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CASE REPORT

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A rare case report of iatrogenic Lutembacher's syndrome with cyanosis and hemodynamic insights

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ABSTRACT

Background: Iatrogenic Lutembacher's syndrome is a rare condition defined by the presence of an acquired atrial septal defect (ASD) along with rheumatic mitral stenosis, frequently resulting from transseptal interventions like balloon mitral valvotomy (BMV).

Case Presentation: We report the case of a 39-year-old woman with rheumatic heart disease who presented with breathlessness, central cyanosis and peripheral oedema for 10 days. She had previously undergone BMV twice. Clinical evaluation indicated signs of right heart failure and cyanosis. An ECG revealed atrial fibrillation with a rapid ventricular response, and chest radiography showed cardiomegaly. Echocardiography indicated moderate mitral stenosis, significant tricuspid regurgitation, tricuspid stenosis, right ventricular dysfunction, and two residual iatrogenic ASDs with bidirectional shunting, predominantly from right to left. The patient's condition improved with conservative management. Although a closure procedure was initially planned, the heart team decided to postpone the intervention due to the severe right ventricular dysfunction. The patient has been under regular follow-up for the past one and a half years, during which her symptoms have been well-controlled.

Conclusions: Persistent iatrogenic ASDs can result in significant hemodynamic consequences, including hypoxemia, especially when there are elevated pressures on the right side of the heart. This case highlights the hemodynamic importance of persistent iatrogenic ASDs and underscores the need for individualized decision-making based on a collaborative heart team approach in their management.

KEY WORDS

Iatrogenic Lutembacher's syndrome; Atrial septal defect; Balloon mitral valvotomy; Right-to-left shunt; Cyanosis

ARTICLE HISTORY

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Introduction

Iatrogenic Lutembacher's syndrome is characterized by the rare association of mitral stenosis alongside an acquired ASD, which typically arises from transseptal interventions like BMV, catheter ablation, or left atrial appendage occlusion [1–3]. The incidence of iatrogenic ASDs is well established, with reported rates varying from 16% to 89%, influenced by the procedural method and the imaging techniques employed [4,5]. Although these defects are generally small and associated with left-to-right shunting that frequently resolves on its own, some may remain persistent and develop significant hemodynamic effects [6]. In certain cases, especially when there is an elevation in right atrial pressure persistent ASDs can lead to right-to-left shunting, which can cause systemic desaturation. This is particularly relevant in patients who have concomitant tricuspid valve disease, pulmonary hypertension, or right ventricular (RV) dysfunction.

We are presenting a rare case of iatrogenic Lutembacher's syndrome in a patient who had rheumatic mitral stenosis and significant tricuspid valve disease. This patient experienced central cyanosis as a result of bidirectional shunting through the residual iatrogenic ASDs. This case highlights the crucial role of echocardiography in diagnosing and managing complications that can arise after cardiac interventions.

Case Presentation

A 39-year-old woman with a history of chronic rheumatic heart disease presented with progressive breathlessness, central cyanosis, bilateral lower limb edema, and abdominal distension for 10 days. She had undergone BMV twice, 15 and 5 years prior to this presentation. No other significant medical, family and psycho-social history.

On physical examination, she had an irregularly irregular pulse, central cyanosis with an SpO_2 of 78%, bilateral pitting pedal edema, and no clubbing. Jugular venous pressure was elevated 15 cm above the sternal angle. Cardiovascular examination revealed a grade 3/6 systolic murmur best heard at the tricuspid area.

Electrocardiography showed atrial fibrillation with a fast ventricular rate. Chest radiography revealed cardiomegaly with features of grade 2 pulmonary venous hypertension.

Transthoracic echocardiography (TTE) demonstrated moderate mitral stenosis with a mitral valve area of 1.6 cm^2 and a transmitral gradient of 12/4 mmHg at a heart rate of 74 beats per minute, along with trivial mitral regurgitation. The left atrium was dilated (LA volume index: 76.7 mL/m2), and the anterior mitral leaflet (AML) exhibited doming in diastole

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with turbulent flow across the mitral valve. Thickened aortic valve leaflets were noted, with mild aortic regurgitation, and the coronary sinus appeared dilated (Figure 1).

The interatrial septum was bulging toward the left atrium. Two residual iatrogenic ASD, each measuring approximately 4–5 mm and located near the fossa ovalis, were visualized with bidirectional shunting, predominantly right-to-left (Qp/Qs = 0.85) (Figure 2).

Torrential organic tricuspid regurgitation was noted, with a vena contracta measuring 22 mm on the apical four-chamber view. Additionally, systolic flow reversal in the hepatic vein confirmed the severity of regurgitation (Figure 3). Continuous-wave Doppler across the tricuspid valve revealed a gradient of 13/7 mmHg and an estimated right ventricular systolic pressure (RVSP) of 45 mmHg (Figure 4). Tricuspid valve morphology demonstrated leaflet thickening with restricted diastolic opening, consistent with organic (rheumatic) tricuspid stenosis. The right ventricle was dilated and dysfunctional, with a tricuspid annular plane systolic excursion (TAPSE) of 11 mm (Figure 5). The right atrium was markedly enlarged (indexed area 42.0 cm^2/m^2), and the inferior vena cava was dilated (3.0 cm) and non-collapsing. Left ventricular function was normal.

A final diagnosis of Iatrogenic Lutembacher's syndrome with organic torrential tricuspid regurgitation, mild-to-moderate tricuspid stenosis, right ventricular dysfunction, mild pulmonary hypertension, bidirectional atrial shunt, atrial fibrillation with fast ventricular rate, and right heart failure was established. The patient was hospitalized and managed with intravenous loop diuretics (furosemide), followed by oral maintenance therapy. A mineralocorticoid receptor antagonist (spironolactone) was added for sustained volume control. Rate-control agents were administered for atrial fibrillation, and long-term oral anticoagulation (warfarin) was continued, resulting in significant symptomatic improvement. Given the presence of significant bidirectional interatrial flow, percutaneous closure of the residual defects was initially contemplated. Following evaluation by the multidisciplinary heart team, it was concluded that intervention would carry prohibitive risk in the context of advanced right ventricular dysfunction. The patient has remained on regular follow-up over the past one and a half year, with satisfactory control of right heart failure symptoms on optimized medical management. A chronological summary of her clinical course, from initial diagnosis to current stable condition, is presented in Figure 6.

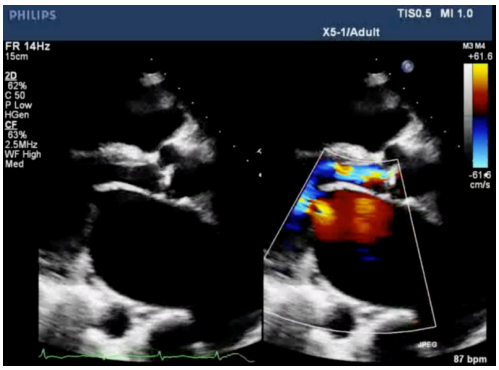

Figure 1. Parasternal long axis view showing dilated left atrium (volume: 115 mL), thickened anterior and posterior mitral leaflets with diastolic doming of the anterior leaflet, turbulent mitral inflow, thickened aortic valve leaflets with mild aortic regurgitation, and dilated coronary sinus.

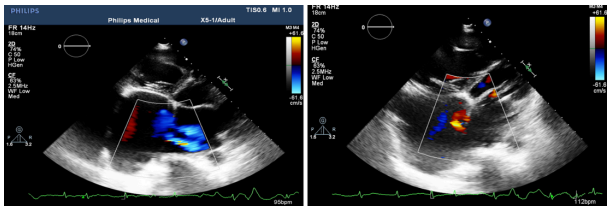

Figure 2. Apical four-chamber view showing markedly dilated right atrium with interatrial septum bulging leftward. Two iatrogenic ASDs (4–5 mm) near the fossa ovalis show bidirectional shunting, upper panel: predominant right-to-left shunt; lower panel: early systolic left-to-right shunt.

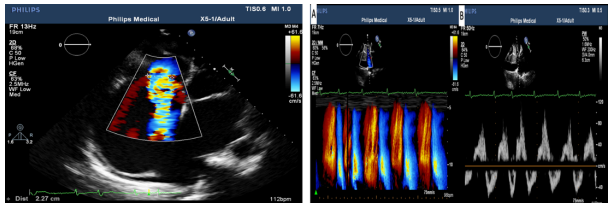

Figure 3. Upper panel - Apical four-chamber view showing torrential tricuspid regurgitation with a vena contracta of 22 mm, Lower panel - (A) Colour Doppler image and (B) pulse-wave Doppler tracing demonstrating systolic flow reversal in the hepatic vein, indicating the severity of tricuspid regurgitation.

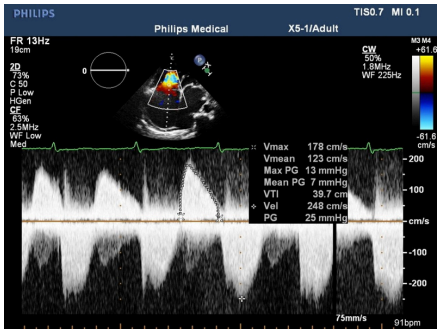

Figure 4. Continuous-wave Doppler across the tricuspid valve demonstrating a pressure gradient of 13/7 mmHg and an RVSP of 45 mmHg.

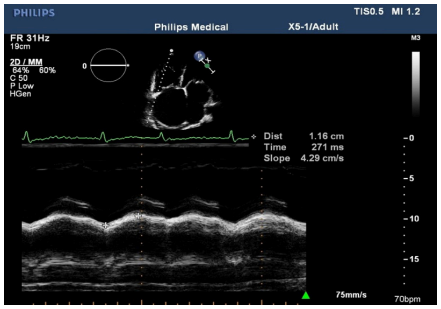

Figure 5. M-mode echocardiography across the tricuspid annulus showing reduced tricuspid annular plane systolic excursion (TAPSE) of 11 mm.

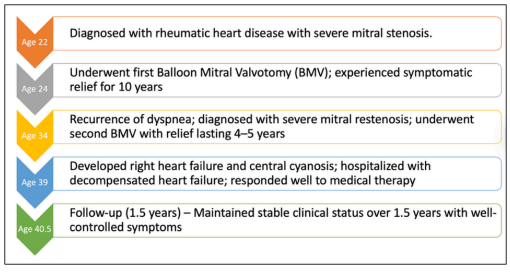

Figure 6. Clinical timeline of the patient.

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Discussion

Iatrogenic ASDs are increasingly recognized following transseptal interventions. Although most are hemodynamically insignificant and close spontaneously, persistent ASDs may alter atrial pressure dynamics and result in clinically significant outcomes. In the presence of mitral stenosis, the shunt is typically left-to-right due to elevated left atrial pressures. Although Lutembacher's syndrome is defined by mitral stenosis and ASD, our patient had only moderate MS (MVA 1.6 cm^2). The dominant physiology was driven by torrential TR, TS, and RV dysfunction, which primarily determined right atrial pressure elevation and shunt direction. However, when right atrial pressure exceeds that of the left, such as in the setting of torrential tricuspid regurgitation, tricuspid stenosis, pulmonary hypertension, or RV dysfunction, a right-to-left shunt may develop [1–3].

The prevalence of iatrogenic ASDs varies depending on procedure type and imaging modality. Manjunath et al. reported an incidence of 66.5% immediately after BMV, with only 8.7% persisting at six months [5]. Linhart et al. observed persistent ASDs in 37% of patients at a median follow-up of 2.9 years following cryoballoon ablation, though none required closure [6]. These findings indicate that while spontaneous resolution is common, a subset of iatrogenic ASDs may persist and become hemodynamically significant, particularly in the presence of elevated right-sided pressures.

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Figure 7 illustrates the typical left-to-right shunting pattern due to residual mitral valve gradient. However, when right atrial pressure transiently or persistently exceeds left atrial pressure during Valsalva maneuvers, coughing, or in the setting of chronic right heart failure reversal of shunt direction may occur. In our patient, the cyanosis mechanism is depicted in Figure 8, where RV dysfunction and torrential tricuspid regurgitation resulted in right atrial pressure elevation and septal bowing toward the left, facilitating right-to-left shunting.

Diagnosis of such cases requires a high index of suspicion. While TTE can demonstrate the defect and shunt direction, transesophageal echocardiography (TEE) and intracardiac echocardiography (ICE) offer superior visualization and flow assessment [9,10]. Invasive hemodynamic studies may be required when noninvasive imaging is inconclusive [11].

Management strategies depend on the size and hemodynamic impact of the defect. Small, asymptomatic left-to-right shunts may not require closure. However, in symptomatic patients with right-to-left shunting and hypoxemia, closure should be considered following optimization of right-sided pressures through medical therapy, including diuretics and rate control [12–14]. Closure has been shown to improve oxygenation and symptoms in such patients [15,16].

The procedural technique also influences ASD formation. Use of dual sheaths through a single puncture has been shown to increase the risk of persistent iatrogenic ASDs compared to single-sheath or separate punctures [17]. Preventive measures include using the smallest necessary sheath, minimizing septal trauma, and routine echocardiographic follow-up in high-risk patients.

Nonetheless, not all patients are suitable for defect closure. The heart team opted for conservative management as a definitive approach due to poor RV function and elevated right-sided pressures. Liver function was mildly impaired, consistent with congestive hepatopathy. No surgical or rhythm interventions were planned, given procedural risk and limited benefit. Surgical risk assessment (EuroSCORE II: 14.8%) further supported this decision [6,13]. The patient had already shown favourable clinical improvement with conservative therapy, including intravenous diuretics, rate control and anticoagulation (warfarin) [12–14]. Hence, a tailored, non-interventional approach was adopted in line with the patient's individual risk profile and response to medical management [13,14].

In the presence of torrential tricuspid regurgitation, tricuspid stenosis, and significant right ventricular dysfunction, the residual ASD in our patient may have served a compensatory role, facilitating right-to-left shunting to decompress the overloaded right atrium. In such scenarios, the ASD functions as a "pop-off" valve, preventing excessive right atrial pressure buildup and subsequent systemic venous congestion. Closure of these defects may paradoxically worsen right heart failure by increasing right-sided pressures and precipitating RV decompensation [6,13]. Additionally, the mixed obstructive-regurgitant physiology of the tricuspid valve reduces effective forward flow, further limiting the RV's ability to tolerate changes in preload or afterload. Given the patient's clinical improvement with optimized diuretic therapy, rate control agents and anticoagulation, the heart team deemed the risks of intervention to outweigh the benefits. Thus, conservative therapy was selected as the most appropriate and individualized management strategy, emphasizing the importance of tailoring decisions to each patient's unique hemodynamic context [13,14].

This case underscores the importance of individualized, heart team–based decision-making. While closure may be beneficial in selected patients [15,16], conservative management remains a valid and often preferable strategy in high-risk individuals, provided there is symptomatic control and close follow-up [13,14]. Moreover, this report reinforces the importance of recognizing residual iatrogenic ASDs as a potential cause of cyanosis after transseptal procedures, especially in the setting of advanced right-sided cardiac disease [7,17].

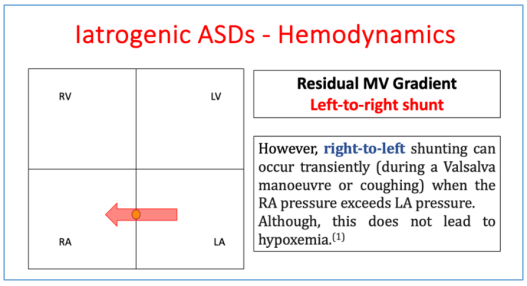

Figure 7. Hemodynamics of iatrogenic ASDs: Left-to-right shunting is typical due to residual mitral valve gradient. However, right-to-left shunting may occur transiently (e.g., during Valsalva or coughing) or persistently when right atrial pressure exceeds left atrial pressure.

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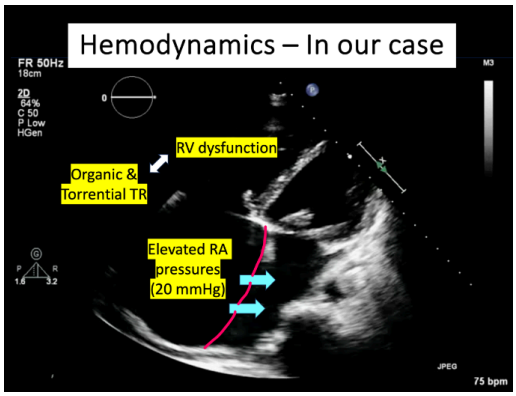

Figure 8. Mechanism of cyanosis in our patient: Organic tricuspid regurgitation and right ventricular dysfunction resulted in elevated right atrial pressure. This pressure shift caused septal bowing towards the left and right-to-left shunting through residual iatrogenic ASDs.

Patient's Perspective

I have been having heart problem since many years. When I was 24, doctors did a balloon surgery through my groin (BMV). After that, I felt fine and lived a normal life for a long time. But again, I started having breathing trouble and weakness, so I had to undergo the same balloon surgery one more time. It helped me for a few more years. One and a half year back, I got swelling in my legs, became very tired, and my lips turned blue. I got admitted to hospital. Doctors told me there was pressure buildup in my heart and a small hole from earlier surgeries was still there. They treated me with medicines and I started feeling better. They said another surgery was not possible now as my heart had become weak. I am continuing with medicines and regular checkups. I gave my full consent for sharing my story in this medical paper. I feel happy that other doctors can read about my case and learn something, because this is a very rare condition. I thank all my doctors who took care of me and are still guiding me.

Conclusion

Iatrogenic ASDs are a recognized a complication of transseptal procedures such as BMV. While most of the ASDs are small and clinically insignificant, persistent ASDs can become hemodynamically significant, especially in the presence of severe tricuspid valve disease, pulmonary hypertension, or right ventricular dysfunction. These conditions may cause right-to-left or bidirectional shunting, resulting in systemic desaturation and cyanosis. A thorough understanding of a patient's hemodynamics is essential for accurate diagnosis and treatment planning. Although closing the ASD might benefit some patients, it may not be appropriate for those with advanced right ventricular dysfunction, where the ASD acts as a decompressive outlet. In such cases, conservative management with medication and monitoring can maintain clinical stability while reducing procedural risks. This case underscores the importance of individualized, heart team–approach and highlights the role of comprehensive hemodynamic assessment in optimizing outcomes after transseptal interventions.

Declarations

Ethics approval and consent to participate

Ethical approval was not required, and written informed consent was obtained from the patient and the relative.

Consent for publication

Written informed consent was obtained from the patient.

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