

Clinical Profile of Pediatric Patients with Acyanotic Congenital Heart Disease in Dr. Soetomo General Hospital Surabaya

Kartika Hardiyani¹, Taufiq Hidayat², Alit Utamayasa², Mahrus Abdur Rahman², Teddy Ontoseno²

¹Resident, ²Professor, Department of Child Health, Faculty of Medicine, Universitas Airlangga / Dr. Soetomo General Hospital, Surabaya, Indonesia

Abstract

Congenital Heart Diseases (CHD) are classified as Acyanotic or Cyanotic CHD. Prevalence of CHD according to various studies is 8-10 per 1000 live births worldwide, 10% of cases became infant mortality in developing countries. This research aims to study the characteristics and clinical presentations in children with acyanotic CHD in Dr. Soetomo General Hospital. A retrospective observational study from children with acyanotic type of CHD from medical record outpatient clinic in the Division of Neonatology and Pediatric Ward at Dr. Soetomo General Hospital from June to December 2016. Age, sex, heart defect, comorbid and nutritional status were taken from the medical record. The most dominant clinical profile was female with a mean age of 4 years. Atrial Septal Defect was the most common type of left-to-right shunt, followed by Ventricular Septal Defect, and Patent Ductus Arteriosus. Pulmonary Stenosis was the most common type of obstructive lesion, followed by Aortic Stenosis. Complications of acyanotic CHD were dominated by recurrent respiratory tract infections followed by growth and developmental disorder, and heart failure. The most common nutritional status was moderate malnutrition.

Key Words: *Acyanotic, CHD, ASD, VSD, clinical profile.*

Introduction

Congenital Heart Disease (CHD) is a disease marked with abnormalities of the heart structure or cardiac circulation function that is present at birth.¹ CHD is the most common congenital abnormality, almost 1/3 of cases of congenital abnormalities are cases of congenital heart disease.² The prevalence of CHD worldwide ranges from 6-10 per 1.000 births. The distribution depends on the demographics. Currently, of the 220 million population of Indonesia, it is calculated that there are 6.600.000 newborn babies and 48.800 of them are diagnosed with CHD.³

CHD is classified into 2 groups, namely acyanotic and cyanotic congenital heart disease.⁴ Cyanotic CHD is more complex and is characterized by cyanosis due to a right-to-left shunt so that blood from systemic veins that contain low oxygen will return to the systemic circulation. This acyanotic CHD has no symptoms or signs of cyanosis, but there was a left-to-right shunt or ventricular outlet obstruction.^{3,4} The number of patients with cyanotic CHD is much greater than that of cyanotic

by 3-4 times, but cyanotic CHD causes higher morbidity and mortality than acyanotic.³

The incidence of CHD is reported to be around 8-10 babies out of 1.000 live births and 30% of them have developed symptoms in the first weeks of life. If not detected early and not treated properly, 50% of deaths will occur in the first month of life. In developed countries, almost all types of CHD have been detected in infancy even at the age of less than 1 month, whereas in developing countries many are only detected after the child is older, so that in some severe types of CHD many children may have died before being diagnosed. In certain types of CHD, early recognition and diagnosis are needed so that the necessary treatment and surgery can be given immediately. To improve services in Indonesia, apart from the provision of funds and an adequate pediatric cardiology service center, it is also necessary to have early detection capabilities for CHD and knowledge of optimal referral times by general practitioners who are first dealing with patients.

Materials And Methods

This study uses a descriptive design. The research data were taken from all medical records of patients with Congenital Heart Disease in the Division of Neonatology and Pediatric Ward of Dr. Soetomo General Hospital Surabaya from January 1, 2016, to December 31, 2016. Incomplete medical records were excluded from this study. The sampling method is a total sampling. The data were entered into the Statistical Software Program for Social Science (SPSS) and analyzed using the chi-square and t-test.

Results and Discussion

From 355 children with acyanotic CHD, 158 incomplete medical records were excluded. The remaining 198 children were predominantly female with 127 female subjects (64.1%) and the mean age was 4.0 years old (SD 3.2). The characteristics of the research subjects are described in table 1.

Table 1 Level of Knowledge regarding computer ergonomics among desktop users N=130

Level of Knowledge	Actual Range of Score	f (%)
Very Good	28-35	4(3.1)
Good	23-27	35 (26.9)
Average	18-22	50 (38.5)
Below Average	0-17	41(31.5)

Table 1: Subjects characteristics

Characteristics	Case n (%)
Age	
· 1 year old	33 (16.7)
· 2 years old	36 (18.2)
· 3 years old	31 (15.7)
· 4 years old	31 (15.7)
· 5 years old	25 (12.6)
· 6 years old	18 (9.1)
· 7 years old	11 (5.6)
· 8 years old	1 (0.5)
· 9 years old	3 (1.5)
· 10 years old	1 (0.5)
· 12 years old	1 (0.5)
· 13 years old	1 (0.5)
· 14 years old	1 (0.5)
· 15 years old	1 (0.5)
· 17 years old	2 (1)
· 26 years old	2 (1)
Sex	
· Female	127 (64.1)
· Male	71 (35.9)
Nutritional Status	
· Good nutritional status	94 (47.5)

Cont... Table 1: Subjects characteristics

·	Moderate malnutrition	95 (48)
·	Severe malnutrition	9 (4.5)
Heart defect		
·	Atrial Septal Defect	97 (49)
·	Ventricular Septal Defect	72 (36.4)
·	Patent Ductus Arteriosus	22 (11.1)
·	Pulmonary Stenosis	4 (2)
·	Aortic Stenosis	3 (1.5)
Complication		
·	Recurrent respiratory tract infections	68 (34.5)
·	Growth and developmental disorder	48 (24.3)
·	Heart failure	37 (18.7)
·	Asymptomatic	45 (22.5)

Atrial Septal Defect (ASD) was the most common type of left to right shunt, seen in 97 cases (49.0%). ASDs are associated with Mendelian inheritance, aneuploidy, transcription errors, mutations, and maternal exposures. Besides, ASDs have been associated with familial genetic disorders and conduction defects. Transcription factors important during the atrial septation include GATA4, NKX2-5, and TBX5. More recent epidemiologic data suggest that ASDs occur in 1.6 per 1,000 live births.⁵ Congenital heart disease is diagnosed more commonly in patients in developed countries who have higher incomes.⁶ This explains ASD as the most common type of left-to-right shunt. Ventricular Septal Defect (VSD) seen in 72 cases (36.4%) was the next most common cause of left-to-right shunt. VSD develops when there is a developmental abnormality or an interruption of the interventricular septum formation during the complex embryologic heart morphogenesis. VSDs are frequently isolated; however, they can occur in association with other congenital heart defects such as atrial septal defects, patent ductus arteriosus, right aortic arch, and pulmonic stenosis. Several genetic factors have been identified to cause VSD including chromosomal, a single gene, and polygenic inheritance.^{7,8} Pinto *et al.*⁹ stated that isolated VSD accounts for 37% of all congenital heart disease in children. The incidence of isolated VSD is about 0.3% of newborns. Because as many as 90% may eventually close spontaneously; the incidence is significantly lower in adults. VSDs have no gender predilection. The percentage of each type is presented in the pathophysiology section. This is in accordance with

the findings in this study. The third most common cause of left-to-right shunt is Patent Ductus Arteriosus (PDA) 22 cases (11.1%). Iwashima *et al.*¹⁰, stated that in at least 10% of other congenital heart disorders, a patent ductus may be present. This statement supports the findings of this study.

Pulmonary Stenosis (PS) was the most common type of obstructive lesion seen in 4 cases (2%), followed by 3 cases Aortic Stenosis (AS) (1.5%). Samánek *et al.*¹¹ dan Stephensen *et al.*¹² stated that pulmonic stenosis accounts for 7%-10% of congenital heart diseases. There is a slight female predominance, and 2% of familial occurrences are without a genetic cause.¹³ The differences in the findings of this study could be due to different research methods. Differences could also be caused by differences in the number and characteristics of the study samples in which the research conducted by Samánek *et al.*¹¹ has subjects of 779 children and were examined at least four times during infancy and at the age of three and four years. A study by Hoffman *et al.*¹⁴ concluded that aortic stenosis accounts for approximately 3–6% of congenital heart defects. This is consistent with the findings in this study. The differences that arise could be caused by differences in the number of samples where Hoffman *et al.*¹⁴ have as many as 19,044 children as research subjects.

Complications of acyanotic CHD were dominated by recurrent respiratory tract infections with 68 cases (34.5%). Some kind of CHD which leads to increased

pulmonary flow (e.g. VSD, PDA) or CHD with desaturation (e.g. TOF, DORV, truncus arteriosus) carries a higher risk of recurrent respiratory tract infections and increased frequency of hospitalizations.¹⁵ This supports recurrent respiratory tract infections as the most common complication of CHD. The second most common complication is growth and developmental disorder with as much as 48 cases (24.3%). A study by Mehrizi and Drash¹⁶ stated that patients with ASD or PS were more likely to develop growth retardation. This is in contrast to the findings in this study where cumulatively, 51% of patients experienced ASD or PS. This difference could be because the research by Mehrizi and Drash¹⁶ was carried out in 1962 where perhaps advanced nutrition feeding was still inadequate. Heart failure is the third most common complication in CHD with 37 cases (18.7%). Heart failure is more common in adult CHD.¹⁷ In a Dutch registry of 10,808 CHD patients followed for 21 years (median), the incidence of HF-admissions was 1.2 per 1,000 patient and the median age at first HF-admission was 46.7 years.¹⁸ The most common CHD induced heart failure route is purely acquired and mechanical with no genetic element. This includes incomplete or palliative correction of a lesion leading to a chronic state of hemodynamic stress and subsequent heart failure.¹⁹ The other route is congenital malformation and cardiomyopathy and reactivation of fetal genes that are in mutated pathways in the setting of CHD.¹⁹ This supports the findings in this study which states that heart failure is not the most common complication of CHD.

The most common nutritional status was moderate malnutrition in 95 cases (48%). Malnutrition occurs among children with congenital heart disease, irrespective of the nature of the cardiac defect, and the presence or absence of cyanosis.²⁰ Children with congenital heart disease (CHD) are more prone to malnutrition for several reasons including decreased energy intake, increased energy requirements, or both.²¹ This study and other studies show that patients with CHD are more prone to malnutrition.

The limitation of this study is that a large number of samples were excluded due to incomplete data on patient characteristics. Further research is needed to determine the main cause of a particular defect can be the most common type of left-to-right shunt or obstructive lesion.

Conclusion and Acknowledgement

Atrial Septal Defect (ASD) was the most common type of left-to-right shunt, followed by Ventricular Septal Defect (VSD), and Patent Ductus Arteriosus (PDA). Pulmonary Stenosis (PS) was the most common type of obstructive lesion, followed by Aortic Stenosis (AS). Complications of acyanotic CHD were dominated by recurrent respiratory tract infections followed by growth and developmental disorder, and heart failure. The most common nutritional status was moderate malnutrition.

We wish to thank our staff in the Division of Neonatology and Pediatric Ward, the doctors, nurses, and the administrator for granting us the permission and necessary support to conduct our research.

Conflict of Interest: Nil

Source of Funding: Not Applicable

Ethical Clearance: Given by Dr. Soetomo General Hospital, Surabaya, Indonesia.

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