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Congenital heart disease in adults and its problems

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ABSTRACT There were 40 adult congenital heart disease (CHD) patients seen in the Cardiology Division during 1 year (February 1993 - February 1994). The most frequently seen defect was atrial septal defect; however there were also cases with patent ductus arteriosus, pulmonary stenosis, ventricular septal defect, and tetralogy of Fallot. Hemodynamic disorder, serious hindrance to education achievement, and occupational threat due to limited physical capabilities as well as malnutrition are some of prominent issues to be closely anticipated. In general the older the patients the more serious hemodynamic disorder they suffer due CHD. It is worth thinking how to improve the quality of life of CHD patients who succeed to live their adult lives and minimize any possible fatal complication risks. **[Paediatr Indones 2001; 41: 237-240]**

***Keywords**: congenital heart disease, adult, congestive heart failure*

In developing countries, including Indonesia, a large number of patients with congenital heart disease (CHD) are left unoperated due to a variety of causes. This makes most of them live their lives as true natural survivals with various complications and poor quality of life. Some of them even deceased in early age, or have their medical attention when they are already in adult age. Under such circumstances, the prognosis and problems will be totally different from patients with CHD diagnosed in early age.

In developed countries CHD is seldom found among adults as most patients have either palliative or total corrective surgery in childhood. Some of them will have special problems in regard with adult cardiology, e.g., (1) the presence of residual anatomic and hemodynamic disorders due to heart defect such as conduction defect in transposition of the great arteries (TGA), aortic regurgitation in ventricular septal defects (VSD), or non-cardiovascular residua such as mental retardation, or (2) sequels due to unavoidable anatomic and hemodynamic consequences of surgery such as conduction defect due to cut during surgery, pulmonary regurgitation due to infundibulum reparation in tetralogy of Fallot (TF), and prosthetic material capable of inducing thromboemboli or endocarditis.[1,2]

The phenomena in both developing and developed countries encourage further studies on CHD among adults for assuring better treatment and determining priorities in cardiologic services. We reviewed the clinical features of patients with CHD older than 12 years of age who had never undergone surgery.

Methods

We reviewed all medical records of patients hospitalized at Soetomo General Hospital, Surabaya, from February 1993 until February 1994. Nomenclature and criteria for Diagnosis of Diseases of the Heart and Great Vessels - The Criteria Committee of the New York Association 1979 were applied. We defined adult as patients older than 12 years of age. Age when initial symptoms occurred

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and time at the first assessment for heart disease were recorded. Malnutrition was defined as body weight <90% of the normal NCHS chart. Hemoglobin level was measured by means of Sahli method. Polycythemia was identified when hematocrit exceeded 65% with hemoglobin level of more than 18g/dl. Pulmonary hypertension was judged when auscultation revealed prominent P2, chest X-ray showed pulmonary arterial dilatation with narrowing of distal segments, echocardiogram showed negative EF slope, midsystolic notching, and absence of an A wave. Cardiomegaly was classified according to cardio-thoracic ratio (CTR) as follows: mild (CTR 51-55%), moderate (CTR 56-60%) and severe (CTR >60%). Level of education was the highest formal attended education. Hemodynamic status was estimated to be mild when there was no complaints, moderate when there were complaints after exertion, and severe when there were always complaints even at rest.

Results and discussion

There were 46 adult congenital heart disease patients observed during the study, but only 40 of them could be analyzed. Twenty-four (60%) patients were males and 16 (40%) females. None of them had undergone any surgery. Based on types of CHD, it was found that secundum atrial septal defect (ASD) was the most frequently seen, i.e., 21 out of the 40 patients; their age range was 18-30 years old. All of them had cardiac failure that initially presented when they were more than 18 years old. They had been practically asymptomatic and enjoyed their school years. On examination 14 of them had cardiac failure and malnutrition. Electrocardiogram showed typical features, i.e., right atrial abnormalities, right axis deviation, and incomplete right bundle branch block. Cardiomegaly with CTR >50% and pulmonary hypertension was also found in all cases, no mitral valve abnormalities were detected on echocardiograph. Cardiac catheterization was done in 6 patients, confirming that all suffered from severe pulmonal hypertension.

Secundum ASD is a type of CHD commonly found during childhood and the patients are frequently able to live their adulthood. This is due to very low-pressure gradient between left and right atrium causing blood pressure and flow to lungs to run slowly. Complaints are clearly identified in adulthood, where dysrhythmia, pulmonary hypertension, or decreased left ventricle distensibility due to systemic hypertension or coronary insufficiency develop. After 40 years of age, the life expectancy of such patients is 50%.[3,4]

Isolated VSD was found in 8 patients aged between 12-18 years. The symptoms usually appeared before the age of 12 years and the patients were aware of the disease during childhood. They went to elementary schools, had some degree of malnutrition, and moderate to severe hemodynamic changes and cardiomegaly with severe pulmonary hypertension in all patients as confirmed by cardiac catheterization. Two patients also showed aortic regurgitation. The echocardiography showed moderate - non-restrictive defect in all cases.

Isolated VSD is a type of CHD commonly found among children. Hemodynamic disorders depend on defect location and size, pulmonary vascular resistance, and associated lesions. In isolated VSD, the presentation in adults can be small restrictive defect. Most small perimembranous VSD will spontaneously close at some time before 6 years old. This disorder is easy to be detected due to harsh pansystolic murmur in left lower sternal border. Some patients with VSD may develop complications including endocarditis and aortic regurgitation; the latter is usually found in doubly committed subpulmonic VSD. Muscular VSD is seldom found among adults because almost all have closed spontaneously in childhood. Large VSD may develop into Eisenmenger complex.[5,6]

Tetralogy of Fallot (TOF) was found in 4 female patients aged between 12-18 years and hospitalized due to cyanotic spells and hemopthysis. The major complaints were cyanosis of the lips and fingertips, especially following exercise. All patients with TOF actually had symptoms before the age of 6-8 years. All showed polycythemia with the hemoglobin level of more than 18g/dl and hematocrit between 60%-75%. On chest X-ray no cardiomegaly was observed, but all showed a typical 'couer en sabout' appearance with collaterals in lungs. Echocardiograms confirmed that all patients had moderate pulmonary stenosis.

More than 3% of the patients with TOF can live their adult lives by establishment of cardiopulmonary collateral circulation, or if the pulmonary stenosis is

of mild degree, or if the duct remains open. Polycythemia, relative anemia, and disorders of hemostatic factors become major problems associated with chronic hypoxia. Stroke and cerebral abscess may develop at any time in unoperated TOF. When associated with systemic hypertension or aortic calcification, it will generate biventricular failure that makes pulmonary blood flow decrease and causes the patients' condition get worse.[5,7,8]

Patent ductus arteriosus (PDA) was found in 5 patients (3 males and 2 females) aged between 12-18 years. All had their complaints when they were less than 12 years old. All patients showed malnutrition and moderate cardiomegaly. In 2 patients the duct was successfully closed by 'double umbrella closure'.

PDA is commonly found among babies and children; depending on the severity of the disease, patients may be totally asymptomatic or show frank cardiac failure with failure to thrive. Early death is usually due to cardiac failure, endocarditis, calcification and rupture of the duct. Small PDA only causes mild hemodynamic disorder and may close spontaneously.[5,9,10]

Valvular pulmonary stenosis was found in 2 patients - 1 male and 1 female aged between 30-36 years old. They did not have real complaints. Incidentally it was found when they were 18-23 years old and had health examinations for higher school admission. Light cardiomegaly was observed with normal pulmonary vascularity. Interventricular septum and pulmonary valve as well as tricuspid valve were thinckened. Symptoms in patients with pulmonary stenosis depends on the degree of obstruction, progressiveness, and RV adaptation ability. In general, but not always, the older the patient the higher the degree of obstruction. It is because of subpulmonal secondary hypertrophy or PV calcification. Death in such patients usually due to RV failure or endocarditis [1,5,11,12]

It appears that the older the patient, the more severe were the symptoms. See **Table 1**. Only 2 out of 12 patients in 13-18 year age group showed severe hemodynamic problem, in contrast to 20 out of 24 patients in 19-30 year age group and both patients in those older than 31 years old.

TABLE 1. RELATIONSHIP BETWEEN AGE AND HEMODYNAMIC DISORDERS

	Hemodynamic disorder			
Age (yr)	Mild	Moderate	Severe	Total
13 –	2	8	2	12
19 -	1	3	20	24
31-	1	1	2	4
Total	4	12	24	40

Table 2 shows that the younger the symptoms appeared, the more severe the clinical manifestations. To some extent it should be related to the severity of the defect; the more severe the defect the earlier the symtomatology. For those who had symptoms before the age of 12 years, 8 out of 12 had severe hemodynamic disorder while of 22 patients who had their first symptom at the age of 13-18 years only 33 showed severe disorder.

This study identified a number of CHD types commonly found among babies and children who are mostly capable of living their adult lives (ASD, PDA, PS). Besides, there are some CHD types customarily found among babies and children who are barely able to live their adult life. Hemodynamic disorder, serious hindrance to education achievement, and occupational threat due to limited physical capabilities as well as malnutrition status are some of prominent issues to be closely anticipated. The older the patients the more serious hemodynamic disorder they suffering from CHD. The earlier the symptom is identified, the faster to asses that one is suffering from CHD. It is worth thinking how to improve the living quality of CHD patients who succeed to live their adult lives and minimize any possible fatal complication risks (infection, malnutrition, anemia, water and electrolyte imbalance.

TABLE 2. RELATIONSHIP BETWEEN AGE AT THE FIRST SYMPTOMS AND HEMODYNAMIC DISORDER

	Hemodynamic disorder			
Age at the first symptoms appears (yr)	Mild	Moderate	Sever	Total
<12	1	3	8	12
13 - 18	3	6	13	22
> 18	0	3	3	6
Total	4	12	24	40

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