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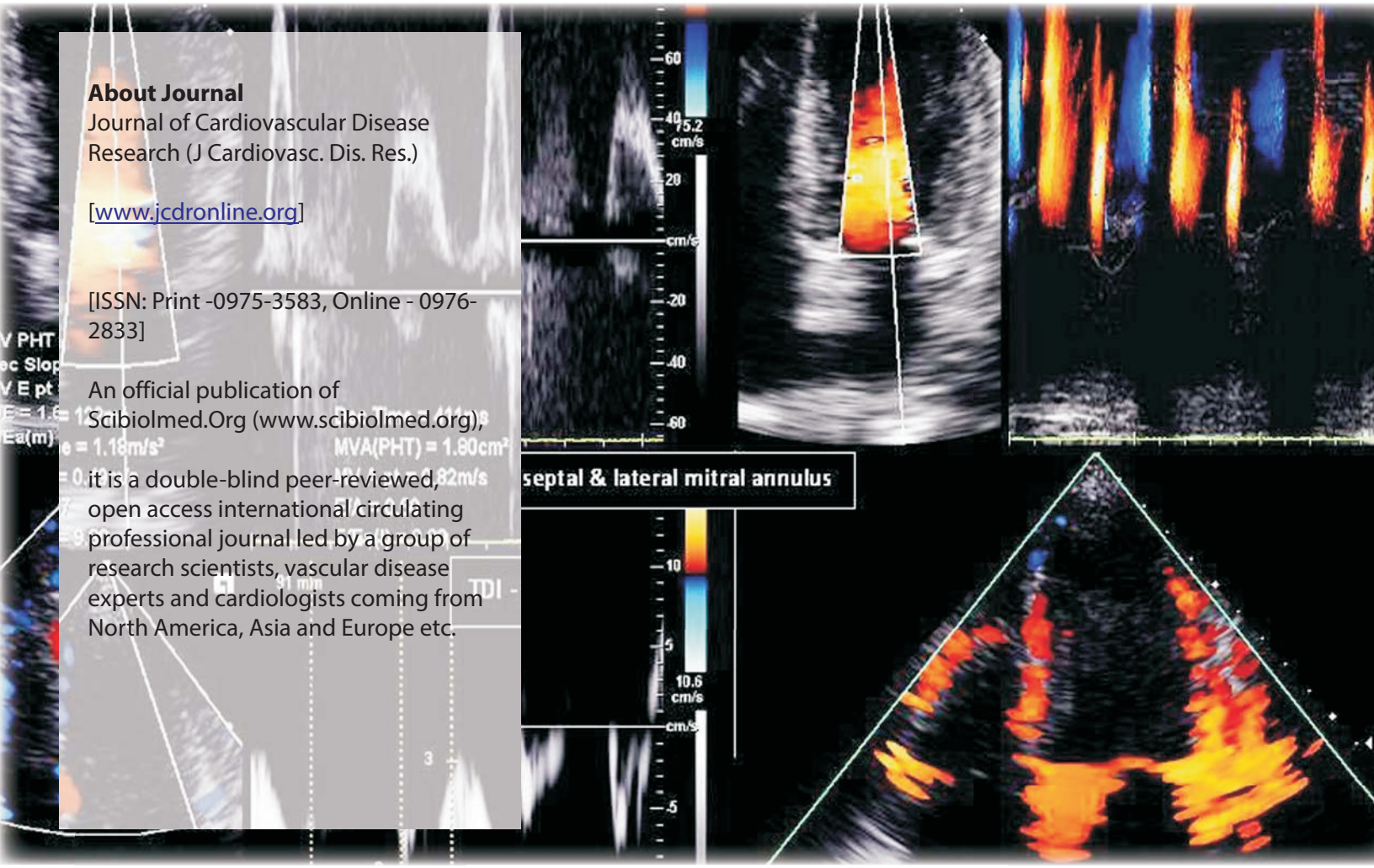

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Case Report

Lutembacher's Syndrome: A Rare Cause of Right Heart Failure

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ABSTRACT

The Lutembacher's syndrome is a rare, complex congenital heart disease, consisting of a congenital defect in the atrial septum (ostium secundum) along with acquired mitral stenosis. It is well established that early diagnosis and timely surgical treatment has good prognosis, while feasibility and safety of percutaneous treatment is demonstrated in few case reports.

Key words: Mitral stenosis, Atrial septal defect, Congenital heart disease, Heart failure.

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INTRODUCTION

Lutembacher's syndrome is an uncommon form of complex congenital heart disease characterized by combination of congenital atrial septal defect (ASD) and acquired mitral stenosis (MS). This rare form of disease may remain asymptomatic until late in life and diagnosis often missed leading to fatal outcome. Strong clinical suspicion is important for timely diagnosis to prevent the untoward complication from this complex disease especially in developing countries like India where prevalence of rheumatic heart disease is still high. Here, we will present a case of a middle aged lady who was admitted with right heart failure, diagnosed to be a case of Lutembacher's syndrome by echocardiography and managed conservatively with a plan for surgical correction later.

CASE HISTORY

A 43 years old married female was admitted with chief complaint of respiratory distress of 7 days. She was having breathing discomfort since her young age which was aggravated during her first pregnancy ten years back but was well tolerated and she delivered a normal healthy baby by cesarean section. Since then, she started having intermittent respiratory tract infection and progressive dyspnea. In the last 2 years, she developed several episodes of respiratory distress with mild bilateral leg swelling, easy fatigability, palpitation and orthopea. Dyspnoea was gradually progressive, non-seasonal, exertional, more on lying down and of NYHA (New York Heart Association) grade three. It was associated with mild productive cough with scanty expectoration. There was also history of gradually progressive distension of abdomen and bilateral lower limb swelling but no history of accompanying chest pain, fever or hemoptysis. There was no history of hypertension, diabetes mellitus, chronic obstructive airway disease, bronchial asthma and tuberculosis. She denied any history of rheumatic or congenital heart disease in the past.

On examination, she was afebrile with presence of pallor and bilateral pedal edema. Jugular Venous pressure was raised. Her pulse was 110 beats/min, low volume, irregular in rate and rhythm. Blood pressure was 100/70 mm Hg, with respiratory rate of 26 breaths/minutes. On cardiac examination, there was loud S1 with variable intensity and wide fixed splitting of S2 with opening snap. There was low pitched mid-diastolic rumbling murmur in mitral area without any radiation. There was an early systolic non radiating murmur in pulmonary area. There was no parasternal heave. Respiratory system examination revealed bilaterally equal normal breath sounds with bilateral lower zone fine crepitations. Abdominal examination revealed presence of ascites and mild tender hepatomegally.

Blood biochemistry revealed mild anaemia and congestive hepatitis, rest of the haematological profile was normal (Table 1). The anti nuclear antibody (ANA) and Rheumatoid Factor were negative. The viral markers for HIV-AIDS and hepatitis B & C were non reactive. Blood culture (three samples taken within 12 h of hospitalization), as well as urine culture were sterile. The sonography of abdomen revealed mild ascites and hepatomegally. Ascitic fluid was transudative in nature with high serum ascites-albumin gradient (SAAG) and low adenosine deaminase level. Electrocardiography showed Incomplete Right bundle branch block (RBBB), atrial fibrillation with controlled ventricular rate (Figure 1). Chest X Ray was showing cardiomegally with straightening of left cardiac border, double contour of left atrial border, dilated pulmonary artery with early feature of pulmonary venous congestion and right sided minimal pleural effusion. (Figure 2).

2D Echocardiography revealed dilated left atrium, right atrium, right ventricle and main pulmonary artery. The mitral valve was calcified with significant restriction of leaflet motion and thickened submitral apparatus. The aortic, tricuspid and pulmonary valves appeared to be structurally normal. Planimetry revealed a mitral valve area (MVA) of 1.3 cm^2 (Figure: 3A). The MVA calculated by spectral Doppler using the pressure half-time method revealed a MVA of 1.3 cm^2 (Figure 3B). Apical four-chamber view with color flow mapping showing left-to-right shunt flow and thickened rheumatic mitral leaflets (Figure 3C). Subcostal image was showing a discrete left to right shunt consistent with a secundum ASD of approximately 18 mm size(Figure 3D). There was also associated moderate tricuspid valve regurgitation with pulmonary arterial pressure of 60 mm Hg. With these findings, a diagnosis of Lutembacher's syndrome (ASD plus MS) was established.

She was managed conservatively with intravenous furosemide (40 mg twice daily), digoxine (0.25 mg daily, five days in a week) and subcutaneous lowmolecular weight heparin (40 mg twice daily) and later followed with oral diuretic (Furosemide and spironolactone), beta blocker (Metoprolol), oral anticoagulant (warfarin) and digoxin. She was also given the standard prophylaxis for rheumatic fever with injection benzathine Penicillin 12 lakh units deep intramuscular (after negative skin test) once every 3 weeks. She was discharged in hemodynamically stable condition.

Table 1: **Showing Laboratory findings in the patient with Lutembacher's syndrome**

Laboratory Parameter (units)	Report		Reference values
	On admission	At discharge	
Haemogram			
Haemoglobin (gm/dL)	10.6	10.9	12-18
Total Leucocytic count (x103/mm3)	5.00	8.60	4.0–11.0
Differential leucocytic count (%)			
Neutrophil	66	80	40-75
Lymphocyte	24	18	20-45
Monocyte	06	02	2-10
Eosinophil	04	00	1-6
Basophil	00	00	≤1
Platelet count ($x10^3/mm_3$)	200	280	150- 400
Erythrocyte Sedimentation rate (mm/h)	45	34	0-20
Liver Function Tests			
Bilirubin (mg/dL)			
Total	0.6	0.9	0.3–1.3
Direct	0.2	0.5	0.1-0.4
Indirect	0.4	0.4	0.2-0.9
ALT (U/L)	126	43	7-41
AST (U/L)	86	35	12-38
Alkaline Phosphatase (IU/L)	143	98	30-120
Protein, total (g/dL)	7.8	7.4	6.3-8.2
Albumin (g/dL)	4.4	4.2	3.5-5.0
Globulin (g/dL)	3.4	3.2	1.5-3.0
Coagulation Profile			
Prothrombin Time (s)	18.1	24.6	12.7–15.4
INR	1.47	2.3	1.34
APTT (s)	41.2	45.4	26.3–39.4
Renal Profile			
Serum Creatinine (mg/dL)	1.5	1.0	0.5- 0.9
Blood Urea (mg/dL)	15	38	10-50
Sodium (meq/L)	136	138	135-145
Potassium (meq/L)	3.6	3.8	3.5-5.5
Calcium (mg/dL)	9.2	9.3	8.7–10.2
Chloride (meq/L)	101	102	90-110
Blood Glucose, Random (mg/dL)	103	108	70-140
Blood Culture	Sterile	Sterile	
Urine Culture	Sterile	Sterile	
HIV I	Non reactive	Not done	
HBsAg	Negative	Not done	
Anti HCV	Negative	Not done	

ALT=Alanine Aminotransferase; AST=Aspartate Aminotransferase; INR=International Normalised Ratio;

APTT=Activated Partial Thromboplastin Time.

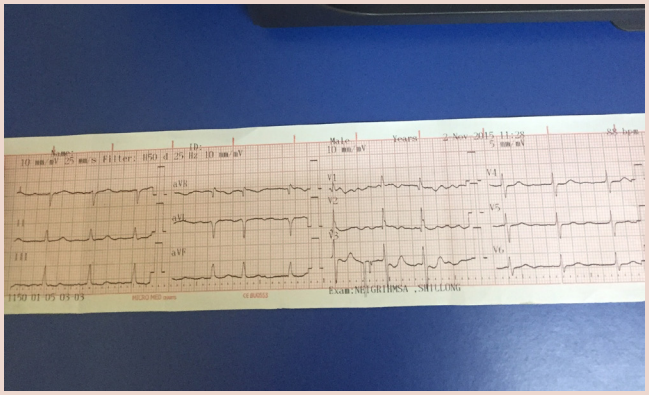

Figure 1: Electrocardiography showed Incomplete Right bundle branch block (RBBB), atrial fibrillation with controlled ventricular rate

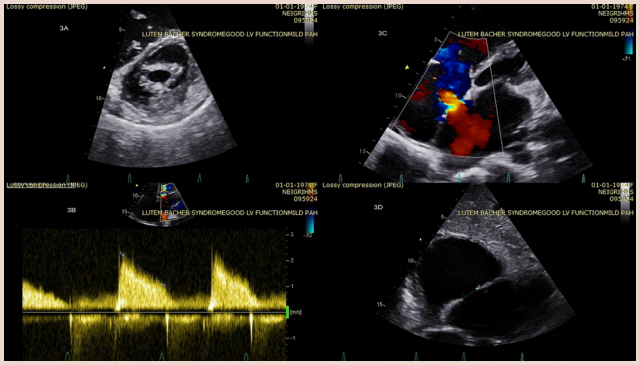

Figure 3: 2D Echocardiography revealed dilated cardiac chambers and main pulmonary artery. The mitral valve was calcified and thickened. Planimetry revealed a mitral valve area (MVA) of 1.3 cm^2 (Figure: 3a). The MVA calculated by spectral Doppler using the pressure half-time method revealed a MVA of 1.3 cm^2 (Figure 3b). Apical four-chamber view with color flow mapping showing left-to-right shunt flow and thickened rheumatic mitral leaflets (Figure 3c). Subcostal image was showing a discrete left to right shunt consistent with a secundum ASD of approximately 18 mm size (Figure 3d)

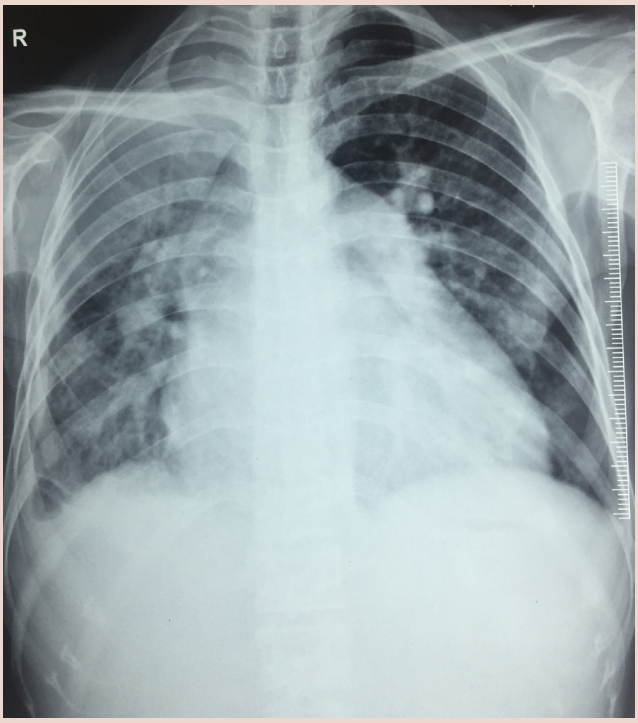

Figure 2: Chest X Ray was showing cardiomegally with straightening of left cardiac border, double contour of left atrial border, dilated pulmonary artery with early feature of pulmonary venous congestion and right sided minimal pleural effusion

She was discharged in hemodynamically stable condition with a plan for high risk surgical correction.

DISCUSSION

The association of ASD and MS was originally described by Corvisart in 1811 and was later in 1916 it was published as a combination syndrome by Lutembacher in a 61 year old lady who had seven normal deliveries before.[1] The disease is more common in females because ostium secundum atrial septal defect and rheumatic mitral stenosis are both more prevalent in females. The incidence rate of MS in ASD has been estimated at 4% while incidence rate of ASD in MS has been estimated at 0.6-0.7%. The incidence rate of coexisting rheumatic Mitral Stenosis depends on the geographic prevalence of rheumatic fever. In the underdeveloped countries, a history of rheumatic fever has been reported in 40% of patients with this condition.[2,3]

Lutembacher's syndrome is a rare, complex congenital heart disease. There are limited epidemiological data in medical literature on this rare syndrome The current consensus is that Lutembacher's syndrome consists of a congenital defect in the atrial septum (ostium secundum) along with acquired mitral stenosis. When these disorders coexist, each modifies the hemodynamic and clinical expression of other. Mitral stenosis increases the resistance to flow from left atrium into left ventricle and thus augments the Left to Right shunt in proportion to the severity of mitral stenosis.[3] The mitral valve is normally the only exit from the left atrium. In Lutembacker's syndrome, ASD constitute an alternative exit that decompresses the left atrium and in doing so, reducing the haemodynamic severity of MS by diminishing the trans mitral gradient, and thus masking symptomatology of MS albeit at the cost of increased haemodynamic severity of ASD**.** In brief the MS increases the shunt flow across an ASD and an ASD decreases the gradient across a stenotic mitral valve.[4] Since the right ventricle is more compliant than the left ventricle, blood flows to right atrium through ASD instead of going into pulmonary veins, thus preventing pulmonary edema. Early diagnosis and surgical treatment have good prognosis.[7,8] If patient develops pulmonary hypertension and heart failure, prognosis become poor.[5] Other dreaded complications which can be fatal are cardiac arrhythmias and thrombo-embolic cerebrovascular disease.[6] If the patient is diagnosed at an early stage before development of complication like pulmonary hypertension or heart failure-ASD closure with mitral valve repair or replacement bears a good prognosis and prolong survival.[9]

CONCLUSION

As our patient is having late presentation with hepatomegaly and ascites (ominous feature of RV failure) ethical dilemma and unanswered QUESTION remain there regarding her long term prognosis and definitive

management strategy. Should she be offered any intervention, surgical or percutaneous, later being less invasive OR should be said NO for any intervention and managed with medical treatment only considering her poor surgical outcome?

ABBREVIATION USED

ASD: Atrial Septal Defect; **MS:** Mitral Stenosis; **NYHA:** New York Heart Association**; ANA:** Anti Nuclear Antibody; **MVA:** Mitral Valve Area; **RBBB:** Right Bundle Branch Block.

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