Case Report
Simultaneous Transcatheter Pulmonary Balloon Valvuloplasty and Atrial Septal Defect Closure in Adult Patient

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ABSTRACT

Background: Percutaneous pulmonary balloon valvuloplasty (PBV) has become the first choice of management in patients with valvular pulmonary stenosis, while transcatheter closure in secundum atrial septal defect (ASD) is an alternative therapy besides surgical closure. The combination of PBV and transcatheter ASD closure is an action that is possible to do, but rarely done.

Case Illustration: We present a case of 26-year-old woman with severe pulmonary stenosis and secundum ASD. Echocardiographic examination revealed the presence of severe pulmonary stenosis (mean gradient 99.19 mmHg) and defects in the intra atrial septum (with a size of 1.3 cm). Cardiac catheterization revealed right ventricular pressure of 160 mmHg. This patient then successfully corrected with both procedures ASD closure and PBV simultaneously.

Discussion: These procedures have high feasibility, good safety, and shows good effectiveness. There were no complications during the procedures, and a satisfying immediate outcome was obtained.

Conclusion: Pulmonary stenosis and atrial septal defects are rare forms of congenital heart disease. Transcatheter pulmonary balloon valvuloplasty and atrial septal defect closure performed simultaneously are promising alternatives, apart from surgical correction.

Introduction

Pulmonary stenosis (PS) is the second most common congenital heart defect after ventricular septal defect (VSD) [¹]. The prevalence of pulmonary stenosis is about 8-12% of all congenital heart defects. This disorder can be found together with other congenital heart defects, including atrial septal defect (ASD). However, severe PS is uncommon to be concomitant with ASD [²].

Recent developments in interventional cardiology techniques have provided alternative therapeutic modalities for patients with complex congenital heart defects. Percutaneous pulmonary balloon valvuloplasty (PBV) has become the first choice of management in patients with valvular PS, while transcatheter closure in ASD secundum is an alternative therapy besides surgical closure [³]. The combination of PBV and transcatheter ASD closure is an action that is possible to do, but rarely done.

Case Presentation

A 26-year-old woman came with complaint of shortness of breath for the last 5 years. The
shortness of breath was experienced during light daily activities. The patient had been diagnosed to have congenital heart disease since 5 years ago by a cardiologist, but refused to undergo further medical workup for surgery. The symptoms became worse in the last two months that she eventually agreed to be referred to Dr. Soetomo General Hospital. The patient also experienced cyanosis on her lips and fingertips during heavier activity. Birth history: the patient was born through vaginal delivery; there is no family history of congenital heart disease.

Physical examination on June 26, 2019 showed blood pressure of 90/70, pulse rate of 72 beats/minute, breathing frequency of 20 times/minute, temperature of 36.7 °C, and oxygen saturation of 91% with free oxygen. Auscultation showed hardened second heart sound and a systolic murmur in the left sidelines of II, grade III / VI. Clubbing fingers were seen on hands and feet. There was no increase in jugular venous pressure, crackles and wheezing on lung examination, or leg edema.

Electrocardiography (ECG) examination showed regular sinus rhythm 75 bpm, frontal RAD axis, horizontal counterclockwise rotation axis, right atrial enlargement, right ventricular hypertrophy (Figure 1.A). Examination of the chest radiograph revealed a prominent pulmonary conus and an inverted comma sign (Figure 1.B). Echocardiographic examination revealed the presence of severe pulmonary stenosis (mean gradient 99.19 mmHg) (Figure 4.A), defects in the intra atrial septum (with a size of 1.3 cm) (Figure 6.A), and dilation of the right atrial and right ventricle.

Cardiac catheterization revealed right ventricular pressure of 160 mmHg. Right ventricular angiography showed severe pulmonary valvular stenosis and post stenotic dilatation (Figure 2-3).

The pulmonary annulus diameter was 10 mm. The contrast injected after catheter crossing from RA to LA revealed the presence of ASD. Measurement using balloon showed its size of 14 mm in diameter.

The procedure is performed under local anesthesia and guided by transthoracal echocardiography. Pulmonary balloon valvuloplasty was performed using a 12 mm Tyshak II Balloon. Balloon inflation was carried out 5 times. Measurement of the right ventricle pressure following valvuloplasty showed a pressure drop of 44 mmHg. ASD closure using 16 mm Septal Occluder Amplatzer was done afterwards (Figure 5). The whole procedure lasted for 90 minutes with fluoroscopy time 36 minutes. There were no complications during the procedure.

Transthoracal echocardiographic evaluation was performed immediately after the procedure and one day after the procedure. It showed a significant decrease in pressure gradient (mean PG 33.79 mmHg) without pulmonary regurgitation (Figure 4.B). The device position and function were satisfactory with no residual flow (Figure 6.B). The day after the procedure, clinical evaluation revealed that complaints of dyspnea during talking or light activity was much reduced. Oxygen saturation of the peripheral artery was improved (SpO2 98%). The patient then went home with 100 mg of aspirin therapy once a day for 6 months.

Discussion

The main goal of PS management is to overcome RVOT obstruction so that the right ventricular pressure load decreases. Initially, the only therapy for patients with valvular PS is surgical. Until 1982, Kan et al. introducing pulmonary balloon valvuloplasty (PBV) techniques [4], and since then, PBV has become the first choice for PS management in all age groups, because of its low risk and good short-term and long-term outcomes [5].
PBV is indicated in patients with pressure gradients on the pulmonary valve >50 mmHg. Patients with pressure gradients <50 mmHg are usually asymptomatic and have a good long-term prognosis. Conversely, patients with pressure gradients >50 mmHg can progressively lead to right ventricular hypertrophy. Patients in this group tend to experience increase of the pressure gradient up to 9 mmHg annually [6]. Our patient showed a pressure gradient of 99.19 mmHg, which indicated PBV should be performed on her.

Mild PS should be observed and followed up every two years to assess the worsening of stenosis. Moderate PS should be followed up at least once a year, and if there is an enlarged right ventricle or the appearance of symptoms, therapy should be done. In contrast, severe PS (which was found in our patient) must be treated immediately [7].

Lateral view on right ventriculography was used to measure the diameter of the pulmonary valve annulus in our patient. The maximum diameter of the balloon (in our case, we used the Tyshak II balloon with a size of 12 mm) was chosen based on 10-20% greater than the pulmonary valve annulus, which was 10 mm in our patient. Several studies state that the most effective balloon/annulus ratio for PBV was 1.1 to 1.2 [8]. Balloons with this size may overcome obstruction in the pulmonary valve yet did not cause significant pulmonary regurgitation [7]. Pulmonary artery dilatation in post-stenosis area may occur due to the hemodynamic “jet-effect”, which may be a good predictor for PBV success [9].

There are various types of balloon catheters available for PBV. The success of PBV depends on the ability to choose the right balloon to achieve full and stable inflation. Rao et al. advocating the use of a low profile balloon that is able to pass as little vascular access as possible in pediatric patients, while balloons with higher inflation pressure can be used in adult patients [10,11].

Tyshak serial balloons: Tyshak, Tyshak II, and Tyshak mini are designed for PBV, and currently, balloon modifications are still being carried out to reduce the size of the catheter, but while maintaining resistance to balloon rupture. In adult patients where the size of vascular access is not a problem, there is a fairly wide choice for PBV balloons, which is balloons with higher inflation pressures. Balloons for PBV currently available include serial Z-med, Diamond, Ultrathin, XXL balloons, Maxi-LD, Opta Pro, and Powerflex [11].

PBV can be done using single-balloon or double-balloon techniques. The use of a double-balloon allows blood flow between the two balloons when full inflation is carried out so that only a slight hemodynamic change occurs. Rao et al. compared the results of a single-balloon with a double-balloon, which showed that PBV with a double-balloon gave excellent results, but was not superior compared to a single balloon if the balloon/valve annulus ratio is precisely determined [10,12]. In addition, the use of a double-balloon has its disadvantages: the procedure takes longer and requires additional vascular access [11]. Based on these considerations, the single-balloon technique was chosen to be performed on our patients.

PBV has various advantages over surgical therapy. It is minimally invasive, does not cause scarring, has lower mortality and morbidity, has shorter hospitalization period, and has good cost-effectiveness [9], PBV has immediate and satisfying long-term outcomes [8].

PBV rarely cause complications. Pulmonary regurgitation can occur due to the mechanism of opening the valve using a balloon catheter that allows the separation of commissures, ruptures, or
leaflet tears, which was not the case in our patient. However, the incidence of pulmonary regurgitation after PBV in a series of publications ranges from 74-100% [13]. In addition, bradycardia and transient hypotension have been reported during balloon inflation, right bundle branch block or atrioventricular block, and balloon rupture were all have been rarely reported [7].

The incidence of restenosis in several case series ranges from 3.8-21%, which is related to the use of inappropriate balloons size [13]. In patients with significant residual PS, repeat PBV can be performed with a larger balloon size. If this does not work, surgical therapy needs to be done [7].

Transcatheter atrial septal defect (ASD) closure is currently widely used for secundum ASD management. When compared with surgical closure, transcatheter closure is less invasive, requires a shorter recovery period and smaller psychological impact [5]. King and Mills reported the first case in 1976, but percutaneous ASD closure entered the clinical area broadly after the introduction of the Amplatzer septal occluder (ASO) device [14]. ASO is a self-expanding device, with a double-disc and a short connector in the middle. ASO has been accepted as a device for percutaneous closure of ASD by U.S. Food and Drug Administration since 2011, and to date, the closure of percutaneous ASD has been the treatment of choice for secundum ASD [15].

At present, almost 85-90% of all secundum ASD can be closed by transcatheter procedures. Studies show that transcatheter ASD closure can be performed in hemodynamically significant secundum ASD patients, which has a pulmonary-systemic ratio of ≥1.5:1. In secundum ASD patients with defects ≥38 mm, or not have enough rim length, the transcatheter procedure is not an option [15]. In our patient, ASD diameter was identified as 13 mm using echocardiography (14 mm using balloon sizing), thus transcatheter closure became the first choice of intervention.

We choose to do PBV before proceeding with ASD closure. The decision of which action is taken first, whether PBV or ASD closure is crucial and controversial. If ASD closure is done first, it can eliminate the left-to-right shunt and volume overload, and may also reduce the pressure gradient through the pulmonary valve, making it possible to reassess the degree of stenosis. On the other hand, if PBV is carried out first, it will be able to minimize the peri-procedural risk for dislocation from ASD occluder devices [5].

**Conclusion**

Pulmonary stenosis and atrial septal defects are rare forms of congenital heart disease. Performing transcatheter pulmonary balloon valvuloplasty and atrial septal defect closure simultaneously is a promising alternative, apart from surgical correction. We present a case of severe pulmonary stenosis and secundum ASD, which we have successfully corrected with both procedures done simultaneously. These procedures have high feasibility, good safety, and shows good effectiveness. There were no complications during the procedures, and a satisfying immediate outcome was obtained.

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There is no conflict of interest.

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Suplementary Data

Figure 1. (A) ECG examination showed right atrial enlargement and right ventricular hypertrophy; (B) a prominent pulmonary conus and an inverted coma sign was shown on chest x-ray examination.

Figure 2. Angiography pre-pulmonary balloon valvuloplasty showed narrow blood jet through stenotic pulmonary valve.
Figure 3. (A) Tyshak II balloon being inflated during pulmonary balloon valvuloplasty; (B) Balloon with full inflation.

Figure 4. Echocardiographic evaluation of pulmonary stenosis: (A) Before PBV procedure (meanPG 99.19 mmHg); (B) After PBV procedure (meanPG 33.79 mmHg)

Figure 5. Amplatzer septal occluder implanted.
Figure 6. Echocardiographic evaluation of ASD. (A) Pre ASD closure: visible defects at IAS; (B) Post ASD closure: position and function of amplatzer was good with no residual flow.