Case report

Atrial Septal Defect Closure In A 70 Year Old Woman: Case Report And Literature Review


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Time was when it was thought that Atrial Septal Defects (ASDs) diagnosed in adulthood, especially in the elderly should only be managed conservatively. The trend is gradually changing and most “elderly ASDs” are getting closed, especially percutaneously if they are the ostium secundum type. The Ostium primum, sinus venosus and coronary sinus ASDs still need open-heart surgery. We present the case of a woman who was diagnosed with an ostium primum ASD at the age of 65 years and had repair at 70 years. This is the oldest case of ASD closure done in this centre. The post operative and follow-up periods have been uneventful.

Key words: Atrial septal defect, Ostium primum Atrial Septal Defect, Open-Heart surgery, percutaneous transcatheter repair.

Introduction

Atrial septal defect (ASD) is the commonest congenital heart disease found in the adult. The consensus is for the repair of the ASD in childhood. When the diagnosis is made in adulthood, especially in the elderly, controversy arises as to what should be done. We present the case report of ASD closure in a 70 year old woman, and review of the literature.

CASE REPORT

This woman was diagnosed 5 years earlier when she was 65 years old. She presented with easy fatigability and occasional palpitations for over a year. She was in New York Heart Association class I (NYHA I). The chest X-ray showed a cardiomegaly of cardiothoracic ratio of 68% (Fig1).

Fig. 1. CXR before the operation
The ECG showed a sinus rhythm, left axis deviation and left ventricular hypertrophy. The echocardiographic finding was a 1.6 cm ostium primum ASD with a cleft in the anterior mitral leaflet, causing a mild mitral incompetence. The main pulmonary artery (MPA) was dilated to over 1.5 times the size of the aorta: MPA 4.9cm, aorta 3.0cm. She was put on anti-failure medications and advised to have surgery. She defaulted due to lack of finances for the surgery. When she was ready for surgery 5 years later, she was still healthy (NYHA I).

There were no comorbidities. The echocardiography showed that the MPA was still getting dilated: MPA 5.4cm, aorta 3.0cm. The other parameters were essentially the same. Swan-Ganz catheterization was done which showed normal central venous, right ventricular and pulmonary artery pressures. There was an oxygen saturation step-up in the right atrium indicating the presence of a shunt, most likely from the ASD. She was scheduled for surgery. There were differences in opinion as to whether she still needed the surgery since she had been stable all this while, considering the risks of open-heart surgery. The points in favour of surgery were that she was healthy, there were no comorbidities, and the increasing dilatation of the main pulmonary artery will increase the flooding of the lungs, eventually leading to pulmonary hypertension and Eisenmenger’s syndrome. The surgery was done. This was through a median sternotomy, under cardiopulmonary bypass (CPD), with moderate hypothermia. The findings were: cardiomegaly, dilated main pulmonary artery, a 2cm ostium primum ASD and a cleft in the anterior mitral leaflet. The mitral valve was repaired and the ASD was closed with a Goretex patch. The post operative period was uneventful and she was discharged home on the 7th post operative day. Subsequent reviews up to 2 years were uneventful. On the repeat CXR the cardiomegaly had reduced to 65% (Fig2).

**Discussion**

An ASD is a defect in the atrial septum that consistently permits inter-atrial communication. It is a common congenital heart disease with an incidence of 6.4 per 10,000 births and a 2:1 female to male prevalence. It is the second commonest congenital heart disease (12%) next to VSD (30%). It may be an isolated lesion, be associated with other lesions or as part of a syndrome. Atrial septal defect is the commonest congenital cardiac lesion found in the adult. Historically the first successful intracardiac surgical repair of a congenital heart disease using cardiopulmonary bypass was the repair of an ASD in 1953 by John Gibbon.

ASDs are commonly classified according to their anatomic location in the atrial septum. There are 4 main types. The first is the ostium secundum ASD, located in the central part of the septum in the area of the fossa ovalis. It is considered the only true defect. It forms the majority (74% – 80%) of ASDs. It is a defect reduction in the size of the main pulmonary artery and was now actually smaller than the aorta: MPA 2.6 cm, aorta 2.9 cm of the septum primum. The second type is the ostium primum ASD, located in the inferior part of the septum close to the mitral valve. It is considered an A-V canal defect. It accounts for 15 – 20% of ASDs. It is usually associated with a cleft in the anterior leaflet of the mitral valve which may lead to mitral incompetence (this is the type this patient had). The third form of ASD is the sinus venosus type (5%). In this type, the defect is located near the orifice of either the superior or inferior vena cava in the right atrium. It is associated with partial anomalous pulmonary venous connection. The right superior pulmonary vein drains into the right atrium or the superior vena cava and the right inferior pulmonary vein drains into the right atrium or the inferior vena cava. The fourth type, called coronary sinus ASD (1%) is rare and it is due to a defect in the roof of the coronary sinus leading to a free communication between the coronary sinus and the left atrium. Rarely, the entire atrial septum fails to develop, or there may be just a rim of tissue in place of the septum resulting in a common or single atrium.

Pathophysiologically, the ASD leads to a shunt between the two atria, with the blood flowing from the left to the right atrium due to the relative higher pressure on the left. The shunt consequently leads to increased blood flow to the lungs resulting in pulmonary vascular congestion and recurrent respiratory infections. Chronic pulmonary congestion from a long standing ASD can lead to pulmonary occlusive vascular disease, pulmonary hypertension and reversal of the shunt with cyanosis (Eisenmenger’s syndrome). By the age of 40 years 35 – 50% of patients with ASDs will have pulmonary...
hypertension\textsuperscript{3}. A patient with pulmonary hypertension and shunt reversal is no longer a candidate for surgery since the repair of the ASD immediately leads to right heart failure, making it difficult to wean the patient off cardiopulmonary bypass. This scenario often leads to an intra-operative or early postoperative mortality.

Clinically, most patients are asymptomatic. Those who have symptoms complain of dyspnoea on exertion, easy fatigability and palpitations. Physical examination might not reveal much. There may be fixed splitting of the second heart sound, a soft systolic murmur loudest at the left sternal edge in the 2\textsuperscript{nd} or 3\textsuperscript{rd} intercostal space. The chest x-ray may be normal or show pulmonary plethora or cardiomegaly. The ECG may be normal, show right ventricular hypertrophy, or right axis deviation. Echocardiography is the mainstay of diagnosis. It shows the location (and therefore type) of the atrial septal defect, direction of blood flow and any other anomalies. Cardiac catheterization is usually not needed unless there is some ambiguity about the diagnosis, or to confirm or rule out pulmonary hypertension.

The management of an ASD is closure of the defect. The best time to close the ASD is in childhood between the ages of 3 – 5 years\textsuperscript{[1,2,3]}. Spontaneous closure of an ASD after the age of 2 years is rare\textsuperscript{3}. Another indication for surgery is when the ratio of pulmonary blood flow to systemic blood flow (Qp:Qs) exceeds 1.5 :1. The surgical procedure involves the repair of the ASD under CPB, through a median sternotomy or right antero-lateral thoracotomy. The ASD is usually closed with a patch of pericardium, Goretex, or dacron. Small ostium secondum ASDs can be closed primarily with a suture. Percutaneous transcatheter device closure of ostium secundum ASDs using the Amplatzer Septal Occluder (ASO) is gradually becoming the trend\textsuperscript{[4,6,7]}. However, the other three types of ASDs (comprising 15 – 20\% ) still need open-heart surgery.

It is the general consensus that ASDs must be repaired in childhood. The controversy arises when the diagnosis is made in adulthood. Should the ASD be closed, or should the patient be left alone? Murphy\textsuperscript{8} analysed the outcome after ASD closure in adult patients and stated that “When repair was performed in older patients, late cardiac failure, stroke, and atrial fibrillation were significantly more frequent”. He concluded that “Among patients with surgically repaired atrial septal defects, those operated on before the age of 25 have an excellent prognosis, but older patients require careful, regular supervision”. Other studies were of different opinion. Gatzoulis\textsuperscript{9} found immediate reduction in the QRS duration on the ECG and reduction in the cardiothoracic ratio within 6 months. He also found improvement in exercise tolerance. He concluded that “Today, operation for atrial septal defects in adults can be performed with no mortality and low morbidity and results in symptomatic improvement in the majority of patients. Clinical improvement was seen even in patients who considered themselves asymptomatic preoperatively. We advocate closure of atrial septal defects in adult patients with symptoms or significant atrial shunts”. Helber\textsuperscript{10} also analysed data after ASD closure in adults and concluded that “Although preoperative cardiopulmonary capacity in adult patients with nonrestrictive ASD was significantly decreased, some improvement was seen at 4 months postoperatively, with complete restitution to normal at 10 years after shunt closure”. Jemielty\textsuperscript{11} found improvement in the NYHA status, reduction in pulmonary congestion, but also more atrial fibrillation. Echocardiographic examination showed a significant reduction in the right ventricular dimensions. He concluded that “Surgical closure of atrial septal defects in patients over 40 years old can improve their clinical status and prevent right ventricular dilatation and insufficiency”.

Our patient had normal right ventricular dimensions but with an increasing main pulmonary artery, right ventricular dilatation was a sure outcome. We decided to close the ASD in other to prevent right ventricular dilatation and insufficiency, as noted by Jemielty\textsuperscript{11}, and also to forestall the consequent development of pulmonary hypertension and Eisenmenger’s syndrome. Other studies also found improvement in right ventricular dimensions and remodeling\textsuperscript{12,13,14} and stroke volume and cardiac output\textsuperscript{15}. However, closure of ASDs in the elderly is not without problems. Notable ones are atrial fibrillation and acute left ventricular failure leading to acute pulmonary oedema. In the elderly patient with impaired left ventricular systolic and diastolic function (left ventricular restriction), the sudden closure of the ASD by the percutaneous transcatheter device method leads to a sudden increase in the left ventricular preload, leading to left ventricular failure and acute pulmonary oedema. This complication is prevented by preconditioning the left ventricle\textsuperscript{16,17}. To identify the patients in whom preconditioning was indicated, temporary balloon occlusion of the ASD was done for 15 min and those in whom the left atrial pressures rose more than 10mmHg were then selected. The preconditioning was done with intravenous dopamine, milrinone and frusemide for 48–72 hours and the ASD was closed at the next session. For those without the indication for preconditioning the ASD was closed at the first transcatheter session.

Conclusion

Closure of ASD in the elderly is gradually becoming routine. Most of them are ostium secundum ASDs and so are closed by the percutaneous transcatheter device method. This spares the patient (and the management team) and the numerous side effects of cardiopulmonary bypass. However, ostium primum, sinus venosus and coronary sinus ASDs still require
repair through open-heart surgery under cardiopulmonary bypass.

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