



## Case Report

# Diagnosis Misinterpreted - MS With TR (or) MS with ASD (Lutembacher Syndrome)

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### Abstract

A Male in his mid 40's with breathlessness on exertion and facial puffiness since 2 months. On evaluation, he was diagnosed with moderate mitral stenosis (Rheumatic, mitral valve area 1.9sqcm) and a large ostium secundum atrial septal defect (40mm) with bidirectional shunt and severe PAH (105mmhg) and newly developed atrial fibrillation. He was initially managed by medical management and later was advised for surgical intervention for valve replacement and defect closure. (Indian J Cardiol 2022;25 (3-4):63-65)

### Background

Johann Friedrich Meckel was the first one to describe Lutembacher<sup>1</sup> syndrome in the year 1750, and described his first case in a 61 year old woman who was having the mitral valvular lesion to congenital mitral stenosis<sup>2</sup>, LS is defined as Mitral Stenosis combination with Atrial septal defects<sup>3</sup>, in a typical case of LS the ASD is usually more than 15mm in size, incidence rate of congenital ASD in patients with MS is 0.6% to 0.7%.

### Case report

A male who is moderately built in his mid 40's presented with complaints of breathlessness which is moderate in grade since 1 month, On grading he had NYHA grade- III dyspnoea, He also had a complaint of palpitations and generalized weakness since 1 month, He later started developing facial puffiness which is moderate in nature, On examination he was having 2+ pallor and grade 3 B/L pedal pitting edema his vitals were as follows BP- 160/99 mmhg,

PR- 101/min which is irregularly irregular in nature and his Jvp was raised, tapping Apex beat and a parasternal heave were also present, Apex pulse deficit was present, On Auscultation- first heart sound was loud and variable and second heart sound was wide and split, mid diastolic murmur was heard on apex and a systolic murmur was heard on left 3rd ics and a systolic murmur in tricuspid area, His ecg shows (Figure 1) Atrial fibrillation, 2D-Echo finding shows - thickened AML and PML with thickened and fixed annulus (Figure 2), Large ostium secundum ASD (40mm) with Bidirectional shunt, Paradoxical motion of septum, Mild MR, severe TR (90mmhg), severe PAH (105mmhg), Grossly dilated RA / RV.

### Investigation (other)

The patients LFT, RFT, CBC, Prothrombin time were normal whereas APTT was slightly raised (38.0 sec), His ECG showed Atrial Fibrillation (Figure 1).

2D-Echo shows thickened AML, PML, and Annulus, Hockey Stick sign (Figure 2), Fish Mouth

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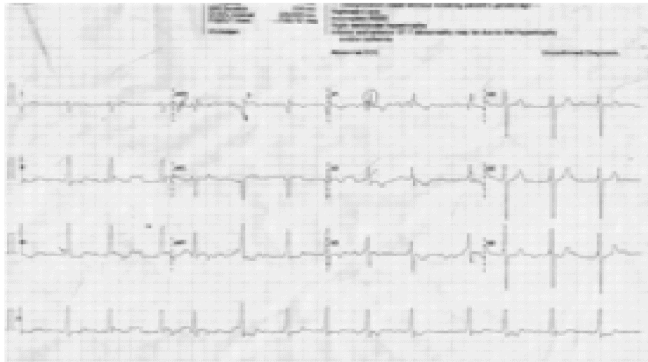


Fig. 1 : ECG showing Af1 / fibrillatory waves with absent p wave

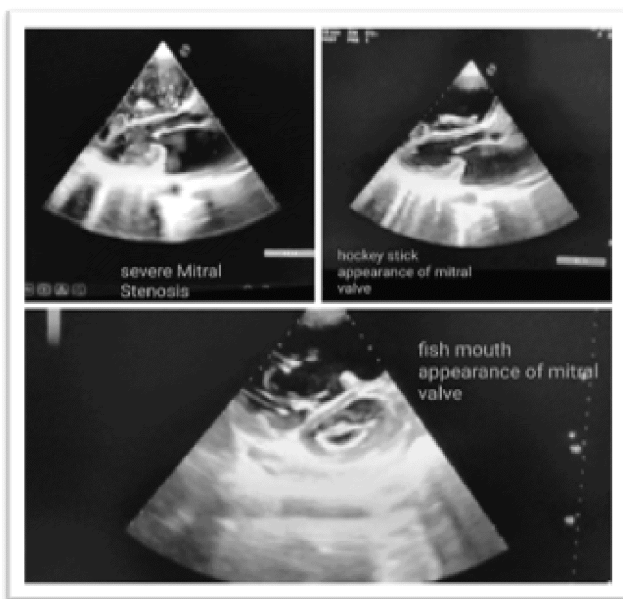


Fig. 2 : 2d - echo showing signs of severe mitral stenosis ; (A) coloured doppler showing severely stenosed mitral valve B) classic hockey stick appearance of mitral valve C) Fish mouth shaped mitral valve suggesting Severe Mitral stenosis

appearance of mitral valve (Figure 2), Dilated and non-collapsible IVC (Figure 4), A large ostium secundum ASD (Figure 3), with Bidirectional Shunt (Figure 5).

#### Treatment

Initially he was managed using diuretics to treat CHF, oral antiplatelet therapy, oral anti coagulation therapy to prevent clot formation, Digoxin therapy and betablocker and angiotensin receptor blockers and later he was referred to a CTVS surgeon for further surgical management.



Fig. 3 : 2d-echo image showing a large Atrial Septal Defect.



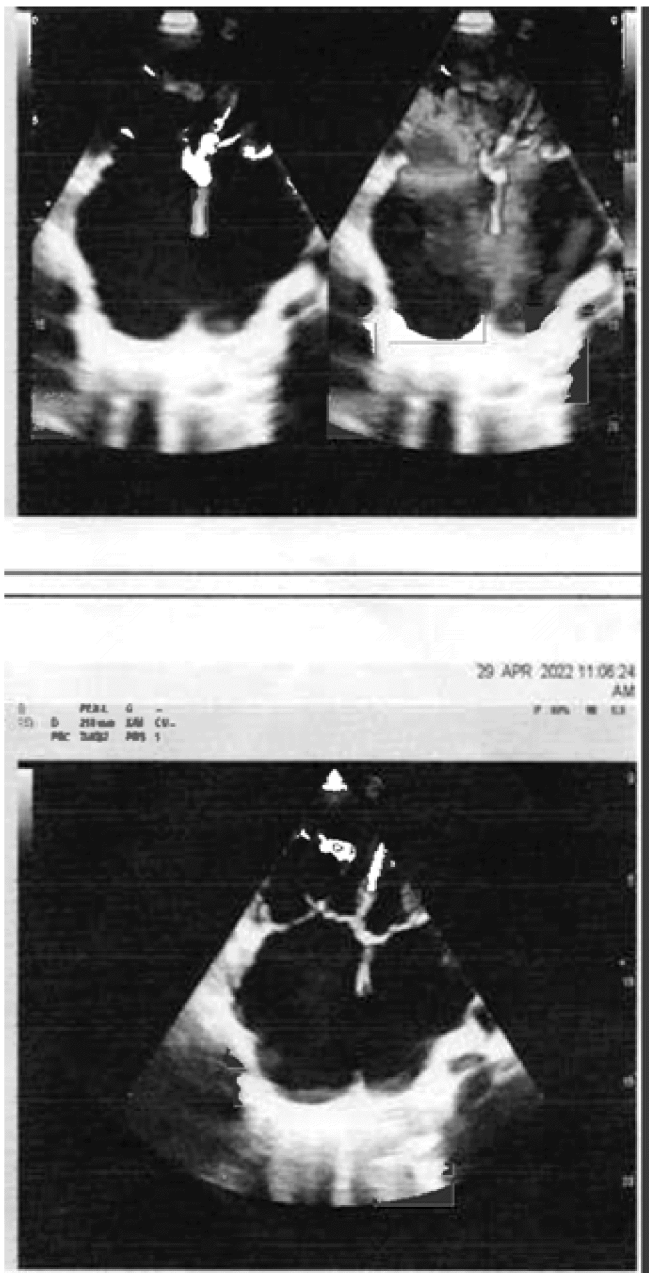
Fig. 4 : Echo findings showing Dilated IVC

#### Outcome and Follow-up

Patient was referred to a CTVS surgeon for valve replacement and atrial septal defect closure for which she got operated and recovered well (on follow up with patients attendants).

#### Discussion

Patient presented with dyspnea, irregularly irregular pulse with MDM at mitral area with systolic murmur at tricuspid area leading towards the diagnosis of mitral stenosis with TR with right



**Fig. 5 : Echo showing a Large Ostium Secundum ASD with Bidirectional Shunt**

ventricular failure, When echocardiography was done although MS was there but a new lesion came out that was mimicking TR and it was ASD (ostium secundum type), So clinical diagnosis was deferred to other diagnosis on echo.

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