Balloons atrial septostomy procedures for cyanotic congenital heart defect in Sanglah Hospital

Ni Ketut Mena Epiani,* Eka Gunawijaya, Ni Putu Veny Kartika Yantie

ABSTRACT

Balloons atrial septostomy (BAS) is a technique used to enlarge a hole between the right and left atrium. Balloon atrial septostomy is an initial life-saving treatment in particular cyanotic congenital heart defect especially transposition of the great arteries-intact ventricular septum (TGA-IVS) and pulmonary atresia-intact ventricular septum (PA-IVS). This procedure aimed to create an atrial septal defect that will enhance the bidirectional mixing of pulmonary and systemic venous blood, hence improving oxygen saturation in this type of heart defect. Complications of BAS consist of arrhythmia, heart block, valve dysfunction, myocardial ischemia, neurodevelopmental abnormalities, stenosis or occlusion of the coronary artery. In our center, during April 2015 – June 2016, there were 4 BAS procedures and one re-ballooning. The range of ages was 1-15 days in BAS patients. The NuMedZ-5® balloon catheter number 13.5 was used. The diameter of atrial communication and oxygen saturation was increased significantly after the procedure.

Keywords: balloon atrial septostomy, cyanotic congenital heart defect
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INTRODUCTION

Cyanotic congenital heart defects such as dextroposition of the great arteries (d-TGA), tricuspid atresia, pulmonary atresia (PA), total anomalous pulmonary venous drainage (TAPVD), and hypoplastic left heart syndrome (HLHS) require obligatory and non-restrictive mixing of blood at the atrial level to sustain life.1-2 The presence of a small and restrictive atrial septal defect (ASD) will lead to an inadequate mixture of blood and cause persistent hypoxia and metabolic acidosis.2 Palliative procedure to create ASD or increasing the size of existing patent foramen ovale (PFO) will provide a second mixing site at the atrial level.2 Balloon atrial septostomy remains a necessary palliative interventional procedure in certain forms of congenital heart disease.1,3 Now, this procedure is known as Rashkind balloon atrial septostomy that extensively used to improve atrial mixing in neonates with transposition of great arteries and has been extended to treat atrial level obstruction in patients with tricuspid atresia, pulmonary atresia-intact ventricular septum (PA-IVS), TAPVD, and HLHS including mitral atresia.2-4 Typically it is performed in the cardiac catheterization laboratory under fluoroscopic guidance with hemodynamic monitoring.5 This report aims to describe our experience using BAS in several cases.

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CASE ILLUSTRATION

During April 2015 - June 2016, there were 4 BAS procedures and one re-ballooning procedure due to severe pulmonary stenosis (PS) performed in our center. All patients had general anesthesia and used femoral access. The NuMedZ-5™ (Numed Inc, New York, USA) balloon catheter number 13.5 was used (Figure 1). The diameter of atrial communication and oxygen saturation increased significantly after the procedure. Complications persist during the process in 3 patients, but no significant difficulty found during procedures. The procedures were under general anesthesia. The vascular access was obtained in the femoral vein. The shaft size was 6 F's. Once, the position of the catheter confirmed in the left atrium and away from the pulmonary veins and mitral valve, the balloon was then inflated with 2 to 2.5 milliliters of normal saline. Finally, while holding to close to the point of entry in the body, the catheter was pulled back suddenly in a jerky manner, using wrist joint to produce jerks while the elbow joint being stabilized and fixed at the same time. Two to three pull was attempted in each patient. The resultant size of the ASD was measured, and the flow across it was confirmed echocardiographically using 2-D, as well as color Doppler mode. A list of the heart defects and procedures detail showed in table 1.

First case

BD, a 5 days old girl, was referred to our hospital due to central cyanosis since 2 hours of life. She was born full-term and vigorous through elective caesarian section, with bodyweight 2.8 kg and 49 cm in length. No history of heart disease in the family. Antenatal care was within normal and no history of mother illnesses during pregnancy. Physical examination revealed calm centrally cyanose. Heart rate was regular, and there was no respiratory distress. The first heart sound was normal with a single second heart sound; the continuous murmur was found on the upper left sternal border. A chest x-ray showed an egg-shaped heart. Echocardiography revealed TGA – IVS, small ASD Secundum, small PDA. Balloon atrial septostomy performed on 1st day of life with Nu-Med Z-5 balloon catheter number 13.5. The atrial septal defect and oxygen saturation increased after the procedure. The patient got better after 7 days of antibiotic course and referred to Jakarta for arterial switch operation (ASO).

Third case

BH, a 1-day-old boy, was born in our hospital and central cyanosis found after 4 hours of life. He was born full-term and vigorous through elective caesarian section due to placenta previa, with bodyweight 2.6 kg and 47 cm in length and no history of heart disease in the family. Antenatal care was within normal and no history of mother illnesses during pregnancy. Physical examination revealed a calm cyanotic baby. Heart rate was regular, and there was no respiratory distress. Chest examination showed first heart sound was normal with single second heart sound; the continuous murmur was found on the upper left sternal border. Balloon atrial septostomy procedure performed a few hours later due to worsening conditions using misoprostol with Nu-Med Z-5 balloon catheter no 13.5. The atrial septal defect and oxygen saturation increased after the procedure. The patient died 10 days after the procedure due to sepsis.

Fourth case

BP, a 15-days-old boy, was referred to our hospital due to central cyanosis since 8 hours of life. Cyanosis found on the mouth and extremities. She was born full-term and vigorous through elective caesarian section, with bodyweight 2.8 kg and 53 cm in length. No history of heart disease in the family. Antenatal care was within normal and no history of mother illnesses during pregnancy. Physical examination revealed irritable. Heart rate was regular, and there was no respiratory distress. Chest examination showed first heart sound was normal with single second heart sound; the continuous murmur was found on the upper left sternal border. No oxygen support and misoprostol was readily given at that time. Balloon atrial septostomy performed on 15 days old with Nu-Med...
Z-5 balloon catheter number 13.5. The atrial septal defect and oxygen saturation increased after the procedure. There was a minor complication that occurred after BAS as transient supraventricular tachycardia-induced by catheter and resolved spontaneously, and the patient discharged a few days after.

Fifth case
BD, the first case, came again on 1 year old as a regular patient to our policlinic with chief complaint worsening of central cyanosis since one month. She had a history of BAS and ASO during the newborn period. Physical examination revealed cyanosis children with clubbing fingers. The chest examination showed visible ictus cordis on the left anterior axillary line of the fourth intercostals space. No thrill or right ventricle heave found. A systolic murmur was detected on the mid clavicle line of the 3rd intercostals space. Routine blood examination showed a slightly increased hemoglobin and hematocrit. Catheterization balloon performed with two procedures. The first procedure was percutaneous transluminal balloon valvuloplasty (PTBV) with Run-through NS 0.014. The second procedure was re-ballooning with Saphire II 4.0/20 mm balloon catheter no 16 and 14 to repair severe PS. Saturation was increased around 85-90% after the procedure. No complication found after procedure. The characteristic of each case could be summarized in Table 1.

Table 1  Characteristic of patients balloon atrial septostomy

<table>
<thead>
<tr>
<th>No</th>
<th>Identity</th>
<th>Heart defects</th>
<th>Instrument</th>
<th>Complication</th>
<th>Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>BD, 5 d, girl</td>
<td>TGA-IVS, small ASD, PDA, mild TR, smallish LV</td>
<td>NuMed Z-5 no 13.5</td>
<td>VT induced catheter</td>
<td>Survive</td>
</tr>
<tr>
<td>2</td>
<td>BK, 1 d, boy</td>
<td>TGA-IVS, small ASD secundum, small PDA</td>
<td>NuMed Z-5 no 13.5</td>
<td>Temporary VES</td>
<td>Survive</td>
</tr>
<tr>
<td>3</td>
<td>BH 1, 1 d, boy</td>
<td>Pulmonary atresia-IVS, small tortuous PDA, PFO,</td>
<td>NuMed Z-5 no 13.5</td>
<td>None</td>
<td>died due to sepsis</td>
</tr>
<tr>
<td>4</td>
<td>BP, 15 d, boy</td>
<td>TGA, PFO, PDA</td>
<td>NuMed Z-5 no 13.5</td>
<td>Transient SVT</td>
<td>Survive</td>
</tr>
<tr>
<td>5</td>
<td>BD, 1 year, girl</td>
<td>Post ASO, severe RPA stenosis, small ASD II</td>
<td>Sapphire II 4.0/2.0 mm no 16</td>
<td>No complication</td>
<td>Survive</td>
</tr>
</tbody>
</table>

Abbreviation: TGA-IVS (Transposition of Great Artery-Intact Ventricular Septum); ASD (Atrial Septal Defect); PDA (Patent Ductus Arteriosus); TR (Tricuspid Regurgitation); LV (Left Ventricle); PA-IVS (Pulmonary Atresia-Intact Ventricular Septum); PFO (Patent Foramen Ovale); ASO (Atrial Switch Operation); RPA (Right Pulmonary Atresia); VT (Ventricular Tachycardia); VES (Ventricular Extrasystole); SVT (Supraventricular Tachycardia)

Table 2  The outcome of patient with balloon atrial septostomy

<table>
<thead>
<tr>
<th>No</th>
<th>Identity</th>
<th>Saturation before BAS</th>
<th>Saturation after BAS</th>
<th>Diameter of ASD before BAS</th>
<th>Diameter of ASD after BAS</th>
<th>Treatment before BAS</th>
<th>Treatment after BAS</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>BD, 5 d, girl</td>
<td>65-68%</td>
<td>72-78%</td>
<td>2 mm</td>
<td>3.9 mm</td>
<td>No oksigen, Misoprostol 15 mcg every 6 hours (oral)</td>
<td>Misoprostol 15 mcg every 6 hours (oral)</td>
</tr>
<tr>
<td>2</td>
<td>BK, 1 d, boy</td>
<td>72-76%</td>
<td>83-87%</td>
<td>2.4 mm</td>
<td>4.9 mm</td>
<td>No oksigen, Misoprostol 15 mcg every 6 hours (oral)</td>
<td>Misoprostol 15 mcg every 6 hours (oral)</td>
</tr>
<tr>
<td>3</td>
<td>BH, 1 d, boy</td>
<td>64-67%</td>
<td>75-79%</td>
<td>2.2 mm</td>
<td>3 mm</td>
<td>No oxygen, Misoprostol 12.5 mcg every 4-6 hours</td>
<td>Ventilator support, Misoprostol 12.5 mcg every 4-6 hours.</td>
</tr>
<tr>
<td>4</td>
<td>BP, 15 d, boy</td>
<td>68-71%</td>
<td>85-88%</td>
<td>2.6 mm</td>
<td>6.9 mm</td>
<td>No oksigen, Misoprostol 12.5 mcg every 4-6 hours.</td>
<td>Ventilator support, Misoprostol 12.5 mcg every 4-6 hours.</td>
</tr>
<tr>
<td>5</td>
<td>BD, 1 year, girl</td>
<td>65-68%</td>
<td>85-87%</td>
<td>2.1 mm</td>
<td>3.5 mm</td>
<td>CPAP support, Captopril 3 mg every 12 hours, spironolacton 6.25 mg every 12 hours, furosemid 3 mg every 12 hours, dopamine 7.5 mcg/kg/min ~ 0.7 mL/hour</td>
<td>Ventilator support, Captopril 3 mg every 12 hours, spironolacton 6.25 mg every 12 hours, furosemid 3 mg every 12 hours, dopamine 7.5 mcg/kg/min ~ 0.7 mL/hour</td>
</tr>
</tbody>
</table>

Abbreviation: BAS (Balloon Atrial Septostomy); mcg (Microgram); mL (Milliliter)
CASE ILLUSTRATION

DISCUSSION

Congenital heart defect (CHD) define as anatomic malformation of the heart or great vessels which occur during intrauterine development, irrespective of the age at presentation. Congenital heart defect may be classified as acyanotic, and cyanotic depends on whether patient clinically exhibits cyanosis or not. Tetralogy of Fallot (TOF), TGA, and tricuspid atresia are the three most common cyanotic heart defects in the children. Duct dependent systemic circulation consists of PA, essential PS, TOF and tricuspid atresia. Duct dependent systemic and pulmonary circulation is the transposition of the great arteries.

Rearrangement of the great arteries is the most common CHD defect presented in the newborn period. It constitutes 5% from all CHD, and 10% of all neonatal cyanotic CHD. This defect will lead to systemic venous blood does not get oxygenated, and the pulmonary venous blood does not get delivered to the body. The infants will not survive unless there are inter-circulatory shunts such as an atrial or ventricular septal defect or patent ductus arteriosus. In this case, there were 3 cases of TGA and 1 case of pulmonary atresia that required BAS procedure during early life as palliative non-surgical therapy before surgical therapy performed.

Clinical features of cyanotic CHD are varied depending upon the anatomic type. Transposition of great arteries with intact ventricular septum (type I TGA) patients usually severely cyanotic but without respiratory distress, until severe hypoxemia and acidosis develop. The right ventricular impulse often increases with a single second heart sound. Murmur is rare and if persist, can be found as a non-specific ejection systolic murmur with low grade (grade I-II/VI). Patients with TGA and VSD (type II) usually showed symptoms of congestive heart failure (tachypnea, tachycardia, sweating, and poor feeding) during 4-8 weeks of life, but cyanosis is minimal. Physical examinations can reveal hepatomegaly, increase right and left ventricular impulses, single second heart sound, and an III-IV/VI grade of holosystolic murmur at the left lower sternal border. Patients with TGA, VSD, and PS or type III TGA usually showed similar symptoms with TGA with intact septum, TGA with VSD, or TOF depends on the degree of mixing and severity of PS. In this case, all patients showed cyanosis symptoms without respiratory distress few hours after birth, and none of them had signs of heart failure.

Accurate diagnosis with echocardiography makes invasive studies (catheterization and angiography) becomes unnecessary. Balloon atrial septostomy is needed for rapid relief of hypoxemia and acidosis before arterial switch procedure. Sometimes BAS and dilatation of pulmonary valve must perform together in patients that require Rastelli type of repair. In this case, echocardiography performed to establish the diagnosis, but angiography were not performed.

Balloon atrial septostomy can enhance atrial mixing and to decompress the left atrium, and essential to augment the cardiac output in the
right side of obstruction lesion. This procedure can perform in the catheterization laboratory or neonatal intensive care unit with angiographic or echo-guidance. Vascular access can be obtained through the umbilical vein or femoral vein.10-11 This procedure is an option for children under 6 weeks of age, and if BAS has done in older infants, it could increase the thickness of the atrial septum.9,10 In this case, four patients had BAS before 6 weeks of life. All of the patients were performed in the catheterization laboratory under echocardiography and fluoroscopic guidance.

Prostaglandin E1 (PGE1) can be a life-saving drug for infants born with ductus-dependent congenital heart disease.11,12 Therapy with PGE1 can effectively maintain an infant's cardiovascular function until palliative or corrective cardiac surgery performed.13 Prostaglandin E is currently recommended as initial therapy for infants with an isolated defect that restricts pulmonary blood flow (PS and PA), poor arterial-venous mixing (TGA), and conditions that interfere with systemic circulation (interruption or coarctation of the aorta).14,15 Endogenous prostaglandins, primarily PGE, and PGE3, are produced within the vessel lumen during gestation to keep the ductus patent. At the time of birth, an increment of arterial oxygen saturation and a decrement of endogenous prostaglandin stimulate an alteration of vascular integrity and promoting closure of the duct. Exogenous prostaglandins can be used to extend the patency of the ductus.15 Prostaglandin E must be administered as a continuous intravenous infusion because of its rapid extraction and metabolism by the lungs. The metabolites, 13,14-dihydro-PGE1, is pharmacologically active. The elimination half-life of PGE1 is approximately 42 seconds. Many of the adverse effects of PGE1 are apnea, hyperthermia, flushing, fever, bradycardia, and/or hypotension may indicate excessive prostaglandin effect and need to dose reduction.16

Misoprostol is a synthetic PGE, analog, structure misoprostol consist of 15-deoxy-16-hydroxy-16-methyl PGE1. The advantages of misoprostol are widely available, stable at room temperature, can be given oral, cheap and have few side effects. Misoprostol differs structurally from PGE1 by the presence of methyl ester at C-1, it can increase the anti-secretory potency and duration of action of misoprostol, whilst the movement of the hydroxyl group from C-15 to C-16 and the addition of methyl group at C-16 improves oral activity, increases the duration of action, and improves the safety profile of the drug.17 In this case, to maintain ductus patency, we use misoprostol because the PGE1 is not available in our center.

The complication of BAS procedure were graded as major and minor. These were ranked as major if some intervention was required in reverted the condition to normal, or if it resulted in permanent damage, debility or death. Minor complication if did not require any intervention to revert back to normal, or it didn't result in permanent injury or debility.7,8 Complications during atrial septostomy could be classified into mechanical, traumatic, embolic, and electrical.16 Mechanical complication includes the rupture of the balloon with or without embolization of balloon fragments, failure in balloon deflation and inflation of the balloon in an inadequate position/place. Traumatic complication includes cardiac damage with rupture of the atrial appendage, mitral valve injury or vascular injury of the pulmonary veins or the inferior cava vein. The embolic complexity is a stroke. Electrical complication includes transitory rhythm disturbance (arrhythmia).9,10 Adverse neurological outcomes in infants with cyanotic CHD that performed BAS has been an issue.

**SUMMARY**

Upon diagnosis, some CHD will require immediate treatment or stabilization and subsequent corrective palliative surgical therapy, although arterial switch operation for transposition of great vessels is typically performed within days after birth and opening of ductus arteriosus can be maintained with PGE1. Balloon atrial septostomy is the first option regarding the dilatation of a PFO in the newborn with an indication for atrial septostomy, although other new techniques have developed. It is performed almost routinely in all cases of TGA and many times with only using echocardiographic guidance. Complications are rare, and benefits are beyond.

**REFERENCES**


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