Trans-Catheter Therapy of Lutembacher Syndrome: A Case Report

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Abstract- Lutembacher syndrome refers to the rare combination of a congenital atrial septal defect and acquired mitral stenosis. Traditionally, Lutembacher syndrome has been corrected by surgical treatment. We describe two patients treated percutaneouly with a combined Inoue balloon valvuloplasty and septal defect closure using the Amplatzer septal occlusion device.

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Introduction

Lutembacher syndrome, first described by Lutembacher in 1916 (1), is a combination of a congenital atrial defect (ASD) and acquired mitral stenosis (MS). Mitral stenosis serves to augment the left-to-right atrial shunt through the ASD, but decompression of the obstructed left atrium by the ASD attenuates the symptoms of MS (2).

The condition is usually treated surgically (3); however, both abnormalities are amenable to percutaneous transcatheter therapy (4-8). Combined transcatheter therapy was first described by Ruiz *et al*. (9) in 1992.

Herein we describe two cases of patients with Lutembacher syndrome who benefited from combined transcatheter therapy of balloon valvuloplasty for MS and device closure for ASD with an Amplatzer septal occluder.

Case Report

Case 1

A 50-year-old woman was admitted to the hospital complaining of dyspnea on exertion, palpitations, and chest tightness. The physical examination revealed accentuation of the first heart sound with an opening snap and a grade 2/6 mid-to-late diastolic rumbling murmur at the apex. Additionally, there were features suggestive of ASD, such as fixed splitting of the second heart sound with a grade 3/6 ejection systolic murmur at the pulmonary area and an early diastolic murmur at the lower left sternal border (tricuspid area). The exercise capacity was NewYork Heart Association class III.

An electrocardiogram showed atrial fibrillation, incomplete right bundle branch block, and right atrial hypertrophy. A chest radiogram revealed cardiomegaly with increased pulmonary vascularity, a prominent main pulmonary artery, and a cardiothoracic ratio of 0.64. Transthoracic and transesophageal echocardiography showed a medium-sized ASD secundum and moderate mitral stenosis. The mitral valve area was 1.24 cm^2 by planimetry and Doppler methods, and 1.48 using the Doppler pressure halftime method. There was trivial mitral regurgitation. The diameter of the ASD was estimated to be 28 mm.

Color flow mapping revealed a left-to-right shunt across the ASD and trivial mitral regurgitation. As both the ASD and MS appeared suitable for percutaneous treatment, concurrent transcatheter therapy was planned.

After informed consent was obtained, a routine right and left heart catheterization was performed. The pulmonary-to-systolic flow ratio (Qp/Qs) was 4.8/1 s, as measured by Fick's principle. The pulmonary pressure was 40/10 mmHg with a mean of 20 mmHg. The mean pressure of the left atrium was 25 mmHg, and the mean diastolic pressure gradient across the mitral valve was 21 mmHg.

The stretched diameter of the ASD was determined to be 30 mm using a sizing balloon catheter (Figure 1). The mitral valve was crossed and dilated with an Inoue balloon catheter. Two inflations of 25 and 26 mm were performed under transthoracic echocardiography and fluoroscopic guidance (Figure 2).

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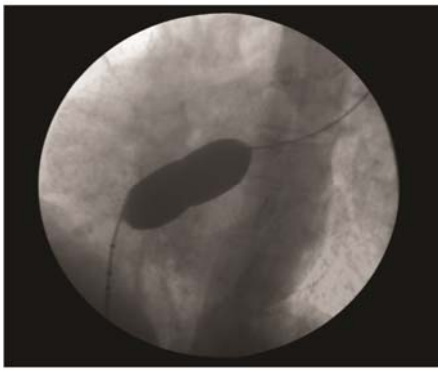

Figure 1. Shows measurement of the stretched diameter of the atrial septal defect with a sizing balloon catheter.

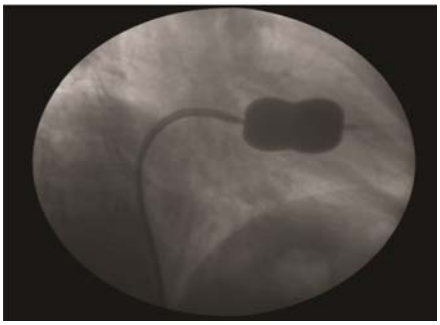

Figure 2. Shows balloon mitral valvutomy using an Inoue balloon, disappearance of the waist indicates commissural separation and an excellent result.

After valvuloplasty, the mean pressure of the left atrium decreased to 13 mmHg and the mean diastolic mitral pressure gradient decreased to 6 mmHg. The Qp/Qs decreased to 3.3/1. Under transthoracic echocardiography and fluoroscopy guidance, the ASD was closed with a 32 mm Amplatzer septal occluder (Figure 3).

Subsequently, the right catheterization and oximetry run was repeated.

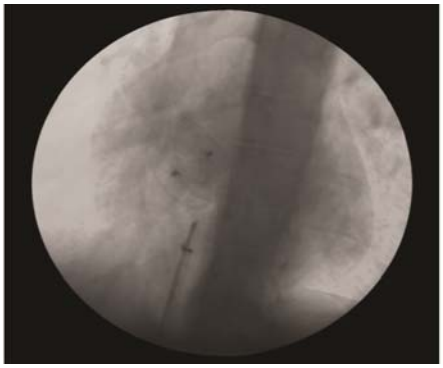

Figure 3. Left anterior oblique view showing the Amplatzer septal occluder was fully deployed and was released by unscrewing

There was trivial O_2 step-up at the right atrial and right ventricular levels. Immediately after the procedure, transthoracic echocardiography showed a trivial mesh leak residual shunt. The fluoroscopy and procedure times were 17 and 64 minutes, respectively. Transthoracic echocardiography repeated 24 hour after the procedure revealed no residual atrial shunt, appropriate deployment of the Amplatzer device, and a mitral valve area of 2.5 cm. The patient was discharged uneventfully the following day. There were no complications during 35 months of follow-up.

Case 2

A 52-year-old female was referred to the Department of Cardiology for evaluation and catheter therapy for MS and ASD. She complained of a recurrent cough, palpitations, and dyspnea on exertion. She had several episodes of bronchopnemonia during the recent two years. The physical examination revealed features of MS and ASD.

A chest x-ray revealed cardiomegaly and pulmonary plethora. Transthoracic and transesophageal echocardiography showed a medium-sized secundum ASD (26 mm) moderate MS, and mild mitral regurgitation (MR). The mitral valve was pliable and without calcifications. The mitral valve area was 1.2 cm^2. Right femoral vien and artery access were obtained and coronary angiography was performed, revealing no significant coronary stenosis. The pulmonary artery pressure was 43/15 mmHg with a mean pulmonary artery pressure of 25 mmHg and a Qp/Qs of 3.4/1. After arterial access, 5000 unit of intravenous heparin was given and percutaneous balloon mitral valvuloplasty was performed using a 26 and 28 Inoue balloon. The mitral valve was crossed and dilated twice with balloon catheters of 26 and 27 mm in diameter. The post-valvuloplasty mitral valve area increased from 1.2 to 2.5 cm^2 and there was mild MR by transthoracic echo cardiography. The transmitral valve gradient fell from 18 to 11 mmHg. Right and left catheterization was then performed and calculation of the left-to-right shunt across the ASD using oximetry yielded a Qp/Qs of 2.7/1. The stretched diameter of the ASD was determined using a sizing balloon and was found to be 30 mm.

Under transthoracic echocardiography and fluoroscopy guidance, ASD closure was successfully performed using a 32 mm Amplatzer septal occluder. The fluoroscopy and procedure times were 15 and 59 minutes, respectively. Transthoracic echocardiography revealed appropriate deployment of the Amplatzer

device with mild MR and no residual shunt. There were no complications during 25 months of follow-up.

Discussion

The original case describing Lutembacher syndrome involved a 61-year-old woman who had been pregnant 7 times (1). Female predominance has been noted in both ASD and MS, and thus Lutembacher syndrome has a predilection for females. The incidence of MS in patients with ASD is 4% and conversely, the incidence of ASD in patients with MS is 0.6- 0.7% (10).

In Lutembacher syndrome, MS augments the left-to-right shunt through the ASD, while the non-restrictive ASD decompresses the left atrium, reducing the diastolic mitral pressure gradient (11). The pressure half-time method consistently overestimates the mitral valve area. The extent of overestimation is greater in patients with a larger atrial shunt. The hemodynamic pressure half-time is independent of the mitral valve area, chamber compliance, and peak transmitral gradient. The hemodynamic pressure half-time is dependent on the magnitude of the atrial shunt, although the correlation obtained is only fair (r=0.61), indicating that the Doppler pressure half-time method is an inaccurate measure of the mitral valve area whenever an atrial shunt coexists with MS. Planimetry and the Doppler continuity equation methods yield an accurate mitral valve area in the Lutembacher syndrome (12, 13).

The experience with transcatheter treatment of the Lutembacher syndrome is small. (2,3,9,14-17) The use of percutaneuus treatment of the Lutembacher syndrome was first described by Ruiz *et al*. (9) in 1992.

Combined umbrella closure of ASD with Lock's Clamsher occluder in conjunction with mitral and aortic balloon valvotomies was carried out as a palliative rescue procedure in a 43-year-old female with Lutembacher syndrome and severe pulmonary hypertension to improve the risk prior to surgical repair. She refused surgery and died suddenly a few weeks after the percutaneous repair. Subsequent reports demonstrated the feasibility of percutanous balloon valvuloplasty and ASD closure using a variety of balloon catheter techniques and an ASD closure device. Successful combined transcatheter therapy was first described by Joseph *et al*. (14) in 1999. They used the Amplatzer septal occluder for ASD and the Joseph mitral balloon catheter for MS. Since then, several cases have been reported (2,3,15,16,18-20).

Currently, the Inoue balloon is most widely used for percutaneous balloon mitral valvuloplasty, and the Amplatzer atrial septal occluder for closure of an ASD (17). The successful combined use of the two devices was first described by Hau *et al*. (15).

The advantages of percutaneous correction over traditional surgical correction include the avoidance of complications associated with open heart surgery, general anesthesia, and blood transfusion, and quicker recovery with a shorter hospital stay.

One obvious consequence of this approach is that repeat transseptal procedures cannot the performed. In the event that a patient develops mitral valve restenosis, either surgery or a transarterial retrograde non-transseptal technique in which entry into the left atrium is achieved retrograde via the ventricle (17).

The cases described herein are the first and second cases of patients with Lutembacher syndrome who received combined transcatheter therapy in Iran. We used the Inoue balloon antegrade through an existing ASD to dilate the stenotic mitral valve and closed the ASD with an Amplatzer septal occluder.

In summary, combined transcatheter therapy of Lutembacher syndrome is safe and effective, with a short hospital stay and without the complications of open heart surgery.

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