ANESTHETIC MANAGEMENT IN ATRIAL SEPTAL DEFECT WITH SMALL LEFT VENTRICLE AND PULMONARY HYPERTENSION

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eISSN: 2828-4070
Proceedings ICE on IMERI. 2024.
Received: November 28th, 2024
Accepted: December 23rd, 2024
Published online: January 17th , 2025

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Abstract

Atrial septal defect (ASD) can remain undiagnosed throughout adulthood and frequently develop complications. We report major challenges caused by late onset ASD including small left ventricle and pulmonary hypertension. We report the successful management of ASD closure in a 24-year-old man presented with a large secundum ASD with a diameter of 57 mm and bidirectional shunt, and a small left ventricle (LV). Moderate mitral regurgitation (MR) and tricuspid regurgitation (TR) were also found. Right heart catheterization showed high flow, low resistance pulmonary hypertension (PH). Despite surgery went well with short period of cardiopulmonary bypass (CPB) time. We found decreased function in both ventricles when weaning from cardiopulmonary bypass machine, which required epinephrine 0.2-0.4 mcg/kg/min, milrinone 0.375 mcg/kg/min, norepinephrine 0.05 mcg/kg/min, and dobutamine 5 mcg/kg/min to stabilize the hemodynamics. Post-operative transesophageal echocardiography (TEE) showed a left to right shunt small ASD with a diameter of 2 mm, trivial TR, moderate MR, no LV D-Shape, and small LV. The ejection fraction was 59%, with a high level of hemodynamic support, cardiac output of 3.6 l/m, and stroke volume of 41 mL. The patient was able to be weaned off hemodynamics supports and ventilator within five days. The patient was discharged 12 days after surgery. The management of ASD can be challenging in the presence of small LV, pulmonary hypertension, and arrhythmia.

Keywords

Atrial septal defect, small left ventricle, pulmonary hypertension

Introduction

Atrial septal defect (ASD) is an anatomical structural abnormality that causes the shunting of blood between the atrial chambers. It is the second most common type of congenital heart defect, affecting five to six newborns out of every 1000 live births, and accounts for approximately 5–10% of all congenital heart defects.[1,2] Clinically silent ASD can remain undiagnosed until adulthood.[3]

Despite the higher volume strain on the right ventricle (RV), isolated ASDs are typically asymptomatic during infancy and childhood. Heart failure commonly occurs during the second or third decade of life because of persistent RV volume overload. Pulmonary hypertension (PH) occurs in up to 13% of unoperated patients under the age of ten; nevertheless, progression to Eisenmenger syndrome is seldom. The risk of arrhythmia rises with increasing shunt volume and atrial dilation.[2]

We report successful management of patient with large ASD with pulmonary hypertension and small left ventricle.

Case Report

A 24-year-old man presented with dyspnea and exercise intolerance. Physical examination revealed pansystolic murmur and electrocardiogram showed atrial fibrillations (AF) and pansystolic murmur. Echocardiography showed a large secundum ASD with a diameter of 57 mm, bidirectional shunt, and small left ventricle (LV) with a left ventricle internal diameter diastolic of 42.2 mm (Figure 1 and 2). Moderate mitral regurgitation (MR) with an eccentric jet and severe tricuspid regurgitation (TR) with a pressure gradient of 70 mmHg were also detected. Aside from dilated right atrial and ventricular chambers, ventricles were well-contracting, with an EF of 64% and TAPSE of 2.8 cm. Right heart catheterization showed high flow, low resistance pulmonary hypertension (PH): pulmonary artery pressure (PAP) was 48/14(26) mmHg, aortic pressure (AoP) was 65/37(44) mmHg, flow ratio (FR) 3.9, and pulmonary vascular resistance (PVR) 0.95 WU with an index of 1.5 WU/m^2. The patient was scheduled for elective ASD closure surgery under general anesthesia, with invasive monitoring of central venous pressure (CVP), arterial blood pressure, and transesophageal echocardiography (TEE).

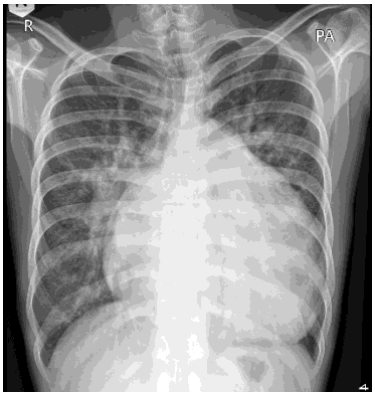

Figure 1. Chest X-Ray Before Surgery

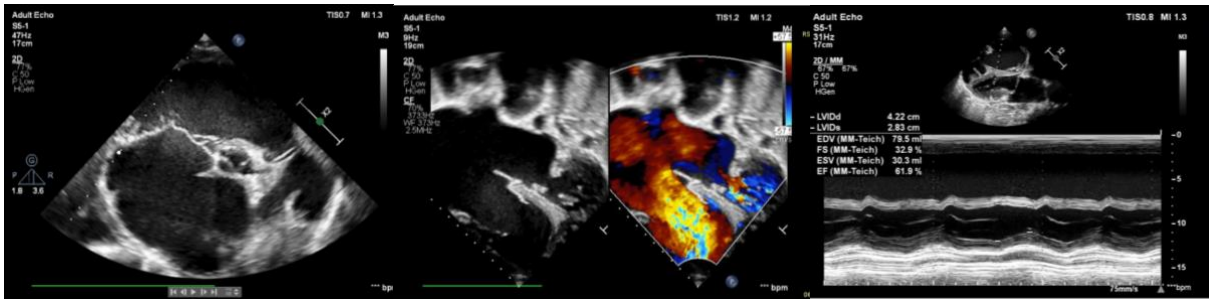

Figure 2. Preoperative Transthoracic Echocardiography

Pre-incision hemodynamics were stable with a CVP of 22 mmHg. The presence of large ASD with small LV, giant RA, and dilated RV was confirmed with pre-operative TEE, with the diameter of the pulmonary artery twice the size of the aorta (Figure 3). Severe TR with dilated tricuspid annulus and moderate MR were also seen. Surgical closure was performed with small ASD remains superiorly 3 mm and tricuspid valve repair with De-Vega annuloplasty. The cardiopulmonary bypass time was 90 minutes, while the cross-clamp time was 46 minutes. We found decreased function in both ventricles when weaning from cardiopulmonary bypass, which required epinephrine 0.2-0.4 mcg/kg/min, milrinone 0.375 mcg/kg/min, norepinephrine 0.05 mcg/kg/min, and dobutamine 5 mcg/kg/min to stabilize the hemodynamics. Post-operative TEE showed an L-R shunt small ASD with a diameter of 2 mm, trivial TR, moderate MR, no LV D-Shape, and relatively small LV with left ventricle internal diameter diastolic was 48.3 mm (Figure 4). The ejection fraction was 59%, with a high level of hemodynamic support, cardiac output of 3.6 l/m, and stroke volume of 41 mL. The patient was able to be weaned off hemodynamics supports and ventilator within five days. The patient was discharged 12 days after surgery.

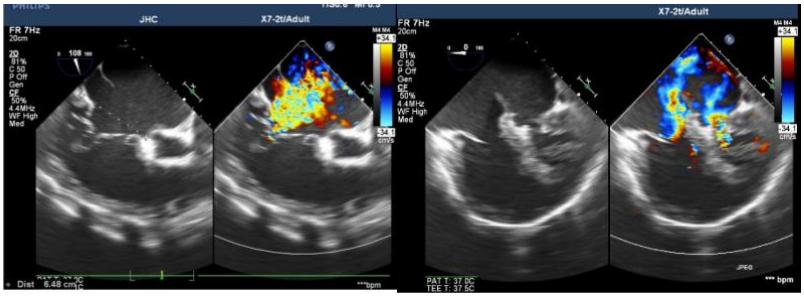

Figure 3. Pre-incision Transesophageal Echocardiography

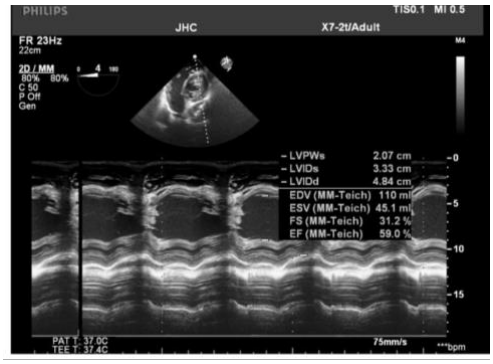

Figure 4. Transesophageal Echocardiography after weaned from Cardiopulmonary Bypass Machine showing Ejection Fraction and Left Ventricle Internal Dimention

Discussion

Large ASD generates a left-to-right shunting of the blood, resulting in RV volume overload and then PH, which leads to AF, heart failure, and Eisenmenger syndrome if there is shunt reversal.[4]

The shunt volume is determined by the compliance of both ventricles, the extent of the defect, and the pressure of the left and right atrium.[2,3] The greater compliance of the RV compared to the left is responsible for the resulting L–R shunt. Defects larger than 10 mm may result in a significant shunt, leading to RV volume overload and pulmonary overcirculation.[3]

The increased afterload of RV resulting from the increased PVR leads to RV distention and, ultimately, dysfunction. The right-sided ventricular failure is the most closely associated, prevalent, and significant contributor to morbidity and mortality in PH. As the RV expands, a paradoxical 'squashing' of the LV, in addition to a lower preload due to the high PVR, results in lower cardiac output, aggravating the morbidity and mortality of these patients. Measures to sustain RV function by promptly increasing pulmonary vasodilators are critical to a patient's outcome.[5]

Pulmonary vasodilation improves RV function by reducing the afterload. The core keys of RV support are as follows: reducing afterload, optimizing volume, increasing contractility, and maintaining LV contractility and pressure. Additionally, this approach necessitates proper LV function. The most suitable agent or combination for inotropic support of RV contractility is not known, although in practice, the phosphodiesterase type 3 inhibitors are preferred. Milrinone is preferred over β1 agonists as it improves ventricular contractility and RV output in adult patients with PH. However, it also decreases PVR and SVR, which may result in systemic vasodilation and necessitate the use of vasopressors. Vasopressors raise SVR and systemic blood pressure, which improves RV output by increasing the LV pressure. Improvement of systemic pressure can also improve coronary perfusion. Norepinephrine increases LV output, systemic pressure, and pulmonary blood flow while decreasing the pulmonary-to-systemic pressure ratio.[6]

Another finding in our case was the relatively small size of LV. This results from altered LV geometry, known as the "reverse Bernheim's effect", in which the septum expands into and interferes with the LV chamber. The suggested pathophysiology states that for larger shunts, right volume overload leads to the expansion of the right-sided chamber. Echocardiography shows a diastolic flattening of the interventricular septum, with an end-diastolic D-shaped LV and a circular shape in systole. These findings imply that while the atrioventricular valves are open, pressures are equal in both the atrium and ventricles, whereas when the atrioventricular valves are closed, the septal morphology reflects the ratio of left and right ventricular pressures. As a result, interventricular interdependence is impaired, and the LV ends up underfilled due to the dilated RV, causing decreased cardiac output.[7]

Nonetheless, various factors influence the LV diastolic pressure-volume relationship, including innate diastolic rigidity, extrinsic constraint effects, relaxation, and continuous filling characteristics. Following interatrial shunt suppression, external constraints and ongoing filling characteristics are lowered or normalized. Following interatrial shunt suppression, external constraints and ongoing filling characteristics are lowered or normalized. As a result, only two main causes, left ventricular diastolic stiffness and aberrant relaxation, may be responsible for immediate and, in most cases, transitory heart failure following shunt closure.

Aging has been associated with LV stiffness, which increases the possibility of heart failure when the defect is closed. Lower LV performance has been reported in cases of RV volume overload due to reduced preload, following reduced left ventricular end-diastolic volume, which, as a result, reduces LV stroke volume, according to Starling's law.[7]

According to the recommendations by the American Society of Echocardiography, a small chamber size is defined as an end-diastolic diameter of less than 42 mm for men and 37.8 for women. A large retrospective cohort study by Leibowitz et al. in 2024 discovered that the presence of a small LV has a statistically significant association with mortality.[8]

A long-standing significant shunt leads to chronic volume overload in the right-sided chamber, which leads to myocardial injury. Right heart enlargement increases the myocardial oxygen demand then hypoperfusion of the myocardium. The increase of myocardial remodeling markers may be related to increased myocardial stiffness and, eventually, diastolic heart failure.[7]

In this case, we discovered a long-standing complication of a large ASD: PH, RV failure, and a relatively small and stiff LV, resulting in a poor cardiac output state. Any variables that could cause shunt reversal or increased shunt fraction were carefully prevented. In our case, PH was managed by continuously monitoring the ventilator and avoiding conditions that could exacerbate the condition, such as acidosis, hypercarbia, and atelectasis. We maintained adequate preload and cardiac contractility, near normal heart rate, SVR, and PVR through appropriate monitoring, including TEE.

Competing Interests

There is no conflict of interest to declare.

Acknowledgments

This article was presented at the 9th International Conference and Exhibition on Indonesian Medical Education and Research Institute (9th ICE on IMERI) 2024, Faculty of Medicine, Universitas Indonesia. We are grateful for the outstanding assistance provided by the committee of the 9th ICE on IMERI 2024 throughout the preparation of the manuscript and the peer-review process.

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