

Transcatheter Treatment of Lutembacher's Syndrome

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ABSTRACT

Lutembacher's syndrome is a rare combination of atrial septal defect (ASD) and rheumatic mitral stenosis. Traditionally, this condition is treated surgically. We present a case of Lutembacher's syndrome that was successfully treated with percutaneous transcatheter mitral commissurotomy (PTMC) using the Inoue balloon. Closure of ASD was successfully achieved with an Amplatzer atrial septal occluder under transthoracic echocardiogram (TTE) guidance without general anesthesia.

Key words: *Lutembacher's syndrome. Atrial septal defect. Percutaneous transcatheter mitral commissurotomy.*

INTRODUCTION

Lutembacher's syndrome is a rare combination of acquired mitral stenosis and congenital atrial septal defect (usually an ostium secundum type), resulting in left-to-right shunt.¹ It occurs predominantly in women and in young adults but may also present in the elderly. Patients may remain asymptomatic for many years and may have history of rheumatic fever. Symptoms vary according to the size of ASD. Symptoms are usually due to pulmonary congestion and due to right heart failure. These patients have predisposition to atrial arrhythmias and atrial fibrillation is very common. Symptoms caused by mitral stenosis are less frequently seen in Lutembacher's syndrome, than in isolated mitral stenosis but are more common in Lutembacher patients with a small ASD.

CASE REPORT

A 17 years old female presented to the institute with history of shortness of breath and decreased exercise tolerance for 02 years. On examination, she had small volume, regular pulse of 96/ minutes. Examination of precordium showed left parasternal heave and tapping apical impulse. On auscultation, she had loud first heart sound, opening snap loud P₂ component of second heart sound and grade II/IV mid-diastolic rumbling murmur. Transthoracic echocardiogram (TTE) showed severe non-calcific pulse mitral valve stenosis with mitral valve area 0.9 cm² by planimetry. The Wilkins echo score was 6. Transesophageal echocardiogram

(TEE) showed the diameter of ASD of 11 x 14.5 mm. Cardiac catheterization revealed left to right shunt at atrial level with a ratio of pulmonary to systemic flow of 2.6:1. Peak systolic pulmonary artery pressure was 42 mmHg. Mitral valve was dilated using Inoue balloon catheter with single inflation of 24 mm. End diastolic trans mitral gradient reduced from mean 24 mmHg to 01 mmHg. Mitral valve area was increased to 1.9 cm². After balloon dilatation, no mitral regurgitation was seen. The stretched diameter of ASD (secundum) was estimated to be 14.5 mm. An amplatzer ASD occluder of 20 mm was deployed successfully under TTE guidance with no residual left to right shunt. At three months follow-up, she showed good symptomatic improvement. The mitral valve area by planimetry was 1.9 cm² with no significant regurgitation or left to right shunt at atrial level and normal pulmonary artery pressure.

DISCUSSION

Lutembacher's syndrome refers to a rare combination of congenital ASD and acquired rheumatic mitral stenosis. This combination was first described by Rene Lutembacher, a French physician, in 1916. Subsequently, few cases were described with severely acquired mitral stenosis with high left atrial pressure and presumed stretching of a patent foramen ovale leading to left-to-right shunting.²

The hemodynamic effects of these co-existing conditions are very interesting. Mitral stenosis augments the left to right shunt through the ASD, which decrease the left atrial pressure resulting in decrease in trans-valvular mitral gradient, resulting in an ameliorating effect on the clinical expression of mitral stenosis. In this condition, mitral valve area should be determined by planimetry because the Doppler half-time method is inaccurate and may lead to underestimation of the severity of mitral stenosis in patients with Lutembacher's syndrome.³ This condition has traditionally been treated by open heart surgery. With the introduction of

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transcatheter closure of ASD and percutaneous balloon, mitral valvuloplasty, now Lutembacher's syndrome can be treated percutaneously.¹

The use of percutaneous treatment of Lutembacher's syndrome was first described by Ruiz *et al.* in 1992.⁴ Patient underwent combined umbrella closure of ASD with a lock clamshell occluder for 20 mm ASD along with a single balloon aortic valvuloplasty and a double balloon mitral valvuloplasty. This therapy was used as a bridge to surgery but patient suddenly died after few weeks of percutaneous treatment before surgical repair could be done. Subsequently, the feasibility of percutaneous balloon mitral valvuloplasty and ASD closure, using different types of balloon catheter techniques and ASD closure devices, were demonstrated.⁵ Currently, the Inoue balloon is most widely used for mitral valvuloplasty and Amplatzer closure device for percutaneous ASD closure. The successful combined use of these two devices was first described by Chau *et al.*⁶ We performed device closure of ASD under TTE guidance and local anesthesia. The advantages of percutaneous correction over surgical correction includes the avoidance of complications of open heart surgery, quick recovery and short hospital stay, but if patient develops mitral valve restenosis, either surgery or retrograde percutaneous therapy would be required.

Iatrogenic Lutembacher's syndrome is also a well-known entity that was first described by Dr. John Ross Jr, and colleagues.⁷ Later on, iatrogenic Lutembacher's syndrome was seen in patients who had balloon valvuloplasty of the mitral valve by transatrial septal approach.^{8,9} There appear to be two causes of a persistent ASD after balloon valvuloplasty of mitral valve by transatrial septal approach, that is, inadequate procedure success, resulting in persistently increased left atrial pressure and a large initial hole size. A large initial septal hole size might be related to certain

technical misadventures. First, tail of the balloon might be across the atrial septum during inflation of the balloon, resulting in further enlargement of the atrial septostomy. Cequier *et al.*⁹ also noted that small left atrium is more likely to be associated with persistent ASD. Secondly, inadequate deflation of the balloon before withdrawing it may also result in persistent ASD after the procedure.

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