Characteristics of Cyanotic Congenital Heart Disease at Dr. Soetomo General Hospital Surabaya

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Abstract

Background. *Cyanotic congenital heart disease (CHD) is an abnormality of the heart that causes blood with low oxygen content to circulate directly to the body. Objective. To evaluate the profile of cyanotic congenital heart disease at Dr. Soetomo General Hospital Surabaya.*
Methods. *This is a descriptive study using medical records of cyanotic CHD patients aged ≤18 years old from June 2018 – May 2020. Gender, age, nutritional status, sign and symptom, type of cyanotic CHD, complication and treatment were taken as the variables.*
Result. *Out of 116 patients in this study, the majority were male (58.62%). Most patients first diagnosed at the age of 0 – <1 year (76.72%) with normal nutritional status (51.72%). Tetralogy of Fallot (TOF) is the most common type (42.24%). Murmur was the most common sign and symptom (92.24%). Frequently observed complications were pneumonia (37.07%).*
Conclusion. *Patients with cyanotic CHD are mostly male, diagnosed at the age of 0 – <1 year with normal nutritional status. TOF is the most common type. Most patients present with murmurs. Predominant complication is pneumonia.*

Keywords: *Congenital Heart Disease, Cyanotic, Pediatric*

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Profil Penyakit Jantung Bawaan Sianotik di Rumah Sakit Umum Daerah Dr. Soetomo Surabaya

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Abstrak

Pendahuluan. *Penyakit jantung bawaan (PJB) sianotik merupakan suatu kelainan struktur maupun fungsi jantung yang menyebabkan darah dengan kadar oksigen rendah kembali beredar ke sistemik.*
Tujuan. Untuk mengetahui profil penyakit jantung bawaan sianotik di RSUD Dr. Soetomo Surabaya.
Metode. *Penelitian ini adalah penelitian deskriptif menggunakan rekam medik pasien usia ≤18 tahun periode Juni 2018 – Mei 2020. Jenis kelamin, usia, status gizi, tanda dan gejala klinis, tipe PJB sianotik, komplikasi dan tatalaksana merupakan variabel penelitian ini.*
Hasil. *Dari 116 pasien PJB sianotik yang menjadi subjek penelitian ini, didapatkan mayoritas pasien laki-laki (58.62%) dan terdiagnosis pertama kali pada usia 0 – <1 tahun (76.72%). Status gizi terbanyak adalah gizi baik (51.72%). Tetralogy of Fallot (TOF) merupakan jenis PJB sianotik yang paling banyak ditemukan (42.24%). Tanda dan gejala klinis yang terbanyak adalah murmur (92.24%) dan komplikasi terbanyak adalah pneumonia (37.07%).*
Kesimpulan. *Pasien PJB sianotik kebanyakan laki-laki, berstatus gizi baik, terdiagnosis pertama kali pada usia 0 – <1 tahun. TOF merupakan jenis terbanyak. Tanda dan gejala klinis yang terbanyak adalah murmur. Pneumonia adalah komplikasi yang dominan.*

Kata kunci: *Penyakit Jantung Bawaan, Sianotik, Anak*

Introduction

Congenital heart disease (CHD) is defined as a disorder in the structure or function of the cardiovascular system that appears since birth even though it may be detected later in life.[1] Congenital heart disease (CHD) occurs in about 8-10 babies out of 1000 live births.[2] In Indonesia, the incidence of CHD is estimated to be 9 in 1000 live births every year.[3] Cyanotic CHD has a lower incidence than cyanotic CHD, with a ratio of 1:4 events. However, cyanotic CHD causes higher morbidity and mortality than cyanotic CHD.[4]

Congenital heart disease is generally classified into cyanotic and acyanotic CHD. In cyanotic CHD, there is a shunt of blood from the right heart chamber to the left, resulting in a mixture of blood from the systemic veins with lower oxygen levels and blood from the pulmonary veins with higher oxygen levels. Mixed blood with inadequate oxygen levels will then circulate into the systemic circulation and cause cyanosis.[5] The most common type of cyanotic CHD is Tetralogy of Fallot (ToF).[4,5]

Previous studies on the profile of CHD in Indonesia still provide mixed results and could not describe specifically the profile of cyanotic CHD. In addition, signs and symptoms of patients with cyanotic CHD do not always appear immediately after birth, but some appear later in life according to the severity of the lesion and the obstruction that occurs.[6] This results in a delayed diagnosis of cyanotic CHD, which can lead to a significant increase in morbidity and mortality.[7]

This delay in diagnosis and management can increase the risk of morbidity and mortality as well as the risk of developing complications in children with cyanotic CHD. Therefore, it is important to know the clinical

profile of cyanotic CHD. This study aims to determine the profile of patients with cyanotic congenital heart disease in RSUD Dr. Soetomo Surabaya.

Method

We conducted a descriptive study that examines the profile of cyanotic CHD patients. Data was gained from the Medical Records Installation Unit at RSUD Dr. Soetomo and the Department / SMF of Children's Health Sciences RSUD Dr. Soetomo Surabaya using medical record data of all patients with cyanotic congenital heart disease aged below 18 years old during June 2018 to May 2020. Data will be excluded if the medical records were incomplete or damaged.

Nutritional status assessment in this study used the weight/height index and the 2006 WHO growth chart for children aged 0 – 5 years. Assessment by the percentage of actual weight / ideal body weight according to actual height and CDC 2000 growth chart is used for children aged >5 – 18 years. The nutritional status of the children was then categorized according to 2006 WHO criteria for 0-5 years of age and Waterlow's criteria for >5 – 18 years of age.[8]

The data obtained were processed and analyzed descriptively and then presented in the form of a frequency distribution table. Data processing regarding the nutritional status of patients was carried out using the WHO Anthro Survey Analyser and PediTools software.[9] This research has been reviewed and obtained research permission from the Research Ethics Committee of RSUD Dr. Soetomo, Surabaya.

Result

The subjects in this study were 116 patients with cyanotic congenital heart disease at Dr. Soetomo in the period June 2018 – May 2020. Most of the subjects were male (58.62%), diagnosed for the first time in the infant age group (76.72%), and had normal nutritional status (51.72%) [Table 1]. The most common type of cyanotic CHD was Tetralogy of Fallot (42.24%), followed by Transposition of the Great Arteries (24.14%) and pulmonary atresia (18.97%). Other types found were tricuspid atresia (8.62%), Ebstein anomaly (6.03%), truncus arteriosus (6.03%), Total Anomalous Pulmonary Venous Return (3.45%), and single ventricle (2.59 %) [Table 2].

Table 1. General Characteristics of The Patient

Characteristics	n	%
Gender		
Male	68	58,62
Female	48	41,38
Age		
Infant (0 – < 1 years old)	89	76,72
Toddler (1 – < 5 years old)	23	19,83
Child (5 – < 10 years old)	4	3,45
Teen (10 – 18 years old)	0	0,00
Nutritional Status		
Severe malnutrition	32	27,59
Malnutrition	23	19,83
Normal	60	51,72
Overweight	1	0,86
Obesity	0	0,00

Table 2. Distribution of Cyanotic CHD Types

Types of Cyanotic CHD	n	%
Tetralogy of Fallot (TOF)	49	42,24
Transposition of the Great Arteries (TGA)	28	24,14
Pulmonary Atresia	22	18,97
Tricuspid Atresia	10	8,62
Ebstein Anomaly	7	6,03
Truncus Arteriosus	7	6,03
Total Anomalous Pulmonary Venous Return (TAPVR)	4	3,45
Single Ventricle	3	2,59

The most common signs and symptoms were murmur (92.24%), cyanosis (86.21%), shortness of breath (76.72%), cough (43.10%), and chest retraction (41.38%) [Table 3]. The most common complications were pneumonia (37.07%), followed by heart failure (33.62%), malnutrition (27.59%), respiratory failure, (13.80%) and anemia (12.07%) [Table 4]. The treatment given to patients with cyanotic congenital heart disease at Dr. Soetomo Surabaya was administering medication to 116 patients (100%), interventional cardiology to 22 patients (18.97%), palliative surgery to 10 patients (8.62%), and corrective surgery to 3 patients (2.59%) [Table 5].

Table 3. Distribution of Signs and Symptoms in Patients with Cyanotic CHD

Signs and Symptoms	n	%
Murmurs	107	92,24
Cyanotic	100	86,21
Dyspnea	89	76,72
Cough	50	43,10
Chest retraction	48	41,38
Fever	41	35,34
Rhonchi	25	19,83
Clubbing finger	16	13,80
Seizure	15	12,93
Conjunctival anemic	14	12,07
Diarrhea	10	8,62
Vomiting	9	7,76
Gallop	7	6,03
Wheezing	4	3,45

Table 4. Distribution of Complications in Cyanotic CHD Patients

Complications	n	%
Pneumonia	43	37,07
Heart Failure	39	33,62
Malnutrition	32	27,59
Dyspnea	16	13,80
Anemic	14	12,07
Cyanotic spell	13	11,21
Pulmonary Hypertension	12	10,34
Septic	11	9,48
Acute Kidney Injury	7	6,03
Metabolic Acidosis	5	4,31
Gastroenteritis	4	3,45
Endocarditis	3	2,59
Brain Abscess	2	1,72
Atelectasis	2	1,72

Table 5. Distribution of Management in Patients with Cyanotic CHD

Management	n	%
Medicine	116	100,00
Cardiology Intervention	22	18,97
Palliative Operation	10	8,62
Corrective Operation	3	2,59

Discussion

The majority of cyanotic CHD patients were male, as many as 68 patients (58.62%), while the number of female patients was 48 patients (41.38%). This finding is similar to the study by Varma et al.[10] in India with a sample of 80 cyanotic CHD patients, which also found that most cyanotic CHD patients were male (51.25%). Meanwhile, another study in Egypt with a sample of 50 cyanotic CHD patients also found that the majority of patients were male (78%).[11] Regarding the type of cyanotic CHD, research by Zhao et al.[12] mentioned that the most common types of cyanotic CHD, namely Tetralogy of Fallot (TOF) and Transposition of the Great Arteries (TGA), were more common in male patients. Infants with cyanotic CHD can show clinical manifestations immediately after birth or appear later in life. This is influenced by the type of cyanotic CHD experienced, lesion severity, obstruction severity, and the presence or absence of shunt.[6,13] This study found that most patients with cyanotic CHD were diagnosed for the first time in the infant age group (0 – < 1 year) as many as 89 patients (76.72%).

This phenomenon might happen due to early diagnosis and a good referral system or because most patients have severe lesions that show signs and symptoms early on. Similar study by Meshram and Gajimwar in India also found that most patients were in the <1 year age group (56.28%).[14]

Babies with CHD generally have normal weight and length at birth because their nutritional needs come from the placenta, and there is a shunt in the normal circulation in the womb.[15] In this study, most subjects aged <1 year had normal nutritional status (51.72%). This is similar to a study by Balogun and Omokhodion in Nigeria, which found that 70.6% of cyanotic CHD patients had normal nutritional status while 29.4% were malnourished.[16]

Unmet nutritional needs in children with CHD are due to having difficulty eating and being unable to receive large amounts of food intake.[17] Furthermore, with increasing age, there will be an increase in metabolism and energy requirements.[15] These things explain the occurrence of malnutrition in 32 patients (27.59%) and malnutrition in 23 patients (19.83%) with cyanotic CHD in this study. The occurrence of malnutrition in children with CHD affects the mortality rate, occurrence of infection, and length of stay in the hospital. Therefore, children with CHD need

more attention on their nutritional condition.[15]

The incidence of TOF is estimated at 10% of all CHD events; hence TOF is the most common type of cyanotic CHD.[4] The second most common type of cyanotic CHD is TGA, with a prevalence of about 7% of all CHD events TGA cases.[5] Studies by Zhao et al.[12] in China, Meshram and Gajimwar[14] in India, and Khasawneh et al.[18] in Jordan found that TOF and TGA had the highest prevalence, respectively. This study had the same results as the theory and several previous studies.

Clinical signs and symptoms experienced by patients with cyanotic CHD can be caused by CHD itself or complications experienced by the patient. This study found that the most common clinical signs and symptoms found in cyanotic CHD patients were murmurs (92.24%) and cyanosis (86.21%). Khasawneh et al. also mentioned that murmurs and cyanosis are the most common clinical signs and symptoms and are the main referral indications in cyanotic CHD patients.[18,19]

Cyanosis is a bluish discoloration of the skin and mucous membranes due to an increase in the concentration of reduced hemoglobin in the bloodstream.[20] In patients with cyanotic CHD, a shunt occurs from right to left and there is a mixing of oxygen-rich blood with carbon-dioxide-rich blood so that the blood flowing throughout the body has decreased oxygen saturation, resulting in manifestations of cyanosis in the patient.[21]

Other clinical signs and symptoms found in cyanotic CHD patients were shortness of breath (76.72%), cough (43.10%), and chest retractions (41.38%). Shortness of breath can occur when the child is active because of the increased demand for oxygen and blood pressure in the pulmonary veins.[20] Cough is usually caused by complications such as respiratory infections, heart failure or pulmonary hypertension.[20,21] In addition to coughing, cyanotic CHD patients with left heart failure or significant pulmonary impairment may experience chest tightness and retractions.[20]

CHD can affect the condition of various systems in the body because of the dynamic interaction of the cardiovascular system with various other systems in the body. This can cause complications in children with CHD that can complicate the process of patient care and recovery, affect disease progression, and even affect response to treatment.[22] The most common cyanotic CHD complications found in this study were pneumonia (37.07%), heart failure (33.62%), and malnutrition (27.59%). A previous study by Manopo, Erling, and Adrian in Manado found that pneumonia was the most common complication, with a prevalence of 48.15%.[23]

Children with CHD are prone to infection by Respiratory Syncytial Virus (RSV), so they often experience complications in respiratory disorders such as pneumonia.[24] The pathophysiology of respiratory tract infections in children with CHD varies but is dominated by airway compression, changes in lung function, and increased airway responsiveness.[24] In addition, cyanotic CHD that causes a decrease in blood flow to the lungs, such as TOF, will cause impaired development of structures in the lungs and damage to the respiratory tract mucosa as well as impaired lung immune system so that children are more susceptible to respiratory infections.[25]

Heart failure that often occurs in children with CHD is associated with ventricular dysfunction, increased volume loading, excess pressure on the heart or the great vessels associated with the heart.[26] In addition, the occurrence of myocardial dysfunction due to perfusion disorders causing myocardial ischemia or disturbances in myocardial architecture, heart rhythm disturbances, occurrence of systemic arterial hypertension, and pulmonary hypertension, can also underlie the occurrence of heart failure in patients.[27]

Management of CHD consists of conservative therapy with medication, palliative therapy (interventional cardiology and palliative surgery), and definitive therapy in corrective surgery.[28] This study showed that in all patients with cyanotic CHD were given medical therapy before underwent intervention such as interventional cardiology (18.97%), palliative surgery (8.62%), and corrective surgery (2.59%). Similar to this study, a study in 50 cyanotic CHD patients by Abou-Taleb, Abdelhamid, and Bahkeet in Egypt, also found that 100% of patients were given drugs (PGE1, inotropic, drugs to treat heart failure, vitamin K), antibiotics, and sodium bicarbonate). Moreover, 26% of patients underwent interventional cardiology in the form of Balloon Atrial Septostomy (BAS).[11]

Medication is given as the initial treatment to reduce the clinical signs and manifestations experienced by the patient so that it is given to all patients with cyanotic CHD. When the patient does not respond to medication, palliative therapy can be performed in non-surgical (interventional cardiology) or surgical (palliative surgery) ways.[4] Corrective surgery is carried out as definitive therapy in

children with CHD to correct anatomical heart problems, which should be done as early as possible to prevent distortion of heart growth and pulmonary hypertension due to CHD.[29]

The treatment given to the patient is adjusted to the patient's condition. Babies must be large enough in terms of age, weight, and body length to be able to undergo definitive therapy (corrective surgery) to treat CHD, usually in children aged 1-2 years.28 In this study, most patients were < 1-year-old, so more patients were given medical management, interventional cardiology, or palliative surgery than corrective surgery.

Conclusion

Patients with cyanotic congenital heart disease were more common in males (58.62%), most patients were diagnosed for the first time in the infant age group (0 – <1 year), which was 76.72%, and most of the patients had good nutritional status, namely 60 patients (51.72%). Tetralogy of Fallot (42.24%) was the most common type of cyanotic congenital heart disease at RSUD Dr. Soetomo. The most common clinical signs and symptoms were murmurs (92.24%), and the predominant complication was pneumonia (37.07%). The treatment given to patients with cyanotic congenital heart disease at Dr. Soetomo Surabaya is given medication (100%), interventional cardiology (18.97%), palliative surgery (8.62%), and corrective surgery (2.59%).

Conflict of interest

None declared.

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