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Original Research

# Pulmonary Hypertension in Patient with Acyanotic Congenital Heart Defects in Abdul Wahab Sjahranie General Hospital, Samarinda from 2015-2016

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ABSTRACT

**Background.** Congenital Heart Defects (CHD) is the most common congenital disease that affects 8 out of every 1000 births. CHD can be classified as cyanotic and acyanotic. Acyanotic is the most frequently diagnosed CHD. The most common complication in acyanotic CHD is pulmonary hypertension. **Aims.** This descriptive study was based on medical records of acyanotic CHD patients with pulmonary hypertension in RSUD Abdul Wahab Sjahranie Samarinda during 2015 to 2016 who were diagnosed by cardiologist and confirmed with echocardiography. **Methods.** There were 62 patients diagnosed with acyanotic CHD and pulmonary hypertension which 58% of patients were diagnosed with Atrial Septal Defect (ASD), Ventricular Septal Defect (VSD) (21%), and Patent Ductus Arteriosus (PDA) (21%). Defect sizes distribution were mostly large (66%), then medium (29%), and small (5%). There were 56% patients diagnosed before 18 years old and 44% diagnosed after they reached 18 years old. There were 68% female patients and 32% male patients. **Results.** Pulmonary hypertension severity distribution were mild (52%), moderate (26%), and severe (22%). The most diagnosed acyanotic CHD was ASD. Defect size in acyanotic CHD was mostly large, and mostly found in female patients. The severity of pulmonary hypertension was mostly mild.

## Introduction

Birth defect is the leading cause of infants' death and it contributes around 20% of infants' death [1]. Congenital Heart Defects (CHD) is the most common birth defects found in infants [2]. CHD is known as a problem within heart structure or abnormalities in heart circulation, presents at birth due to a failure or disruption of the heart structure development in the early phase of fetal growth [3].

CHD is the most common congenital disease that affects 8 out of every 1000 births or around 40,000 births every year in the United States [2].

Prevalence of neonates born with CHD compared to total birth rate increases each year, accompanied by an increase in adult patients with CHD. According to its clinical manifestations, CHD is divided into acyanotic and cyanotic type.

Cyanotic type is the most common case found in CHD [4]. The three most common incidence of acyanotic CHD are Ventricular Septal Defect (VSD) (30%), Atrial Septal Defect (ASD) (19%), and Patent Ductus Arteriosus (PDA) (19%). This shows that the incidence of acyanotic CHD is way more common than cyanotic CHD with a ratio of acyanotic CHD reaching 79% of all CHD, or about 3 to 4 times more than cyanotic CHD [5].

People who were born with acyanotic CHD are generally more susceptible to disease and have a risk of developing CHD complications such as pulmonary hypertension than people who were born without it [6]. Pulmonary hypertension is a condition when the mean Pulmonary Artery Pressure (mPAP) is >25mmHg at rest or >30mmHg during exercise [7]. Pulmonary hypertension is divided into mild, moderate and severe based on mPAP estimated by hypertension in echocardiography [8]. Pulmonary hypertension often developed in patients with CHD and considered as the most common complication that occurs in patient with acyanotic CHD [9].

According to French National Registry, pulmonary hypertension prevalence reached around 15 cases per million people [10]. In 2012, a report from United Kingdom National Pulmonary Hypertension Audit shows that around 5% of adult CHD patients developed pulmonary hypertension and the estimated pulmonary hypertension cases as a complication of CHD can reach around 1.6 to 12.5 cases per each million population [11]. A research conducted in a Scottish Hospitals in 1986 to 2001 shows a record of 374 patients diagnosed with pulmonary hypertension, and there were a total of 88 patients diagnosed with pulmonary hypertension due to CHD [12].

There is no available data in Indonesia that shows pulmonary hypertension caused by CHD prevalence yet, but a study conducted in Abdul Wahab Sjahranie Hospital in Samarinda from 2013- 2015 shows that pulmonary hypertension was the most common complication of CHD, affected 20% or 18 patients out of 94 patients with CHD [13].

## Methods

This descriptive study were based on medical records of acyanotic CHD patients with pulmonary hypertension in Abdul Wahab Sjahranie General Hospitals in Samarinda during 2015-2016 who were diagnosed by cardiologist and confirmed with echocardiography.

All patients with acyanotic CHD (ASD, VSD, or PDA) who developed pulmonary hypertension and underwent an echocardiography test during 2015-2016 in Abdul Wahab Sjahranie General Hospital were included. Patients with more than one type of CHD were excluded from this study. The variables studied included the type of acyanotic CHD, defect size, age, sex, and pulmonary hypertension severity.

## Results

A total of 267 patients with acyanotic CHD underwent echocardiography test in Abdul Wahab Sjahranie General Hospital during 2015 to 2016, and only 62 patients developed pulmonary hypertension. The distribution of patients across the various type of acyanotic CHD is given in Table 1. More than half of enrolled patients (58%) presented with ASD, followed by VSD (21%), and PDA (21%).

Table 1. Distribution of acyanotic CHD types

| Types | Number of Cases (%) |
|---|---|
| ASD | 36 (58%) |
| VSD | 13 (21%) |
| PDA | 13 (21%) |
| Total | 62 (100%) |

**ASD : Atrium Septal Defect**
**VSD : Ventricle Septal Defect**
**PDA : Patent Ductus Arteriosus**

The defect size of CHD was divided into three groups (Figure 1), small (≤0.5cm), moderate (0.6-1cm), and large (>1cm). The distribution of defect sizes is shown in Table 2, where 41 patients (66%) have large defects, 18 patients (29%) with medium defects, and only 3 patients (5%) have small defects. The largest defect size of acyanotic CHD patient with pulmonary hypertension during 2015 to 2016 was 3.94 cm, while the smallest size was only 0.3cm.

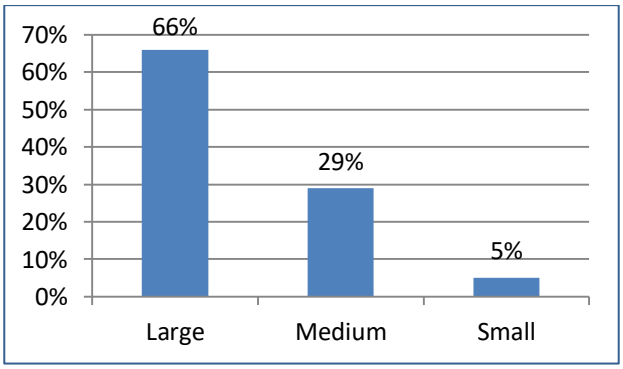


Figure 1. Distribution of size defect

As seen in Table 2, age distribution was predominantly in children (56%) than adults (44%). The youngest patient who was diagnosed with acyanotic CHD and developed pulmonary hypertension was only 2 months old. The oldest patient who enrolled to our hospitals with pulmonary hypertension due to acyanotic CHD was 63 years old.

Table 2. Age distribution

| Age | Number of Cases (%) |
|---|---|
| Child (<18 years old) | 35 (56%) |
| Adult (18≥ years old) | 27 (44%) |
| Total | 62 (100%) |

A significant proportion of CHD patients who developed pulmonary hypertension were female (68%), around 2 times more than male (32%) as seen in Figure 2.

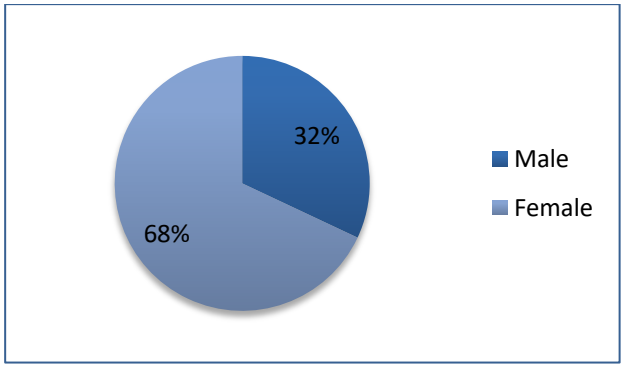


Figure 2. Sex distribution

Pulmonary hypertension severity was divided into (Table 5) mild (mPAP 26-40mmHg), moderate (mPAP 41-55mmHg), and severe (mPAP >55mmHg). Out of 62 cases, half of patients (52%) showed mild pulmonary hypertension, followed by moderate (26%) and severe (22%). The highest mean pulmonary artery pressure registered during our study was 71mmHg.

Table 3. Distribution of pulmonary hypertension severity

| Severity | Number of Cases (%) |
|---|---|
| Mild | 32 (52%) |
| Moderate | 16 (26%) |
| Severe | 14 (22%) |
| Total | 62 (100%) |

## Discussion

This study result showed that pulmonary hypertension mostly developed from a large size defect, which happened in 41 cases (61%). An increase in both vascular resistance and blood flow will result in pulmonary hypertension. The size of the defect in acyanotic CHD has important role in the magnitude of the shunt. A small defect will create a large resistance for the left-to-right shunt. On the contrary, in medium- large defects, smaller resistance was formed. A small resistance will allow a greater blood flow to passes through the defect and will eventually cause a more significant overload compared to the small-sized defects. The greater the pulmonary blood flow, the greater the increases in the mean pulmonary arterial pressure [14,15].

Acyanotic CHD with pulmonary hypertension were found more common in children that adults. Although there are many theories who suggest that pulmonary hypertension as a complication of acyanotic CHD more often found in adulthood, the results in this study showed a slightly different age distribution. This result might be biased by the fact that acyanotic CHD patients, especially those with medium-large defect, usually did not survive due to some difficulty. Most patients with medium to large defects die at young age due to their inability in reaching referral hospitals or inability in covering high expense care, making a lessen number in grown up CHD patients [16,17].

In addition, developments in pediatric cardiology and surgery and have led to not only an improvement in early identification and treatment, but also in preventing the development of pulmonary hypertension in most cases [11].
This study showed a significant proportion of CHD patients who developed pulmonary hypertension was female (68%), around 2 times more than male (32%). A study in Turkey showed a similar sex distribution where the incidence of CHD with pulmonary hypertension usually 2 to 3 times more in female [17].

In addition, a cohort study on patients with pulmonary hypertension performed in France showed more female developed pulmonary hypertension than male, with 67.1% female and only 32.9% male [10]. Another study conducted in Scotland showed distribution of sex in CHD patients who developed pulmonary hypertension was divided into 68% (60 cases) female and 32% (28 cases) male [12]. The difference in distribution among pulmonary hypertension severity can be due to patients' age distribution which is still quite young so that the burden of the shunt is still mild [14].

## Conclusion

The most common type of acyanotic CHD with pulmonary hypertension during 2015 to 2016 was ASD (58%), followed by VSD (21%) and PDA (21%). The most commonly found defect in CHD was large (66%), medium-sized (29%), and small (5%). Most acyanotic CHD patients with pulmonary hypertension were children (56%) and only 44% of them were adults. Acyanotic CHD with pulmonary hypertension was diagnosed more in female (68%), around 2 times more than male (32%). The distribution of pulmonary hypertension severity were mostly consists of mild pulmonary hypertension (52%), moderate (26%), and severe (22%).

## Acknowledgement

There is no conflict of interest.

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