

Acute Changes on Pulmonary Pressure Following Percutaneous Closure of Secundum Atrial Septal Defect

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ABSTRACT

Pulmonary hypertension is a common complication of congenital heart disease due to systemic – pulmonary circulation shunt which if left uncorrected leads to increased pulmonary artery pressure, vascular remodeling and further increase of pulmonary vascular resistance. Percutaneous closure of the defect interrupts this shunt thus reducing right heart and pulmonary circulation load and pulmonary artery pressure. In this paper we present two cases of percutaneous secundum atrial septal defect closure complicated by pulmonary hypertension along with echocardiographic evaluation of cardiopulmonary hemodynamic changes before and shortly after device closure. Forty years old and thirty three years old females presented to our clinics with classical symptoms of atrial septal defects, assessment revealed TVG of 37 mmHg and 30 mmHg, shortly after the procedure patient was re-evaluated and revealed TVG of 39 mmHg and 23 mmHg respectively. From these cases we conclude that changes in pulmonary artery pressure is not constantly found after device closure. However both patients display improvements in functional capabilities.

Keywords: Atrial Septal Defects; Percutaneous ASD Closure; Pulmonary Hypertension

INTISARI

Hipertensi pulmonal adalah komplikasi umum pada kelainan jantung kongenital yang diakibatkan oleh *shunt* sirkulasi sistemik-pulmonal yang jika dibiarkan tidak dikoreksi akan menyebabkan peningkatan tekanan arteri pulmonal, perubahan vaskular dan peningkatan resistensi pembuluh darah pulmonal. Penutupan defek menghentikan aliran melalui *shunt* sehingga mengurangi beban sirkulasi jantung kanan dan tekanan arteri pulmonal. Dalam makalah ini kami mempresentasikan dua kasus penutupan defek septum atrium dengan hipertensi pulmonal disertai evaluasi ekokardiografi terhadap perubahan hemodinamika kardiopulmonal sebelum dan sesaat setelah penutupan defek. Perempuan berusia 40 dan 34 tahun berkunjung ke klinik kami dengan gejala klasik defek septum atrium, pemeriksaan menunjukkan TVG 37 mmHg dan 30 mmHg. Setelah penutupan defek, re-evaluasi menunjukkan TVG 39 mmHg dan 23 mmHg. Dari kasus ini kita dapat menyimpulkan bahwa perubahan tekanan arteri pulmonal setelah penutupan defek tidak selalu didapat, namun demikian kedua pasien menunjukkan perbaikan klinis.

INTRODUCTION

Pulmonary Hypertension (PH) can be found in 9 to 35% of patients with atrial septal defect (ASD). It is a consequence of increased left to right shunt resulting in volume overload of

the right heart and pulmonary circulation.¹ If left untreated pulmonary hypertension might further develop into right heart failure, arrhythmia, and thromboembolic events.² Percutaneous closure has become a preferred treatment in patients

with secundum type ASD. It does not only increases exercise tolerance, it also improves heart hemodynamics.³ These cases evaluate the hemodynamic effects of percutaneous atrial septal defects closure on pulmonary hypertension.

CASE PRESENTATION

Case 1

Forty years old female came to our clinics with complaint of chest pain, palpitations and dyspnea on effort since last 1 year. On examinations, she had wide fixed splitting heard best at 3rd ICS. Chest X-ray showed cardiomegaly and peripheral pruning. Electrocardiogram (ECG) showed sinus rhythm with incomplete right bundle branch block. Transthoracic echocardiography (TTE) revealed inferior vena cava (IVC) dilatation

and IVC collapse of less than 50%, right atrial and ventricular dilatation, no left ventricle (LV) D-shaped. Tricuspid valve pressure gradient (TVG) was 37 mmHg with estimated mean pulmonary arterial pressure (mPAP) of 37 mmHg and estimated right systolic ventricular pressure of 57 mmHg. Tricuspid regurgitation (TR) Vmax was 3.03 m/s with PV Acceleration Time of 94 ms. There was no anomalous pulmonary connection. Right heart catheterization confirms the presence of pulmonary hypertension with PA pressure 26/24 mmHg. Three months later evaluation with transesophageal echocardiography (TEE) was done, there was 1.6-2.3 cm secundum ASD with left to right shunts and pulmonary hypertension with TVG 35mmHg. Rim assessment proved suitable for device closure. She underwent device closure procedure with amplatzer

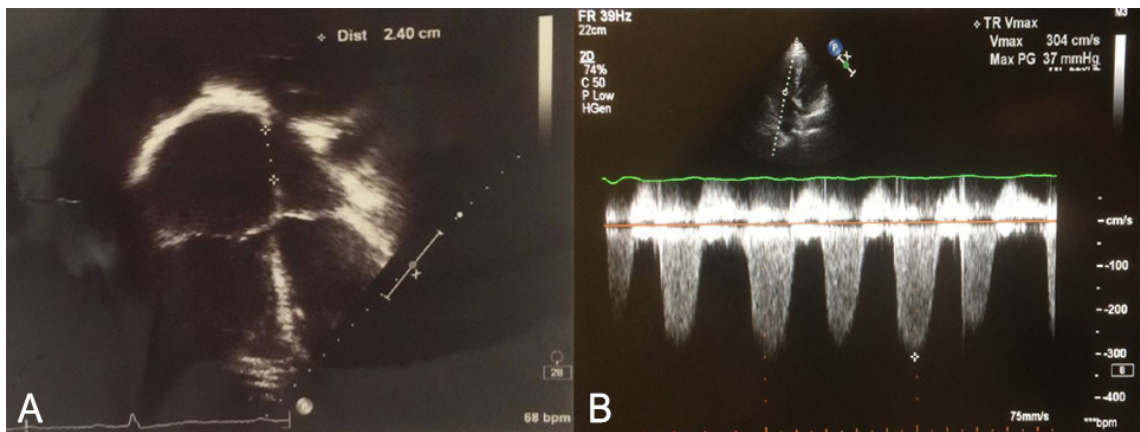


Figure 1. Case 1 Pre ASO evaluation. A. Atrial Septal Defect. B. Transvalvular Gradient

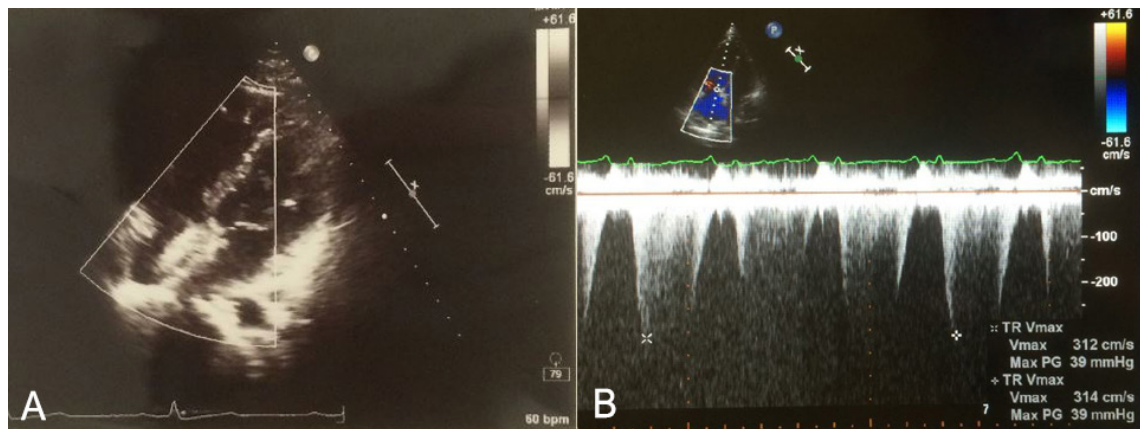


Figure 2. Case 1 Post ASO evaluation. A. Atrial Septal Defect. B. Transvalvular Gradient

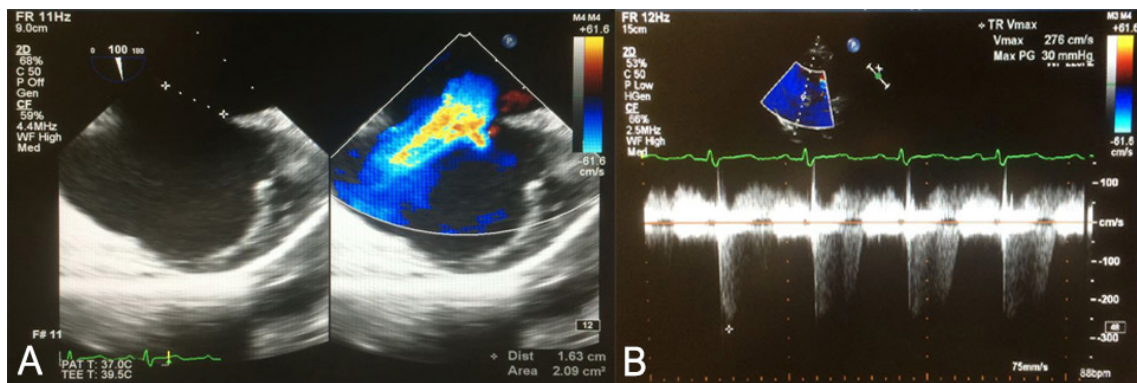


Figure 3. Case 2 Pre ASO evaluation. A. Atrial Septal Defect. B. Transvalvular Gradient

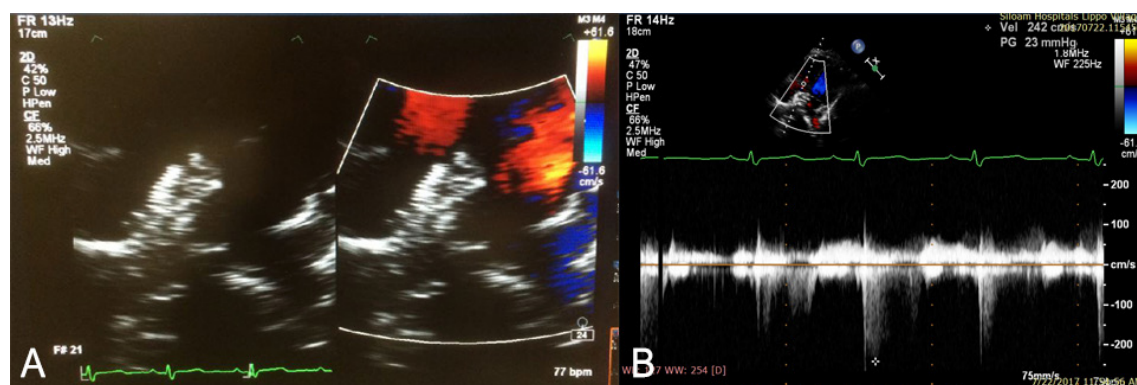


Figure 4. Case 2 Post ASO evaluation. A. Atrial Septal Defect. B. Transvalvular Gradient

septal occluder (ASO). After the procedure, heart hemodynamics was re-evaluated with transthoracic echocardiography. Estimated mPAP was 36 mmHg, TVG was 39 mmHg, TR Vmax 3.13 m/s, and PV Acceleration Time 88 ms. On follow up, patient feel better with increased functional class.

Case 2

A thirty four years old female came to our clinics with a complaint of palpitation since 1 year ago and recurrent dyspnea for the past three months and exercise intolerance. On examinations, she had wide fixed splitting with ECG showing incomplete right bundle branch block. There was no history of paradoxical embolism. TEE was performed confirming diagnosis of secundum Atrial Septal Defects with left to right shunts and an estimated diameter of 1.6-2.3cm. TVG was 30 mmHg. As rim was suitable for ASD closure, she was then planned

to undergo TEE guided ASD closure. After closure, assessment showed good placement of septal occluder device, with TVG 23 mmHg and there was no IVC dilatation, IVC collapse was less than 50%. On follow up there was no complication. On follow up patient no longer experience dyspnea and there was no other complaint.

DISCUSSION

Pulmonary hypertension is a progressive disease characterized by increased of mPAP of ≥ 25 at rest and an elevation of pulmonary vascular resistance (PVR) of more than 3 Woods Unit (WU) with normal PA wedge pressure of ≤ 15 mmHg.⁴ Definitive diagnosis is to be made by right heart catheterization (RHC). Although it is a relatively safe procedure, RHC is invasive and thus a noninvasive alternative is desirable for early diagnosis of PH. Echocardiography

has been used to estimate presence of PH and to assess morphological and functional consequences of PH. It can be used to assess several variables that had significant prognostic values and plays a role in monitoring patient's response to therapy.⁵

PH associated with congenital heart disease (CHD) is classified in Pulmonary Arterial Hypertension (PAH) - Group I according to ESC guideline along with idiopathic PH, familial PH, drugs and toxin induced PH, PH associated with connective tissue disease, HIV infection, portal hypertension and schistosomiasis.⁶ Five to ten percent of patients with PAH have congenital heart disease, and one of the most common CHD is atrial septal defects.⁴

Atrial septal defect maybe single or multiple and can be located anywhere along the atrial septum. Embryologically, atrial septation involves septum primum, septum secundum and atrioventricular (AV) canal septum. Normal development of the atrial septum results in the formation of fossa ovalis bounded by septum secundum. Secundum atrial defect is the most common congenital heart defect that causes systemic - pulmonary circulation shunting and is frequently due to deficiency of septum primum.⁷ Patients with isolated ASD are usually asymptomatic during the first two decades of life. As patients reaches adulthood, around the fourth decades of life, the degree of left to right shunt increases as ventricular compliance decreases accompanied by increase in systemic vascular resistance. These changes lead to several clinical manifestations such as exercise intolerance and palpitation that frequently occur as a result of right heart chamber enlargement, pulmonary hypertension, RV failure, tricuspid regurgitation and atrial arrhythmias. Which if left untreated will progress into right sided heart failure. Life expectancy is not normal in patients with untreated ASD, mortality increases by 6% per year after forty years old.⁸

With increased left to right shunt, there is increased flow to the right heart and pulmonary circulation, and thus increased pulmonary artery flow. At this point, correction of pulmonary artery hypertension is still reversible. However, prolonged exposure to increased flow to the pulmonary artery leads to remodeling of the pulmonary vasculature. Intimal fibrosis and hypertrophy of medial smooth muscle layer of pulmonary artery cause narrowing of lumen. These changes increases PVR and thus increased pulmonary artery pressure. This creates pressure overload in the right ventricle. Pressure overload increases the work of ventricle leading to right ventricular and right atrial dilatation, which further progress to right ventricular dysfunction, abnormal inter-ventricular septal motion and dilatation of the superior and inferior vena cava and hepatic veins, as well as pericardial effusion.^{5,9,10} With progressive increase of the right heart pressure and pulmonary vascular resistance, eventually reversal of shunt may arise, developing Eisenmenger syndrome. Eisenmenger syndrome is the most advanced form of PAH-CHD which was defined in 1958 as pulmonary hypertension at systemic level due to high pulmonary vascular resistance with reversed or bidirectional flow through a septal defect. With reversal of shunt directions, symptoms arise from hypoxemia. Reduced hemoglobin saturation stimulates bone marrow production of red blood cells (erythrocytosis). This can cause hyperviscosity manifesting as fatigue, headache, and stroke due to cerebrovascular occlusion. Hemoptysis may occur in case of pulmonary artery infarct/rupture.^{11,12} When Eisenmenger syndrome developed, there is no remedies to turn back the disease process, although some treatment strategies can be applied, the only effective long-term strategy for severely affected patients is lung / heart-lung transplantation.¹² However this is limited to the scarcity of donor organs and

lack of organ specificity in developing countries.¹¹ Fortunately, with early detection and correction of congenital heart defects, Eisenmenger syndrome has become less common.¹²

Small defects often close spontaneously within the first few years of life. However, significant ASD (Qp/Qs of > 1.5) seldom do, therefore they are closed surgically or by interventional catheter techniques before school age or at the time of diagnosis later in life.¹³ Closure of interatrial septal defects closes the left to right shunt, resulting in reduction of volume overload on the right heart and pulmonary circulation. Thus pulmonary vascular disease might be prevented or reversed with early correction of the defects.¹ Surgical closure of the defects has been practiced for over 45 years, but over the past three decades transcatheter device closure has evolved and is increasingly used in recent years. A wide variety of device has been developed for transcatheter closure of ASD, in this study we demonstrate the use of Amplatzer septal occluder (ASO) device. ASO is one of the commonly used device, safe, easy to use and has high success rate (98.4%). On comparison, success rate for both surgical or transcatheter device closure is not statistically different. However, complication rate was lower and length of stay was shorter in device closure patients.¹⁴ It is also easily visualized on TEE and fluoroscopy. In the last decade it has become the treatment of choice for device closure. Successful closure is complete closure with residual shunts of less than 1-2 mm, while long-term success is associated with reduced RV pressure and PH. Possible complications include gastrointestinal or intracranial bleeding due to use of antiplatelet/ anticoagulant, device embolization, atrial arrhythmia (2-4%), catheter related thrombus formation, air embolism, pericardial effusion, and transient ischemic attack or stroke.¹⁵

In this paper we evaluate changes in pulmonary hypertension after device closure by echocardiography. Estimation of PA pressure by echocardiography can be made by assessing estimating tricuspid valve pressure gradient. Aside from that, RA pressure estimation can be used to evaluate right heart hemodynamics. Estimation of RA pressure can be made with IVC diameter and degree of inspiratory collapse.^{1,16,17} The first case displayed no changes in TVG while having increased clinical performance. This could be caused by a long standing pulmonary hypertension leading to pulmonary vascular remodeling causing increased pulmonary vascular resistance and reduced compliance.¹⁸ Or because of the reduced compliance of the left ventricle in comparison to right ventricular compliance.¹⁹ Meanwhile, the second case displayed changes in TVG after device closure and clinical improvements. This difference in TVG evaluation result might be caused by a ten year age difference. Studies have shown that aging correlates with increased pulmonary systolic arterial pressure.²⁰ In older patients, ASD closure resulted in further deterioration of LV relaxation impairment and increased LV stiffness.²¹ Despite the difference in PH changes after percutaneous ASD closure, both patients display clinical improvements.

CONCLUSION

From the cases above, acute hemodynamic changes of the pulmonary artery is not constantly found subsequent to percutaneous closure of atrial septal defect. Even so, without significant changes of pulmonary artery pressure, our patients had improved functional capabilities. However due to limitations regarding the small sample size, this conclusion is prematurely made, thus further study is required.

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