*Case Report*

# Lutembacher's Syndrome in a young female treated Surgically: A Case Report

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**Abstract:**

**Lutembacher's syndrome is a rare clinical condition presenting with a combination of congenital atrial septal defect (ASD) and acquired mitral stenosis (MS). Lutembacher's syndrome is more prevalent in developing countries and its prevalence depends on the prevalence of rheumatic fever in that geographical area. The syndrome can present at any age but is usually more commonly observed in young female adults. Echocardiography remains the gold standard for diagnosis and evaluation of Lutembacher's syndrome (LS). Now a days many treatment options are available for LS – either percutaneous intervention or surgical correction. But pericardial patch closure of atrial septal defect and prosthetic mitral valve replacement is the treatment of choice for Lutembacher's syndrome.**

***Keywords:*** *Lutembacher's Syndrome, Mitral Stenosis, Atrial Septal Defect, Surgical Correction.*

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**Introduction:**

Lutembacher's Syndrome is defined as a rare cardiac abnormality presenting with a combination of congenital atrial septal defect (ostium secundum type) and acquired mitral stenosis.[1]

Mitral stenosis can be either congenital, or acquired in origin, most commonly due to rheumatic mitral valve disease. However, the current consensus defines Lutembacher's Syndrome (LS) as any combination of ASD (congenital or iatrogenic) and Mitral stenosis (congenital or acquired) [2]. In a typical case with Lutembacher's Syndrome, the atrial septal defect (ASD) is usually more than 15 mm in size. However, in the current era of percutaneous balloon mitral valvuloplasty for acquired mitral stenosis (MS), residual iatrogenic ASD secondary to trans septal puncture is more common than congenital ASD. Physicians refer to this as iatrogenic Lutembacher's Syndrome (LS) [3]. Lutembacher's Syndrome is more prevalent in developing countries where the incidence of rheumatic fever is high and a history of rheumatic fever has been reported in 40% of patients with Lutembacher's Syndrome. It is more common in females than males. There is a predilection for females because ASD and rheumatic MS are both more prevalent in females. The hemodynamic features and the natural history of the patients with LS depend upon the size of the atrial septal defect, severity of the mitral stenosis, compliance of the right ventricle and the degree of pulmonary vascular resistance [4]. Patients may remain asymptomatic for many years with this syndrome. Signs and symptoms vary according to the

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size of the atrial septal defect (ASD). Palpitation, shortness of breath and fatigue, are common presenting symptoms and appear early in patients with LS due to increased left to right shunt and decreased systemic cardiac output. Echocardiography is the gold standard method to establish the diagnosis of Lutembacher's Syndrome. Many treatment options are available now for LS – either percutaneous intervention or surgical correction. Here, we will present a case of Lutembacher's Syndrome with large atrial septal defect (ostium secundum type) with moderately calcified mitral stenosis treated surgically.

**Case Report**

A 28 years old female diagnosed as a case of Lutembacher's Syndrome, was admitted in cardiac surgery unit from cardiac out patient department. For almost one and half years, she complained of palpitations and chest discomfort on exertion. She was not diabetic or hypertensive. She had no previous symptoms of chest discomfort, syncope, orthopnea, paroxysmal nocturnal dyspnea, or limb edema.

She had experienced rheumatic fever when she was ten years old.

She was fairly built, with a consistent pulse rate of 76/min, blood pressure of 110/60mmhg, and a respiratory rate of 20/min on physical examination. On auscultation-S2 was wide and fixed splitted and there was a mid-diastolic murmur in the mitral region, but no crackling or wheezing in the lungs. There was no evidence of ankle oedema or hepatomegaly. Her electrocardiogram revealed normal sinus rhythm and RVH. Chest radiograph showed cardiomegaly and pulmonary plethora. All biochemical tests were within normal limit.

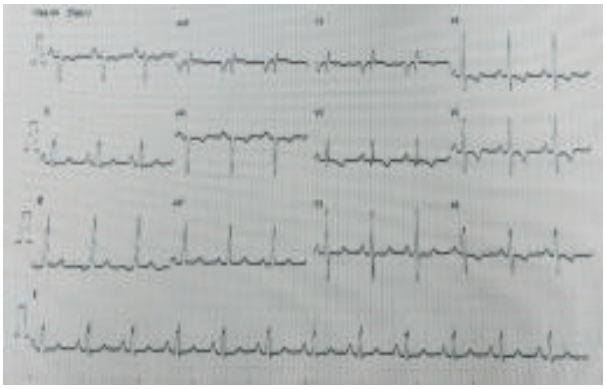

**Fig.-1:** *ECG showed normal sinus rhythm and features of RVH.*

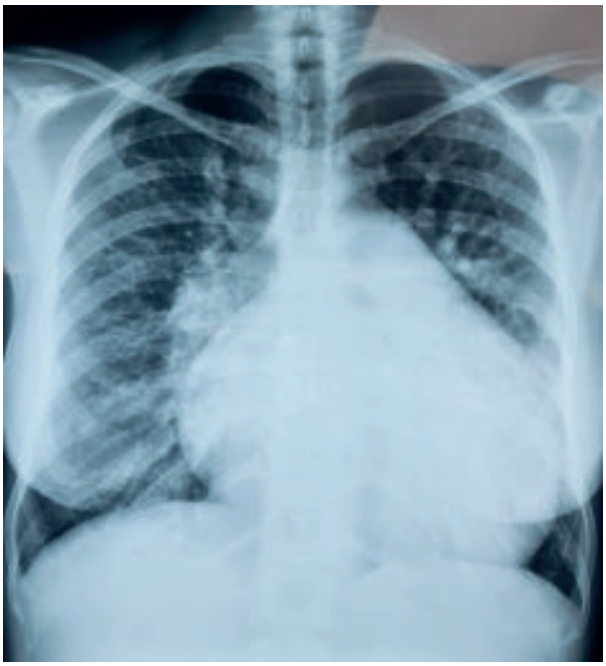

**Fig.-2:** *Chest radiograph showed cardiomegaly and pulmonary plethora.*

Later on echocardiographic evaluation (Both 2D & Doppler) was done which revealed large atrial septal defect (Ostium Secundum type) with left to right shunt with moderate mitral stenosis.

**Fig.-3:** *Transthoracic Echocardiography*
(Apical 4C view) showing atrial septal defect and mitral stenosis.

Left atrium, right atrium and right ventricle were dilated. Size of ASD was 43 mm with deficient posterior, superior and IVC rims. There were thickening of both mitral valve leaflets. There was systolic doming of anterior mitral leaflet with restricted mobility of posterior mitral leaflet. Mitral valve area (MVA) was 1.3 cm2 which was calculated by 2D planimetry method, mean trans mitral pressure

gradient was 5.70 mmhg, pulmonary artery systolic pressure was 45 mmhg.

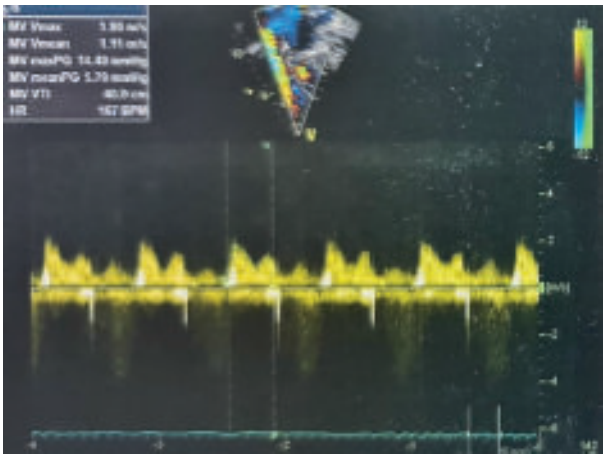

**Fig.-4:** *CW Doppler of mitral inflow of patient with Lutembacher's Syndrome*

All other valves morphology appeared to be structurally normal. Left ventricular ejection fraction was 60%. Coronary angiogram was not performed. After evaluation of the patient, elective surgical procedure was done under general anesthesia in supine position. After a median sternotomy , cardiopulmonary bypass (CPB) was established using aortic and standard bicaval cannulation with moderate hypothermia. After cross clamping , heart was arrested with antegrade cold blood cardioplegia through aortic root along with topical myocardial cooling using cold normal saline. The right atrium was opened obliquely (RA tomy) and large ostium secundum type of ASD was visualized. Both the leaflets of mitral valve was visualized which were thickened & calcified, both commissures were fused. So, the decision was made for surgical correction. Then mitral valve was excised and sized.

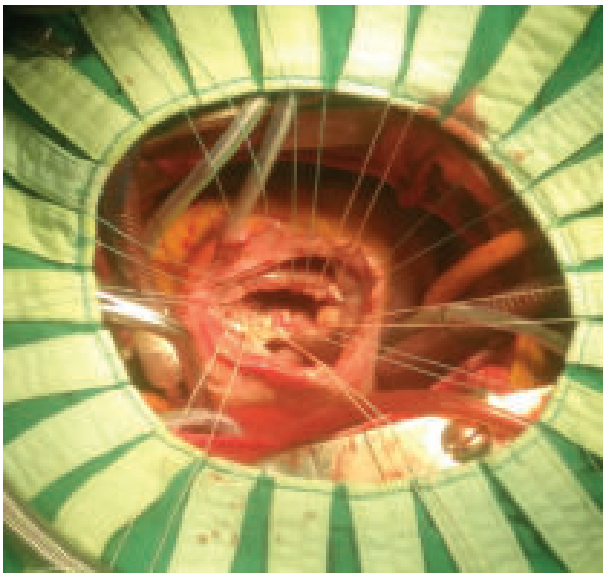

**Fig.-5:** *After excising diseased mitral valve.*

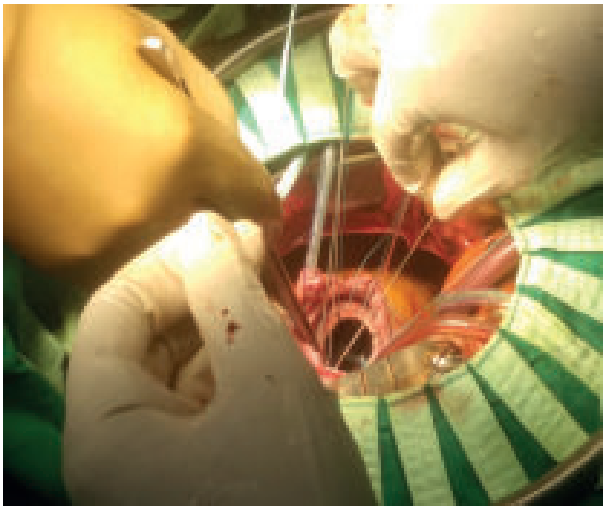

**Fig.-6:** *Metallic mitral valve replacement done.*

After that, the mitral valve was replaced with 25mm Medtronic bileaflet mechanical heart valve (BLMV). ASD was closed using autologous pericardial patch.

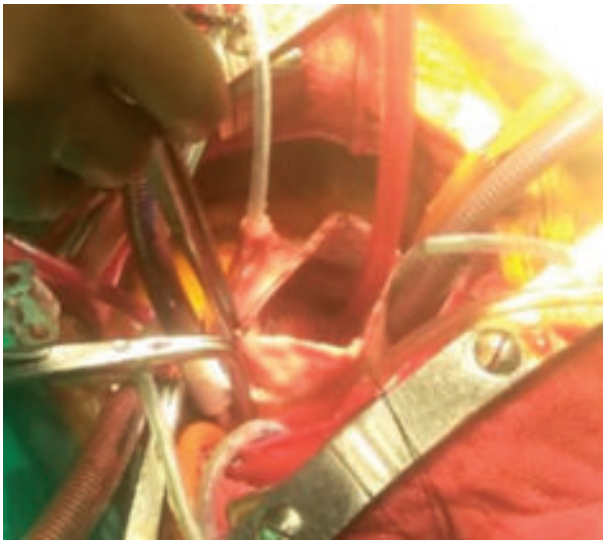

**Fig.-7:** *ASD closed with autologous pericardial patch .*

RA-tomy was closed, heart was deaired and gradually weaned from CPB. Decannulation done. Heparin was reversed with protamine sulfate. After maintaining proper hemostasis, wound was closed in layers, leaving two chest drain tubes and two RV pacing wires in situ. Patient

was shifted to ICU with stable hemodynamics with dobutamine 3 mics. She was extubated on 1st post-operative day and subsequent post-operative period was uneventful. Post-operative biochemical investigations were within normal limits. Dobutamine was tapered off on 3rd POD. Patient was discharged on 10th POD with smooth recovery.

**Follow Up**

At one year follow up, patient was asymptomatic and maintaining normal daily activities and echocardiogram showed 60% LVEF, well seated and functioning mitral prosthesis, no paravalvular leakage. ASD patch was intact with no residual flow.

**Discussion**

In 1750, Lutembacher syndrome (LS) was first described in a letter by anatomist Johann Friedrich Meckel. Corvisart who first described the association of mitral stenosis (MS) and atrial septal defect (ASD) in 1811. Rene Lutembacher, a French physician born in 1884, described his first case of this syndrome in a 61 year old women who had been pregnant 7 times before and published the first data described as LS in 1916[5,6,7]. Lutembacher's Syndrome was described as a rare combination of congenital ostium secundum defect type of ASD and acquired mitral stenosis. Congenital MS is rare. The current consensus is that LS consists of a congenital defect in the atrial septum upon which acquired MS is imposed. The incidence of MS in patient with ASD is 4% while the incidence of ASD in MS is 0.6-0.7% [8]. Its prevalence depends on the prevalence of rheumatic fever in that geographical area[9]. The exact prevalence of LS is not known. It is more prevalent in areas with higher prevalence of rheumatic heart disease[10]. In developing countries, a history of rheumatic fever has been reported in 40% of patient with LS. This condition can present at any age but more commonly found in young adult female patients. The hemodynamic effects of this syndrome are the result of the interplay between the relative effects of atrial septal defect and mitral stenosis. The hemodynamic features depend upon the size of the ASD, severity of MS, compliance of the right ventricle and the degree of pulmonary vascular resistance. When mitral stenosis (MS) is severe and atrial septal defect (ASD) is non-restrictive, left atrium (LA) finds another exit through the septum in addition to the mitral valve (LA decompression). Therefore, LA pressure does not rise in proportion to the severity of MS. For this reason, pulmonary venous hypertension takes a long time to develop resulting increased left to right shunt across the ASD. Right atrium (RA) and right ventricle (RV) are progressively dilated with increased pulmonary blood flow. In untreated cases, the pulmonary vascular resistance continues to increase which leads to right ventricular failure.[11] In contrast, if the ASD is restrictive, the shunt across the defect will be less and hence, the patient will follow the course of isolated MS. Patient with restrictive ASDs and moderate to severe MS, present much earlier and usually with features of pulmonary congestion from MS.[12]

Two-dimensional echocardiography with color flow doppler is the diagnostic modality of choice in patients with Lutembacher's Syndrome (LS). The type and size of ASD and severity of MS are accurately estimated by this technique[13].

Typically, ASD in LS should have a diameter of more than 15mm. Planimetry by 2D/3D echo is the more reliable method to assess MVA and severity of MS in patients with LS[3].

The patient with Lutembacher's Syndrome is managed either by open heart surgery or percutaneous interventional techniques. But open heart surgery is the gold standard treatment for LS. Many limitations are found for percutaneous intervention like large ASD, with lack of margin of ASD, mitral valve restenosis, presence of left atrial thrombus, presence of anomalous pulmonary drainage, MR(Gr-III) or higher, bi-commissural calcification and finally lack of expertise[14]. The classical LS can be treated satisfactorily where as the acquired LS usually need early surgical intervention as they are more prone to deteriorate with the development of severe pulmonary hypertension and right heart failure[15].

Prognostic factor of this syndrome include pulmonary vascular resistance, right ventricle (RV) compliance, size of ASD and severity of MS.

**Conclusion**

Lutembacher's Syndrome is a rare cardiac abnormality. Early diagnosis and corrective surgical treatment including, ASD closure with mitral valve replacement, is associated with a good outcome and prolongs survival. Preoperative assessment and effective management depend on the better outcome of surgical procedures.

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