*Case study*

# Lutembacher's syndrome: a case report from hospital IBN Rochd of Casablanca, Morocco


## ABSTRAT

Lutembacher's syndrome (LS) refers to the uncommon combinaison of acquired mitral stenosis (MS) with congenital atrial septal defect (ASD). Other forms described in the literature include iatrogenic LS and reverse LS. LS is a very rare entity with a female predominance, it has been either overdiagnosed or misdiagnosed. The prognosis is good before the onset of pulmonary hypertension and right heart failure. We report the observation of a 62-year-old female presented with a one year history of progressive shortness of breath. Cardiac ultrasound lead to the diagnosis of this rare clinical syndrome. The patient was referred for mitral valve replacement with ASD closure.



Keywords: Mitral stenosis (MS); Atrial septal defect (ASD); Lutembacher'syndrome (LS); Morocco.


## INTRODUCTION

Most cardiovascular conditions are either acquired or congenital in origin, but in rare instances, a combination of both is found. Lutembacher syndrome (LS) refers to the uncommon combinaison of acquired mitral stenosis (MS). Other forms described in the literature include iatrogenic LS and reverse LS (1). LS is a very rare entity with a female predominance (2, 3). Symptoms present at any age. The prognosis is good before the onset of pulmonary hypertension and right heart failure. Surgical and percutaneous trans-catheter therapies with balloon valvuloplasty and septal closure using an Amplatzer closure device have proven to be beneficial (4). We report the case of an adult female who presented with this rare clinical syndrome.

## CASE REPORT

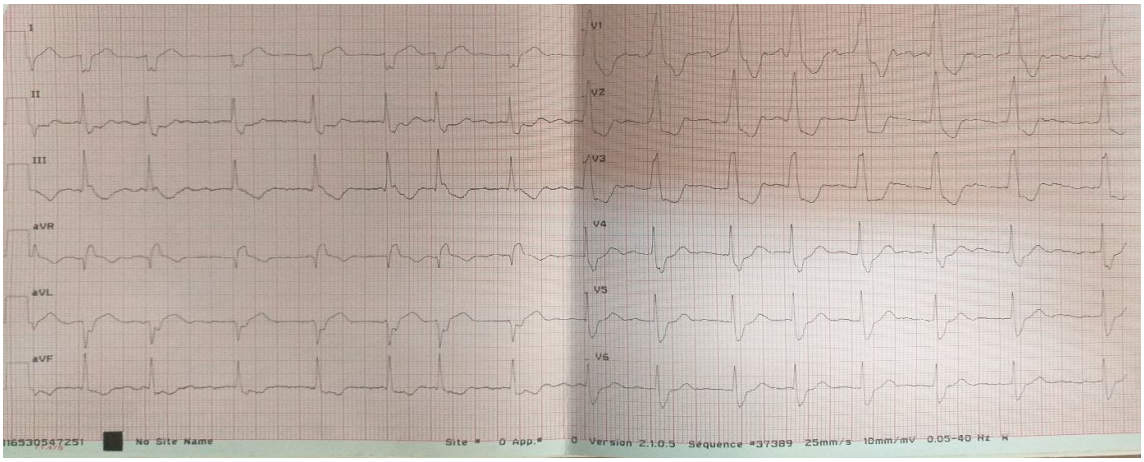
A 62-year-old female presented with a one year history of progressive shortnesss of breath on extertion and fatigue, which had worsened recently. Three weeks prior to consultation, she reported worsening dyspnea. There was no documented history of acute rheumatic fever in childhood. On examination, she was afebrile with normal volume regular pulse of 81 beats/min and blood pressure 130/70 mm of Hg, with respiratory rate of 20 breaths/min. In cardiac examination, there was loud mitral $S_1$, and wide fixed split pulmonary $S_2$ with opening snap in mitral area. There was III/IV mid diastolic murmur in mitral area and early systolic murmur, and non-radiating short murmur in pulmonary area. There was no parasternal heave. Respiratory system examination revealed bilaterally equal normal breath sounds with bilateral lower zones end-inspiratory fine crackles.

Her investigation reports were as follows:

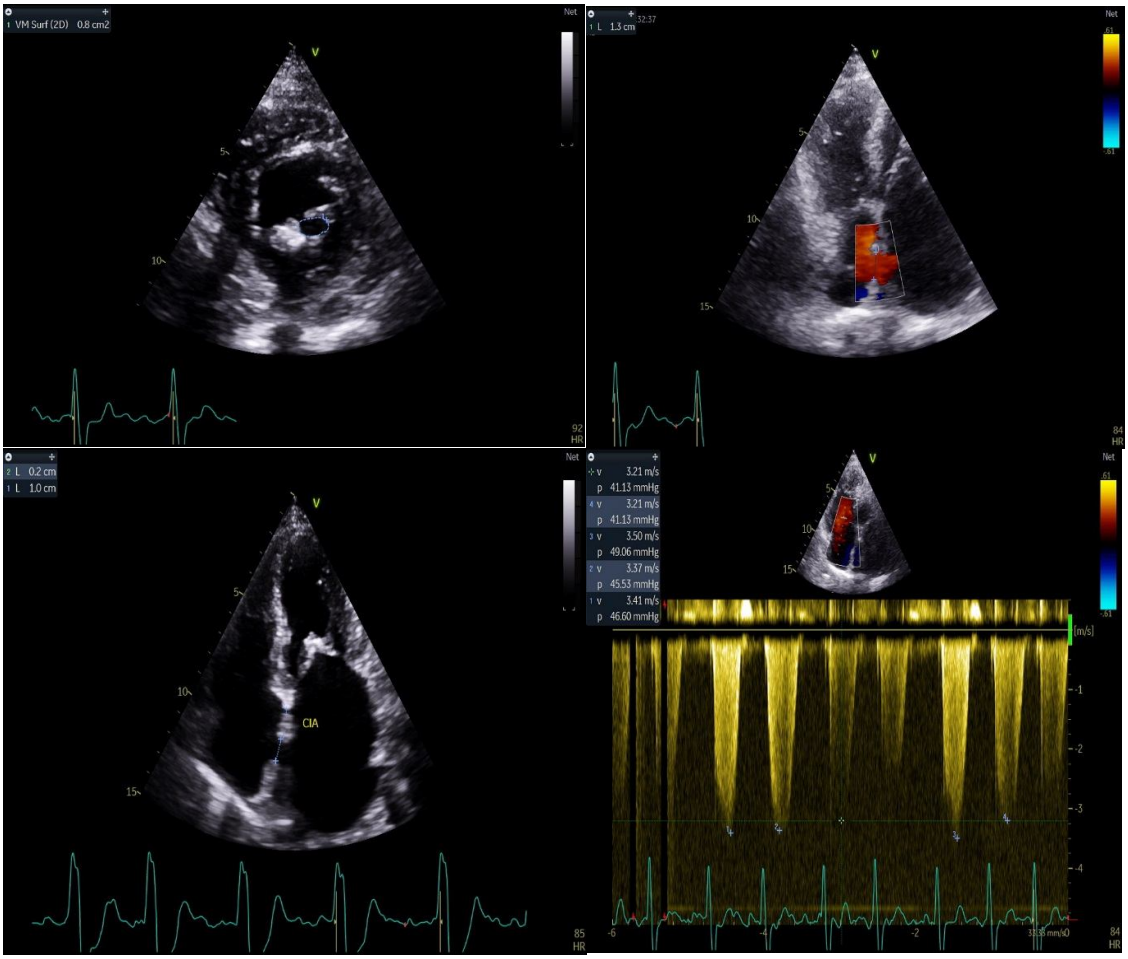
- Hemoglobin (Hb) :14.2 g/dl;
- White blood cells (WBC) : 10.270/cu mm of blood;
- Polymorphs : 54%
- Lymphocytes :44%;
- Eosinophils : 02%;

- Platelet count : 2.86 lakh;
- Random blood sugar : 76 mg/dl;
- Blood urea : 0.25 g/dl; [normal value→20 – 40 mg/dl]
- Serum creatinine : 9.5 mg/dl; [normal value→0.8 -1.6 mg/dl]
- Serum sodium : 137 mEq/L; [normal value→135 – 145 mEq/L]
- Serum potassium : 3.6 mEq/L. [normal value→3.5 – 5.5 mEq/L]

Electrocardiogram [Figure 1]→ Atrial fibrillation Rhythm; Incomplete Right Bundle Branch Block (RBBB); Left Atrial Enlargement; Normal PR Interval/Normal Qtc.




2D-Echocardiography [Figures [Figures 2 and 3,4,5] → Very Tight Mitral stenosis; Mild Mitral regurgitation; mitral valve area 0.70-0.80 $cm^2$. Large Ostium Secondum atrial septal defect With Left to Right Shunt; Mild pulmonary arterial hypertension.




Other Valves- Normal. No Clots/No Effusion/No Vegetations.

The patient is put on diuretics, antiarrythmics and anticoagulants.

She was referred for mitral valve replacement with ASD closure.

**DISCUSSION**

In Lutembacher's syndrome, initially, high left atrial pressure due to mitral stenosis was thought to stretch open the patent foramen ovale, causing left-to-right shunt and providing another outlet for the left atrium. Now ASD in this syndrome, like mitral stenosis, is recognized as being either congenital or acquired, as already described. The haemodynamic effects of this syndrome are a result of the interplay between the relative effects of ASD and mitral stenosis. In its initial description, the ASD was typically large in Lutembacher syndrome, thus providing another route for blood flow. The direction of blood flow is determined largely by the compliance of left and right ventricles. Normally, the right ventricle is more compliant than the left ventricle. As a result, in the presence of mitral stenosis, blood flows to the right atrium through the ASD instead of going backward into the pulmonary veins, thus avoiding pulmonary congestion. This happens at the cost of progressive dilatation and, ultimately, failure of the right ventricle and reduced blood flow to the left ventricle.

Development of Eisenmenger syndrome or irreversible pulmonary vascular disease is very uncommon in the presence of large ASD and high left atrial pressure because of mitral stenosis.

The incidence of this condition is very rare. In one study published in American Heart Journal in 1997, it is found that the incidence of Lutembacher's syndrome is- 0.001/10,00000.[2] The ameliorating role of the ASD in MS was evident in Lutembacher's original report of 1916; the patient was a 61-year-old woman who had been pregnant seven times.[3] An earlier case report in the literature in 1880 (and referred to by Perloff)[1] was of a 74-year-old woman who had endured 11 pregnancies. Survival to advanced age has also been reported;[4] in one instance an 81-year-old woman experienced no symptoms related to her heart disease until she reached 75 years of age.[4] These favourable reports, however, should not obscure the fact that the long-term natural history of ASD is unfavourably influenced by MS, which augments the left-to-right shunt and predisposes to atrial fibrillation and right ventricular failure.[5] The presence of MS, especially when accompanied by mitral regurgitation, increases susceptibility to infective endocarditis, in contrast to the low incidence of infective endocarditis in uncomplicated ASD,[1] just like in our case. Early diagnosis and surgical treatment bears a good prognostic value. If patient is diagnosed at late stage, pulmonary hypertension and heart failure develops and the prognosis is bad.[6] If the patient is diagnozed earlier before the development of pulmonary hypertension and heart failure, ASD closure with mitral valve replacement bears a good prognosis and prolongs survival.

## CONCLUSION

LS remains a rare clinical entity. If diagnosed early, patients benefit from surgical or percutaneous trans-catheter therapy. The outcome is better if treated before the onset of heart failure and pulmonary hypertension. However, surgical and percutaneous trans-catheter therapy is costly and not readily available in low-income settings in developing countries.

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