Percutaneous Treatment of Pulmonary Valvular Stenosis and Secundum Type Atrial Septal Defect in a Young Adult

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ABSTRACT

Pulmonary valve stenosis (PS) and secundum type atrial septal defect (ASD) are common forms of congenital heart disease. While traditional treatment for PS and ASD is open surgery, percutaneous treatment of both diseases are widely used with high rates of success and lower complication. However, the combination of PS with ASD in the same patient is relatively rare, and the optimal strategy is debated. We performed consecutive percutaneous balloon pulmonary valvuloplasty and ASD occlusion in a young adult patient with severe congenital PS associated with a secundum ASD.

Key Words: Atrial septal defect; pulmonary valve stenosis

Genç Erişkinde Pulmoner Kapak Stenozu ve Sekundum Tip Atrial Septal Defektin Perkütan Tedavisi

ÖZET

Pulmoner kapak stenozu (PS) ve sekundum tip atrial septal defekt (ASD), konjenital kalp hastalıklarının genel formlarıdır. PS ve ASD'nin geleneksel tedavisi açık cerrahi iken, her iki hastalığın da perkütan tedavisi yüksek başarı oranları ve daha düşük komplikasyon oranları ile yaygın olarak kullanılmaktadır. Ancak aynı hastada PS ile ASD kombinasyonu nispeten nadirdir ve optimal stratejisi tartışmalıdır. Vakamızda sekundum ASD ile ilişkili şiddetli konjenital PS'si olan genç erişkin bir hastada ardışık perkütan balon pulmoner valvüloplasti ve ASD kapatması gerçekleştirdik.

Anahtar Kelimeler: Atriyal septal defekt; pulmoner kapak stenozu

INTRODUCTION

Among congenital heart diseases, pulmonary valve stenosis (PS) and secundum type atrial septal defect (ASD) are more common than others. ASD and PS were treated as open surgery in the past. Nowadays, transcatheter interventional therapy of ASD and PS are performed with high success rates[1-5]. However, the combination of PS with ASD in the same patient is relatively rare, and the optimal strategy is debated. We performed consecutive percutaneous balloon pulmonary valvuloplasty (PBPV) and ASD occlusion in a young adult patient with severe congenital PS associated with a secundum type ASD.

CASE REPORT

A 22-year-old male patient followed up with known pulmonary valve stenosis was admitted to our hospital with dyspnea and worsening exercise intolerance. Physical examination revealed systolic ejection murmur (grade 3) at the mesocardiac and pulmonary area. Electrocardiography showed sinus tachycardia with incomplete right bundle branch block. Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) revealed ejection fraction as 60%, dilated right chamber sides of heart, secundum type atrial septal defect (ASD) and severe pulmonary valve stenosis (max gradient: 74 mmHg). First, we planned to perform right heart catheterization (RHC) to verify pulmonary valve gradient and measure pulmonary vascular resistance. Then RHC was performed via the right femoral vein. In RHC, peak to peak pulmonary valve gradient was 70 mmHg, and PVR was 2 Wood units, with mean pulmonary artery pressure as 15 mmHg. Then in the same procedure, we

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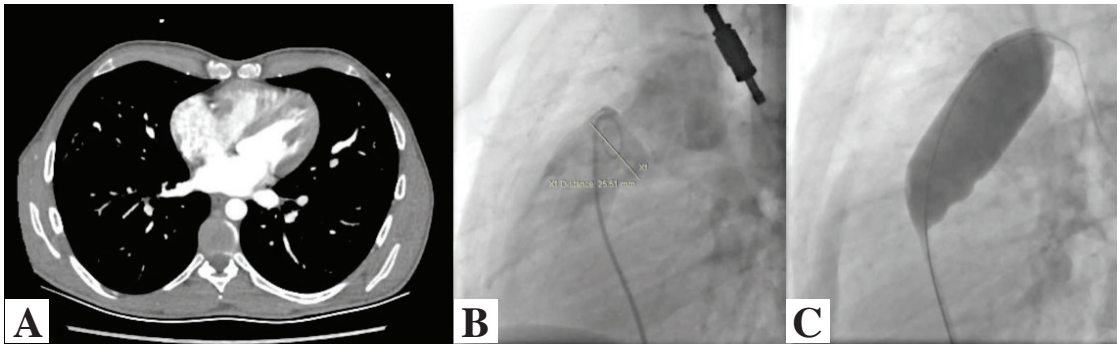

Figure 1. A. Computed tomographic image at the level of the pulmonary valve. **B.** Angiographic image of the pulmonary valve. **C.** Percutaneous balloon pulmonary valvuloplasty with a 2.5 x 15 mm pulmonary balloon.

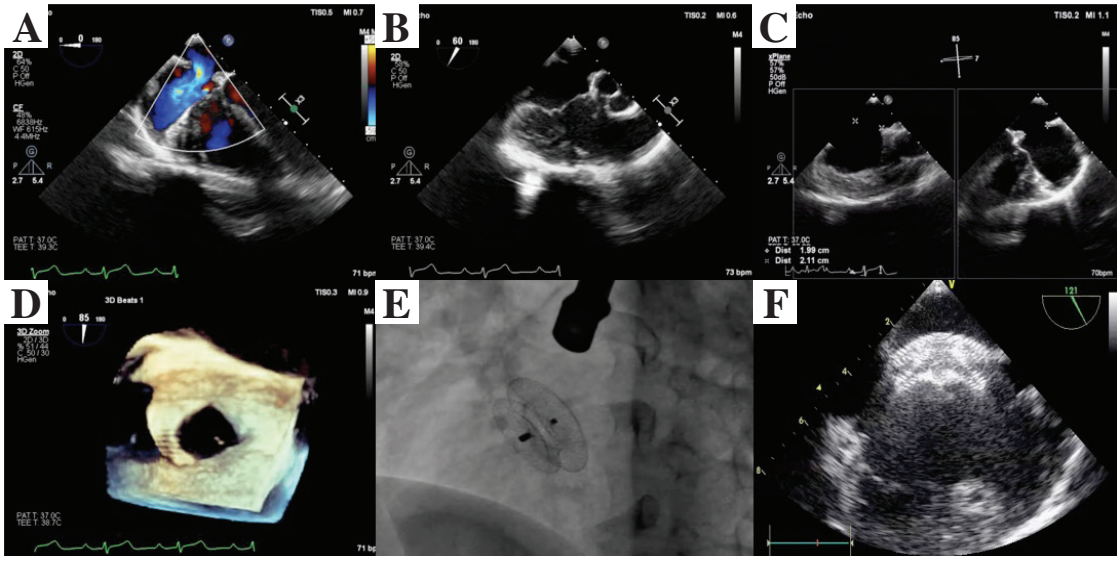

Figure 2. A-C. Transesophageal echocardiography imaging of the atrial septal defect. **D.** 3D transesophageal echocardiography imaging of the atrial septal defect. **E.** Angiographic image after percutaneous ASD closure. **F.** Transesophageal echocardiography imaging after atrial septal defect closure.

decided to perform PBPV. A pigtail catheter was placed in distal left pulmonary artery. Amplatz Superstiff wire was advanced through the pigtail to the distal pulmonary artery. The pigtail catheter was withdrawn, and a 2.5 × 15 mm pulmonary balloon was inflated in the appropriate position on the pulmonary valve. The procedure was terminated without any complications. Control TTE examination was performed after the procedure. There was a mild pulmonary regurgitation, and maximum gradient of the pulmonary valve was detected 18 mmHg. TEE examination was performed a month later after the procedure. Twenty-one mm secundum type ASD with a significant left to right shunt and mild pulmonary regurgitation were observed. Percutaneous ASD closure was planned to perform. The right femoral vein was cannulated with 6F sheath. A multipurpose catheter and hydrophilic guidewire were advanced through from the right atrium to the left atrium and left upper pulmonary vein. The Amplatz Superstiff wire was advanced in the pulmonary vein and the multipurpose catheter was removed. MemoPart occluder delivery system was advanced to the left atrium over the Amplatz Superstiff wire. Afterwards, a complete device occlusion of ASD was performed with a 22-mm Amplatzer septal occluder device. The patient was discharged from the hospital without any complications one day after the procedure, and is still followed up as asymptomatic.

DISCUSSION

The valvular type of PS is generally associated with ASD. The main pathological change of this disease is obstruction of the outflow tract of the right ventricle. When these two conditions are present simultaneously in a patient, a significant left-to-right shunt from ASD is often blocked due to the increased right ventricular pressure that originates from the outflow tract obstruction. It results in nearly normal pulmonary vascular pressure until adulthood[6,7]. Nowadays, percutaneous intervention techniques have become the method of choice for the treatment of these abnormalities in all age groups. PBPV

has become the initial choice for the treatment of PS in all age groups. When we look at the literature, we see two different approaches for transcatheter implementation. One of them is a combined approach in PBPV and ASD closure[8,9]. The other one is sequential PBPV and ASD closure. There is no clear consensus on this issue. Significant left to right shunt can increase pulmonary valvular gradient and results in pseudo-severe pulmonary stenosis. Thus, before performing PBPV, we should be sure that the valvular PS is clearly severe. On the other hand, severe PS can result in right to left shunt from ASD due to the increased right ventricle pressure. After performing PBPV, direction of the shunt and pulmonary vascular resistance should be evaluated carefully. In addition to having high success and long term clinical follow- up of both ASD closure and PBBV seperately, simultaneous procedures may be preferred compared to two-stepped procedures in the future. When we confirm combined adult congenital heart diseases and exclude pseudo-severe pulmonary stenosis, we need to consitute a specific algorithm in order to ensure cost-efectiveness and lower complication rates. After performing PBBV successfully ,we may have a opportunity to close ASD with the guidance of TEE and RHC at the same session. On the other hand, operator experience is quite important and requires more clinical cases. However,there are not enough clinical series regarding this issue. In our patient, we evaluated valvular PS as severe with the help of TEE, computed tomography and RHC. After performing PBPV, we performed RHC again and detected a significant left to right shunt from ASD with a low PVR. Then we performed percutaneous ASD closure. However, comparative studies are needed on this subject. During the procedure, whether or not there is any difference between the two groups in terms of complications should be checked. At the same time, it should be checked if there is any difference in the short and long-term clinical results between these two stepped or simultaneous procedures. We think that large-scale studies are needed to evaluate long-term results of different percutanoeus treatment modalities.

CONCLUSION

Percutaneous treatment of PS and ASD are widely used with higher success and lower complication rates. However, the management of combined adult congenital heart diseases is still controversial. Combined treatment may be performed simultaneously or as stepped procedures. According to patient characteristics and clinical determinants, there is still no consensus about optimal percutaneous treatment strategies.

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