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Case Report

Surgical Repair of Atrial Septal Defect with Severe Pulmonary Hypertension: A Case Report

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Abstract:

This case report details the successful surgical repair of an atrial septal defect (ASD) in a 28-year-old female with severe pulmonary arterial hypertension (PAH), who presented with dyspnea, fatigue, and palpitations. The patient had a large secundum ASD (26 mm size) with bi-directional shunt and severe PAH, with a Pulmonary Arterial Systolic Pressure (PASP) of 107 mmHg and Mean Pulmonary Arterial Pressure (mPAP) of 57 mmHg, as revealed by echocardiography. The patient was optimized medically with diuretics, oxygen, and pulmonary vasodilators (Sildenafil & Ambrisentan), and was deemed eligible for surgical closure of the ASD. A pericardial patch was used to close the defect, and the patient's postoperative recovery was uneventful. Follow-up echocardiography showed a significant reduction in mPAP and right ventricular size. Although surgical closure of an ASD with severe PAH is a high-risk procedure, it can be safely performed in carefully selected patients with appropriate preoperative evaluation, medical optimization, and careful surgical technique. Timely closure of the defect can result in a considerable improvement in the patient's hemodynamic status and quality of life.

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Introduction:

An atrial septal defect (ASD) refers to a congenital heart defect characterized by an absence of continuity in the atrial septal wall. If left untreated or not closed promptly, it can lead to the development of severe pulmonary hypertension (PH). Surgical repair is the preferred treatment option for patients with ASD and severe PH, but it is associated with increased morbidity and mortality risks.

Case Report:

A female patient, aged 28, with a medical history of recurring respiratory infections and exercise intolerance, presented with symptoms of progressive dyspnea, fatigue, and palpitation. A diagnostic echocardiography conducted on January 24, 2021, revealed the presence of a large secundum ASD measuring 26 mm, with bidirectional shunt, and severe PAH with a PASP of 107 mmHg and a mPAP of 57 mmHg. Due to the high risk associated with surgical repair, the patient was treated with sildenafil and ambrisentan for pulmonary vasodilation. One year later, a cardiac catheterization report on February 20, 2022, indicated the presence of a secundum ASD and supra systemic PA pressure with PASP of 120, Pulmonary Arterial Diastolic Pressure (PADP) of 30, and mPAP of 60 mmHg, Pulmonary Vascular Resistance (PVR) of 10.84 Wood unit, and Qp: Qs ratio of 1.50:1. After administering oxygen for 10 minutes, PA pressure reduced to 110/20/50 mmHg, post-oxygen PVR was 4.95 Wood unit, and Qp: Qs ratio was 2.49:1. Although the patient was advised to undergo urgent surgery, she declined and continued with medical management. One year later, another echocardiography conducted on January 9, 2023, revealed the presence of a

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large secundum ASD with bidirectional shunt and severe PH with PASP of 69 mmHg and PADP of 42 mmHg and mPAP of 51mmHg. After being admitted to Chittagong Medical College Hospital (CMCH), the patient was treated with diuretics, oxygen, and pulmonary vasodilators to optimize her condition before undergoing surgery.

Surgical procedure: The patient underwent surgical correction on February 16, 2023. Initially, a median sternotomy was performed, followed by thymectomy and pericardiotomy. A hugely dilated pulmonary artery was observed. Direct measurement of the right ventricular and pulmonary artery pressures was performed. The following pressures were found: 80/25/44 mmHg in the RV body, 80/29/41mmHg in the RVOT, and 81/50/60 mmHg in the MPA. Cardiopulmonary bypass was established using bi-caval cannulation, and cardiac arrest was achieved with hypothermic antegrade blood cardioplegia after aortic cross clamping. Right atrium was opened with an incision parallel to and one cm away from the Atrioventricular groove. Intracardiac inspection revealed a large defect in the interatrial septum. Mitral valve was tested and found normal. All four pulmonary veins were seen draining normally in left atrium. The ASD was closed using a fresh autologous pericardial patch. The right atrium was closed. The cross clamp was released and cardiopulmonary bypass was discontinued. The cross clamp time was 30 minutes, and extracorporeal circulation time was 64 minutes. Hemostasis was ensured and two mediastinal drain tubes were placed - one retrosternal and the other in the retrocardiac position - before closing the chest wound in layers. During the procedure, the patient received two units of FFP and one unit of whole human blood. After eight hours in the ICU, the patient was extubated and started on a medication regimen of Tab. Ambrisentan (5 mg) in the morning and Tab. Sildenafil (50 mg) twice daily. The patient had an uneventful stay in the ICU, and her SPO2 remained at 100%. She was discharged on the eighth postoperative day with instructions for regular follow-up. After one month, the patient's overall condition was stable, and plans were made to gradually withdraw her from pulmonary vasodilators following the surgery.

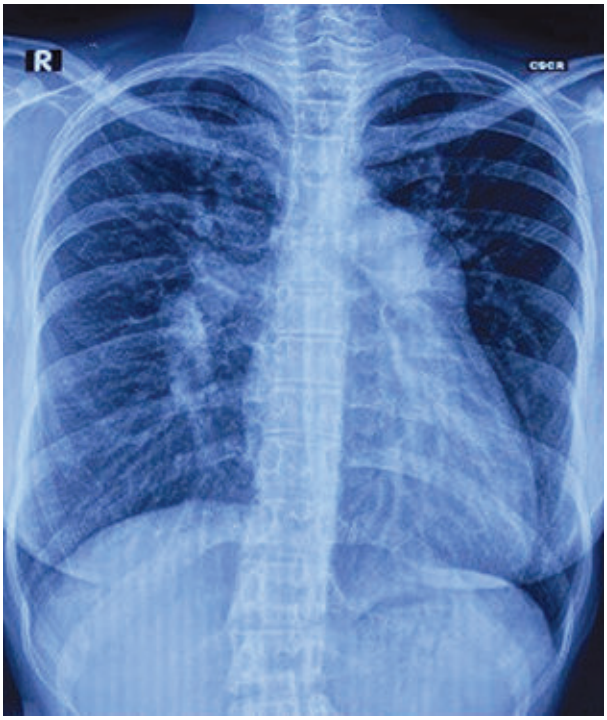

Figure 1: *Pre opt x-ray chest P/A view*

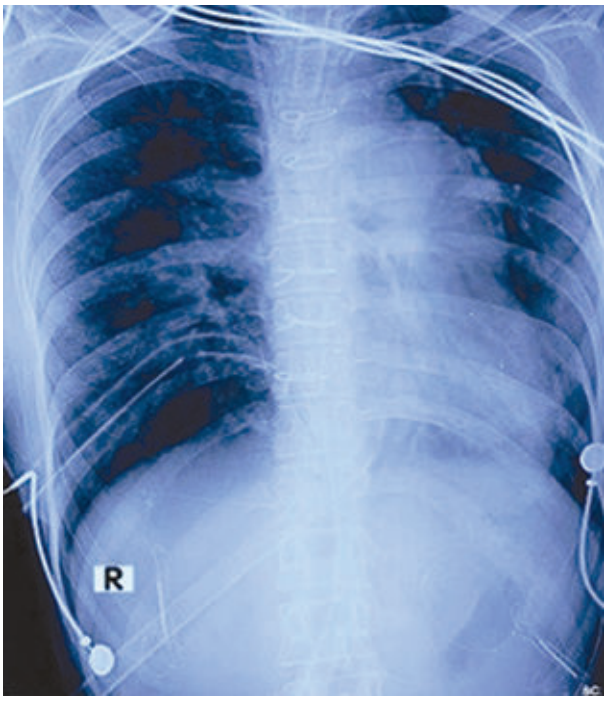

Figure 2: *Post opt X-ray chest A/P view*

Discussion:

ASD patients who have an uncorrected left-to-right shunt are vulnerable to developing pulmonary arterial hypertension (PAH). This condition arises due to prolonged exposure to

increased blood flow and pressure, leading to vascular remodeling and dysfunction that causes a progressive elevation of pulmonary vascular resistance (PVR) and mean pulmonary arterial pressure (mPAP).[1] As per the 2022 ESC/ERS Guidelines on the diagnosis and treatment of pulmonary hypertension, PH is now defined by a mean pulmonary arterial pressure >20 mm of Hg at rest, whereas the diagnosis of PAH requires a PVR >2 Wood Units and pulmonary arterial wedge pressure d"15 mm Hg. The severity of PAH is classified as mild, moderate, or severe based on the mPAP, which ranges from 20 to 40 mm Hg, 41 to 55 mm Hg, and >55 mm Hg, respectively.[2]

A pulmonary vasodilator test is typically conducted in patients with shunt-related PAH to evaluate the potential reversibility of the condition. If there is a reduction of mPAP or PVR that is less than 20% after the test, it indicates that the PAH is irreversible.[3] Patients who have severe or irreversible PAH are deemed unsuitable for defect closure since this procedure can lead to a decline in cardiac output, a rise in right-sided heart failure, and mortality in such cases.[4]

There have been several reported cases that support the use of a treatment and repair approach for patients with ASD and severe pulmonary hypertension. A study by Jung et al. treated ASD patients with severe PAH with oral bosentan therapy for one year, resulting in clinical improvement after shunt closure following long-term vasodilator therapy.[5] In a case described by Young Hwue Kim and colleagues, a 41-year-old woman with large ASD and severe pulmonary hypertension was managed conservatively with tab. Sildenafil 50 mg BD as corrective repair was deemed impossible. After two years of treatment, the patient was re-examined by cardiac catheterization, revealing a reduction in PVRI from 25 to 12.63 WU/m2 and pulmonary/systemic vascular resistance ratio to 0.43. The patient underwent surgical repair and showed NYHA class I symptoms at 6 months post-surgery. At 1 year 8 months post-repair, the dose of Sildenafil was tapered to 50 mg daily, and all medications, including Sildenafil, were discontinued at 3 years post-repair. Four years after corrective repair, the patient had no symptoms, and the 6-minute walk test result was 550m.[6] Lastly, Hoetzenecker and colleagues reported on a 71-year-old woman with a PVR of 5.8 WU, reduced to 3.0 WU with NO, who showed a decrease in pulmonary pressure after treatment with Bosentan, following which the defect was surgically repaired.[7]

Sildenafil is a potent and selective inhibitor of phosphodiesterase type 5, which is responsible for degrading cyclic guanosine monophosphate (cGMP) in the pulmonary artery, resulting in relaxation of the vessel's smooth muscles. Several randomized controlled trials have established the safety and effectiveness of sildenafil in improving mPAP, PVR, the cardiac index, and exercise tolerance in patients with PAH.[8]

Ambrisentan is a medication that belongs to the class of endothelin receptor antagonists. It works by blocking the action of endogenous endothelin peptide, which would otherwise cause constriction of the smooth muscles in blood vessels, leading to increased blood pressure. By inhibiting this process, ambrisentan promotes relaxation of the blood vessels, allowing for a reduction in blood pressure.[7]

The operability of patients with ASD and pulmonary arterial hypertension is determined based on various factors, including signs and symptoms such as dyspnea, cyanosis, SP02, palpable P2, and loud P2, X-ray findings such as prominent MPA and reduced pulmonary vascularity, echocardiogram findings such as severe TR with high peak velocity, and cardiac catheterization report which may indicate high PVRI with reduced oxygen responsiveness and a borderline PVRI and SVRI ratio. If these criteria suggest a low likelihood of benefit from permanent closure, corrective repair may not be recommended.[9] However, in the case being discussed, treatment with sildenafil and ambrisantan was effective in reducing mPAP and reversing PAH, even though the patient's baseline hemodynamic findings were borderline. After surgery, the patient experienced a significant improvement in symptoms and exercise tolerance, with no residual leakage observed on follow-up Trans thoracic Echocardiography. The right atrium and right ventricle returned to almost normal function and diameter as well.

Conclusion:

The use of long-term combined therapy with sildenafil and ambrisentan may provide a new option for closing ASD with severe PAH that were previously considered unsuitable for surgical repair. This finding also suggests that such therapy may be beneficial in managing patients with shunt-related PAH.

Conflict of Interest - None.

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