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Percutaneous Intervention of Pulmonary Supravalvular Stenosis in Two Patients

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ABSTRACT

Introduction: Balloon angioplasty has been successfully used to dilate vascular obstructions. However, its value in some conditions such as supravalvular pulmonary stenosis is still controversial. Although some surgical techniques have been reported to achieve acceptable short- and long-term successes, they are invasive and technically challenging and may result in higher morbidity and mortality compared to percutaneous procedures.

Case Presentation: Successful balloon dilatation was reported for two patients with supravalvular stenosis. One of them had supravalvular pulmonary stenosis developed after pulmonary artery debanding and the second one had supravalvular pulmonary stenosis due to an obstructive congenital web at that site.

Conclusions: The use of balloon dilatation to treat supravalvular pulmonary stenosis is still controversial. However, experiences in two patients showed successful results.

1. Introduction

Balloon angioplasty was successfully used to dilate vascular obstructions. Intimal and medial disruption was shown in histological examination in successful experimental procedures (1). However, its value in other conditions such as supravalvular pulmonary stenosis is still controversial. Use of balloon dilatation to treat supravalvular pulmonary stenosis developed after anatomical correction for complete transposition of the great arteries has been reported (1). Although it has been reported that some surgical techniques have achieved acceptable short- and long-term successes, these techniques are invasive, technically challenging, and may result in higher morbidity and mortality compared to percutaneous procedures. Unsuccessful reports of surgery for supravalvular pulmonic stenosis included an attempted arteriotomy and restenosis after patch graft placement (2, 3).

2. Case Presentation

Balloon dilatation was performed for two patients with supravalvular stenosis. The first patient was a 19-year-old female with a history of complete atrioventricular septal defect who had undergone pulmonary artery banding before total correction. She had a surgery for total correction of the complete atrioventricular septal defect and pulmonary artery debanding when she aged six years. Then, she presented to the clinic with the complaint of chronic exertional dyspnea at New York Hear Association (NYHA) functional class II-III at the age of 19 years. The general appearance was normal. During cardiac examination, the heart rhythm was regular, the first heart sound was within the normal limits, and a grade IV/VI left basilar systolic murmur was auscultated with mild diastolic murmurs. The electrocardiograms revealed normal sinus rhythm and right ventricular hypertrophy pattern. In addition, chest x-ray showed normal cardiothoracic ratio and decreased vascular pattern. Moreover, transthoracic and transesophageal echocardiography demonstrated normal left ventricle size, mild systolic dysfunction, ejection fraction=50%, severe right ventricle enlargement, mild systolic dysfunction, severe right ventricular hypertrophy, normal biatrial size, mitral valve and tricuspid valve being at the same level, prolaptic anterior mitral valve leaflet, no mitral stenosis,

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moderate central mitral regurgitation, no aortic stenosis and regurgitation, thickened and dome-shaped pulmonary valve with localized supravalvular narrowing (diameter = 8 mm), severe increased gradient (Peak Pressure Gradient (PPG) = 70 mmHg, Mean Pressure Gradient (MPG) = 50 mmHg mostly due to Supravalvular Pulmonary Stenosis (SPS), mild pulmonary insufficiency, main pulmonary artery = 2 cm, at least moderate highly eccentric tricuspid valve regurgitation (tricuspid valve regurgitant gradient (TRG) = 72 mmHg), and small residual inlet ventricular septal defect (4 mm) with left to right shunt. The stenosis site was located seven mm from the valve. Considering the symptomatic severe supravalvular stenosis, the patient was an appropriate candidate for catheterization and percutaneous transcatheter pulmonary balloon angioplasty. Right catheterization was performed via the femoral vein by hemodynamic evaluation and measurement of cardiac chambers pressure. In the injection of the right ventricle and pulmonary artery, the location of supravalvular stenosis as a discrete waist was noted. Right Ventricular Systolic Pressure (RVSP) was 80 mmHg, and pulmonary artery pressure at the distal part of the band was 25 mmHg. The internal diameter of the stenotic lesion was estimated to be 12 mm at its narrowest point, while the pulmonary valve annular diameter was 24 mm. The size of the balloon was chosen at least twice the diameter of the stenosis and smaller than the Pulmonary Valve (PV) annulus. Next, an Atlas balloon 20*40 mm was advanced across the stenotic lesion and was rapidly inflated. Loss of the waist occurred with continued inflation (Figure 1). The subsequent inflation failed to show a discrete waist, suggesting the successful alleviation of the stenosis. After dilation, the balloon catheter was removed, pressure and oxygen saturation measurements were repeated, and a post dilation right ventriculogram and pulmonary artery angiogram were performed using a pigtail catheter. The pressure gradient after successful balloon dilation revealed a systolic pressure gradient of 25 mmHg and RVSP = 50 mmHg, showing a 50% reduction in trans stenotic gradient. Post procedural Trans Thoracic Echocardiography (TTE) revealed mild pulmonary insufficiency (Figure 2).

The second patient was a 50-year-old man with no history of congenital heart disease with chronic dyspnea on exertion who was referred to the hospital for further examinations. During the physical examination, the heart rhythm was regular, the first heart sound was normal, a grade III/VI systolic murmur was heard in the left upper sternal border, and there was no diastolic murmur. The electrocardiograms showed normal sinus rhythm and right ventricular hypertrophy pattern. The chest x-ray also indicated normal cardiothoracic ratio and decreased vascular pattern. The transthoracic echocardiography showed TRG = 70 mmHg and severe pulmonary supravalvular stenosis with PPG = 60 and MG = 45 mmHg due to a web that was equidistant from the pulmonary valve annulus and pulmonary artery bifurcation and approximately 10 mm from the PV. The web thickness was measured to be about 3 mm based on echocardiography. Cardiac catheterization followed by balloon angioplasty was prescribed for the patient due to being symptomatic (Figure 3). Percutaneous cannulation of the femoral vein

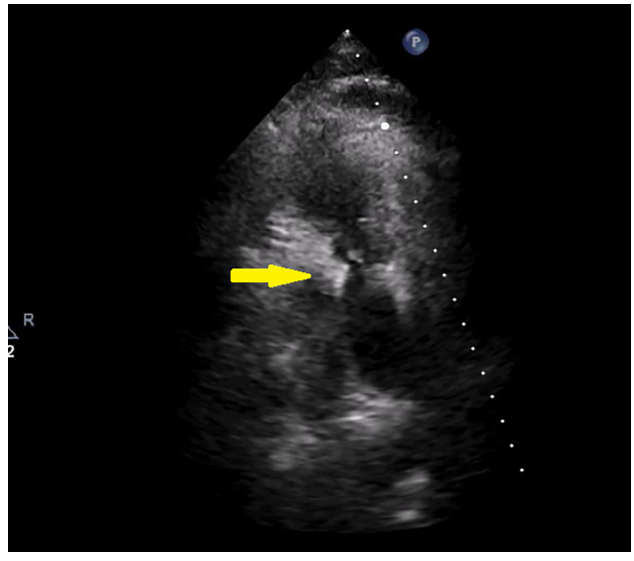

Figure 1. Pulmonary Supravalvular Stenosis in the First Patient

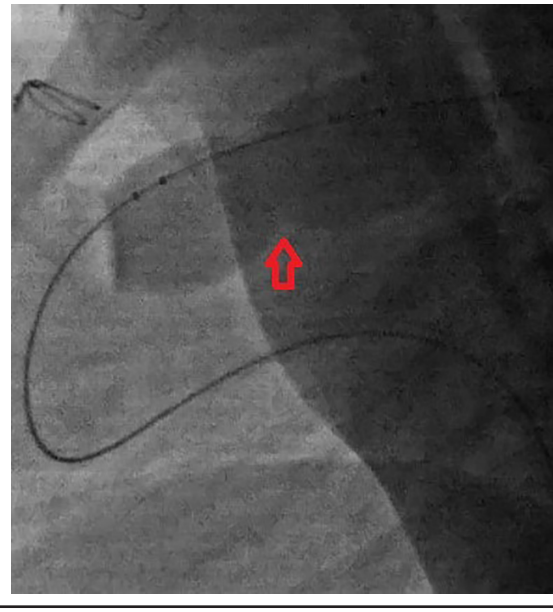

Figure 2. Supravalvular Pulmonary Stenosis in the Second Patient before the Intervention

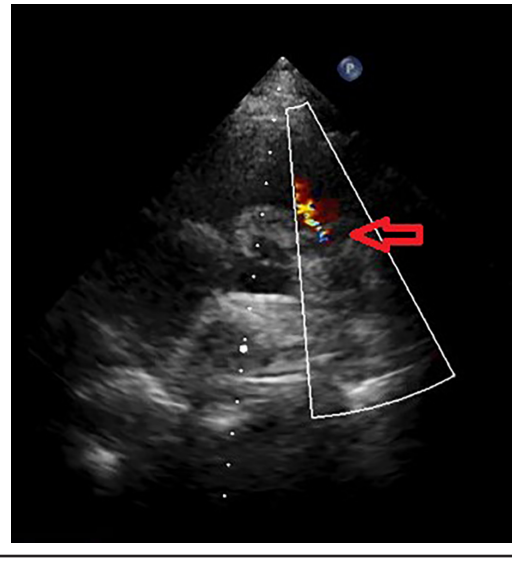

Figure 3. Pulmonary Insufficiency in the First Patient after the Procedure

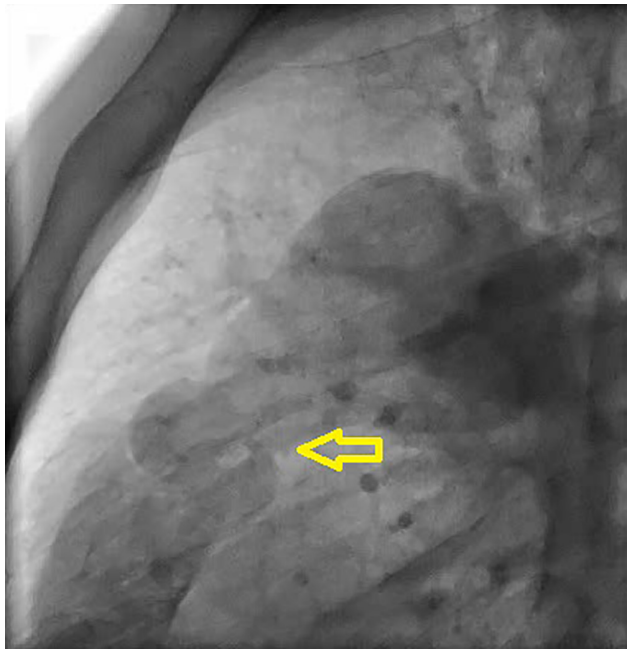

Figure 4. Mild to Moderate Pulmonary Insufficiency in the Second Patient after the Procedure

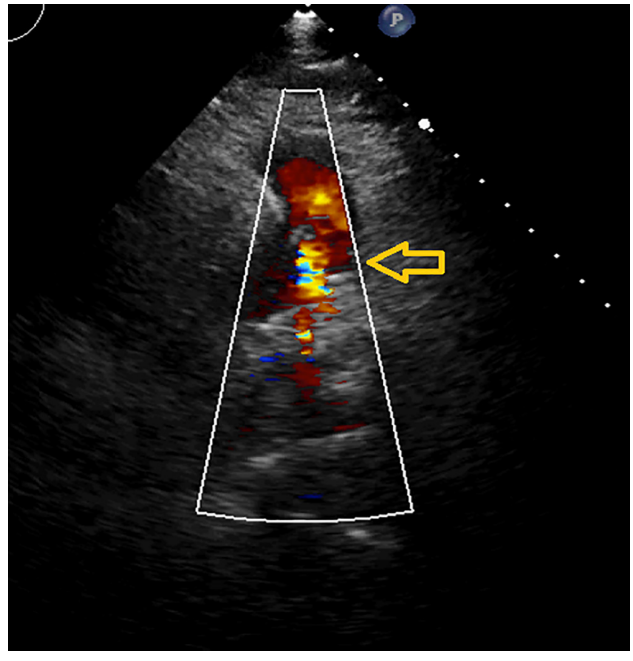

Figure 5. 5-2D Echocardiography of the Stenosis Site in the Second Patient after the Intervention

was performed. The internal diameter of the stenotic lesion was estimated to be 14 mm at its narrowest point, while the pulmonary valve annular diameter was 27 mm. The size of the balloon was chosen at least twice the diameter of the stenosis and smaller than the PV annulus. Then, a 24*40 mm Zmed balloon was used and inflated with the dilute contrast medium under pressure for approximately 20 seconds. Therefore, the waist of the supravalvular pulmonary stenosis disappeared. Using negative pressure, the balloon was deflated. The inflation was repeated twice, during which any air still in the balloon was aspirated into the inflating syringe. After balloon dilatation repetition, right ventricular and pulmonary artery systolic pressures were recorded. The right ventriculogram was obtained under the same conditions of the pre-dilatation angiogram. The gradient decreased from 45 to 20 mmHg. Post procedural TTE revealed mild to moderate pulmonary insufficiency (Figures 4 and 5).

3. Discussion

Pulmonary stenosis is a narrowing part between the right ventricle and the pulmonary artery that is divided into three categories: (a) valvular, (b) subvalvular, and (c) supravalvular (1). SPS is very rare and congenital, and can occur at one or several places, with the second one being more common. SPS may result from a range of congenital or acquired disorders including congenital rubella, Noonan, Williams, DiGeorge, and Down syndromes. Sometimes, SPS may be a consequence of surgeries such as mechanical pulmonary valve replacement that requires additional intervention therapy and Arterial Switch Operation (ASO) for transposition of great arteries. The incidence of post-operative SPS is soaring. Depending on the applied surgery technique and patient's age, the incidence of SPS has been reported to range from 17% to 55% in various studies. In addition, it seems that SPS has male predominance, with a male-to-female ratio of 1.8:1. The mean age of patients at operation was 13 ± 33 days (4).

Over the years, balloon dilatation procedures have been widely used to treat different congenital anomalies, which resulted in lower surgery rates and, in some particular cases, delay in surgeries. Nowadays, balloon dilatation is a known intervention for certain defects such as pulmonary valve stenosis (5). Due to the elasticity and recoil of the sinotubular constriction, balloon dilation of congenital SPS may be unsuccessful (1).

In the present study, balloon angioplasty reduced the gradient from 55 to 25 mmHg for the first patient. For the second patient, the gradient reduced from 45 to 20 mmHg with improved angiographic appearances. Hence, these two cases of SPS had balloon valvuloplasty accompanied by successful reduction in gradient. The day after the procedure, echocardiography was done for both patients and a moderate increase was observed in the gradient of the supravalvular pulmonary stenosis site (PPG = 50 mmHg, MPG = 28 mmHg). In addition, a mild to moderate pulmonary insufficiency was reported for the first patient. For the second patient, a moderate gradient increase in the supravalvular pulmonary stenosis site (PPG = 45 mmHg, MPG = 25 mmHg) and mild to moderate pulmonary insufficiency were observed. Both patients were treated with propranolol 20 mg daily. The symptoms of dyspnea were significantly reduced in both patients at two-week, two-month, and six-month follow-up visits.

Overall, the results of the present study indicated that balloon angioplasty as a widely available modality was an appropriate therapeutic option for congenital SPS. There were not many complications in this procedure.

3.1. Ethical Approval

A known therapeutic option was used.

3.2. Informed Consent

Written informed consent forms were obtained from the

patients.

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Authors' Contribution

Study concept and design: Z.K. and M.K.H. Critical revision of the manuscript for important intellectual content: M.K.H. and M.N. The original idea and the protocol: Z.K. and A.F. Abstracting and writing the manuscript: M.K.H. and M.N. Guarantor: M.N. and F.J. Development of the protocol and preparation of the manuscript: M.N. and F.J.

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Financial Disclosure

The authors declare that there is no conflict of interest.

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