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Lutembacher's Syndrome Presenting in the Third Trimester: A Multidisciplinary Challenge in a Low-Resource Setting

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Case Report

Keywords: Lutembacher's Syndrome, pregnancy, labour, Atrial septal defect, mitral valve stenosis, low resource setting

Posted Date: December 8th, 2025

DOI: https://doi.org/10.21203/rs.3.rs-8061405/v1

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Additional Declarations: No competing interests reported.

Version of Record: A version of this preprint was published at BMC Cardiovascular Disorders on January 26th, 2026. See the published version at https://doi.org/10.1186/s12872-026-05550-0.

Abstract

Background

Lutembacher's Syndrome is a rare case, defined as Mitral valve stenosis in association with atrial septal defect (ASD). Both defects, Mitral stenosis and ASD may be congenital or acquired in origin. Poses significant challenges during pregnancy. The prevalence of this syndrome is not well-documented, but it is considered rare.

Case presentation:

We report the case of a 40-year-old Afghan pregnant woman presenting with NYHA class IV heart failure symptoms one day before labour. Echocardiography revealed severe mitral stenosis, a large secundum ASD with left-to-right shunt, severe pulmonary hypertension, and multivalvular regurgitation. She also had atrial fibrillation, anemia, and fetal congenital heart defects. Medical stabilization allowed successful assisted vaginal delivery of a preterm infant. Despite the need for urgent postpartum surgery, intervention was refused due to financial and cultural constraints.

Conclusion

This first reported case of Lutembacher's syndrome in Afghanistan underscores the critical impact of late diagnosis, particularly in pregnant women, where the physiological burden of pregnancy can exacerbate undetected cardiac disease. In resource-limited settings, economic problems and cultural barriers often delay both diagnosis and timely intervention. Such delays increase the risk of severe maternal and fetal complications, making pregnancy management significantly more complex.

Background

Lutembacher's Syndrome is a remarkably uncommon medical condition described by René Lutembacher (1884–1968) of Paris for the first time in 1916, referring to a congenital or acquired mitral valve stenosis associated with an atrial septal defect, either congenital or iatrogenic, following a transseptal mitral valvoplasty. Although individually both ASD and MS are well-characterized, their combination in Lutembacher's Syndrome results in a distinctive hemodynamic interaction influenced by factors such as the duration of disease progression, the size of the ASD, and the severity of the mitral valvopathy [3]. The increased left atrial pressure from mitral stenosis facilitates left-to-right shunting across the ASD, often leading to right atrial and ventricular volume overload. This shunting can paradoxically mask symptoms of pulmonary congestion, delaying clinical recognition and diagnosis. Its clinical manifestations can vary from subtle exertional dyspnea and fatigue to overt signs of right-sided heart failure or pulmonary hypertension [4]. While the precise prevalence of Lutembacher's syndrome is elusive, its incidence is more remarkable in regions where rheumatic valvular heart disease is prevalent, limited to the consistency of ASD incidents [5]. In areas like Afghanistan, the dual burden of untreated congenital heart disease and rheumatic heart disease (RHD) remains significant due to limited access to routine

screening and preventive care [6]. It's noteworthy that no documentation of LS cases is present in Afghanistan.

Here we report for the first time Lutembacher's Syndrome in an Afghan pregnant woman, masked even until 32 weeks of her second pregnancy, diagnosed one day before her labour.

Case report

A 40-year-old pregnant woman in her last trimester, a day before her labour, presented to the cardiology department of Ariana Medical Complex, with shortness of breath, New York Heart Association (NYHA) class IV, associated with fatigue, weakness, dry cough on lying down, and lower extremities swelling. The patient has no prior medical history of Hypertension, diabetes mellitus, or significant lung diseases. She has a history of a caesarean section 3 years ago with an alive baby.

The patient had been experiencing shortness of breath worsening day by day from dyspnea with mild exercises to dyspnea during the lying position (orthopnea) associated with severe pedal edema. Upon initial assessments, her vital signs were as follows: a blood pressure of 110/80 mmHg, pulse rate of 118 bpm (irregular-irregular), oxygen saturation of 96%, and temperature of 37,5 degrees Celsius.

On physical examination, she was found to be anxious, pale, with no signs of icterus, cyanosis, or clubbing, except for bilateral three positive pitting edema. On cardiac auscultation, a grade-III rumbling diastolic murmur at mitral level and early to mid-systolic loud murmur at mitral and tricuspid levels were audible. Respiratory system examination revealed bilaterally equal normal breathing sounds at the apex and mid with absent respiratory sound in the left basal area. Abdominal examination revealed, caesarean section scar, Linea nigra, mild hepatosplenomegaly, striae gravidarum, symphysial fundal height of 32 weeks, longitudinal lie and breech presentation

Electrocardiography (ECG) showed Atrial fibrillation with Rapid ventricular response subsequently, her *Echocardiogram* depicted dilatation of Right atrium, right ventricle, pulmonary artery and left atrium, mitral valve leaflets were thickened, calcified, causing moderate to severe mitral valve stenosis (area of 1.19 cm^2) , *(figure-1,2)*, moderate to severe mitral and tricuspid valves regurgitation, severe pulmonary hypertension, pulmonary artery systolic pressure of 74mmHg and large secondum type ASD measuring 1.5 cm, (*figure-3)*, shunting left to right confirmed by echo-contrast (*figure-4)*, no clot, effusion or vegetation were detected, LV systolic function was in normal range (LVEF= 50-55%).

Abdomin-pelvic Ultrasound showed Congestive liver (*figure-5)*, Bilateral mild hydronephrosis, Mild splenomegaly, single intrauterine pregnancy of $30^{+}/^{-}2$ weeks with breech presentation.

Blood examination represents; Hemoglobin of 7.8g/dl, WBC count of 17.200 cells /ml, platelets were 121.000 cells/ml, Serum albumin of 2.3g/dl and Serum creatinine of 0.8 mg/dl along with normal range of liver function test.

Fetus Echocardiography showed Partial AV-canal defect, Muscular and small apical type VSDs, with patent foramen oval and ductus arteriosus.

Based on all the findings mentioned above, the patient was declared to be in critical condition and required close monitoring and management under the supervision of the cardiology and gynecology teams in the intensive care unit. The initial management focused on stabilizing the patient's condition medically. Integrated management of heart failure, including the use of intravenous diuretics for volume overload, rate and rhythm control, and anticoagulation, was initiated, resulting in clinical improvement for the patient. Following stabilization, the cardiac surgery team recommended that definitive surgical intervention, including atrial septal defect (ASD) closure combined with mitral valve replacement, be performed as soon as possible after delivery. However, the procedure was refused due to financial and cultural constraints.

On the second day of admission, the patient developed worsening abdominal pain indicative of active labor. Examination revealed full cervical dilatation, ruptured membranes, and a breech fetus at zero station, consistent with advanced precipitated labor. Following multidisciplinary consultation, assisted breech vaginal delivery was undertaken with an episiotomy. A preterm infant was delivered successfully and admitted to the NICU for specialized care. The third stage of labor was actively managed, the episiotomy was sutured, and standard measures were implemented to prevent postpartum hemorrhage. Post-delivery, the patient was monitored in the ICU, where she subsequently developed multiple critical complications. These included episodes of malignant cardiac Arrhythmias, which were promptly managed with appropriate antiarrhythmic medications, direct current shock, and temporary pacemaker insertion. Notably, she had multiple episodes of cardiac arrest, each of which was successfully resuscitated with cardiopulmonary resuscitation measures. In addition, the patient suffered from several episodes of seizures, which were effectively controlled with anticonvulsant therapy. Continuous multidisciplinary care was provided throughout the postpartum period to ensure stabilization of her general condition. Following a 14-day stay in the intensive care unit (ICU), the patient was discharged in a vital and stable condition.

Discussion

Lutembacher's Syndrome, the combination of mitral stenosis and ASD, is a rare but clinically significant cardiac syndrome, with a global incidence of just one in a billion individuals [7] and an estimated incidence of just 0.001 per million people in the United States[8]. This rate is notably higher in Southeast Asia than in Western countries, largely due to the greater occurrence of rheumatic heart disease in those regions[1]. The relevance of such epidemiological insights is particularly evident in South Asia, including Afghanistan, where the healthcare system is fragmented and the burden of RHD remains high and continues to be a significant cause of cardiovascular morbidity in low- and middle-income countries [9] [10]. Since Rene tembacher's first described the condition in a 61-year-old woman with congenital ASD and mitral stenosis who had experienced seven pregnancies [4], The literature has consistently reported that the condition tends to affect females more frequently, as Lutembacher's primary components, both

atrial septal defects (ASDs) and mitral stenosis, are more prevalent among women[11]. While the condition can remain asymptomatic for decades of life, some cases have been detected as late as the seventh decade of life[12]; earlier diagnosis, particularly in women of reproductive age, offers significant benefits in preventing complications during pregnancy and childbirth. However, in many low-resource settings, timely diagnosis remains elusive, as seen in this case. In Afghanistan, limited access to healthcare services is compounded by widespread poverty, gender disparities, and poor public health literacy [9]. These structural deficiencies disproportionately affect women, who often rely on male family members for financial support and decision-making and face cultural and economic barriers in seeking healthcare. The challenges faced by women in accessing healthcare in Afghanistan are well-documented, with reports indicating that limited female autonomy and low maternal health literacy substantially hinder the timely diagnosis and management of chronic diseases, including cardiovascular disorders [13]. This situation is further reflected in Afghanistan's consistently low Global Gender Gap Index ranking [14], which underscores the systemic barriers to women's healthcare access and decision-making autonomy. Our case illustrates this challenge. A 40-year-old woman in her third trimester presented with severe dyspnea and atrial fibrillation, indicative of decompensated Lutembacher's syndrome. Notably, the diagnosis was made only one day before delivery, despite her history of a previous cesarean section. This delay highlights a critical diagnostic gap in antenatal care. It underscores how even contact with healthcare systems does not guarantee comprehensive cardiac evaluation, particularly in under-resourced environments where antenatal visits often focus exclusively on obstetric issues. Further complicating her clinical trajectory was her postpartum refusal of surgical correction. Financial constraints, strong cultural opposition to invasive interventions, and lack of personal autonomy played decisive roles in her decision, emphasizing that even when care is available, social determinants of health can act as powerful barriers to treatment. Therefore, what makes our case unique is not just the late diagnosis, but also the sociocultural and healthcare challenges that contributed to the delay. It highlights how gender roles and weaknesses in the health system can affect patient care.

From a clinical perspective, a late diagnosis of Lutembacher's syndrome during pregnancy carries significant risks. The physiological changes of pregnancy, particularly increased plasma volume and cardiac output, can further worsen the compromised cardiovascular function caused by mitral stenosis and atrial septal defect [4]. Without timely intervention, patients are at high risk for complications such as right heart failure, pulmonary hypertension, atrial fibrillation, thromboembolism, and even sudden cardiac death [15]. In our case, the patient experienced atrial fibrillation, episodes of ventricular arrhythmias, and seizures, necessitating intensive care, which further complicated her postpartum course. Although no thromboembolic events were noted, the presence of such severe arrhythmias highlights the potential for rapid decompensation in the peripartum period. This reinforces the need for heightened vigilance, multidisciplinary collaboration, and critical care support in managing similar cases, especially in settings where tertiary-level services are not available or inadequately resourced. This case, to our knowledge, is the first documented Lutembacher's Syndrome being diagnosed during the third trimester of pregnancy in Afghanistan. It offers a rare but powerful lens into the intersection of clinical

complexity and structural inequity. The delayed diagnosis reflects systemic weaknesses that extend beyond the individual and speak to broader failings in healthcare delivery, particularly for women. Addressing such cases requires a multifaceted approach. Improving access to echocardiographic screening for women of reproductive age is essential, especially in high-risk populations. It is also important to include basic cardiovascular assessments in routine antenatal care, a step that could enable earlier identification of structural heart disease in pregnant women. Beyond structural reforms, public health initiatives must aim to raise awareness about RHD and congenital heart disease. This should include community-based education campaigns and school health programs that promote understanding of mouth hygiene, early symptoms of cardiac disease, and the importance of echocardiographic screening in pregnant women. Empowering women to access healthcare services independently is also critical. This will require targeted strategies to enhance maternal health literacy, promote female autonomy in health decision-making, and implement gender-sensitive healthcare policies that recognize and accommodate the unique barriers women face in accessing care.

Conclusion

This case represents the first documented occurrence of Lutembacher's syndrome in Afghanistan, demonstrating the critical consequences of delayed diagnosis of cardiovascular diseases in a low-resource setting, especially during pregnancy. It underscores significant gaps in antenatal cardiovascular screening, especially in regions with a high burden of rheumatic heart disease. Successful maternal and neonatal outcomes were achieved through coordinated, multidisciplinary care despite the patients' multiple complications.

Although there was a clear surgical indication, the patient's refusal of radical surgical correction of cardiac abnormalities in the postpartum period highlights the significant impact of social determinants on healthcare outcomes. Financial limitations, cultural apprehensions about invasive procedures, and restricted female autonomy contributed to this issue, suggesting that the mere availability of medical services does not guarantee their utilization. Adequate healthcare, particularly in resource-limited settings, must, therefore, address sociocultural barriers alongside clinical needs to empower women in healthcare decision-making. Such an integrated approach is essential to prevent diagnostic delays and improve outcomes for women with structural cardiac disease in low-resource environments.

Abbreviations

(ASD): Atrial Septal Defect

(RHD): Rheumatic heart disease

(NYHA): New York Heart Association

(ECG): Electrocardiography

Declarations

Ethics approval and consent to participate

The Institutional Review Board of Ariana Medical Complex approved the study. The patient and her relatives signed informed consent forms before her inclusion.

Consent for publication

The patient gave written informed consent and agreed to her medical information being shared for research purposes and publication.

Availability of data and materials

The data supporting the findings of this study are available from the corresponding author upon reasonable request.

Competing interests

The authors declared no conflicts of interest.

Funding

No funding was received.

Clinical trial number:

Not applicable.

Authors' contributions

AWS was responsible for project conceptualization, supervision, investigation, methodology, and manuscript review.

AR and MN contributed to the investigation and methodology, and were involved in drafting and revising the manuscript.

HRN performed the diagnostic imaging and contributed to the investigation.

MNassery provided gynecological and obstetric consultation, and participated in the investigation and treatment.

KS provided surgical consultation.

KR managed data collection and curation.

All authors reviewed and approved the final version of the manuscript.

Acknowledgements

The authors would like to express their sincere gratitude to Assistant Professor Nommanudien Naibkhil, Department of Biochemistry, Faculty of Pharmacy, Kabul University, and Department of Research, Ariana Medical Complex, and Negara Nabi, Department of Research, Arian Medical Complex, for there valuable guidance, critical review, and continuous support during the preparation and revision of this case report.

AI usage: ChatGPT was used to improve language**.**

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Figures

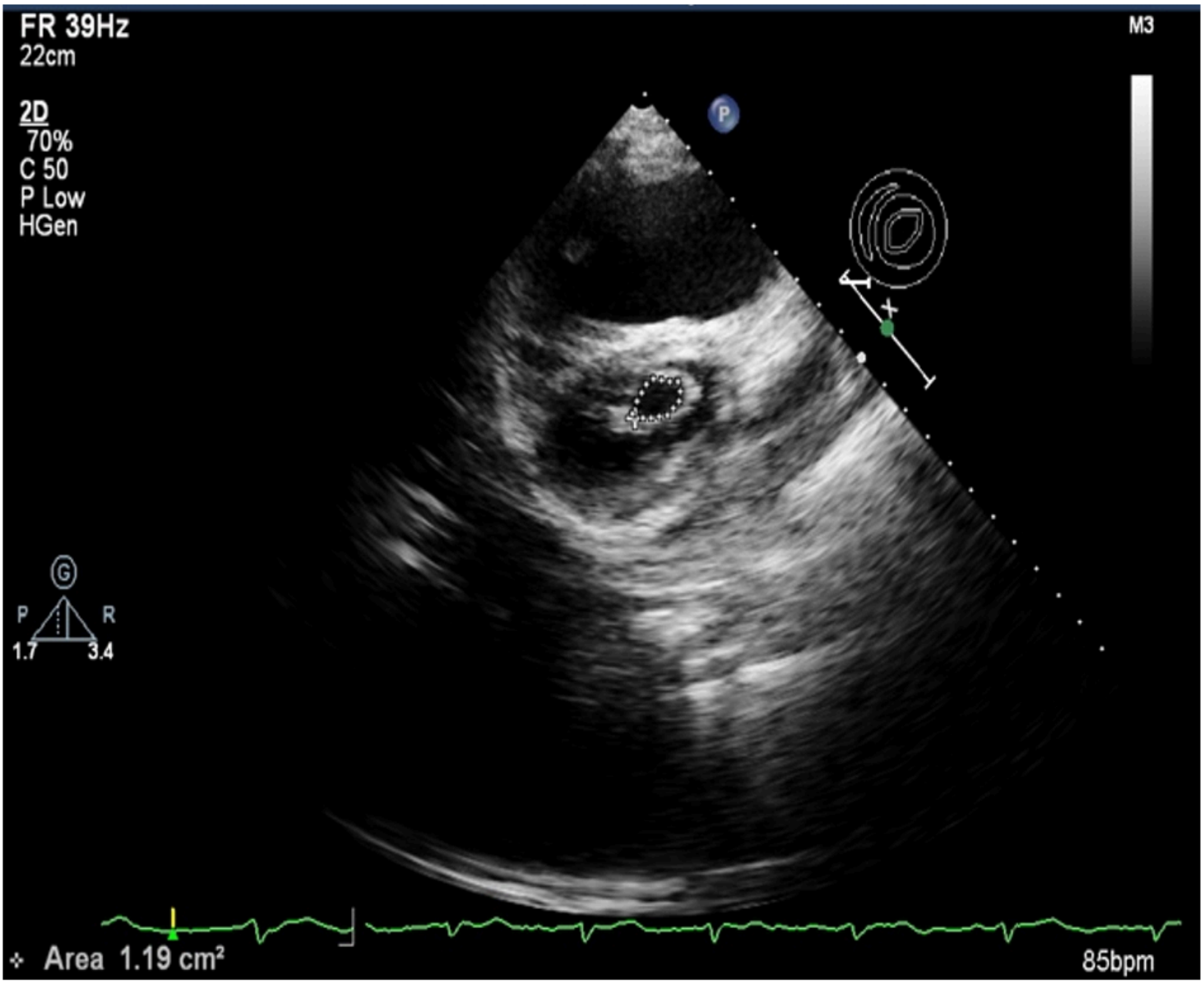


Figure 1

Echocardiography image in the parasternal long-axis (PLAX) view at the mitral valve level demonstrates moderate to severe mitral valve stenosis, with a calculated mitral valve area of 1.19 cm².

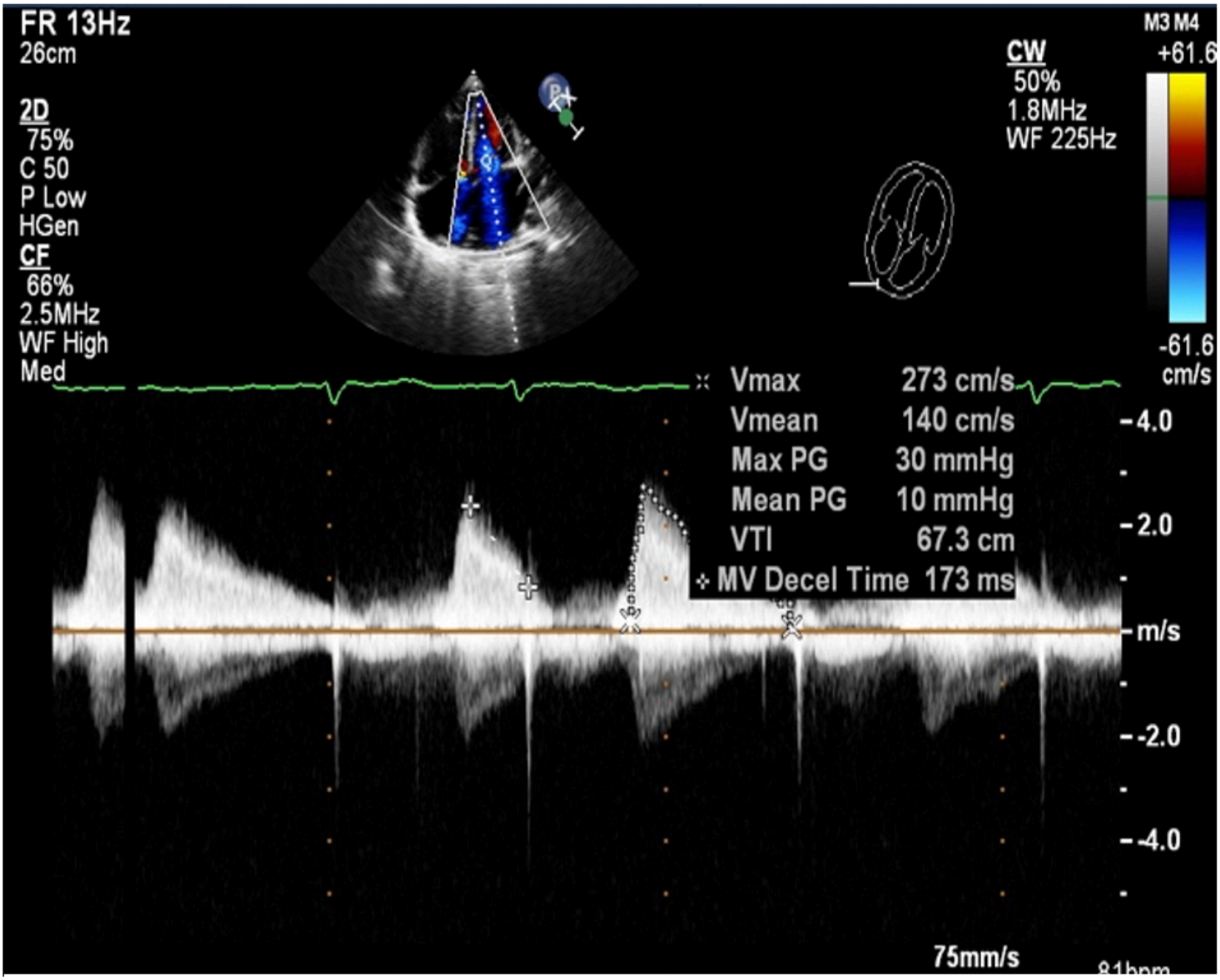


Figure 2

Echocardiography in the apical four-chamber (A4C) view with spectral Doppler assessment of the mitral valve demonstrates moderate to severe mitral stenosis, with a transmitral pressure gradient of 30/10 mm Hg (peak/mean).

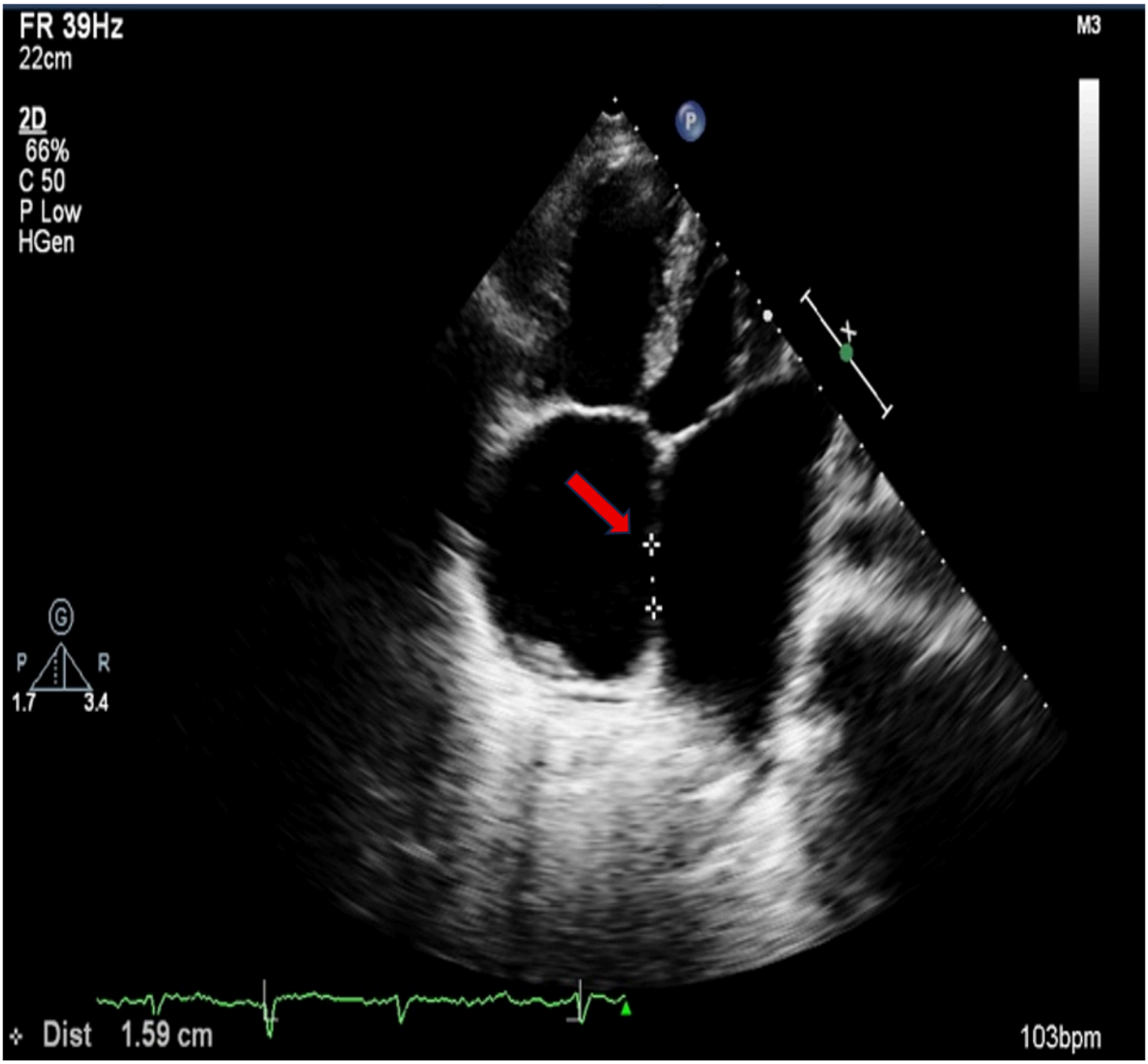


Figure 3

Echocardiography image, apical four-chamber (A4C) view, showing large ASD with measurement 1.5 cm

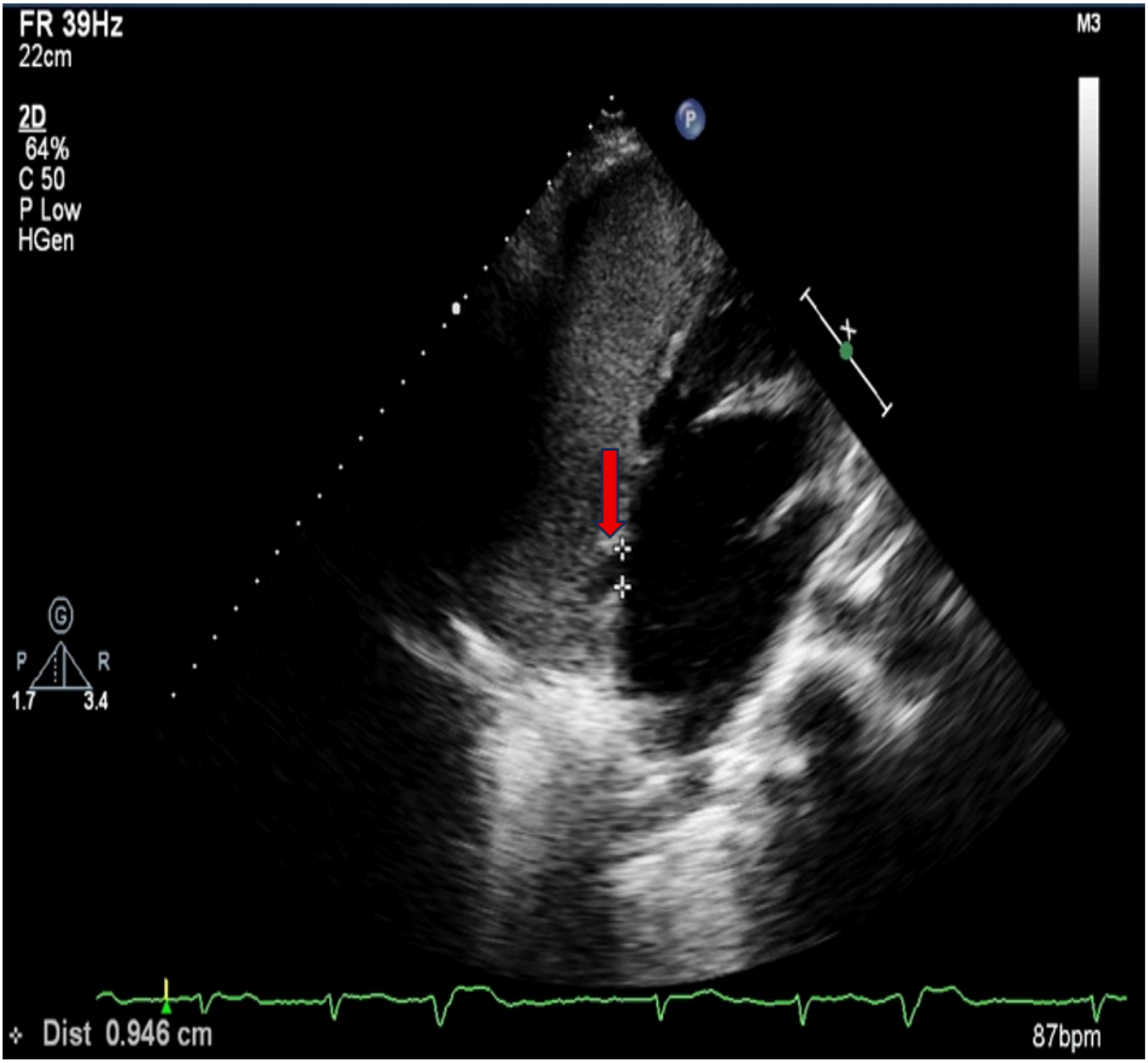


Figure 4

Contrast echocardiography image in the apical four-chamber (A4C) view demonstrates a left-to-right shunt.

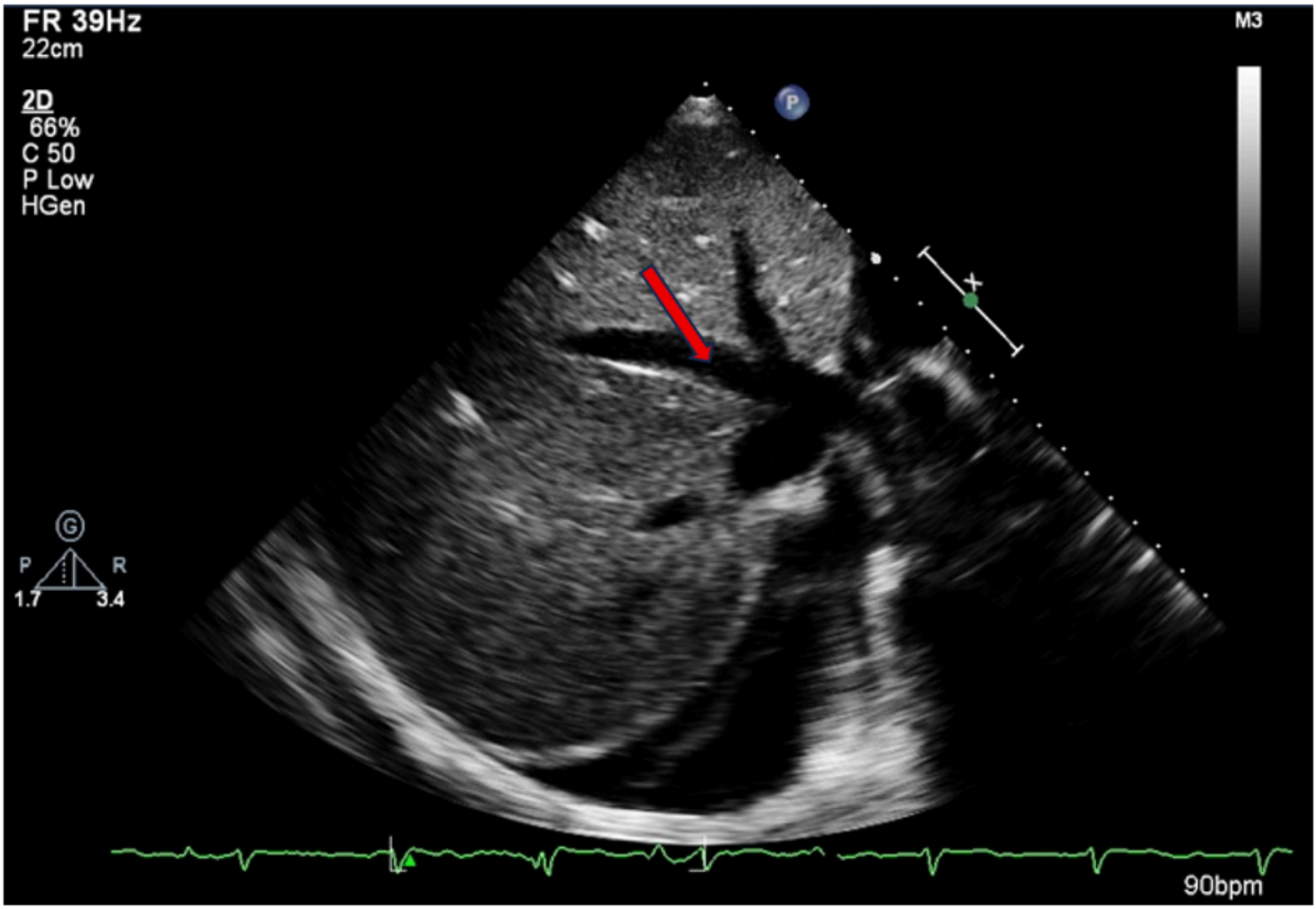


Figure 5

Ultrasound image of the liver demonstrating features of congestion (bunny sign).