

Case Report

Surgical Management of Lutembacher's Syndrome in Congestive Cardiac Failure: A Case Report & Review of Literature

Rajeshwar Yadav¹, Swati Pathak¹, Shubham Kumar Gupta²

¹Assistant Professor, Department of CTVS, Institute of Medical Sciences, Banaras Hindu University, Varanasi

²Resident Department of General Surgery, Institute of Medical Sciences, Banaras Hindu University, Varanasi

Received: 04 November, 2022 Accepted: 05 December, 2022 Published: 08 December 2022

Abstract:

Introduction: Lutembacher's syndrome is a rare clinical entity. In 1916, French physician Rene Lutembacher described his first case of 61 years old woman with a combination of congenital ASD with acquired MS. But he attributed this mitral lesion to congenital mitral stenosis. The definition of this syndrome named after him has undergone many changes since then. The current consensus defines Lutembacher Syndrome (LS) as any combination of ASD (congenital or iatrogenic) and MS (congenital or acquired). The clinical presentation of LS is a result of the interplay between the size of ASD, the severity of MS, pulmonary vascular resistance and RV compliance. Financial constraints also add to the delayed diagnosis and adequate treatment in our part of the world. Here we have discussed a case of LS with severe TR and moderate PAH in congestive cardiac failure which was successfully treated by surgery thereby demonstrating cost-effective and successful surgical management of a rare cardiac disease where vascular resistance, compliance, size and severity of lesions interplay causing cardiac failure.

Case report: A 42 years old thin built, poorly nourished housewife presented with chest discomfort and fatigue for the last 4 years. She was diagnosed with an atrial septal defect four years back but could not receive appropriate treatment due to financial constraints. For the last 15 days, she had breathlessness even at rest and paroxysmal episodes that interrupted her sleep. Icterus and pedal edema was also present. As we evaluated the patient, her clinical examination could not be entirely explained by ASD alone. Soon, our suspicion of LS was confirmed by TTE and the diagnosis of Lutembacher Syndrome with Severe Tricuspid Regurgitation with Moderate PAH was made. As the patient presented to us with atrial fibrillation & heart failure, she was admitted for optimization. The patient was then successfully managed by MVR [#27 SJM, metallic] with pericardial patch closure of ASD with Devaga's Tricuspid Valve annuloplasty and discharged on a postop day 11.

Conclusions: Early diagnosis and surgical treatment bear a good prognosis and prolong survival. If a patient is diagnosed late, pulmonary hypertension and heart failure set in and the outcome becomes poor. The higher cost of surgical intervention and low socio-economic distribution of the disease in developing countries like India should not challenge the patient's right to receive the standard treatment.

Keywords: Mitral stenosis (MS); atrial septal defect (ASD); Congenital heart disease (CHD), Lutembacher syndrome :(LS); Congestive cardiac failure; pulmonary arterial hypertension (PAH).

Introduction

In 1916, French physician Rene Lutembacher described his first case of 61 years old woman with a combination of ASD with acquired MS. But he attributed this mitral lesion to congenital mitral stenosis.[1] The definition of this syndrome named after him has undergone many changes since then. The current consensus defines LS as any combination of ASD (congenital or iatrogenic) and MS (congenital or acquired).

LS is a very rare entity with incidence reported to be 0.001 per million populations with a female predominance (2,3). The clinical presentation of LS is a result of the interplay between the size of ASD, the severity of MS, pulmonary vascular resistance, and RV compliance. The presence of pulmonary arterial hypertension and congestive cardiac failure indicates an unfavorable outcome.

While percutaneous trans-catheter therapies (balloon valvuloplasty with septal closure using an Amplatzer closure device) are an upcoming modality, surgical correction remains the standard of care, especially for those where percutaneous intervention is not possible (4). Here we have discussed a case of LS with severe TR and moderate PAH in congestive cardiac failure which was successfully treated by surgery.

Case report

42 yrs, housewife from Begusarai, Bihar presented with a history of chest discomfort for the last 4 years which was initially on exertion but gradually worsen to persist even at rest.

This was associated with palpitation, generalized fatigability and lethargy. There is no history of fever associated with joint pain following throat infection, rashes or abnormal body

movements in childhood. She has had multiple episodes of LRTI since childhood. She has no other comorbidities. For above said complaints she was referred to SGPGI, Lucknow by her local treating doctor, where she was investigated and diagnosed with ASD and was planned for surgical intervention. But due to financial constraints, she denied surgery.

During the last 15 days, she had breathlessness even at rest and paroxysmal episodes that interrupted her sleep. [NYHA IV] This was associated with a non-productive cough. For these complaints, she presented to CTVS OPD of SSH, BHU from where she was admitted for stabilization and re-evaluation. In the general survey, a thin-built poorly nourished patient with a BMI of 15.8 Kg/m² with a pulse rate of 106 bpm, irregular, hypovolemic recorded in the right radial artery without any radio-radial or radio femoral delay and blood pressure of 90/60 mm Hg (right brachial artery); Respiratory rate: 26 cycles per minute thoracoabdominal. Oxygen saturation was 90% at room air. She had moderate pallor and bilateral pedal edema with raised JVP.

On cardiac examination, the diffuse apical impulse was found in the 6th ICS just lateral to midclavicular line with Grade-2

left parasternal heave. On auscultation, wide and fixed split S2 and an ejection systolic murmur in the left upper parasternal area (flow murmur), Mid-diastolic murmur at the mitral area, pansystolic murmur transmitting to the apex and increasing in intensity on deep inspiration was audible at the tricuspid area (Carvallo's sign). Respiratory System Bilaterally equal normal vesicular breath sounds with fine basal crepts. Per abdomen palpation revealed mild tenderness was noted in the right hypochondrium along with moderate hepatomegaly.

Her routine blood parameters were as follows -

- CBC:Hb-7.2g/dl; TLC-8500/cumm; DLC-N70/L27/E11; PLT-259000/micro-litre
- LFT: SGPT/SGOT-68.4/69.2U/L; Total Bilirubin/Indirect Bilirubin -3.8/2.8mg/dl; ALP-284U/L; TP/ALB - 5.3/2.2 g/dl
- RFT: Urea /Creatinine -24 / 0.9 mg/dl; Serum Sodium / Potassium-142 / 3.6 mEq/L
- PT/INR:13.5/1.08
- HIV1&2-Non Reactive; HBsAg- Non Reactive; Anti-HCV-Non Reactive
- COVID (RT PCR) -Negative

Chest X-ray PA view shows the prominent left pulmonary artery, cardiomegaly predominantly right ventricular enlargement, straightening of the left border of the heart and pulmonary plethora. (Figure 1. A)

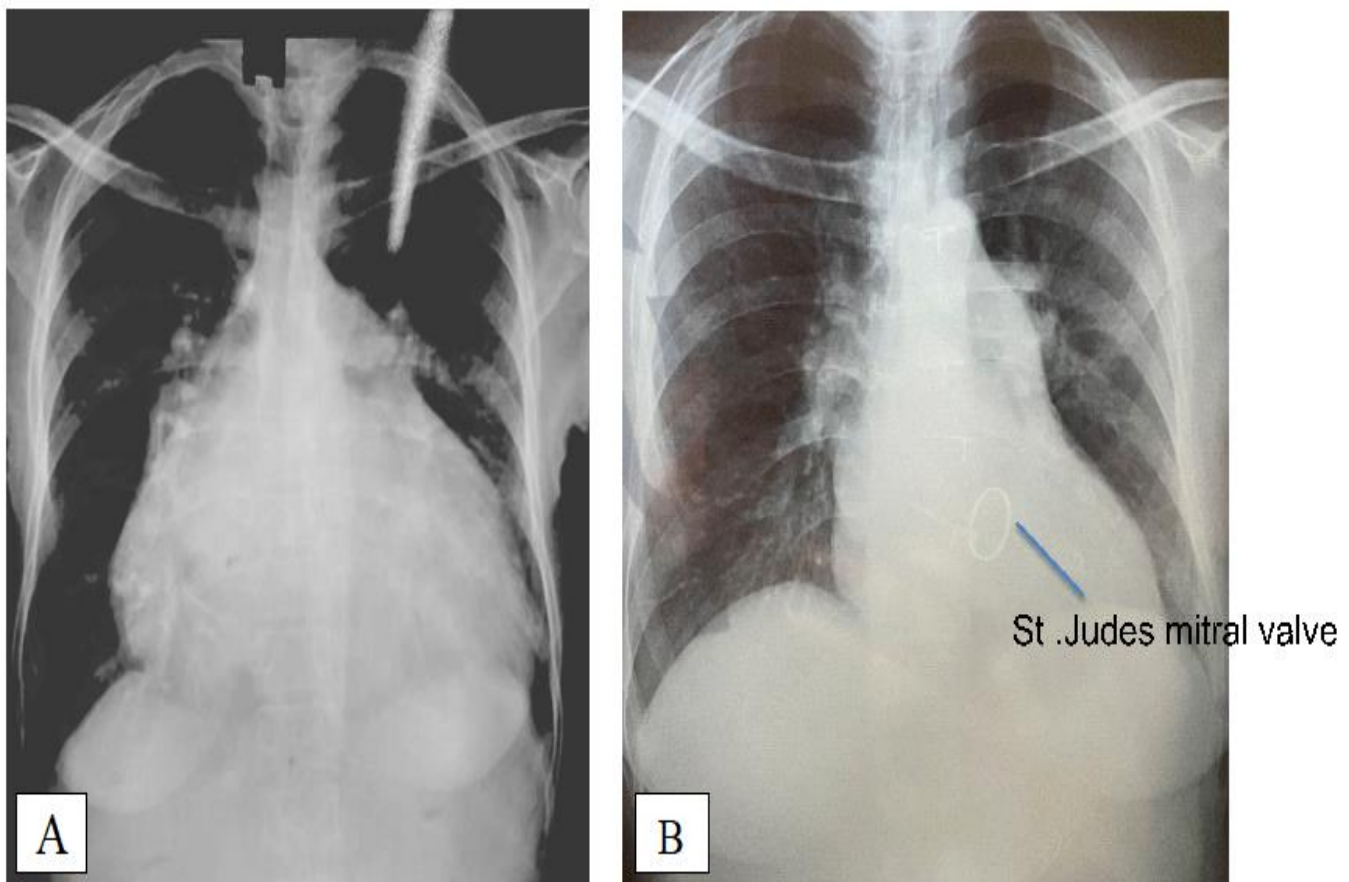


Figure 1 : A. Preoperative Chest X ray of patient showing cardiomegaly predominantly right ventricular enlargement and straightening of left border of heart with prominent left pulmonary artery. B. Post-operative follow-up (at 3 months), chest X ray PA view revealed marked reduction in cardiac size with prosthetic (St. Judes) valve seen in mitral areas.

ECG revealed irregularly irregular rhythm, right atrial and right ventricular enlargement, and right axis deviation.

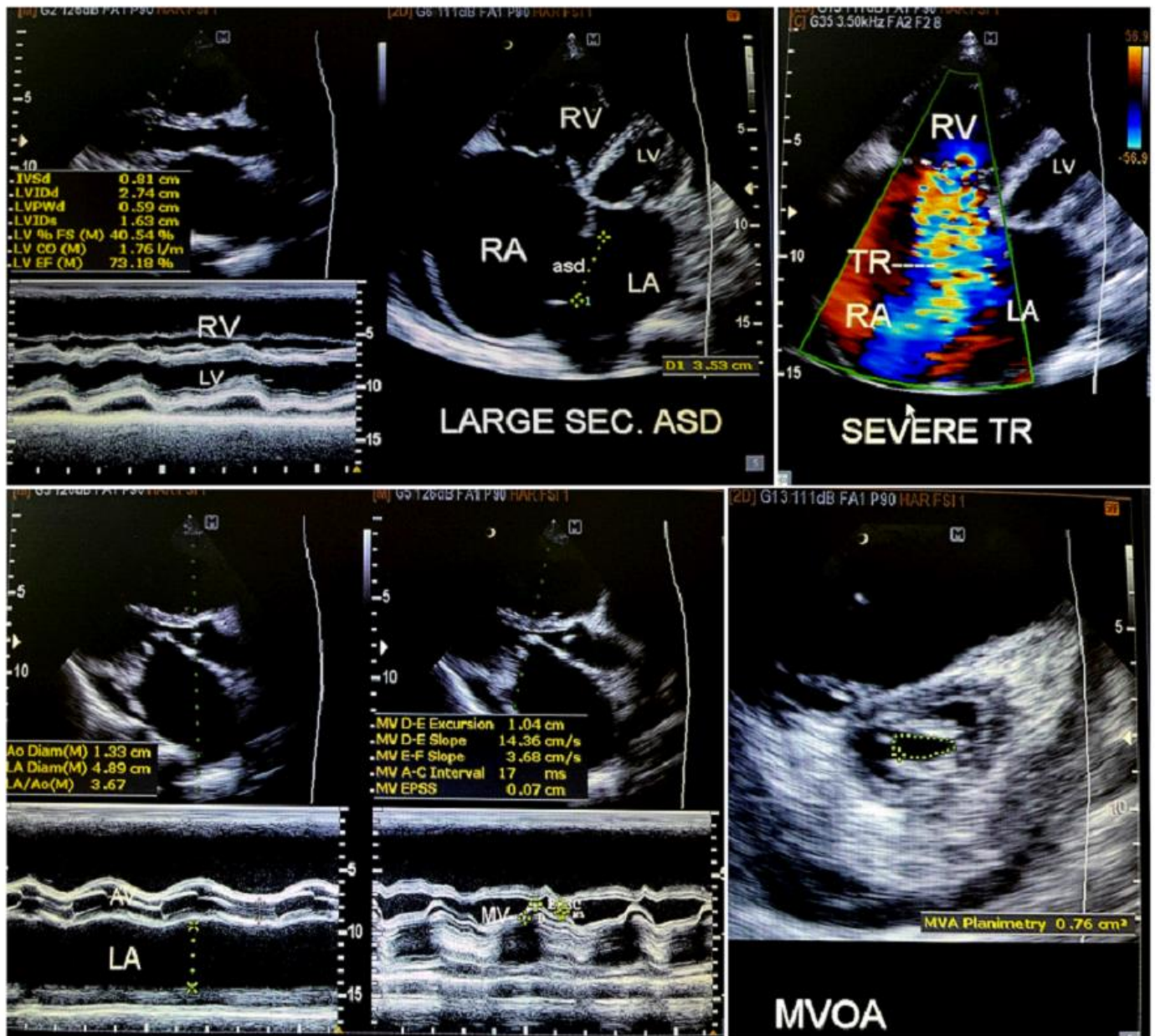


Figure 2 : Preoperative Trans-thoracic Echocardiogram showing acyanotic CHD [Very large ostium secundum ASD(~32 mm) with L→R atrial shunt, Qp/Qs >2.5:1. Marked right side volume overload , preserved biventricular functions. Dilated MPA, moderate to severe rheumatic MS (MVOA <1 cm²), moderate PAH , severe TR , dilated LA , no thrombus in LA/LAA. Dilated IVC and hepatic veins

Her trans-thoracic 2 D Echocardiography revealed-(Figure 2)

- AML and PML are thickened esp. at the tips. AML doming present and PML restricted, motion paradoxical, no significant calcification. Bi-commissural fusion is present with moderate subvalvular thickening.
- Planimetered MVOA<1.0sqcm, flow acceleration and forward turbulence seen at the level of Mitral valve.
- Severe TR and Mild PR were PASP~50 mm Hg from TR jet suggesting moderate (hyperkinetic) PAH.
- Very large Ostium Secundum ASD (~32mm,good rims) is present with non-restrictive L→R atrial shunt (small R→L shunt also seen); Qp/Qs > 2.5:1

- Overall preserved biventricular systolic function, LVEF~70%
- RA, TV annulus, RV and MPA(27mm) are markedly dilated. LA is moderately dilated. LV and aorta appear small (underfilled).

Coronary angiography was normal in this patient.

On reevaluation, the patient was found to have Severe MS along with large OS-ASD [Lutembacher’s syndrome] that was in accordance with the clinical presentation of the patient. As the patient presented to us with the poor general condition and congestive cardiac failure, optimization was essential before major surgical intervention. She was nursed in a semi-recumbent position and with oxygen support, started on

verapamil 40mg thrice daily, intravenous furosemide 40mg twice daily & digoxin 0.125mg once daily along with hemoglobin optimization, PPI, IFA, high protein diet (nutritional supplementation), nebulization and chest physiotherapy. The patient's general condition & vitals improved in about 2 weeks with aggressive supportive therapy. After optimal decongestive therapy patient was planned for elective surgery. Induction was achieved with 0.5 mcg/kg of fentanyl and 0.2 mg/kg of etomidate intravenous. 0.1 mg/kg of vecuronium assisted in tracheal intubation using a 7 mm ID ET tube.

The right atrium, right ventricle, and left atrium were enormously enlarged whereas the left ventricle was small in size. Approaching via right atriotomy AML was excised preserving the subvalvular apparatus. The Mitral valve was replaced with a 27 mm St. Judes bileaflet metallic valve with ethibond 2-0 suture. ASD was closed with an autologous pericardial patch using prolene 4-0 suture. DeVega's TV annuloplasty was done using prolene 2-0 suture (it saved the additional financial burden of the TV ring in a low economy scenario). The closure was commenced and immediately with good ejections size of the right side of the heart showed a marked reduction. The patient was shifted to ICU in stable condition. Postoperatively minimal inotropic support was required. No ET bleed was witnessed postoperatively. The patient's postoperative course was uneventful. Antibiotic, diuretic anticoagulant was started. INR monitoring is routinely performed. Injection enoxaparin with tablet warfarin was started till target INR (2.5-3.5) was achieved. The patient was ambulatory on POD 3 when medicinal drains were removed. On POD 10 patient was discharged with an uneventful postoperative course. Initially, the patient was followed every 15 days for 1 month then monthly for the next 2 months followed by three monthly for one year. On 3 months follow-up, chest X-ray PA view revealed a marked reduction in cardiac size with prosthetic (St. Judes) valve seen in mitral areas. Echocardiographic findings (on follow-up) showed prosthetic mitral valve in situ with normal functioning with no MR and paravalvular leak. Max PG 8mm Hg, Mean PG 5 mm Hg LVEF 50-55%, mild TR. No flow across the intertribal septum during the study. No clot/vegetation/PE.

Discussion

Though the exact prevalence of LS in India is not known, it is more likely to be prevalent in Southeast Asian countries where there is a high prevalence of rheumatic heart disease. However, only up to 40% of patients with LS give a positive history of rheumatic fever in childhood. [5]

Classical LS refers to a congenital atrial septal defect complicated by acquired mitral stenosis as mentioned by Rene Lutembacher in 1916. Though the association of ASD with MS was first described by Corvisart in 1811. The ameliorating role of ASD in MS was evident in Lutembacher's original report of 1916; the patient was a 61 years old woman who had been pregnant seven times. [1]

The clinical presentation has a wide spectrum from incidental findings following recurrent RTI to non-specific symptoms of

chest discomfort, palpitation, and generalized fatigue to breathlessness at rest (NYHA IV) with cardiac failure/ atrial arrhythmia. The size of ASD (restrictive/non-restrictive), the severity of MS, pulmonary vascular resistance, and compliance of the right ventricle are recognized factors that influence the natural history and hemodynamics in patients with Lutembacher syndrome thus dictating the time of diagnosis and outcome of surgical intervention [6].

The interplay between the hemodynamic effects of ASD & MS dictates the clinical presentation of this syndrome. Initially high atrial pressure due to MS is thought to stretch open the patent foramen oval, causing left to right shunt and providing another outlet for the left atrium. This left-to-right shunt decompresses the left atrium in MS, following which the right ventricle dilates and pressure and volume overload develops leading to PAH. If left untreated for a long, the right to left shunt may develop due to severe PAH causing deterioration of symptoms [2].

Presystolic accentuation and opening snap are less commonly seen. Also due to posterior shifting of the left ventricle auscultatory features of MS are missed. Irreversible pulmonary vascular disease is very uncommon in the presence of large ASD and high atrial pressure because of MS. According to the present understanding of this syndrome ASD and MS can be acquired or congenital. In iatrogenic LS ASD diameter ranges from 0.5-1.0 cm (restrictive) whereas in classical LS ASD is large and nonrestrictive.

LS may manifest at any age. But in our case, large OS ASD (non-restrictive) with severe MS delayed the diagnosis till the early fourth decade and went unnoticed even when the patient underwent 2 pregnancies & labor uneventfully. There are reports of LS that have manifested during pregnancy, especially in the third trimester [7]. Non-specific presentations like chest discomfort & fatigue brought her to clinical notice 4 years back. But a diagnosis of ASD was made & that of LS was missed. Their low economic status refrained her to achieve successful treatment and she was lost to follow-up until 15 days back when she presented to us with cardiac failure.

With the help of trans-thoracic echocardiography with color flow Doppler study which is the diagnostic modality of choice, we reached the diagnosis of LS with severe TR and moderate PAH. The subcostal window is the preferred view.

From our understanding of the hemodynamics of non-restrictive ASD, it is clear that doppler pressure half-time is not a reliable method as it overestimates the mitral valve orifice area missing the possibility of MS. Therefore, planimetry is considered a better method for the determination of MVA in LS [8]. Cardiac catheterization is not recommended routinely [9].

Recently, the percutaneous transcatheter intervention has become the most widely accepted therapy, using a balloon mitral valvuloplasty for MS and the Amplatzer atrial septal occluder for closure of an ASD [10]. It is preferred over surgical correction in terms of decreased morbidity and length of hospital stay [11]. Late presentation where PAH and severe TR (functional TR develops due to volume overloaded right

side and severe PAH) have already started to set in, the percutaneous intervention has limited utility. Development of PAH dictates poor outcomes and ineligibility for surgery. The majority of these patients die from heart failure, cardiac arrhythmias, and thromboembolic cerebrovascular diseases [12]. But this is a grey area where the decision to go for surgical correction is not well supported in the literature. This justifies our intention behind reporting this case.

Optimization of the patient before undergoing a surgical correction and post-operative elective ventilator support for a couple of days along with rigorous pre & post-op chest physiotherapy cannot be overemphasized to achieve a favorable outcome. Lutembacher syndrome is either underdiagnosed, missed, or misdiagnosed unless there is a high suspicion of the disease in the mind of the clinician and the echocardiologist while catering South East Asia.

Early diagnosis and surgical treatment bear a good prognostic value. If the patient is diagnosed late, pulmonary hypertension and heart failure develop and the prognosis is bad.

Unfortunately, most patients of low socioeconomic status coming from rural India present to the hospital in advanced stages with severe symptoms, and often the diagnosis is made too late for curative treatment. Together with the high cost of open heart surgery and lack of health insurance coverage, late presentation accounts for poor outcomes and high case fatality. Like in our case, financial constraints should not challenge a patient's right to receive curative treatment, that too at right time.

Conclusions

LS is a rare clinical entity. The size of ASD (restrictive/non-restrictive), the severity of MS, pulmonary vascular resistance, and compliance of the right ventricle are factors that influence the natural history and hemodynamics in patients with LS and therefore dictate the outcome of surgical intervention. Early diagnosis and surgical treatment bear a good prognosis and prolong survival. If the patient is diagnosed late, pulmonary hypertension and heart failure set in and the outcome becomes poor. Financial constraints on the part of patients delay definitive management even though a proper diagnosis is made. This is attributed to The higher cost of surgical intervention and the low socio-economic distribution of the disease in developing countries like India.

Disclaimer: Author's contributions

All the authors have contributed significantly to the content of this article. All the authors have read and approved the submission of this manuscript.

This manuscript has not previously been published in print or electronic form and is not under consideration by another publication.

Compliance with ethical standards was ensured and informed consent was taken from the patient whose case report is being published.

Conflict of interest: The authors declare that they have no conflict of interest.

Funding: No funding received

References

1. Lutembacher R. De la sténose mitrale avec communication interauriculaire. Arch Mal Coeur 1916;9:237-60.
2. Nagamani AC, Nagesh CM. Lutembacher Syndrome (Ch.64) in A Comprehensive Approach to Congenital Heart Diseases. Jaypee Brothers Medical Publisher, 2013: 908-16.
3. Ali SY, Rahman M, Islam M, et al. Lutembacher's Syndrome- A case report. Faridpur Med Coll J 2011;6:59-60.
4. Perloff JK. eds. The clinical recognition of congenital heart disease. 4th ed. Philadelphia: WB Saunders, 1994:323-8.
5. Fadel BM, Hiatt BL, Kerins DM. Isolated Rheumatic Tricuspid Stenosis with Reverse Lutembacher's Physiology. Echocardiography 1999;16:567-73.
6. Bashi VV, Ravikumar E, Jairaj PS, et al. Coexistent mitral valve disease with left-to-right shunt at the atrial level: clinical profile, hemodynamics, and surgical considerations in 67 consecutive patients. Am Heart J 1987;114:1406-14.
7. Kulkarni SS, Sakaria AK, Mahajan SK, et al. Lutembacher's syndrome. J Cardiovasc Dis Res 2012;3:179-81.
8. Aminde LN, Dzudie A, Takah NF, Ngu KB, Sliwa K, Kengne AP. Current diagnostic and treatment strategies for Lutembacher syndrome: the pivotal role of echocardiography. Cardiovasc Diagn Ther. 2015;5(2):122-132.
9. Budhwani N, Anis A, Nichols K, et al. Echocardiographic assessment of left and right heart hemodynamics in a patient with Lutembacher's syndrome. Heart Lung 2004;33:50-4.
10. Behjatiardakani M, Rafiei M, Nough H, et al. Transcatheter therapy of Lutembacher syndrome: a case report. Acta Med Iran 2011;49:327-30
11. Phan, Q. T., Nguyen, H. L., Le, T. D., Lee, W., et al. (2018). Combined Percutaneous Procedure in Patient with Lutembacher Syndrome: A Case Report & Real-World Experience Review. *Cardiology Research*, 9(6), 385-391.
12. Suzuki K, Ho SY, Anderson RH, et al. Morphometric analysis of atrioventricular septal defect with common valve orifice. J Am Coll Cardiol 1998;31:217-23.

Copyright (c) 2021 The copyright to the submitted manuscript is held by the Author, who grants the Clinical Medicine and Health Research Journal a nonexclusive license to use, reproduce, and distribute the work, including for commercial purposes.



This work is licensed under a [Creative Commons Attribution 4.0 International License](https://creativecommons.org/licenses/by/4.0/)