## **Case Report**

DOI: 10.5455/2349-2902.isj20150527

# Lutembacher's syndrome: a study of 3 cases

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Received: 09 April 2015 Accepted: 22 April 2015

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## **ABSTRACT**

Lutembacher's syndrome is rare clinical condition which comprises of congenital atrial septal defect and acquired mitral stenosis. We study three cases of this syndrome admitted over one month in our outpatient department. Successful surgical correction was done by pericardial patch closure and prosthetic mitral valve replacement.

**Keywords:** Lutembacher's syndrome, Surgical correction, Atrial septal defect, Mitral valve stenosis, Pulmonary hypertension

## INTRODUCTION

Lutembacher's syndrome is defined as the rare combination of congenital ASD and acquired mitral stenosis.

The treatment for this condition can be either percutaneous intervention or surgical correction.

We report a series of 3 cases which were treated by surgical correction. In all the three cases atrial septal defect was a large ostium secundum and mitral valve was severely calcified with rheumatic affection. Therefore the surgical correction was opted for.

### **CASE REPORT**

We study three cases of Lutembacher's syndrome admitted from our outpatient department. The various features of these patients are mentioned in the Table 1.

Surgical technique: The average valve area in all the three cases was 0.8-0.9 cm². The valve was also non pliable, severely calcific, subvalvular crowding with Wilkin's score of 11/15. Thus the valve was considered unsuitable for percutaneous balloon mitral valvuloplasty. Therefore the decision for surgical correction was made.

After a midline sternotomy, cardiopulmonary bypass (CPB) was instituted using standard bicaval and aortic cannulation with moderate hypothermia.

After cross clamping cold blood cardioplegia was given alongwith topical myocardial cooling.

The right atrium was opened obliquely and ASD visualized.

The mitral valve was replaced using TTK Chitra valve and posterior mitral leaflet preservation. ASD is repaired using pericardial patch.

Table 1: Three cases of Lutembacher's syndrome admitted from our outpatient department. The various features of these patients are mentioned.

	Patient1	Patient 2	Patient 3
Age (years)	32	28	45
Sex	M	F	F
Symptoms	Dyspnoea NYHA III, Palpitations, Chest pain	Dyspnoea NYHA III, Palpitations,	Palpitations, Chest pain
Medical history		Diagnosed as congenital heart disease during previous pregnancy	
Physical examination	↑ JVP, MDM	↑ JVP, MDM, left parasternal heave	↑ JVP, MDM
Chest radiograph	Pulmonary plethora and left atrial enlargement	Pulmonary plethora and left atrial enlargement	Pulmonary plethora and left atrial enlargement
ECG	Normal sinus rhythm	Atrial Fibrillation	Right bundle branch block
Preoperative 2 D Echo	ASD -35mm, severe MS, calcified mitral leaflets, mod PH	ASD- 23mm, severe MS, moderate MR, mild TR, moderate PH, calcified mitral leaflets	ASD-20 mm, severe MS, moderate PH, calcified mitral leaflets
Coronary angiography	Not performed	Not performed	Normal coronaries
Surgical treatment	Pericardial patch closure of ASD+ 23 mm PMV replacement+ PML preservation	Pericardial patch closure of ASD + 27 mm PMV replacement + PML preservation	Pericardial patch closure of ASD + 23 mm PMV replacement + PML preservation

#### Abbreviations:

PMV - Prosthetic mitral valve (TTK Chitra valve), EF - Ejection fraction, MMPG - Mean mitral pressure gradient, MPAPG - Mean pulmonary artery pressure gradient, LVIDD - Left ventricular internal diameter in diastole, LVIDS - Left ventricular internal diameter in systole

## **DISCUSSION**

In 1916, Rene Lutembacher, a French Physician described his first case of this syndrome in a 61 year old woman who had been pregnant 7 times before. Lutembacher's syndrome was described as a rare combination of congenital atrial septal defect and acquired mitral stenosis. The incidence of this condition is very rare 0.001/1000000 according to one study published in the American Heart Journal in 1997. An earlier case report in the literature in 1880 (and referred to by Perloff) was of 74 year old woman who had endured 11 pregnancies. Survival to advanced age has been reported. In one instance in an 81 year old experienced no symptoms related to heart disease until she reached 75 years of age. 4

The hemodynamic effects of this syndrome are a result of the interplay between the relative effects of atrial septal defect and mitral stenosis. Mitral stenosis augments the left to right shunt through the ASD. This decreases the left atrial pressure gradient resulting in decrease in transvalvular mitral gradient, resulting in an ameliorating effect on the clinical expression of mitral stenosis. However it also unfavourably influences the long term natural history of ASD which augments the left to right shunt and predisposes to atrial fibrillation and right ventricular failure. The presence of mitral stenosis when accompanied by regurgitation enhances susceptibility to

infective endocarditis in contrast to low incidence of endocarditis in ASD.<sup>1</sup> Planimetry is preferred to Doppler half time method as Doppler method is inaccurate and may lead to underestimation of severity of mitral stenosis.<sup>6</sup> The symptoms are dependent on the size of ASD, severity of MS, compliance of right ventricle and pulmonary artery hypertension.

Early diagnosis and surgical treatment bears a good prognostic value. The patient is diagnosed a late stage pulmonary hypertension and heart failure. ASD closure with mitral valve replacement bears a good prognosis and prolongs survival. The success of this operation is closely related to the mitral procedure and perioperative period treatment with effective management of pulmonary hypertension and left ventricular function. 11,12

Funding: No funding sources Conflict of interest: None declared Ethical approval: Not required

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DOI: 10.5455/2349-2902.isj20150527 **Cite this article as:** Marghade SG, Sancheti A, Patwardhan A. Lutembacher's syndrome: a study of 3 cases. Int Surg J 2015;2:264-6.