

Non-surgical correction of Lutembacher syndrome

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ABSTRACT

Lutembacher syndrome is the combination of congenital atrial septal defect and acquired mitral stenosis. The condition is usually treated surgically. We describe a patient treated percutaneously with a combined Inoue balloon valvuloplasty and septal defect closure using the Amplatzer septal occlusion device.

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Lutembacher syndrome is a combination of congenital atrial septal defect (ASD) and acquired mitral valve stenosis.¹ We describe a patient with Lutembacher syndrome who underwent complete percutaneous treatment for this condition.

Case Report. A 42-year-old lady presented with a 2-year history of progressive shortness of breath. On examination, the weight was obese, 105 kg and height 156 cm. The pulse was 64 beats per minute, blood pressure was 143/84 mm Hg, the jugular venous pulse was not elevated. A diastolic murmur was heard in the apical area and no other abnormalities were heard. The chest was clear and there was no lower limb edema. A transthoracic echocardiogram showed a secundum ASD measuring 9 mm. The mitral valve leaflets were thickened, the subvalvular apparatus was mildly diseased and no calcification of the valve was noted. The mitral valve area measured 1 cm² by planimetry and pressure half time and the gradient across the valve was 8 mm Hg. Mild-moderate mitral regurgitation was noted. The left ventricular size and function was normal. The right ventricle measured 39 mm and the right atrium measured 54 mm. The patient was taken to the cardiac catheterization laboratory where, under

general anesthesia, a trans-esophageal echocardiogram (TEE) was performed. This showed the mitral stenosis and a secundum ASD measuring 9 mm with adequate rims on all sides (**Figure 1**). A mitral regurgitation (grade 2 of 4) was noted. Right heart catheterization study was performed. The oxygen saturation was 64% in the superior vena cava and 76% in the main pulmonary artery. The left to right shunt was 1.8:1. The pulmonary artery systolic pressure was 57 mm Hg. The mean left atrial pressure was 20 mm Hg with a trans-mitral gradient of 9 mm Hg.

A 7 French (F) multipurpose catheter was introduced into the left atrium via the right femoral vein and then the right atrium across the ASD. Intra-venous heparin (5000 units) was administered. Using the Inoue wire, a size 28 Inoue balloon catheter (Toray) was placed across the mitral valve and a single dilation of 26 mm was performed. The balloon was removed and hemodynamic measurements immediately following the balloon dilation revealed a significant drop in the trans-mitral gradient, which was reduced to 3 mm Hg, and an increase in the mitral valve area to 1.5 cm². The mitral regurgitation increased from grade 2/4 to grade 3/4. Following the balloon mitral valvuloplasty a 7F multipurpose catheter was placed in the left upper

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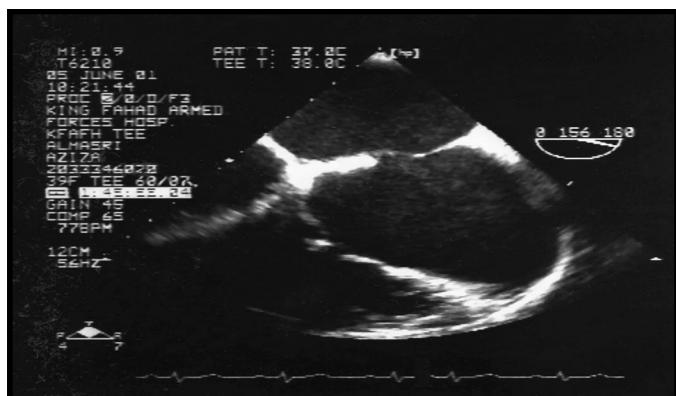


Figure 1 - Trans-esophageal echocardiogram shows the secundum atrial septal defect and the thickened rheumatic mitral valve.

pulmonary vein (LUPV) across the ASD. A 0.035-inch wire was placed into the LUPV. A 24 mm Amplatzer sizing balloon was used to measure the ASD over the wire. The stretched balloon diameter measured 11 mm, however after the balloon dilation, the defect measured 14 mm on TEE. A 17 mm Amplatzer septal occluder (AGA Medical, Golden Valley, MN) was selected to close the defect. An 8F Amplatzer delivery sheath (AGA Medical) was placed over the wire into the LUPV. The Amplatzer septal occluder was deployed across the septum. Complete closure with no shunt was verified by TEE. At 8 months of follow-up the patient was asymptomatic and doing well.

Discussion. In Lutembacher syndrome the combination of mitral stenosis and ASD augments the left-to-right shunt and leads to right sided failure.² Surgical treatment of this condition has been the method of choice. While percutaneous mitral commissurotomy has been well established for a long time, transcatheter closure of the ASD has only recently been described.^{3,4} Percutaneous treatment of Lutembacher syndrome can be performed by combining balloon valvuloplasty and the transcatheter ASD closure. The first such procedure described was in a terminal patient with pulmonary hypertension and severe mitral and aortic valve disease

who ultimately died.⁵ Subsequently Joseph et al⁶ and Chau et al⁷ have described successful transcatheter treatment in 2 young patients.

To our knowledge this is the first case report of a percutaneous treatment of Lutembacher syndrome in the Middle-East by combined Inoue balloon mitral valvuloplasty and Amplatzer device closure of the ASD. Although the mitral regurgitation increased from 2/4 to grade 3/4 after dilation. The decrease in gradient made the patient tolerate the mitral regurgitation well. In carefully selected patients this approach is a less invasive alternative to open heart surgery and avoiding the trauma, scarring and complications associated with thoracotomy and heart-lung bypass machine.

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