The Natural History of Congenital Heart Disease in Young Adults

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Under the Heart Disease Program for elementary school children and junior high school students in Osaka City, the cardiac patients found through mass examination are supposed to be checked annually by the program's staff. After their graduation from junior high school, however, the cardiac patients leave our medical jurisdiction. In order to gather information from those graduates, we have mailed and continue to mail questionnaires to them once every 5 years. By analyzing that information, we sought to describe the natural history of congenital heart disease in young adults.

The study revealed that among graduates older than 20 years of age as of March 31, 1978, 17.5% underwent initial cardiac surgery; 1.8% were found to have no more cardiac lesions due to spontaneous healing; 12.3% answered that they had no significant changes; 2.0% were found to have deteriorated cardiac lesions and 2.1% of the graduates had died. Eight point one per cent of the graduate did not answer, although minimum data as to whether they were alive or not was gathered.

The mortality rates were calculated by using all follow-up data. It was revealed that the mortality rate for cardiac patients under 14 years of age, for those between 15 and 20 years of age and for those older than 21 years of age are 14 times, 6 times and 3 times as high as the general population in each age group, respectively. The mortality rate for those with mild cases of cardiac lesions is 3 times higher than the general population. It is 13 times higher among moderate cases and 77 times higher among severe cases when compared to the general population.

REPORTS on the natural history of congenital heart diseases during adult life are rare in comparison to the number of reports on congenital heart disease during childhood and adolescent.¹⁻¹⁰ This paper seeks to describe the course of congenital heart disease among young adults based upon the study of a recent questionnaire.

Key Words:

Heart disease program
Natural history
Congenital heart disease
Mortality rate of young adult

MATERIALS AND METHODS

The subjects of this study consist of a total of 844 patients who had been in heart disease program. Over 830 of them expected to be 20 years of age or older as of March 31, 1978. Fourteen patients were known to have died after reaching 18 years of age.

Table I shows the types of index disease (the last status before leaving the program due to graduation) by severity and sex. The classification of the types of index disease and severity was done based upon physical, X-ray and ECG

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TABLE I TYPE OF INDEX DISEASE BY SEVERITY AND BY SEX

Type	Severity										
	Post cardiac surgery		Mild		Moderate		Severe		Total		No (%)
Sex	M	F	M	F	M	F	M	F	M	F	Total
VSD	3	8	144	120	18	19	6	6	171	153	324 (38.4)
ASD	16	14	43	58	39	41	0	1	98	114	212 (25.1)
PDA	11	27	12	57	4	16	1	0	28	100	128 (15.2)
PS	0	0	13	12	13	12	2	2	28	26	54 (6.4)
TOF	12	2	0	0	4	2	4	7	20	11	31 (3.7)
Others	9	7	16	7	18	22	8	8	51	44	95 (11.3)
Total	51	58	228	254	96	112	21	24	396	448	
No (%)	109 (12.9)		482 (57.1)		208 (24.6)		45 (5.3)				844 (100.0)

VSD = ventricular septal defect; ASD = atrial septal defect; PDA = patent ductus arteriosus; PS = pulmonary stenosis; TOF = tetralogy of Fallot; M = male; F = female

TABLE II COURSE OF CHD AFTER GRADUATION

Severity	No. of case	Course						
		Operated	Spontaneous closing	Fair	No change	Deterioration	Alive no detail information	Death
		No. (%)	No. (%)	No. (%)	No. (%)	No. (%)	No. (%)	No. (%)
Post-cardiac surgery	107	3 (2.8)	— (—)	14 (13.1)	76 (71.0)	0 (—)	14 (13.1)	0 (—)
Mild	465	47 (10.1)	14 (3.0)	61 (13.1)	289 (62.2)	7 (1.5)	43 (9.2)	4 (0.9)
Moderate	203	74 (36.5)	1 (0.5)	20 (9.9)	87 (42.9)	6 (3.0)	9 (4.4)	6 (3.0)
Severe	44	19 (43.2)	0 (—)	6 (13.6)	9 (20.5)	3 (6.8)	0 (—)	7 (15.9)
Total	819	143 (17.5)	15 (1.8)	101 (12.3)	461 (56.3)	16 (2.0)	66 (8.1)	17 (2.1)

Average age: 24.3 years as of March 31, 1978

findings.

The criteria used are as follows:

Mild: Neither X-ray nor ECG shows evidence of cardiomegaly.

Moderate: Either X-ray or ECG shows evidence of cardiomegaly. In the case of the cyanotic types, the degree of cyanosis is mild or moderate.

Severe: Either X-ray or ECG shows evidence of advanced cardiomegaly.

In the case of cyanotic types, the degree of cyanosis is severe.

X-ray Criteria for Cardiomegaly

Cardiomegaly: $0.65 > \text{CTR} \geq 0.55$

Advanced cardiomegaly: $\text{CTR} \geq 0.65$

ECG Criteria for Cardiomegaly

LVH (left ventricular hypertrophy): Simple high R on left precordial leads is excluded.

RVH (right ventricular hypertrophy): Simple IRBBB is excluded.

CVH (combined ventricular hypertrophy): Suspicious findings are excluded.

Advanced cardiomegaly: LVH, RVH, CVH with ST-T changes.

The method of the study was by questionnaire which was sent and gathered by mail.

RESULTS

Information was finally obtained from 89.2% of the patients which included 14 cases of death already known. Although 91 patients did not return the questionnaire, 66 of them were confirmed to be alive through inquiry at the related city registrar's office. No information was available on the remaining 25 patients (3% of the total subjects).

Table II shows the course of disease of 819 cases excluding the 25 cases about whom nothing

TABLE III ANNUAL MORTALITY RATES OF CONGENITAL HEART DISEASE
(Including deaths related to cardiac surgery)

Severity	Post-cardiac surgery	Mild	Moderate	Severe	Total No. (%)
6 ~ 8	0/449	1/1,594	6/346	10/98	17/2,487 (0.68)
9 ~ 11	3/594	0/1,852	5/385	4/118	12/2,949 (0.41)
12 ~ 14	3/655	2/2,386	5/534	10/159	20/3,734 (0.54)
15 ~ 17	3/663	3/2,228	6/552	7/143	19/3,586 (0.53)
18 ~ 20	0/513	3/1,340	2/510	4/123	9/2,486 (0.36)
21 ~ 23	0/376	1/994	2/371	1/62	4/1,803 (0.22)
24 ~ 26	0/163	0/561	2/206	1/24	3/954 (0.31)
27 ~ 29	0/56	0/233	0/70	0/14	0/373 (—)
30 ~ 32	0/11	0/52	0/9	2/5	2/77 (2.60)
Total No. (%)	9/3,480 (0.26)	10/11,240 (0.09)	28/2,983 (0.94)	39/746 (5.22)	86/18,449 (0.47)

No. of death/No. of subject (April, 1962–March, 1978)

was known after their graduation. The average age at the time of this study was 24.3 years, ranging from 20 to 35 years.

Spontaneous closing was encountered in 15 patients (1.8%). There were 12 cases of mild ventricular septal defect (VSD), one case of moderate VSD and 2 of mild patent ductus arteriosus (PDA). Specialists in cardiology announced these patients' cardiac lesions nearly closed or spontaneously closed.

Table III shows mortality rates by age and by severity of index diseases. The leading cause of death was cardiac surgery being encountered in 37 cases followed by sudden death in 14 cases, chronic congestive heart failure in 14, infective endocarditis (IE) in 7, central nervous system complication in 4, accidents in 4, anoxic spell in 2, diseases other than heart disease in 2, and unknown causes in 2. Deaths related to cardiac surgery, sudden episodes and chronic congestive heart failure were most prevalent among severe cases and less among moderate cases. Deaths due to IE were rather prevalent among mild cases.

DISCUSSION

Because the subjects of the present study consists of the cardiac patients being found through our heart disease program for elementary school children and junior high school students covering almost all of Osaka City, it can be said that the present study is unique for its minimum bias. The problem is that the study was mainly done by questionnaire without medical examina-

tions performed by the program's staff.

One hundred and forty patients underwent their first cardiac surgery after leaving the program. One hundred and seven patients had already had surgery before their graduation. Therefore, a total of 247 patients had gone through cardiac surgery at the time of the study. This constitutes 31% of the total subjects still alive. Among the 558 patients who had not undergone cardiac surgery, it is speculated that almost 130 cases (16% of the living subjects) would benefit from cardiac surgery if almost all of the moderate cases and almost half of the severe cases are operable.¹¹ The above 31% and 16% are almost similar to the recent rate of surgery performed on cardiac patients. Actually the rate of cases having had cardiac surgery has increased to 40 or 50% among children and students under the heart disease program of Osaka City.

Our present data showed that 12 cases of mild VSD (0.8% per annum), one case of moderate VSD (0.5% per annum) and 2 cases of mild PDA (0.6% per annum) appeared with their cardiac lesions nearly closed or closed. Judging from the fact that only 44% of the subjects had any medical examinations after graduation (the rate is 50% even including the cases which appeared for our medical examination after the questionnaire was received), the healing rate should be higher. This speculation is compatible with the other authors' reports.¹⁻⁴

The mortality rate shown in Table III was calculated from the data of graduates and the

children and students under our heart disease program. The rate is 14 times as high as the age corrected rate for general population for those under 14 years of age, 6 times as high as the age corrected rate for general population for those between 15 and 20 years of age, and 3 times as high as the age corrected rate for general population for those older than 21 years of age. The mortality rate for CHD cases showed a decreasing tendency as age advanced. The mortality rate for those with mild cases of cardiac lesions is 3 times higher than the general population. It is 13 times as high among moderate cases and 77 times as high among severe cases when compared to the general population. The rate for post surgical cases is 3.7 times as high as the age corrected mortality rate for the general population.

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