27). Timely and accurate pre-operative diagnosis to demonstrate abnormal anatomy and disturbed haemodynamics is as important as upgrading of surgical skill, anaesthetic expertise and standard of post-operative critical care. In this regard, availability of more catheter time, especially on an emergency basis, is the first pre-requisite. Adequate supply of trained medical, surgical, nursing and ancillary staffs ensures optimal pre-, intra- and post-operative care of children who have undergone major cardiac surgery. Periodic upgrading and maintenance of diagnostic equipments, including the availability of axial cineangiography (28, 29), 2-D echocardiography and special purpose catheters are equally important. Last but not least, critical post-operative care requires reliable monitoring and resuscitative equipments such as continuous cardiopulmonary monitor and appropriate ventilators.

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