Abstract
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Introduction
Lutembacher syndrome for the first time described by Rene Lutembacher in 1916. Percutaneous transcatheter treatment can be done in selected cases, although surgery is the preferred treatment for these patients. In a patient of Lutembacher syndrome with severe pulmonary hypertension, Ruiz et al did first combined transcatheter treatment by ASD closure with Lock’s clamshell occluder and balloon mitral and aortic valvotomies.

Case Report
A 18-year-old male presented to us with six month history of dyspnea on exertion and palpitations. Patient gave history suggestive of rheumatic heart disease, diagnosed at peripheral hospital three years back and was on penicillin prophylaxis. His blood pressure was 94/60 mm Hg with a regular heart rate of 76 beats per minute. On auscultation loud first heart sound, fixed splitting of second heart sound, grade 3/6 ejection systolic murmur at pulmonary area and middiastolic murmur at apex was heard. Routine laboratory investigations were within normal limits. His electrocardiogram showed sinus rhythm, tall P waves, right ventricular hypertrophy and incomplete right bundle branch block. X-ray chest showed cardiomegaly with pulmonary plethora (Figure 1). Transthoracic echocardiogram was performed which revealed dilated right atrium and right ventricle. There was rheumatic mitral stenosis with mitral valve area of 1.1 cm² by planimetry, a large ostium secundum ASD with left to right shunt and adequate rims. Left ventricular function and all other valves were normal. Transesophageal echocardiogram (TEE) was done to rule out left atrial appendage clot and evaluation of ASD. In TEE it was found to be a 30 mm ASD with superior vena cava rim of size 12 mm and inferior vena cava rim of size 7 mm (Figure 2) and no left atrial or left atrial appendage clot. There was no mitral regurgitation (MR).

Diagnosis of Lutembacher syndrome was made and the patient was stabilized with diuretics. Percutaneous transcatheter treatment was planned. Right ventricular catheterization study showed normal right ventricular and pulmonary arterial pressures indicating adequate diuresis. Percutaneous trans-luminal mitral commissurotomy (PTMC) was done using 28 mm Accura balloon catheter introduced through ASD (Figure 3). Pre and post PTMC hemodynamic and oximetry parameters are shown in Table 1. The ASD closure was done with 38 mm size Amplatzer ASD closure device (Figure 4). The whole percutaneous transcatheter procedure of Accura balloon mitral commissurotomy and Amplatzer ASD device closure was done in a single stage under general anaesthesia. Post procedure transesophageal and transthoracic echocardiogram showed ASD device in situ with no residual shunt. Mitral valve was opening well with mitral valve area of 1.8 cm² by planimetry, without mitral regurgitation (Figures 5, 6). The patient was then discharged after two days in stable condition.

Discussion
Lutembacher syndrome is more common in females due to female preponderance of both ASD and MS. The incidence of MS in patients with ASD is 4% and the incidence of ASD in patients with MS is 0.6-0.7%. Altered clinical and hemodynamic features are observed in Lutembacher syndrome because combined ASD and MS affect each other. MS augments the left to

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atrial septal defect and presence of adequate rims (< 5 mm) around the left atrial thrombus, (2) absence of contraindications are: (1) presence of pulmonary hypertension, except in patients with PTMC, (3) any degree of pulmonary valve morphology favorable for symptomatic moderate to severe MS; 8 (4) persistent ASD after mitral occluder probably due to large size; 7 (5) embolization or slippage of septal room for mitral valve assessment; 7 (6) lack of expertise. 5,6

Mitral regurgitation

Heart rate

Sa O₂

Mv O₂

Pa O₂

Mid RA O₂

Qp/Qs

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Pre PTMC</th>
<th>Post PTMC</th>
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<tbody>
<tr>
<td>Mean LA pressure</td>
<td>20 mm Hg</td>
<td>6 mm Hg</td>
</tr>
<tr>
<td>Aorta pressure</td>
<td>110/70 mm Hg</td>
<td>118/70 mm Hg</td>
</tr>
<tr>
<td>LV pressure</td>
<td>100/6 mm Hg</td>
<td>110/6 mm Hg</td>
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<tr>
<td>Mitral valve area</td>
<td>1.1 cm²</td>
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<td>Mitral regurgitation</td>
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<td>Heart rate</td>
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<tr>
<td>Mv O₂</td>
<td>72%</td>
<td>72%</td>
</tr>
<tr>
<td>Pa O₂</td>
<td>91%</td>
<td>86%</td>
</tr>
<tr>
<td>Mid RA O₂</td>
<td>92%</td>
<td>87%</td>
</tr>
<tr>
<td>Qp/Qs</td>
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<td>2.1</td>
</tr>
</tbody>
</table>

Sa O₂, Mv O₂, Pa O₂, and RA O₂ are systemic arterial, mixed venous, pulmonary venous, pulmonary arterial and right atrial blood oxygen saturations, respectively. Qp and Qs are pulmonary and systemic blood flow respectively.

of tail of balloon atrial septum during inflation (increasing atrial septostomy) or inadequate deflation of balloon prior to withdrawing it. 9 Despite these problems, percutaneous treatment of Lutembacher syndrome overcomes the complications associated with open surgery, with rapid recovery and shorter hospital stay. The case reported by us was treated successfully by Accura balloon mitral commissurotomy and Amplatzer ASD device closure in a single stage. Therefore percutaneous transcatheter treatment of Lutembacher syndrome is reasonable and effective in selected group of patients, avoiding complications associated with open heart surgery.

Table 1: Pre and post PTMC hemodynamic and oximetry parameters

Fig. 3: Percutaneous trans-luminal mitral commissurotomy (PTMC) using 28 mm Accura balloon

Fig. 4: Amplatzer 38 mm ASD closure device deployed in ASD

Fig. 5: Post procedure transthoracic echocardiogram shows ASD device in situ with no residual shunt

Fig. 6: Post procedure transthoracic echocardiogram shows mitral valve area of 1.8 cm² by planimetry

References