Journal of Rare Cardiovascular Diseases
ISSN: 2299-3711 (Print) | e-ISSN: 2300-5505 (Online)
www.jrcd.eu
RESEARCH ARTICLE

Successful Pregnancy in a Woman Operated for Lutembacher's Syndrome with Sinus Venosus Type ASD: A Rare Journey

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Article History
Received: 10.11.2025
Revised: 22.11.2025
Accepted: 05.12.2025
Published: 17.12.2025

Abstract: ***Introduction:*** Lutembacher's syndrome is a rare cardiac condition characterised by the coexistence of mitral stenosis and an atrial septal defect, which may be congenital, acquired, or iatrogenic. The interplay between these lesions alters intracardiac hemodynamics, often masking or modifying the severity of each component. Pregnancy poses additional physiological stress, with increased blood volume and cardiac output potentially exacerbating symptoms and complications. Careful preconception counselling, multidisciplinary management, and individualised anticoagulation strategies are therefore crucial to ensure optimal maternal and fetal outcomes. We report a 29-year-old G3P1L1A1 woman with Lutembacher's syndrome (sinus venosus type ASD), who presented at 26 weeks of gestation with fatigue, exertional dyspnoea, palpitations, gestational diabetes, and moderate anaemia (modified WHO 2.0 pregnancy risk classification III, NYHA II). She had a history of rheumatic fever and underwent mechanical mitral valve replacement at 19 years, followed by six thrombotic episodes, all managed with thrombolysis. Her current pregnancy was uneventful, and she delivered a healthy 2.3-kg baby at term via elective LSCS with tubal ligation. ***Conclusion*** Multidisciplinary team care, along with regular antenatal visits that emphasise risk assessment, counselling, and vigilant monitoring, is essential for optimising pregnancy outcomes and ensuring maternal–fetal well-being in cases of Lutembacher's syndrome.

Keywords: Lutembacher's syndrome, mechanical valve, pregnancy outcome, sinus venosus type Atrial Septal Defect, thrombolysis, case report

INTRODUCTION

Lutembacher's syndrome (LS) is an exceptionally rare cardiac anomaly, with an estimated incidence of 0.001 per 1,000,000 individuals [1]. However, its prevalence may be higher in regions where rheumatic fever remains endemic. The syndrome encompasses a spectrum of presentations, all defined by the coexistence of mitral stenosis (MS) and an atrial septal defect (ASD)—most commonly of the ostium secundum type. Either or both lesions may be congenital, acquired, or occasionally iatrogenic, the latter often resulting from a transseptal puncture during mitral valvuloplasty for acquired MS [2]. The most frequent form involves a congenital ASD with an acquired, typically rheumatic, MS component [3]. Among the various ASD subtypes, the sinus venosus variant accounts for only about 5–10% of cases [4], rendering its association with LS particularly uncommon. The following case study aptly demonstrates the importance of vigilance and a customised care bundle in such unique situations. Written informed consent was obtained from the patient after ensuring confidentiality, and approval from the Institutional Ethics Committee was secured.

Case Description

A 29-year-old woman, gravida 3 para 1 living 1 abortion 1 (G3P1L1A1), presented to our tertiary care centre at 26 weeks of gestation with complaints of fatigue, exertional dyspnoea, and palpitations. She had a childhood history of rheumatic fever and was on nicoumalone (2.5 mg once daily), aspirin (150 mg once daily), and carvedilol (10 mg daily) at the time of presentation.

A pre-prosthetic transthoracic echocardiogram (TTE) performed at the age of 19 years revealed a large sinus venosus-type atrial septal defect (ASD) with a left-to-right shunt and dilatation of the left atrium(LA), right atrium(RA), and right ventricle(RV). She underwent mechanical mitral valve(MV) replacement at that time. Subsequently, she developed six episodes of recurrent prosthetic valve thrombosis—three before her first pregnancy, one at four weeks postpartum, and two during the interpregnancy interval. All episodes were managed successfully with conservative thrombolytic therapy. Owing to repeated thrombotic events, persistently elevated mitral valve gradients, and one permanently immobile leaflet, a redo mitral valve replacement was recommended in 2022. However, the patient declined surgical intervention and was discharged on continued medical management following a cardiothoracic (CTVS) consultation.

At the time of the current pregnancy, she appeared poorly nourished with a body mass index (BMI) of 16.2 kg/m². Her modified WHO 2.0 pregnancy risk classification was

III, and she was categorised as NYHA functional class II. Echocardiography showed restricted disc movement of prosthetic MV, normal LV, dilated LA, mild Aortic Regurgitation (AR), and grade II Tricuspid Regurgitation (TR). Left Ventricular Ejection fraction was noted to be 60%. (Figure 1).



Figure 1. 2D Echocardiography with colour Doppler (operated case of Lutembacher's syndrome) showing restricted disc movement of prosthetic MV

Review of literature:

The European Society of Cardiology (ESC) Guidelines for the management of adult congenital heart disease (ACHD), formerly known as Grown-Up Congenital Heart Disease, emphasise that advances in medicine, surgery, and technology have significantly improved survival among individuals born with congenital heart disease (CHD), with more than 90% now surviving into adulthood.[5] This improvement has, in turn, contributed to a rising prevalence of pregnancies complicated by CHD—estimated to have increased from 6.4 to 9.0 per 10,000 hospital deliveries.[6]

Lutembacher's syndrome (LS) is more commonly encountered in young women, with a history of rheumatic fever reported in up to 40% of cases. The most frequent presenting symptoms include fatigue, exertional dyspnoea, and palpitations, though the clinical manifestations and haemodynamic impact depend on the delicate interplay between mitral stenosis (MS) and atrial septal defect (ASD). [1,7]

Diagnosis can be challenging, as the presence of an ASD often masks the classical signs of MS. Two-dimensional transthoracic echocardiography (TTE) remains the gold standard for confirming the diagnosis, assessing severity, and formulating management strategies. If left uncorrected, a large ASD (often >1.5 cm) in the setting of coexisting MS may exacerbate the left-to-right shunt, leading to progressive pulmonary hypertension and, eventually, Eisenmenger's physiology.[2] Rarely, a "reverse Lutembacher's syndrome" has been described, characterised by tricuspid stenosis and predominant right-to-left shunting across the ASD. [8] Definitive correction of LS can be achieved either through open surgical repair or a percutaneous approach, depending on anatomical and haemodynamic considerations.[9]

Preconception assessment is a crucial step in managing women with ACHD. Several validated models—including the CARPREG (Cardiac Disease in Pregnancy) risk index, CARPREG II, the ZAHARA score, and the modified WHO pregnancy risk classification—are used to estimate maternal cardiac risk

based on parameters such as cardiac history, NYHA functional class, and current cardiac status. According to these models, our patient was categorised as mWHO2.0 class III, Bethesda classification: moderate (mitral disease) to simple (repaired sinus venosus ASD without residua), and had a CARPREG II score of 3.[10,11]

Women with mechanical valve prostheses require lifelong anticoagulation due to the thrombogenic nature of these valves. While bioprosthetic valves pose a lower risk of miscarriage, they lack long-term durability and are prone to structural valve deterioration. [.12] [The incidence of valve thrombosis, even under adequate anticoagulation, ranges between 0.5% and 8%, influenced by the type and position of the prosthesis—the cage-ball valve exhibiting maximal thrombogenicity, while bileaflet valves carry a low risk. [13] Recurrent episodes of thrombosis, such as in our patient, typically necessitate urgent surgical intervention. Pillai *et al.* reported a similar scenario wherein a postpartum mitral valve thrombus developed following a preterm caesarean section. [14]

Despite these complexities, most women with cardiovascular disease in pregnancy—both congenital and acquired—can anticipate favourable outcomes, provided that risk factors are recognised early and managed appropriately. Studies by Marghade *et al.* and Adu-Boakye *et al.* have shown that Lutembacher's syndrome carries a good prognosis in the absence of pulmonary hypertension and with timely diagnosis and intervention. [7,8] Kulkarni *et al.* also reported the case of a multiparous woman diagnosed with LS during early labour who successfully delivered vaginally without complications.[15].

Patient Management and Follow-up

The patient was enrolled at our institute under the Very High-risk category and received comprehensive antenatal surveillance. She continued haematinics, multivitamins, and underwent serial obstetric ultrasonography to assess fetal growth and well-being. A repeat echocardiogram at term revealed a normal-sized left ventricle with a preserved ejection fraction of 60%, along with mild pulmonary hypertension. (Figure 2)

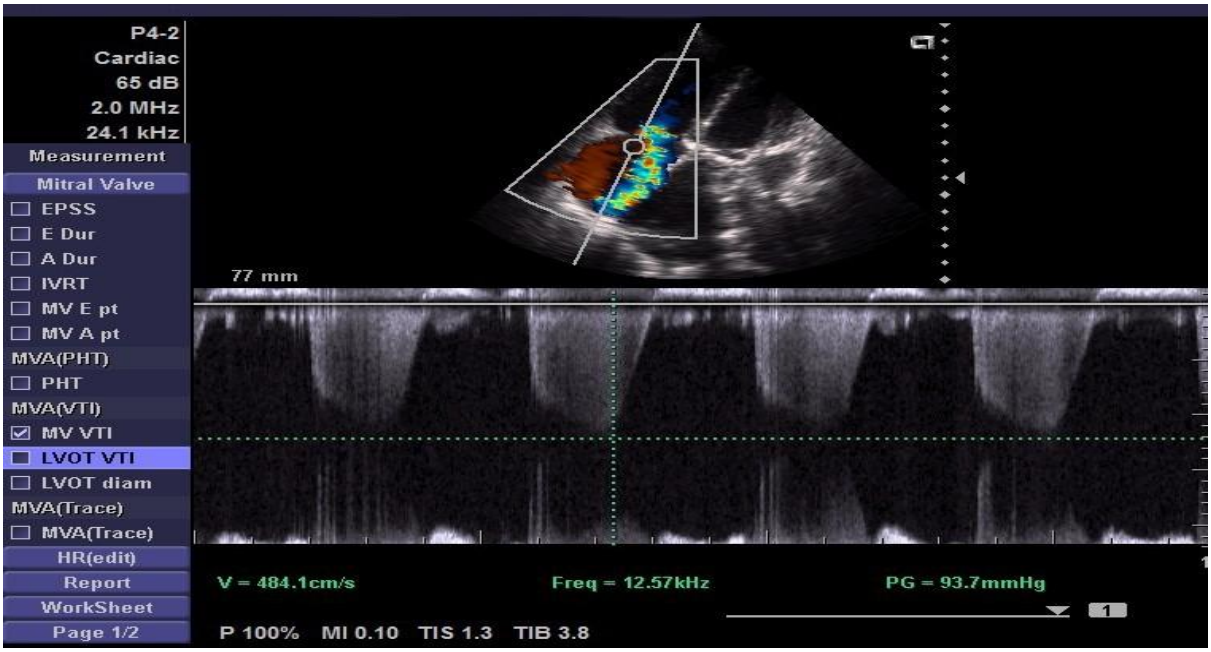


Figure 2. 2D Echocardiography (operated case of Lutembacher's syndrome) showing a normal-sized left ventricle, left ventricular ejection fraction (LVEF) of 60%, and mild pulmonary hypertension

During the course of pregnancy, she developed gestational diabetes mellitus (GDM) and moderate anaemia. Both conditions were managed according to standard clinical protocols—oral iron supplementation for anaemia and oral hypoglycaemic therapy for glycaemic control. She remained under close multidisciplinary supervision throughout pregnancy, with regular antenatal visits and no cardiac complications. At term, an elective lower-segment caesarean section with tubal ligation was performed, resulting in the delivery of a healthy 2.3-kg infant. The postpartum course was uneventful, and both maternal and neonatal outcomes were favourable. She continued regular follow-up in the postpartum outpatient department for six months and appeared clinically stable, maintaining a satisfactory general condition. However, during a routine maternal follow-up inquiry conducted one year later, it was reported that the patient had passed away at home. The exact circumstances surrounding her demise remain unknown.

Clinical Significance: Lutembacher's syndrome in

pregnancy poses a unique hemodynamic challenge because the ASD may initially mask symptoms of mitral stenosis by decompressing the left atrium, delaying diagnosis until pregnancy unmasks decompensation. Prompt recognition and tailored management are crucial to mitigate substantial maternal and fetal morbidity and mortality.

CONCLUSION

This case underscores the pivotal role of a multidisciplinary team approach, integrating preconception risk assessment, comprehensive counselling, and meticulous antenatal surveillance, in optimising the course of pregnancy and ensuring favourable maternal and fetal outcomes in women with Lutembacher's syndrome.

Legends

Figure 1 2D Echocardiography with colour Doppler (operated case of Lutembacher's syndrome) showing restricted disc movement of prosthetic MV.

Figure 2 2D Echocardiography (operated case of Lutembacher's syndrome) showing a normal-sized left ventricle, left ventricular ejection fraction (LVEF) of 60%, and mild pulmonary hypertension.

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