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Atrial septostomy to prevent pulmonary hypertension crisis in children with ventricular septal defect (VSD) and pulmonary hypertension (PH) underwent cardiac surgery: a case series

Haryo Aribowo[1]*, Yoga Arditya[2], Gita Christy Gabriela[3]

[1]Division of Cardiac, Thoracic, and Vascular Surgery, Department of Surgery, Faculty of Medicine, Public Health, and Nursing/Dr. Sardjito General Hospital, Yogyakarta, [2]Division of Cardiac, Thoracic, and Vascular Surgery, Department of Surgery, Faculty of Medicine, Public Health, and Nursing/Dr. Sardjito General Hospital, Yogyakarta, [3]Department of Surgery, Faculty of Medicine, Public Health, and Nursing/Dr. Sardjito General Hospital, Yogyakarta 55281, Indonesia

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ABSTRACT

Severe pulmonary hypertension (PH) was reported in 22.4% of ventricular septal defect (VSD) and it was mainly seen on a large VSD. Atrial septostomy (AS) could improve the hemodynamic condition and long-term survival of PH patients. Here, three VSD and PH cases in children who underwent AS surgery as their early treatment, concomitant with PH-specific pharmacological treatment were reported. Patient's hemodynamic and general condition improved with no further complications during the follow-up period. Atrial septostomy was usually conducted after all PH-specific pharmacological interventions failed. However, a study found that the survival benefit of AS was significantly increased if it was conducted before PH-specific pharmacotherapies. Most of the patients in this case received immediate hemodynamic and functional improvement. In this case series, it was reported that the AS procedure could lower the pulmonary arterial pressure and be safely conducted without further complications or death >24 hr post-surgery. Considering the clinical benefit, safety procedure, and improved pulmonary arterial pressure, performing AS procedure concomitant with PH-specific pharmacotherapy as an early treatment for PH patients is recommended.

ABSTRAK

Sebanyak 22,4% pasien ventrikular septal defek (VSD), terutama VSD berukuran besar, dilaporkan mengalami hipertensi pulmonal (HP) berat. Atrial septostomi (AS) dapat memperbaiki kondisi hemodinamik dan kelangsungan hidup jangka panjang pasien HP. Tiga kasus HP pada VSD anak yang menjalani AS bersamaan dengan terapi farmakologi khusus HP sebagai terapi awal disampaikan dalam laporan kasus ini. Paska prosedur AS, terdapat perbaikan hemodinamik dan kondisi umum tanpa disertai komplikasi. Pada umumnya, prosedur AS dilakukan setelah pemberian seluruh farmakoterapi khusus HP gagal. Namun, sebuah penelitian menunjukkan bahwa tindakan AS dapat meningkatkan angka kelangsungan hidup secara nyata bila dilakukan sebelum farmakoterapi khusus HP. Hampir semua pasien pada kasus ini mengalami perbaikan hemodinamik dan fungsional secara cepat. Pada kasus serial ini, dilaporkan bahwa prosedur AS dapat menurunkan tekanan arteri pulmonal dan aman dilakukan tanpa komplikasi lebih lanjut ataupun kematian >24 jam paska operasi. Dengan mempertimbangkan keuntungan klinis, keamanan prosedur, dan perbaikan tekanan arteri pulmoner, dilakukannya prosedur AS bersamaan dengan farmakoterapi khusus HP sebagai terapi awal pasien HP direkomendasikan.

Keywords:
atrial septostomy;
ventricular septal defect;
pulmonary hypertension;
children;
pharmacotherapy

*corresponding author: haryoaribowo@ugm.ac.id

INTRODUCTION

Ventricular septal defect (VSD) is one of the most typical congenital malformations of the heart (40% of all cardiac anomalies).[1] A large VSD is related to severe pulmonary hypertension and increases the risk of developing pulmonary vascular disease. Severe pulmonary hypertension was reported in 22.4% of VSD and it is mainly seen on a large VSD.[2]

Pulmonary hypertension (PH) treatment has undergone many changes in the last 30 years. Before vasodilator therapy, the mean survival age for children was one year.[3] New PH-specific medicine has improved children's five-year survival rates by at least 80%.[4-6] Improving survival rates in children with PH showed the development of PH treatment from pharmacological aspects, human resources, and understanding of PH pathophysiology.[4] PH treatments improve lung microvascular obstruction with anticoagulants, oral vasodilator, intravenous (IV) prostacyclin, or lung transplant as the last choice.[7] Less than 25% of PH showed an improvement with long-term oral vasodilator therapy, whereas 75% of patients with more severe PH needed long-term IV prostacyclin and lung transplant to receive an optimal condition.[8,9] The application of IV prostacyclin and lung transplant worldwide is limited due to technical and cost difficulties. Therefore, research related to a PH alternative therapy is really needed.

The death of PH patients is dominantly caused by right ventricle heart failure (RVHF).[10] Therefore, many interventions have been developed to fix the right ventricle (RV) without a lung transplant; one was pre-tricuspid valve shunt or Atrial Septostomy (AS), an endovascular intervention to make an artificial shunt between the right and left atrial.[5,10] The rationales for its use are: (1) the aggravating impact of RVHF on patient survival; (2) the unpredictable response to medical treatment; (3) the difference in treatment availability worldwide and limited access to lung transplantation.[11,12] If AS is successfully done, it could decompress RV and left ventricle (LV) 's failing and result in a significant clinical improvement, long-lasting hemodynamic effects at rest, and improved survival rate in patients with PH. Many studies showed that AS was usually done after all PH-specific pharmacological interventions failed,[10,13,14] however, Sandoval *et al.*,[13] found that the survival benefit of AS was significantly increased if it was first performed then, followed by PH-specific drug therapies.

Here, three different cases of VSD and PH in children who underwent AS surgery as their early treatment, concomitant with PH-specific pharmacological treatment were presented. One girl presented in doubly committed sub arterial (DCSA) VSD and PH, anther girl was shown in a perimembranous outlet (PMO) VSD and PH, the last girl showed PMO VSD and patent foramen ovale (PFO).

CASES

Case 1

A 6-y.o. girl was referred to Dr. Sardjito General Hospital, Yogyakarta with a history of repeated cough and difficulty gaining weight. The physical examination showed no signs of right heart failure, and her room air oxygen saturation was good. Transthoracic echocardiography (TTE) examination showed a significant DCSA VSD (subpulmonic) and PH. Heart catheterization examination showed a significant DCSA VSD and PH with high-flow low resistance and reactive

O_2 test. Furosemide 2 x 0.5-1 mg/kgBW/time, captopril 2 x 0.3-0.5 mg/kgBW/time, and sildenafil 3 x 0.5-1 mg/kgBW/time were given before surgery. Open-heart surgery was performed to close 2 cm VSD and create a 3 mm ASD. Firstly, we closed the VSD and then made a hole in the middle of the atrial septum using a puncher with a 3 mm diameter. Post-surgical TTE evaluation showed a reduction of pulmonary arterial systolic pressure from 60mmHg to 14mmHg (TABLE 1). Then, she received inotropic therapy (5-10 mg/kgBW) and was being follow-up in the ICU for the first 48 h. The inotropic therapy was stopped the following three days and moved her to the inpatient ward. She was sent home eight days post-surgery and consumed captopril 2 x 0.3-0.5 mg/kgBW/time, furosemide 2 x 0.5-1 mg/kgBW/time, and sildenafil 3 x 0.5-1 mg/kgBW/time.

Case 2

A 2.5-y.o. girl with a history of repeated cough since nine months old and pneumonia treatment was referred to Dr. Sardjito General Hospital in stable hemodynamic, well-room air oxygen saturation, and no signs of heart failure. TTE examination showed 11 mm PMO VSD and PH. Heart catheterization exam exhibited big PMO VSD and PH with high-flow high resistance and reactive O_2 test. She received captopril 2 x 0.3-0.5 mg/kg BW/time, furosemide 2 x 0.5-1 mg/kg BW/time, sildenafil 3 x 0.5-1 mg/kg BW/time, and surgery. Open-heart surgery was conducted to close 2 cm VSD and create an ASD. Firstly, the VSD was closed and then made a hole in the middle of the atrial septum using a puncher with a 3 mm diameter. Post-surgical evaluation with TTE showed decreasing arterial pulmonary systolic pressure from 47 to 23mmHg (TABLE 1). She was in the ICU for three days post-surgery and received inotropic therapy (5-10 mg/kg BW) for two days. There was an excellent hemodynamic improvement following the surgery. Therefore, she was moved to the inpatient ward on the 4th day. Eight days post-surgery, she was sent home and continued oral therapy with captopril 2 x 0.3-0.5 mg/kgBB/time, furosemide 2 x 0.5-1 mg/kgBW/time, and sildenafil 3 x 0.5-1 mg/kgBW/time.

Case 3

An 8-y.o. girl with a malnutrition history and repeated cough since six years old was referred to Dr. Sardjito General Hospital. Her hemodynamic condition was stable, her room-air oxygen saturation was adequate, and there were no signs of heart failure on the physical examination. TTE showed 8-10mm PMO VSD, PFO, and PH. Heart catheterization examination showed persistent left superior vena cava, big PMO VSD, and PH with high-flow low resistance and reactive O_2 test. She received captopril, furosemide, sildenafil, and surgery. Open-heart surgery was done to close 1.8mm VSD, foramen ovale, and create an ASD. Firstly, the VSD was closed and foramen ovale; after that, we made a hole in the middle of the atrial septum using a puncher. Post-surgical examination using TTE showed decreasing pulmonary arterial systolic pressure from 63 to 25mmHg. She was being followed up in ICU for the first 48h post-surgery, then moved to inpatient care for four days. She received inotropic therapy (5-10 mg/kgBW) for the first three days post-surgery. There were no complications following the surgery, Therefore, she was sent home and continued captopril 2 x 0.3-0.5 mg/kgBW/time, furosemide 2 x 0.5-1 mg/kgBW/time, and sildenafil therapy 3 x 0.5-1 mg/kgBW/time.

TABLE 1. Results of supporting examinations and patient hemodynamics.

Variable	Case 1	Case 2	Case 3
Heart catheterization examination			
• PAP (mmHg)	76/43 (60)	68/27 (47)	77/49 (63)
• MV (%)	77.6	71.53	85.27
• FR	2.51	1.75	3.77
• PARI (WU)	3.13	4.7	1.7
• RA pressure (mmHg)	-	18/14 (13)	14/9 (11)
• RV pressure (mmHg)	-	73/12 (18)	79/13 (14)
• LV pressure (mmHg)	79/5 (18)	79/15 (26)	93/1 (18)
Echocardiography			
• Type of defect	DCSA VSD	PMO VSD 11mm	PMO VSD 8-10mm PFO 2.2mm
TTE post-surgery			
• Ejection fraction (%)	62	59	59
• TAPSE	8	11	9,5
• Efusi pericardium	(-)	(+) 6-9 mm	(+)
• Systolic PA pressure (mmHg)	14	23	25
• TVG (mmHg)	4	15	17
SaO_2 Pre-Op/Post Op (%)	99/96	98/99	97/96

PAP: pulmonary artery pressure; MV: mixed vein saturation; FR: flow ratio; PARI: pulmonary artery resistance index; RA: right atrium; RV: right ventricle; LV: left ventricle; TAPSE: tricuspid annular plane systolic excursion; TVG: tricuspid valve gradient; PA: pulmonary artery.

DISCUSSION

Atrial septostomy is a procedure to create an intracardiac shunt in children with various congenital cardiac defects.[14] It was first conducted by Rich and Lam 20 years ago on a patient with severe PH.[7] In 1964, an animal study by Austin *et al.*[15] showed that inter-atrial communication could decompress the dilated and hypertension right ventricle and augment systemic blood flow, especially during exercise. The rationale for AS procedure was also supported by the fact that PH patients with patent foramen ovale (PFO) and Eisenmenger's syndrome patients have higher survival rates than PH patients without intracardiac shunting.[8,12,16,17] In this case, AS creation during open-heart surgery following the closure of VSD was performed. Firstly, the VSD was closed, then made a hole in the middle of the atrial septum with a certain-sized puncher (3 to 5 mm in diameter). The hole was made in the middle of the atrial septum to ease the hole closure by an amplatzer device.

Previous study reported that the combination of AS and pharmacotherapy could lower the prevalence of WHO functional class IV patients compared to AS alone, although it was not significantly different.[13] In most-reported studies, AS was usually conducted after all PH-specific pharmacotherapy failed. Therefore, the impact of the procedural intervention on long-term survival is lessened.[10-15] However, Sandoval *et al.*[13] reported that the survival benefit of AS is significantly increased if it is performed first, followed by PH-specific pharmacotherapies. Most of his patients who received AS procedure produced immediate hemodynamic and functional improvement. In this case, AS was also performed as an early treatment combined with pharmacotherapy. The

patient's hemodynamic and general condition improved with no further complications during the follow-up period. The difference between this case and the previous study was in the timing of pharmacotherapy and the AS technique. In this case, the pharmacotherapy was given before and after the surgery and performed ASD creation while performing open-heart surgery to close VSD and PFO, while in the previous study, the pharmacotherapy was given only after the surgery and AS was conducted using balloon dilatation technique.[13] After the ASD creation, all patients did not show severe limitations (WHO functional class IV).

All of three patients showed a notable decrease in pulmonary arterial pressure (TABLE 1). It was estimated post-surgical pulmonary arterial pressure count with TTE because heart catheterization was an invasive procedure. Previous studies also showed a decrease in mean pulmonary arterial pressure, though the drop was insignificant.[14,18]

The risk of AS procedure is high. Therefore, it is contraindicated in patients with (1) severe RVHF on cardiorespiratory support, (2) mean right atrial pressure (mRAP) >20mmHg, (3) room-air resting O_2 saturation <90%, (4) left ventricular end-diastolic pressure (LVEDP) >18mmHg because the most common cause of death in this procedure was resistant hypoxia.[10] The higher the right-sided pressure, the more likely the shunt to cause uncontrollable hypoxemia.[10] Our patient's general and hemodynamic condition was stable without cardiorespiratory support, and their room-air resting O_2 saturation was also >90%. Therefore, the AS procedure on the patients was performed.

In this case series showed that the AS procedure could be conducted safely without further complication or death 24 h post-surgery. The patients showed decent improvement in their hemodynamic and general condition with no complications during the follow-up period. Atrial septostomy was conducted to anticipate PH crisis or manifest PH because PH crisis can interrupt blood flow to the lung, which can be fatal. Atrial septostomy creation could decrease pulmonary arterial blood flow, so the obstructed blood flow could stream from right atrial to left atrial. Due to the study design, the number of patients was limited. Therefore, further research is still needed to confirm these findings.

CONCLUSION

Atrial septostomy procedure could lower the pulmonary arterial pressure and be safely conducted without further complications or death >24h post-surgery. Considering the clinical benefit, safety procedure, and improved pulmonary arterial pressure, performing AS procedure concomitant with PH-specific pharmacotherapy as an early treatment for PH patients is recommended.

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