

A case report: Percutaneous transcatheter treatment of Lutembacher syndrome

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ABSTRACT : Lutembacher syndrome is a rare combination of Congenital ostium secundum Atrial Septal Defect (ASD) and acquired Rheumatic Mitral stenosis. The gold standard treatment for this condition is the surgical closure of ASD and mitral valve replacement. Here, we report a case of Lutembacher syndrome treated percutaneously with a combined ASD closure with Cocoon septal occluder and balloon mitral valvotomy.

INTRODUCTION:

Lutembacher's syndrome, a combination of Congenital Atrial Septal Defect (ASD) and acquired Mitral stenosis ^[1], has traditionally been treated by open heart surgery ^[2]. Percutaneous treatments of ASD and that of mitral stenosis, have been separately described more than 15 years ago ^[3,4]. Whereas transcatheter device closure of ASD has yet to gain wide acceptance, percutaneous balloon valvuloplasty has become an established method of treatment of mitral stenosis ^[5-9].

We present a patient with Lutembacher's syndrome in whom definitive percutaneous treatment was successfully accomplished by concurrent transcatheter device closure of a secundum ASD and balloon valvuloplasty of rheumatic mitral stenosis.

CASE REPORT:

A 30 year old female presented with a 6 months history of dyspnoea which was initially on exertion and had gradually progressed to dyspnoea at rest. At the time of admission to hospital she was in New York Heart Association functional class IV. She had no history of any diagnosed cardiac ailment and was not on any medications. Physical examination revealed auscultatory features of mitral stenosis (loud first heart sound, opening snap, and mid-diastolic murmur at the apex). Additionally there were features suggestive of ASD (wide splitting of the second heart sound with systolic ejection murmur at the pulmonary area). The electrocardiogram showed sinus rhythm, right axis deviation, and incomplete right bundle branch block. The chest X-ray revealed cardiomegaly and pulmonary plethora.

Transthoracic echocardiography (TTE) showed a 16 mm ostium secundum ASD with severe rheumatic mitral stenosis [Figure 1,2,3]. The mitral valve area (MVA) was 0.8 cm² by planimetry. The mitral valve had preserved mobility, no calcification and a morphologic score of 7 out of 16. Doppler studies revealed trivial mitral regurgitation, mild aortic regurgitation and moderate pulmonary hypertension. Color flow mapping revealed a left-to-right shunt across the ASD. As both the ASD and MS appeared suitable for percutaneous treatment, concurrent transcatheter therapy was planned.

After informed consent, mild sedation and antibiotic prophylaxis, Right Femoral artery and vein access was secured and 2,500 units of Heparin were administered. Routine left and right heart catheterization was planned.

Right femoral arterial puncture was taken and 5F pigtail catheter was introduced through a 5FRENCH sheath. Mulin sheath was directly crossed in the left atrium through the ASD.

The mitral valve was crossed and with an Accura balloon of 26mm size, mitral valve was dilated using 25.5ml saline with excellent results [Figure 4]. The mean gradient fell from 13 mmHg to 5 mmHg after mitral balloon valvotomy and a mild MR was observed.

The ASD was crossed with 9FRENCH, AGA delivery sheath and balloon sizing was done which revealed a defect of 20mm. A 24m Cocoon ASD device was introduced and deployed across the defect. Post deployment, the placement of the device was checked under fluoroscopy and confirmed through guidance of TEE. The ASD was observed to be totally closed and no shunt was seen in the color Doppler flow image performed with TTE.

There were no complications during the procedure and patient was discharged 2 days later.

Table 1 –

	Pre BMV	Post BMV
Heart Rate	64 / min	70 / min
Aorta	100/60 mmhg	110/70mmhg
Left Atrium	32/16 mmhg	10/6 mmhg
Mitral Regurgitation	Trivial	Mild
Left Ventricle	100/6	100/7

DISCUSSION:

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

In patients with Lutembacher syndrome MS is known to increase the left to- right shunt caused by ASD, whereas ASD plays a role in the improvement of left atrial pressure and decrease in the diastolic mitral pressure gradient. As a result, mitral valve measurements in patients with Lutembacher syndrome should be identified by planimetric or continuity equation; and the Doppler pressure should not be used due to faulty half time.⁽¹¹⁾

Ruiz et al. ⁽¹²⁾ first performed a combined percutaneous treatment on a patient with Lutembacher syndrome in 1992. There is currently no randomized or cross-sectional study which investigates combined percutaneous treatment in patients with Lutembacher syndrome. However, the success rate of combined percutaneous treatment in case studies is very high and no short or long term complications have been reported.

The most important complication of the procedure is embolization of the septal occluder device. The procedure of retrieval and re-implantation is implemented when there is embolization of small devices; however surgical interventions are required for large devices⁽¹³⁾.

The success rate of the other percutaneous intervention with BMV by the transseptal Inoue balloon technique was 99%. The life-threatening complication rate (death, puncture in the left ventricle, and stroke) was very low (0%-0.5%) and the risk of developing important and severe complications (grade 3-4 mitral regurgitation, cardiac tamponade and thromboembolism) were found to be lower when compared to the other procedures⁽¹⁴⁾.

The combined percutaneous treatment technique is reported to reduce the mortality and morbidity risk associated with cardiac surgery, to decrease physiologic trauma due to thoracotomy scar and to reduce the surgery related length of stay in the hospital in patients with Lutembacher syndrome.

We report a successful definitive transcatheter treatment of Lutembacher syndrome in a symptomatic young lady.

In conclusion, transcatheter treatment of Lutembacher syndrome can be the procedure of choice in patients with mitral valve stenosis and ostium secundum ASD suitable for percutaneous treatment with good short and long-term results.

FIGURES:



Fig 1 - Trans thoracic echocardiography (TTE) in apical fourchamber view shows a 16 mm ostium secundum ASD with good margins, with left to right shunt. Mitral valve is thickened

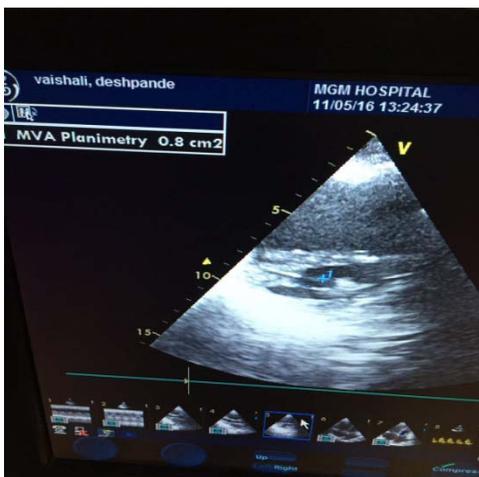


Fig 2 – Severe mitral stenosis (MVA – 0.8 sq cms)



Fig 3 – Mild Tricuspid Regurgitation jet.

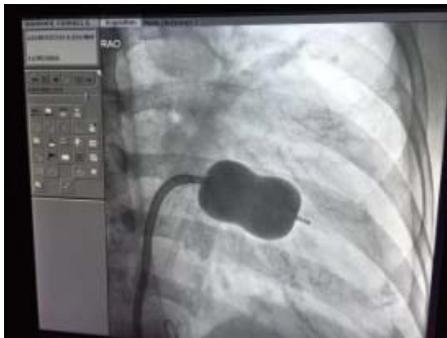


Fig 4 - Stepwise Acura balloon dilatation of mitral valve



Fig 5 - 24 mm ASD occlude device deployed in ostium secundum ASD

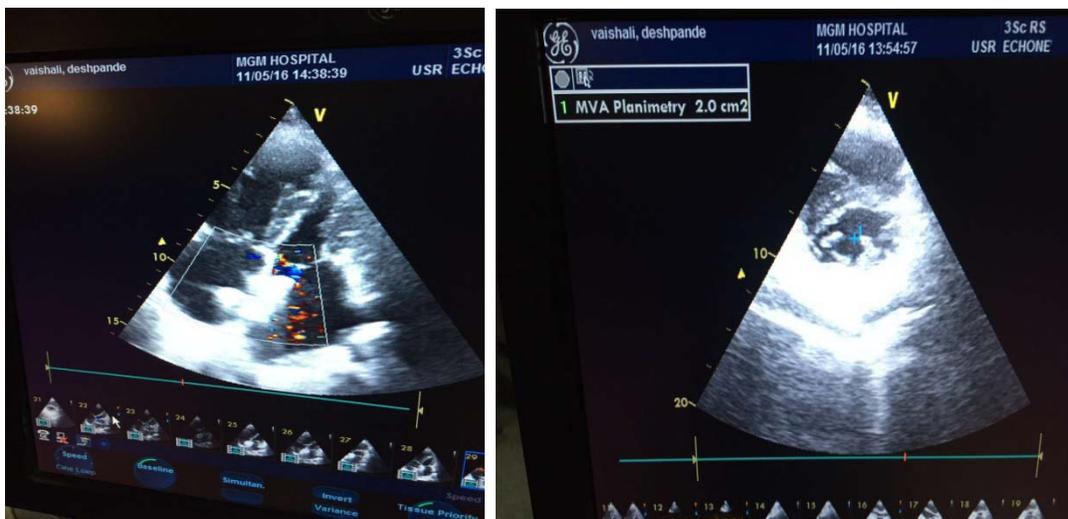


Fig 6 – Post procedure – Successful ASD device closure with no flow seen across the device.
Successful BMV with valve area of 2.0 sq cms (By Planimetry)

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