A patient with large ostium primum atrial septal defect who underwent open heart surgical closure with pericardial patch: a case report

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ABSTRACT

Introduction: Atrial septal defect (ASD) is one of the most common congenital heart diseases and often diagnosed in adulthood. ASD represents a spectrum of disease with symptoms that vary from asymptomatic to right-sided cardiac volume overload, pulmonary arterial hypertension and atrial arrhythmias depending on the size of the defect, size of the shunt and associated anomalies. We reported a patient with large ostium primum ASD who underwent surgical closure with a pericardial patch.

Case Description: A 18 years old man came with chief complaint of abnormal ECG results of the screening examination of police candidates from Bhayangkara Hospital Manado. The patient had no complaints during admission. The patient could still carry out daily activities without any obstacles. The patient also could do high intensity sport without complaints of shortness of breath, heavy chest, or headaches. Electrocardiogram (ECG) examination showed sinus rhythm, heart rate 74 beats per minute, left anterior fascicular block (LAFB), complete right bundle branch block (RBBB) and fragmented QRS at inferior leads. Transthoracic and transesophageal echocardiography revealed large ostium primum ASD L-R shunt. Open heart ASD surgical closure with pericardial patch was done with good results and no residual shunt.

Conclusion: Large ostium primum ASD are associated with significant systemic-to-pulmonary shunts that results in desaturation requiring surgical closure. Large shunt of ASD patients who have a dilated right heart are associated with increased age-related morbidity and mortality. ASD with right heart dilatation should be considered for closure once the diagnosis is established regardless of the patient’s age.

Keywords: atrial septal defect, ostium primum atrial septal defect, atrial septal defect surgical closure, pericardial patch.


INTRODUCTION

Atrial septal defect (ASD) is a prevalent congenital heart condition detected in adulthood. ASD is characterized by an abnormality in the interatrial septum, enabling direct flow of pulmonary veins from the left atrium into the right atrium. ASD encompasses a range of diseases with varying symptoms, ranging from no apparent signs to right-sided cardiac volume overload, pulmonary arterial hypertension, and even atrial arrhythmias. These manifestations depend on the size of the defect, shunt size, and associated anomalies.

There are four primary types of ASD: ostium secundum, ostium primum, coronary sinus, and sinus venosus. Ostium primum ASD arises from incomplete fusion of the septum primum with the endocardial cushion. It constitutes the second most frequent type of ASD, accounting for 15-20% of all cases. Ostium primum ASD is located adjacent to the atrioventricular valves, which undergo deformity and incompetence. Typically, only the anterior or septal leaflets of the mitral valve are displaced.

ASD is more prevalent in women, with a ratio of approximately 2:1. While ASD patients may remain asymptomatic during infancy and childhood, clinical symptoms become evident as they age. By the age of 40, untreated ASD patients will experience symptoms such as exertion-induced shortness of breath, fatigue, palpitations, arrhythmias, and potentially heart failure. In this instance, we present a case of a patient with a large ostium primum ASD who underwent surgical closure utilizing a pericardial patch. Our discussion will focus on the diagnosis and management of patients with ostium primum ASD.

CASE DESCRIPTION

An 18 years old man, lives in Manado, height 166 cm and weight 58 kg, with an ideal body mass index, work as a college student, came to the Cardiac Clinic due to abnormal ECG results of the screening examination of police candidates from one hospital. Patient was advised to have transthoracic echocardiography with suspicion of ASD. Currently the patient has no complaints. The patient could still carry out daily activities without any obstacles. The patient could do high intensity sports such football, badminton or running without complaints of shortness of breath, heavy chest, or headaches. Shortness of breath, paroxysmal nocturnal dyspnea,
orthopnea, chest pain, palpitations, fainting, swollen legs were denied. The patient sleeps only with one pillow. A history of hypertension, diabetes mellitus, dyslipidemia, and asthma were refuted. The patient does not smoke and does not drink alcohol. Currently the patient has no medication. Any past illness or other illness were denied.

On physical examination, he was found mildly ill, compost mentis, with vital signs blood pressure 112/62 mmHg, heart rate 75 times per minute, respiratory rate 20 times per minute, afebrile and oxygen saturation was 97% without supplemental oxygen. Head examinations show anemic conjunctiva and anicteric sclera. Neck examination show normal jugular venous pressure at seven cmH2O with no regional lymph node and thyroid gland enlargement. Chest examination showed symmetrical form and movement of the chest, tactile fremitus was equal for both lungs. Vesicular breath sounds in both pulmonary fields was found with no rales or wheezing. The ictus cordis was palpable at the fifth intercostal space, left midclavicular line. Heart auscultations show normal, regular first and second heart sound. A systolic ejection murmur grade 3/6 was heard at upper left sternal border with wide fixed splitting S2 and diastolic murmur grade 2/4 was heard at lower left sternal border. Other physical examinations were within normal limits. Laboratory examination was unremarkable except the hemoglobin was 17.2 gram/dL. Chest X-ray examination showed no significant abnormalities. Electrocardiogram (ECG) examination showed sinus rhythm, heart rate 74 beats per minute, left anterior fascicular block (LAFB), complete right bundle branch block (RBBB) and fragmented QRS at inferior leads (crochetage sign) (Figure 1).

Transthoracic echocardiography revealed mildly dilated right atrium (RA) and right ventricle (RV), D-shaped LV, no left ventricular hypertrophy (LVH), mitral and tricuspid valves at one level, segmental analysis of left ventricle global normokinetic, normal left ventricle (LV) diastolic function, aorta 3 cusps with no calcification, trivial mitral regurgitation (MR) with no mitral cleft, mild tricuspid regurgitation (TR) with tricuspid valve gradient (TVG) 23 mmHg, low probability of pulmonary hypertension (PH), interatrial septal defect size 1.7-2.0 cm with left to right (L-R) shunt, normal right ventricle (RV) contractility with tricuspid annular plane septal excurs (TAPSE) 2.8 cm, inferior vena cava (IVC) 2.4 cm, collapsibility > 50% with an estimated right atrial pressure (RAP) of 3 mm Hg (Figure 2).

Patients are advised to do a transesophageal echocardiography. A transesophageal echocardiography showed ostium primum ASD with large
defect 1.6 to 2.4 cm L-R shunt (Figure 3). He was diagnosed with large ostium primum ASD L-R shunt and was advised to underwent open heart ASD surgical closure.

Open heart surgical ASD closure was performed on the third with procedure as follows: median sternotomy incision was done. After the pericardium is opened, the heart appeared large with good contractility and aortic size is the same as the pulmonary artery size. Pericardium is then preserved. Right atrium was opened, ostium primum ASD was found with a diameter of 40 mm with mitral cleft. ASD was closed with a pericardial patch. Intraoperative transesophageal echocardiography evaluation found no residual ASD shunt. Installed subternal drain, intrapericardial drain and pigtail. Pericardium is partially closed. The sternum is closed with a sternal wire. Operation complete. The patient was given 1 bag of packed red cells (PRC) and 2 bags of fresh frozen plasma (FFP) during the operation. After hemodynamics were stable, the patient was then transferred to a post-operative intensive care unit.

The patient is then gradually weaned from the ventilator from volume control-continuous mandatory ventilation (VC-CMV) mode to volume control-synchronous intermittent mandatory ventilation (VC-SIMV) mode then to continuous positive airway pressure (CPAP) mode gradually while monitoring the patient’s response every 30 minutes and extubation was done after the patient’s breathing response is good and oxygenation is adequate. Vital sign showed non invasive blood pressure (NIBP) 70/39 mmHg, arterial line blood pressure 71/42 mmHg, pulse rate 95 beats per minute, respiratory rate 24 times per minute, oxygen saturation 98% with additional simple oxygen mask 8 L per minute and central venous pressure (CVP)  4 cmH2O. Loading crystalloid 500 cc and colloid 200 cc was given. NIBP 99/44 mmHg, arterial line BP 112/48 mmHg. Physical examinations were within normal limits, no murmurs were found. Laboratory examination was carried out with significant examination results found hemoglobin 9.7 g/dL, leukocyte 24.400/µL, albumin 2.84 g/dL, and potassium 6.0 mEq/L. The patient was then diagnosed with post open heart ASD closure et causa large ostium primum ASD, hyperkalemia, hypoalbuminemia, anemia, thrombocytopenia, and leukocytosis. The patient was then given additional therapy such as Cefazolin 1 gram t.i.d intravenously, Ca gluconas 1 gram intravenously extra, and Calcium polystyrene sulfonate 1 sachet t.i.d per oral. One unit of packed red cell (PRC) transfusion and 20% Human albumin 1 flash.

The patient was stabilized and medication was continued until the tenth day of treatment. a 6-minute walking test (6MWT) was carried out with a 2 minute walking distance was 84 meters (METs 2.59) and 6MWT was 275 meters (METs 5.86). Phase II cardiac rehabilitation starts with leisure walk with a target of 2x450 meters and is gradually increased by adding 100 to 200 meters per session until it reaches 1x1.800 meters, then continued with a treadmill with an initial speed of 2.4 km per hour. The patient was then sent home with Cefixime 200 mg per 12 hours per oral, Lansoprazole 30 mg per 12 hours per oral, Furosemide 40 mg per 24 hours per oral, Ramipril 5 mg per 24 hours per oral, Bisoprolol 1.25 mg per 24 hours orally and Ibuprofen 400 mg every 8 hours orally. Patients are advised to routinely control the Cardiac Polyclinic and take medications, avoid strenuous activity and gradual physical exercise according to the results of the 6MWT examination.

DISCUSSION

ASD is a prevalent congenital heart abnormality commonly observed in adults. ASD can be associated with additional abnormalities such as anomalous pulmonary venous return (TAPVR) rely on ASD as an alternative blood supply until definitive treatment is possible.

Patients with conditions like tricuspid atresia, transposition of the great vessels, hypoplastic left heart syndrome, pulmonary atresia, or total anomalous pulmonary venous return (TAPVR) rely on ASD as an alternative blood supply until definitive treatment is possible. In certain cases of congenital heart disease, the interaction between the left and right cardiac circulations plays a critical role in patient survival.

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defects, making treatment decisions more complex. 8  

The process of atrial septation begins during the fourth week of pregnancy. It involves the growth of the septum primum from the roof of the primitive atrium towards the endocardial cushion. Mesenchymal cells derived from the embryonic endocardium or mesenchymal cap cover the caudal portion of the septum primum. As the septum primum continues to expand towards the atrioventricular endocardial cushion, it eventually closes the gap between the mesenchymal cap and the atrioventricular cushion, forming the ostium primum. This closure attaches the septum primum anteriorly to the atrioventricular cushion, dividing the primitive atrium into the right and left atria. The septum primum also attached dorsally to the dorsal aspect of the mesenchymal protrusion. Simultaneously, cell death occurs on the dorsal aspect of the septum primum, leading to the formation of the ostium secundum. The septum secundum develops from the roof of the atrium to the right of the septum primum. As the septum secundum progresses caudally and partially covers the ostium secundum, it creates a gap between the septum primum and septum secundum known as the foramen ovale. In the fetal stage, the foramen ovale allows oxygenated blood to bypass the lungs by directing blood from the right atrium to the left atrium. However, after birth, changes in pulmonary vascular resistance and decreased left atrial pressure prompt the septum primum to close the foramen ovale. 9  

Persistent volume overload caused by increased blood flow to the lungs leads to remodeling of the pulmonary vessels. This remodeling results in an increase in the smooth muscle layer of the pulmonary vessel walls, leading to increased resistance in the pulmonary circuit and subsequent elevation in pulmonary pressure, causing pulmonary hypertension. When the pulmonary pressure equals the systemic pressure, the shunt through the ASD is reversed, allowing deoxygenated blood to flow from the right atrium into the left atrium and then into the systemic circulation. This condition is known as Eisenmenger syndrome. 10  

ASD patients often remain asymptomatic until adulthood. Smaller ASD defects, less than 5 mm in size, may not cause symptoms, while ASD defects between 5 and 10 mm can lead to symptoms in the fourth or fifth decade of life. Larger ASD defects may cause symptoms earlier, in the third decade of life. Common symptoms experienced by ASD patients include exertion-induced shortness of breath, fatigue, reduced exercise tolerance, palpitations, supraventricular tachyarrhythmias, and, less frequently, lung infections and signs of right heart failure. 8 Approximately 20% of adult patients have atrial dysrhythmias before undergoing surgery. The presence of a stroke or transient ischemic attack (TIA), especially when accompanied by the discovery of a peripheral blood clot, raises suspicion of ASD. 11 Pulmonary regurgitation often goes unnoticed during auscultation and routine echocardiographic examination due to its low pressure gradient.

ASD is commonly observed as an isolated defect, but it can also be associated with various inherited genetic disorders, aneuploidy, transcription errors, mutations, and maternal exposures. 5,6 Nearly one-third of individuals with ASD exhibit additional malformations, including pulmonary stenosis, ventricular septal defect (VSD), mitral valve prolapse, subaortic stenosis, aortic coarctation, and anomalous pulmonary venous drainage. 12 The presence of ASD is frequently found in individuals with several syndromes, e.g., down syndrome, Treacher-Collins syndrome, thrombocytopenia-absent radii syndrome, Turner syndrome, and Noonan syndrome, which are inherited through Mendelian patterns. 12 Maternal exposure to factors like rubella infection, cocaine, and alcohol can also increase the risk of ASD in the fetus. Furthermore, ASD is associated with familial genetic disorders and conduction defects. 13

In recent times, the life expectancy and survival rates of individuals with ASD have significantly improved compared to a few decades ago. As patients age and pulmonary artery pressure increases, the prevalence of tachyarrhythmias such as atrial flutter and atrial fibrillation also rises. Additionally, there is a potential risk of systemic embolism resulting from paradoxical emboli, atrial fibrillation, and atrial flutter. 8

The electrocardiogram (ECG) plays a crucial role in diagnosing ASD. It can reveal various rhythms in ASD patients, including sinus rhythm, atrial fibrillation, or atrial flutter. Inverted P waves in the inferior leads indicate sinus node deficiency or absence and are commonly observed in venous sinus defects. Right atrial overload is frequently present. First-degree atrioventricular block may suggest a primum ASD, but it can also occur in elderly patients with secundum ASD. The QRS axis generally shifts to the right in secundum ASD, particularly when pulmonary hypertension is present, while ostium primum ASD causes a leftward deviation of the QRS axis. Right ventricular hypertrophy is often present in all types of ASD, typically manifesting as right bundle branch block, with severe cases seen in patients with pulmonary hypertension. The ECG typically exhibits incomplete right bundle branch block (RBBB), right axis deviation (RAD), or superior left axis deviation in partial atrioventricular septal defects. 8

Chest X-ray examination does not provide diagnostic value but can assist in monitoring the clinical condition by identifying cardiomegaly and enlargement of the pulmonary artery. Increased pulmonary vascularity on chest X-ray is frequently overlooked. 14 Cardiac exercise testing can be employed to assess the reversibility of shunt flow, response to activity, and to exclude desaturation in patients with pulmonary arterial hypertension. 11

Diagnostic imaging plays a crucial role in determining the size of the defect and guiding treatment decisions. Echocardiography serves as the primary diagnostic modality, providing information about the diagnosis, quantification of right ventricular volume (which can be an unexpected initial finding in previously undiagnosed ASD cases), and illustrating the hemodynamic relevance of the defect, particularly the shunt ratio. However, echocardiography may have limitations in cases with poor image quality due to scar tissue from previous surgeries, concurrent lung disease, or obesity. 11
Transthoracic echocardiography (TTE) is capable of detecting the size of the defect, blood flow direction, abnormalities involving the endocardial cushion and atrioventricular valves, as well as assessing the structure and function of the heart. It can also estimate pulmonary artery pressure and calculate the ratio of pulmonary to systemic flow (Qp:Qs). Transesophageal echocardiogram (TEE) proves to be a superior imaging modality for diagnosing less common heart defects. It is particularly essential for accurately assessing the size of ASD secundum before closure using a device. TEE examination allows for the evaluation of size, analysis of residual septal morphology, determination of rim size and quality, identification of additional defects, and confirmation of normal pulmonary venous connection. Three-dimensional echocardiography offers visualization of ASD morphology. Pulmonary artery pressure and tricuspid regurgitation are also valuable pieces of information obtained through TEE.

Additional imaging techniques that can be employed include cardiac computerized tomography (CT) scans and magnetic resonance imaging (MRI). Both CT and MRI scans provide detailed images of the heart and surrounding structures within the thoracic cavity. They offer supplementary anatomical descriptions, particularly in cases where major or pulmonary vascular anatomy is less well-defined. MRI is particularly useful in assessing right ventricular volume overload, identifying venous sinus ASDs, quantifying the pulmonary-to-systemic flow ratio (Qp:Qs), and evaluating pulmonary venous connections. Cardiac CT scan serves as an alternative for evaluating pulmonary venous connections.

Frequent late-onset arrhythmias that occur following surgical intervention in individuals under the age of 40 include intraatrial reentrant tachycardia (IART) or atrial flutter, which can be effectively treated with radiofrequency ablation or cryoablation. On the other hand, atrial fibrillation is a more common complication observed in patients aged 40 and above, regardless of intervention. Among individuals over 40 undergoing ASD closure, the prevalence of atrial arrhythmias can reach up to 40% to 60%. Closure of the ASD may lead to restricted access to the left atrium. While sudden cardiac death is rare, older patients with ASD often experience atrial flutter and atrial fibrillation, which can be expected. Prolonged dilation and stretching of the right atrium, aggravated by tricuspid regurgitation, pulmonary arterial hypertension, and heart failure, contribute to the occurrence of atrial tachyarrhythmias or bradyarrhythmias. Even after complete ASD closure, older patients remain at risk of systemic thromboembolism. The pulmonary veins are frequently implicated as sources of arrhythmogenesis and thrombus formation.

**CONCLUSION**

Grown up congenital heart disease is a rapidly growing population due to advances in the diagnosis and treatment of congenital heart disease. Most children with congenital heart disease nowadays are expected to survive into adulthood with or without the aid of surgical correction or palliative therapy. Currently it is estimated that there are approximately one million adult patients with congenital heart disease and this number will continue to increase as advances in diagnosis and therapy. ASD is the most common congenital heart defect and can vary clinically from asymptomatic to pulmonary hypertension, cyanosis and vascular complications such as stroke. Most small ASDs will close spontaneously in the first year of life, but large ASDs are associated with significant systemic-to-pulmonary shunts that result in desaturation and require percutaneous or surgical closure. The development of increasingly sophisticated diagnostic imaging modalities and earlier closure of ASDs have all increased long-term survival in these patients. A holistic approach to diagnosis and treatment is important in patients with congenital heart disease and an important first step is to collect historical data. Certain signs and symptoms should prompt evaluation of adults with congenital heart disease, especially if the patient has progressive symptoms such as syncope and breathlessness on exertion. ASD with right heart dilatation should be considered for closure once the diagnosis of ASD is established, regardless of age.

**AUTHOR CONTRIBUTION**

All authors listed above contributed equally in writing and preparing this case report.

**ETHICAL CONSIDERATION**

This case report has been approved by the Ethical Commission of Faculty of Medicine, Universitas Sam Ratulangi. The patient also has given informed consent to join this case report and is permitted to publish the data.

**CONFLICT OF INTEREST**

All authors declare no conflict of interest to publish this case report.

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**REFERENCES**


CASE REPORT


