
A Patient with Right-to-Left Shunt Atrial Septal Defect Without Pulmonary Hypertension

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Abstract: We present a case of atrial septal defect (ASD) in a 27-year-old young woman who presented with a right-to-left shunt despite normal pulmonary artery pressure without anomalous pulmonary venous drainage and only mild tricuspid regurgitation (TR), but it was associated with right ventricular dysfunction. Considering the dysfunction of the right heart and significant dilatation of the tricuspid annulus, the surgeon still decided to repair ASD and perform tricuspid valvuloplasty. The surgery lasted approximately three hours, and the whole procedure was smooth and uneventful. Her postoperative recovery was uneventful and discharged in good condition. The occurrence of right-to-left (R-L) shunting in patients with ASD is uncommon and is usually an ominous sign, heralding irreversible pulmonary hypertension (PH). There is little information about similar shunts for patients without PH and tricuspid regurgitation (TR), and the majority of these patients have an anatomic anomaly that favors this type of shunt. ASD with R-L shunt without Eisenmenger physiology is rarely reported and may be underestimated due to difficulty in its diagnosis. Intraoperative transesophageal echocardiography (TEE) examination should not only focus on cardiac structure but also the evaluation of ventricular function. Right-to-left shunt is possible in ASD patients even with normal pulmonary arterial pressure.

Keywords: Transesophageal Echocardiography, Atrial Septal Defect, Right-To-Left Shunting, Right Ventricular Function, Pulmonary Hypertension

1. Introduction

ASD is a common congenital heart disease (CHD) [1, 2], which is taught as acyanotic CHD traditionally. In patients with ASD, usually left to right shunt flow is predominant [3]. The occurrence of right-to-left (R-L) shunting in ASD patients even with normal pulmonary arterial pressure is uncommon but is possible which is usually an ominous sign, heralding irreversible pulmonary hypertension (PH) [4, 5]. There is little information about similar shunts for patients without PH and TR, and the majority of these patients have an anatomic anomaly that favors this type of shunt.

Cyanosis due to right -to-left shunt across an atrial septal defect (ASD) brings up lots of questions in the curious mind.

Common reasons for right-to-left shunt with normal pulmonary pressure in ASD include inter-atrial pressure gradient and heart anatomical distortion. What needs us to do are ruling them out based on history, examination, and actively looking for the aetiology. Myocardial biopsy is considered if necessary. Understanding the mechanism of cyanosis in the presence of ASD through comprehensive transesophageal echocardiography (TEE) examination is vital for surgical planning and decision-making.

2. Case Presentation

A 27-year-old young woman presented to our hospital with complaints of mild chest discomfort without dyspnea during exercise. Physical examination revealed cyanosis and a

systolic murmur over the second left intercostal space. There were no other abnormalities on cardiovascular or respiratory physical examination. She denied of a congenital heart disease and hadn't received any clinical examination and treatment previously. A transthoracic echocardiogram (TTE) was performed, which revealed multiple dropouts in the atrial septal, suspicious low-speed shunt, right atrium (RA), and right ventricular (RV) enlarged (54mm and 33mm, respectively), mild tricuspid regurgitation, and normal left ventricular (LV) systolic function (ejection fraction was 69%). Hemoglobin compensatory increased to 18.5 g/dl due to long-term hypoxemia. Preoperative cardiac catheterization showed normal pulmonary artery pressure and without anomalous pulmonary venous drainage. She was scheduled for ASD

repair or closure surgery.

In the operating room, the patient was found hypoxemic, whose pulse oximetry was 84% within room air and increased to 92–95% within 100% oxygen. Central venous pressure was 17 mmHg. After uneventful induction, the TEE probe was placed for preoperative inspection. In the mid-esophageal four-chamber view, TEE revealed an unexpected R-L shunt flow at the level of the atria that was visualized by color-flow Doppler (Figure 1 A), which could explain the hypoxemia in the preoperative. Then, the peak tricuspid regurgitant (mild) velocity was 1.24m/s, PG=6 mm Hg (Figure 1 B), which suggested that the R-L shunt may not be second to pulmonary artery hypertension or tricuspid regurgitation (TR).

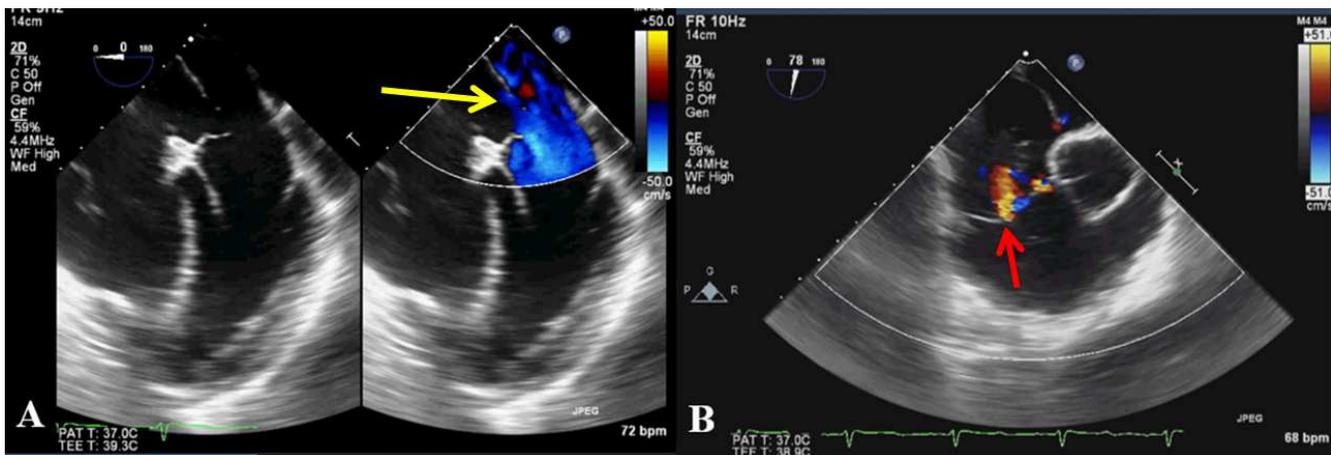


Figure 1. A. The ME-4C view seen here permits imaging of the RA, LA, RV, LV, and Color Doppler echocardiogram demonstrating R-L shunt flow at the level of the atria. Notably, the atrial septum is obviously pushed to the left during the diastolic period, indicating that the pressure of the right atrium is significantly higher than that of the left atrium (as shown by the yellow arrow); B. The ME RV Inflow-Outflow view shows mild tricuspid regurgitation with low-pressure gradient; the diameter of the pulmonary artery is smaller than the aorta (as shown by the red arrow).

There were no echocardiographic signs of evident valve structural abnormalities or any abnormal venous connection. Surgical exploration showed that there was no adhesion in the pericardium, and the heart was enlarged, mainly in the right atrium and right ventricle. The atrial septal defect was ethmoidal and secondary foramen, with a diameter of about 35mm, and there was no abnormal pulmonary venous drainage. Tricuspid annulus dilated obviously, with good texture and mild to moderate insufficiency. Surgical exploration confirmed the presence of ASD and the absence of anatomical distortion, myxomas, and pericardial adhesions. The surgery lasted approximately three hours, and the whole procedure was smooth and uneventful. Post-CPB TEE imaging revealed no evidence of residual shunt. Postoperative TTE showed no residual shunting, while LV systolic function was preserved (ejection fraction 47%). When the ASD was completely repaired, RV systolic function was significantly lower. Her postoperative recovery was uneventful and discharged in good condition. At the six-month follow-up, the patient's TTE showed left ventricular systolic function was normal (ejection fraction 60%), and mild right ventricular systolic dysfunction.

3. Discussion

ASD with R-L shunt without Eisenmenger physiology is rarely reported and may be underestimated due to difficulty in its diagnosis [6]. Common reasons for right-to-left shunt with normal pulmonary pressure in ASD include inter-atrial pressure gradient and heart anatomical distortion [5]. The former can be observed as a result of early atrial systole or during early ventricular systole, right atrial myxomas, adult respiratory distress syndrome, obstructive sleep apnea, lung resection, and mechanical ventilation, particularly with increased pulmonary end-expiratory pressure. This pressure gradient can be exacerbated by changes in posture, inspiration, and Valsalva maneuver. The latter can be seen as a result of abnormal venous connexions, Ebstein's malformation [7], pulmonary stenosis, loculated pericardial effusion [8], kyphoscoliosis or dilated aortic root and so on.

In this case, the above-mentioned abnormalities was ruled out according to the patient's history and intraoperative surgical exploration. And the preoperative TTE showed RA and RV enlarged. Preoperative central venous pressure (CVP) was 17 mmHg, and elevated central venous pressure was

indicative of right ventricular dysfunction (RVD) and/or fluid retention [9]. However, enlargement of the atria occurs when E/A is greater than 1, which is within the normal value but is pseudo-normalization [10]. What's more, we have calculated

the RV fractional area change (FAC) is 27% (Figure 2 B). On the basis of these findings, we can conclude that the R-L shunt in the absence of PH is secondary to RV dysfunction.

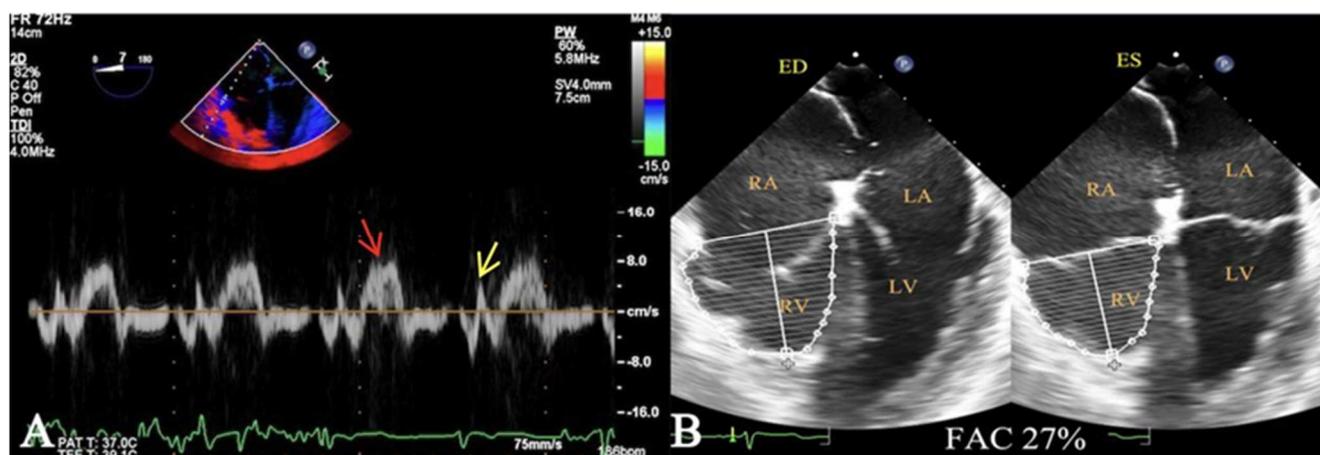


Figure 2. A. Pulsed wave tissue Doppler Imaging from the ME-4C view sampling from the tricuspid valve annulus. Tricuspid annulus tissue doppler: $e' > a'$ (the red arrow represents e' and the yellow arrow represents a'); B. Right ventricular fractional area change (FAC) is 27%.

Whether the patient should repair or occlude the ASD is a dilemma. If the patient has severe right heart dysfunction after the closure of ASD, it is necessary to urgently re-establish an ASD as a pop-off for elevated right atrial pressure (RAP) after cardiopulmonary bypass, including the atrial septal fenestration (4-6mm). If the ASD is directly repaired and performed tricuspid valvuloplasty, we can try to stop the cardiopulmonary bypass to evaluate the right heart function before deciding whether to perform the atrial septal fenestration [11]. Abdelkarim et al [12], partial ASD occlusion in patients with diastolic dysfunction or RV dysfunction and/or PHT is safe and may be better tolerated than complete ASD closure in selected patients. In order to further rule out anatomical abnormalities and observe the morphology of the right ventricular myocardium, ASD repair was performed for her. Unfortunately, the patient's relatives refused myocardial biopsy. After the operation, it was found that the right ventricular systolic function was poor, but it could be separated from CPB. There is no atrial septal fenestration performed.

Cardiac MRI [13], which is considered the noninvasive gold standard for assessing cardiac function, hasn't undergone preoperative. TTE is an essential non-invasive diagnostic tool for CHD, but recent advances in TEE technology have led to its increasing importance in the care of both children and adults with CHD. This case specifically highlights that even though TTE is excellent at detecting CHD, scrutiny during the pre-CPB examination by TEE is necessary to confirm the preoperative TTE findings and identify possible additional pathologic conditions [14]. Additionally, TEE has been shown to provide additional diagnostic information over TTE for specific structural cardiac anomalies and in the perioperative setting, supply the opportunity for confirmation of preoperative diagnosis, and influence the surgical plan if new or different pathology is identified [15].

4. Conclusion

The occurrence of an R-L shunt at the atrial level does not necessarily indicate severe pulmonary hypertension and anatomical structural abnormalities. The R-L shunt may manifest in patients with ASD maybe with potential right ventricular dysfunction. But according to the patient's prognosis, atrial septal occlusion may be the better surgical strategy.

Conflict of Interests

All the authors do not have any possible conflicts of interest.

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