Pulmonary Artery Hypertension in Acyanotic Congenital Heart Disease Underwent Transcatheter Closure at Dr. Soetomo Hospital

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Abstract

Background: Pulmonary artery hypertension (PAH) is common complication of congenital heart disease. Echocardiography before and after transcatheter closure procedure is needed for further evaluation.

Objective: To evaluate the PAH before and after transcatheter closure procedure by echocardiography at Dr. Soetomo Hospital.

Methods: Medical record patients with acyanotic congenital heart disease with PAH and already done transcatheter closure procedure in 2010 – 2014 were reviewed. Data taken were demographic, clinical, and echocardiography. Tricuspid regurgitation pressure gradient (TRPG) was evaluated. Statistical analysis using t test comparative study; P<0.05 was considered significant.

Results: There were 46 patients underwent transcatheter closure, ASD closure 22/46, VSD closure 16/46, PDA closure 8/46 patients. Ten patients with PAH (10/46), 4/10 ASD, 4/10 PDA and 2/10 VSD. Boys were 6/10, median age was 60 (range 4-144 months). Median TRPG before procedure was 32.4 (range 25-43 mmHg). Median TRPG after procedure was 21.5 (range 15-26.9 mmHg). There was significant decreased in PAH after transcatheter closure procedure (P=0.01).

Conclusion: Transcatheter closure procedure in acyanotic congenital heart disease was important for decreasing the pressure gradient of PAH.

Keywords: *Pulmonary artery hypertension, acyanotic congenital heart disease, tricuspid regurgitation, transcatheter closure*

Background

Pulmonary arterial hypertension (PAH) is a common complication of congenital heart disease (CHD), particularly in patients with left-to-right (systemic-to-pulmonary) shunts. Persistent exposure of the pulmonary vasculature to increased blood flow and pressure may result in vascular remodelling and dysfunction. This leads to increased pulmonary vascular resistance (PVR) and, ultimately, to reversal of the shunt and development of Eisenmenger's syndrome.[1] The estimated prevalence of CHD is approximately six to 10 per 1,000 live births and 4–15% of patients with CHD will go on to develop PAH[2,3]. The development of PAH in patients with CHD is associated with increased mortality and high morbidity, reflected in a substantial increase in health service utilisation. While successful early closure of a cardiac defect prevents the development of PAH,have led to a marked decrease in the prevalence of PAH-CHD in western countries, the number of patients with CHD surviving into adulthood has increased.[4,5]

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The interventional approach has become increasingly preferred for the treatment of many congenital heart defects (CHDs), including atrial septal defects (ASDs), ventricular septal defects (VSDs), and patent ductus arteriosus (PDAs). Transcatheter closure of congenital heart defects has been widely accepted as a preferred treatment; however, the high cost of these devices limits their clinical application in some countries.[6] Echocardiography plays crucial role in the safety and efficacy assessment of transcatheter treatment. The advantage of TEE guidance and assistance during the closure procedure is shorter fluoroscopic time. The transcatheter closure of heart defect is nowadays accepted as a safe and effective treatment modality. echocardiography.In the process of selection of eligible defects play crucial role, the closure procedure shoud be performed by TEE guidance and follow-up of closure results by monitoring of TTE. [7]

This study was design to evaluate PAH before and after transcatheter closure procedure by echocardiography at Dr. Soetomo Hospital.

Methods and Materials

The cross sectional study was conducted during January 2010 – Desember 2014 at Division of Cardiology Department of Pediatrics Dr. Soetomo Hospital. Patients were selected based on inclusion and exclusion criteria. The inclusion criteria were 1) patients with acyanotic congenital heart disease 2) underwent transcatheter procedure in Dr. Soetomo Hospital. Patients were excluded if 1) Had multiple congenital anomaly, 2) had a severe infection or severe condition. There were 46 patients acyanotic heart disease diagnosed by echocardiography and underwent the transcatheter closure procedure. The characteristic data including age, sex, chief complaint, physical examination laboratory, chest X-ray and data from echocardiography were collect from medical records. The data taken from echocardiography were include type of congenital defect, size, pressure gradient, and ejection fraction.

A total of ten patients got PAH as complication of congenital heart disease, they were compared pressure gradient before and after trancatheter closure procedure. Statistical Analysis used t- test comparative study, with $P<0.05$ considered significant. All analyses were conducted in SPSS version 21.

Result

During the study period, 46 patients acyanotic congenital heart disease were include in this study. The basic characteristic in this study (table 1) found that sex was dominated by Male (58%), the age of children mostly in >5 (45%) years old and the median age was 5 years. Dyspnea was the most common main complain in our study 73.9%, and followed by recurrent cough 17.3% and fatigue were 8.08%. The atrial septal defect were affected on 22 (47%) patients, ventricular septal defects were 16 (34%) patients and patent ductus arteriosus 8 (17%) patients. The patients were found mostly with malnutrition status (52%), however found well nourish patients (40%), and severe malnutrition (8%). Ten patients had complication pulmonary artery hypertension (PAH). Four patient had PAH with ASD, 4 patient PAH with PDA and two patients less with VSD. Typed Degree of Tricuspid Regurgitation were dominated with mild (90%) and less of moderate (10%).

Table.1. Baseline Characteristics of acyanotic congenital heart disease underwent transcatheter closure

Characteristics	(N=46)
Sex: Female Male	 19 (41%) 27 (58%)
Age (yr): <1 ≥ 1-5 > 5	 10 (21%) 15 (32%) 21 (45%)

Cont... **Table.1. Baseline Characteristics of acyanotic congenital heart disease underwent transcatheter closure**

Diagnosed: ASD VSD PDA	 22 (47%) 16 (34%) 8 (17%)
Main complaint Dyspnea Recurrent cough Fatigue	 34 (73.9%) 8 (17.3%) 4 (8.08%)
Nutrition status Well nourish Malnutrition Severe malnutrition	 18 (40%) 24 (52%) 4 (8%)
Complication PAH Non PAH	 10 (21.7%) 36 (78.3%)
Degree of TR Mild Moderate	 9 (90%) 1 (10%)

Table 2 show median degree of Tricuspid Regurgitation Pressure Gradient (TRPG) before procedure was 32.4 (range 25-43 mmHg). Median TRPG after procedure was 21.5 (range 15-26.9 mmHg). We used t test comparative study to compere degree of TRPG. There was significant decreased in PAH after transcatheter closure procedure (P=0.01) with P< 0.05 considered significant.

Table 2. Median of Tricuspid Regurgitation Pressure Gradient before and after transcatheter procedure

Variable	Before transcateter closure	After transcateter closure	P value*
Median TRPG	32.4 (25-43)	21.5 (15-26.9)	0.01

*p significant < 0.05

Discussion

In this study, the patients who got acyanotic congenital heart disease during January 2010 – Desember 2014 at Division of Cardiology Department of Pediatrics Dr. Soetomo Hospital were 46 patients and most of all was male (58%). Based on study on Kathmandu university of Nepal in 2008 showed that presentation of acyanotic congenital heart disease was dominated with male with male : female ratio = 1.5:1 [8]. The number of age patients was most above than 5 years old (45%) and ASD was the most presentation in our study despite of PDA and VSD. It could be caused by our participants is from tertiary care center as a main referral from all patients in east region of Indonesia. Most of the patients came to our hospital not from earlier age. The former studies are usually passive, in that diagnosis is made in large regional high-quality pediatric cardiology center but relies on referral of patients from local doctors. Thus, if a local physician is comfortable with the management of tiny VSD that patient might not be referred to a center and so not be countered. The relative frequency of acyanotic congenital heart disease shows from the

highest was VSD,ASD, and PDA respectively 35%, 35%, 33%. In research reported that VSD, PDA and ASD Most lesions in infants born alive are 34%, 23.7% and 10.8%, while most lesions on stillbirth is ASD (17.7%), VSD (14.6%), and TOF (13.8%). In addition, some lesion with subtle physical findings, such as ASD might not be detected until they appear in adult life. [9,10]

In our study the patients were found mostly with malnutrition status (52%), however found well nourish patients (40%), and severe malnutrition (8%). Patients with increasing blood flow to the lung and had pulmonary hypertension will suffered from malnutrition and stunted growth associated with hypoxia patients. This situation will facilitate infection that worsen the condition. Anorexia, inadequate nutrition, hypoxemia tissue, hipermetabolik status, acidemia, and cation imbalances, decreased peripheral blood flow, chronic cardiac decompensation,malabsorption and protein loss, tract infections recurrent respiratory, hormonal factors, and genetic will eventually lead to malnutrition condition [11]

Dyspneu was the mostly main complain in our study (73.9%) recurrent cough (17.3) and the less was fatique (8.08%). Most frequently, patients congenital heart disease complain of progressive shortness of breath. For example a studies have shown a reduction in maximum oxygen consumption in the unrepaired ASD population because of the inherent inefficiency of a continuously preload-reduced LV in combination with a volume overload in the pulmonary circulation. After repair of the ASD, exercise capacity improves within days to weeks. [12]

In our study, complication of PAH associated with congenital heart disease was (21.7%). In the French National Registry of PAH, PAH-CHD was the second most commonly associated form of PAH (after connective tissue disease-associated PAH). Data from European registry studies give the overall prevalence of PAH in adult patients with CHD as 4–28% and the prevalence of Eisenmenger's syndrome as ,1–6% [13,14]

Median degree of Tricuspid Regurgitation Pressure Gradient (TRPG) before procedure in our study was 32.4 (range 25-43 mmHg). Median TRPG after procedure was 21.5 (range 15-26.9 mmHg). There was significant decreased in PAH after transcatheter closure procedure. Correction of an underlying congenital heart defect in infancy can prevent the development of PAH-CHD; however, a proportion of patients with left-to-right shunts are not recognized until later in their life, when they already have changes to the pulmonary vasculature and increased PVR. In those patients with increased PAP and Qp, but with a PVR within normal limits or only slightly raised, pulmonary vascular changes are likely to be minimal and the patient may benefit from surgery. Conversely, those patients with high PAP and high PVR are likely to have extensive changes to the pulmonary vasculature and corrective surgery is contraindicated. There remains, therefore, a population of patients with medium-tolarge defects and moderate increases in PVR in whom the extent of pulmonary vascular changes and their potential to be reversed are unknown and so, in whom, the benefits or otherwise of corrective surgery are unclear. Improvements in the diagnosis of CHD and its surgical and medical management have led to a significant increase in the number of patients surviving into adulthood. The best therapy for PAH-CHD remains prevention through a ''timely'' repair of the defect. [15] This study had limitation that not enough sample and participants so need further longitudinal and multicenter study.

Conclusion

In our conclusion, children with acyanotic heart disease commonly associated with complication of PAH. Transcatheter closure procedure in acyanotic congenital heart disease was important for decreasing the pressure gradient of PAH. But this study need further research to identifying, assessing and accurately deciding on management strategies.

Conflict of Interest : None declared.

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Ethical Clearance : Approved by researched ethical committee Dr. Soetomo General Hospital Surabaya**.**

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