

# Successful Percutaneous Device Closure for Giant Atrial Septal Defect with Massive Pericardial Effusion and Pulmonary Hypertension in an Elderly Patient

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## Abstract

An 82-year-old man suffering from shortness of breath, leg edema, and appetite loss visited our outpatient clinic. Although he was diagnosed with atrial septal defect (ASD) requiring surgical repair 40 years ago, he had refused an operation at that time. Echocardiography revealed a 37-mm ASD and massive pericardial effusion. Cardiac catheterization showed significant left-to-right shunt flow with Qp/Qs of 4.6 and pulmonary artery pressure of 93/35/52 mmHg. Pulmonary vascular resistance was calculated as 8.3 Wood units. Surgical treatment was no longer indicated due to his condition, and percutaneous treatment with an Amplatzer septal occluder (ASO) was planned instead. Diuretics, a PDE3 inhibitor and nasal oxygen, were administered preoperatively for 1 month. A 38-mm ASO, the maximum occluder size available in Japan, was implanted successfully. No acute decompensation occurred after ASD closure, and the patient's symptoms improved after ASO implantation. Cardiac catheterization on postoperative day 13 revealed no evidence of residual shunt, and pulmonary artery pressure decreased to 63/20/33 mmHg. As postoperative therapy, a PDE5 inhibitor, endothelin receptor blocker, and PGI<sub>2</sub> analog were administered for residual pulmonary hypertension. Because pericardial effusion did not disappear after pericardiocentesis, surgical pericardiostomy was performed 6 months after ASD closure, which reduced PA pressure to 34/16/24. Appropriate

pre- and postoperative medical therapy, device closure with an ASO, and pericardiostomy were effective in this frail patient with a giant ASD with pulmonary hypertension and massive pericardial effusion.

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## Key Words:

Atrial septal defect • Pulmonary hypertension • Catheter • Intervention

## Introduction

Atrial septal defect (ASD) is a common congenital heart disease in adults. The main indication for ASD closure is the presence of significant shunting as evidenced by right heart volume overload with or without symptoms [1]. Severe fixed pulmonary hypertension is considered a contraindication for surgical repair of an ASD [2]. However, development of percutaneous closure techniques and medical management may make secundum ASD closure feasible and alleviate symptoms [3].

## Case Presentation

An 82-year-old Japanese man suffering from shortness of breath, leg edema, and appetite loss



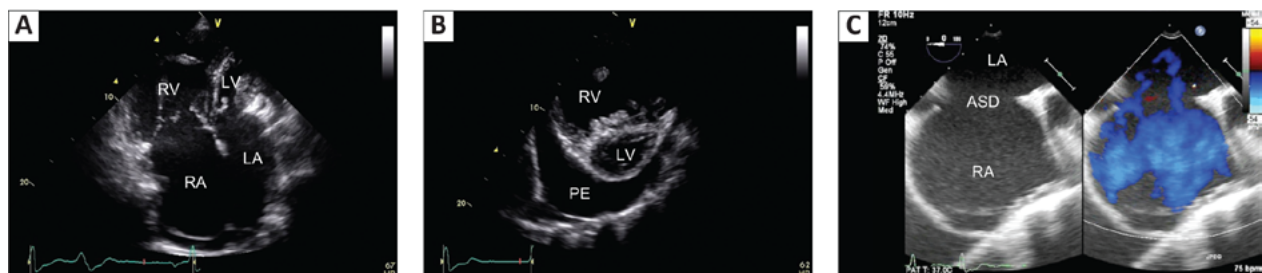
visited our outpatient clinic. He was diagnosed at 43 years of age with secundum ASD requiring surgical repair but had rejected treatment at that time. At 76 years of age, he was referred from a community hospital to our institute due to shortness of breath on exertion. By this time, surgical intervention was no longer indicated because echocardiography revealed severe pulmonary hypertension (estimated right ventricular systolic pressure, 102 mmHg). He was treated conservatively with heart failure medications such as furosemide, digoxin, and carvedilol. However, because his symptoms had worsened considerably over the past several years, he was referred to our institute again and hospitalized while alternative treatments were considered.

Although his vital signs were stable (blood pressure, 125/69 mmHg; pulse rate, 69 beats per minute with irregularity), his transcutaneous oxygen saturation (SatO<sub>2</sub>) in room air had decreased to 95%. During a 6-min walk test, his SatO<sub>2</sub> on 1 l/min of nasal oxygen declined to 88%. On physical examination, his jugular veins were dilated, and facial and pretibial edema were observed. Rales were audible in both lower lung fields. His heart sounds revealed fixed splitting of the second heart sound and a systolic murmur at the second intercostal space and left-sternal border. Laboratory data showed a total bilirubin of 1.3 mg/dl and B-type natriuretic peptide of 407 pg/ml. A chest X-ray revealed cardiomegaly, enlargement of the bilateral pulmonary artery, and pleural effusion (Figure 1). An electrocardiogram showed atrial fibrillation and complete right bundle branch block. Transthoracic echocardiography (TTE) revealed enlargement of the right heart, hypoplasia of the left ventricle (Figure 2A), and

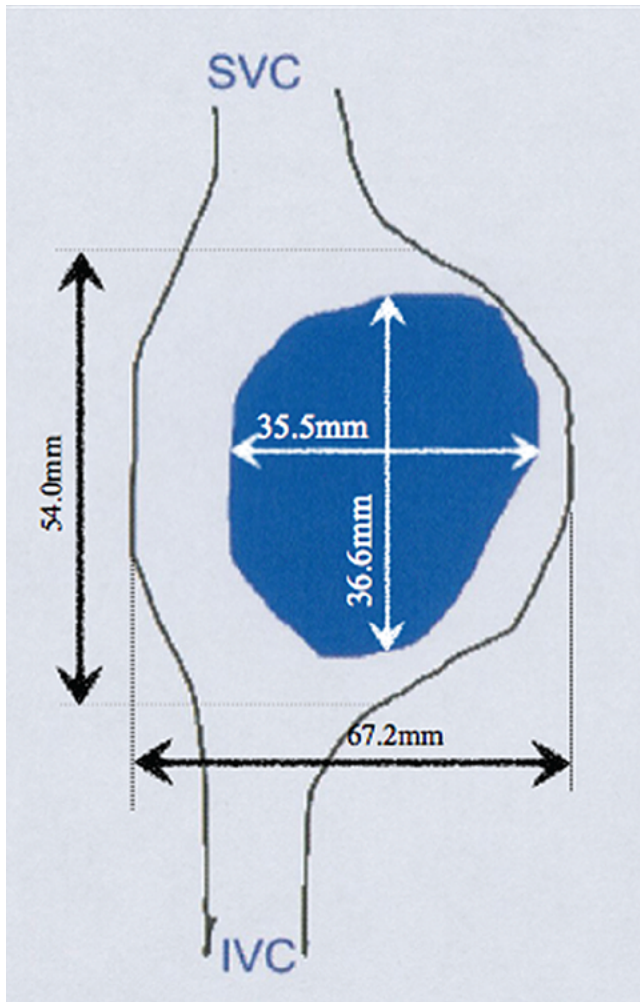
massive pericardial effusion (Figure 2B). In addition, there was an ostium secundum-type ASD with left-to-right shunting and moderate tricuspid regurgitation on transesophageal echocardiography (TEE) (Figure 2C). The size of the defect and the rims were also measured by TEE. His ASD showed a maximal size of 36 × 37 mm, and each rim was long enough for device closure (Figure 3). Cardiac catheterization revealed Qp/Qs of 4.6, pulmonary artery pressure of 93/35/52 mmHg, and pulmonary vascular resistance



**Figure 1.** Chest x-ray on admission. Cardiomegaly, dilatation of the pulmonary arteries, and pleural effusion were observed.



**Figure 2.** Panel A. Four-chamber view of transthoracic echocardiography (TTE). A large right and left atria and ASD were observed. Panel B. Short-axis view of TTE. A dilated right ventricle and hypoplastic left ventricle with massive pericardial effusion were observed. Panel C. Transesophageal echocardiography findings. A dilated right atrium and left-to-right shunt flow through the ASD were observed. ASD = atrial septal defect; LA = left atrium; LV = left ventricle; PE = pericardial effusion; RA = right atrium; RV = right ventricle.



**Figure 3.** Transthoracic echocardiography findings of atrial septal defect (ASD). The ASD occupies large part of the atrial septum. SVC = Superior vena cava; IVC = Inferior vena cava.

(PVR) of 8.3 Wood units. His PVR index was 5.4 Woods units · m<sup>2</sup>, and pulmonary-to-systemic blood pressure ratio was 0.87.

Mainly due to his frailty, percutaneous treatment with an Amplatzer septal occluder (ASO) was planned. Two different diuretics, a PDE3 inhibitor (milrinone 0.2 µg/kin/min) and nasal oxygen, were administered as preoperative therapy in preparation for acute decompensation (Figure 4). After 1 month of medical treatment, we performed percutaneous device closure for atrial septal occlusion. We performed the operation while monitoring intra-cardiac pressure. When we occluded the defect with an ASO

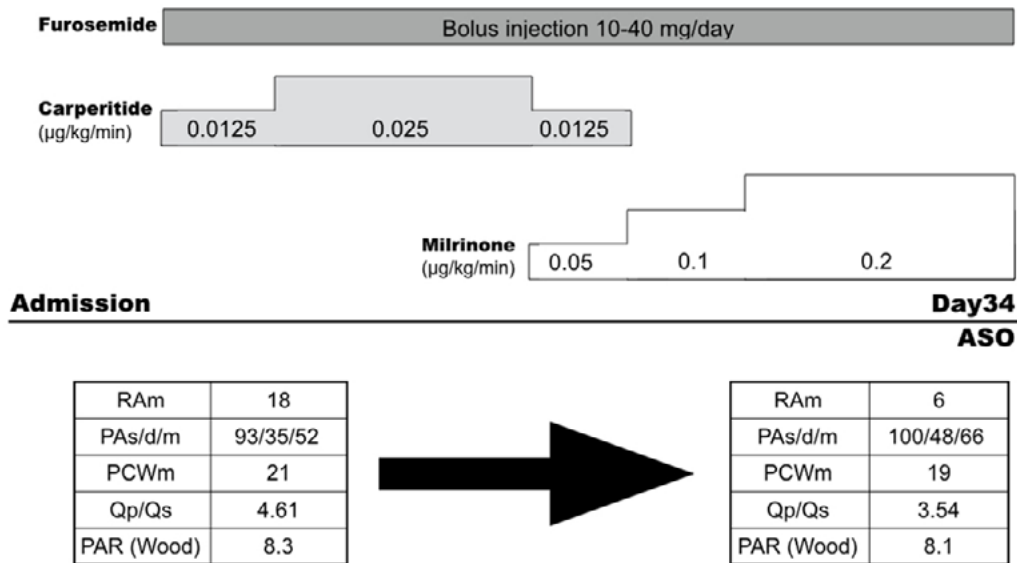
temporarily, we observed that pulmonary capillary wedge pressure and left ventricular end diastolic pressure did not rise markedly. Finally, we successfully implanted a 38-mm ASO, which was the maximum size available in Japan at the time (Figure 5). No pulmonary edema occurred after ASO implantation, and the patient's symptoms improved without oxygen administration. When cardiac catheterization was performed on postoperative day 13, there was no evidence of residual shunting, and pulmonary artery pressure had decreased to 63/20/33 mmHg. As pericardial effusion was still present after successful ASD closure, pericardiocentesis was performed, and 1,500 ml of fluid was removed. A PDE5 inhibitor (sildenafil 60 mg), endothelin receptor blocker (ambrisentan 5 mg), and PGI<sub>2</sub> analog (beraprost 120 µg) were administered to treat the remaining pulmonary hypertension. The patient was subsequently discharged from the hospital on postoperative day 41 and returned home on foot.

Three months after ASO implantation, the patient was re-hospitalized due to dyspnea on exertion because pericardial effusion appeared again, even after pericardiocentesis and intensive medical therapy (Figure 6). His pulmonary artery pressure was still high at 47/26/35 mmHg. Therefore, surgical pericardiostomy was performed 6 months after ASD closure. Finally, his left ventricular shape became round and enlarged (Figure 7), and pulmonary artery pressure decreased to 34/16/24 (Figure 6). Finally, he was discharged again and became asymptomatic.

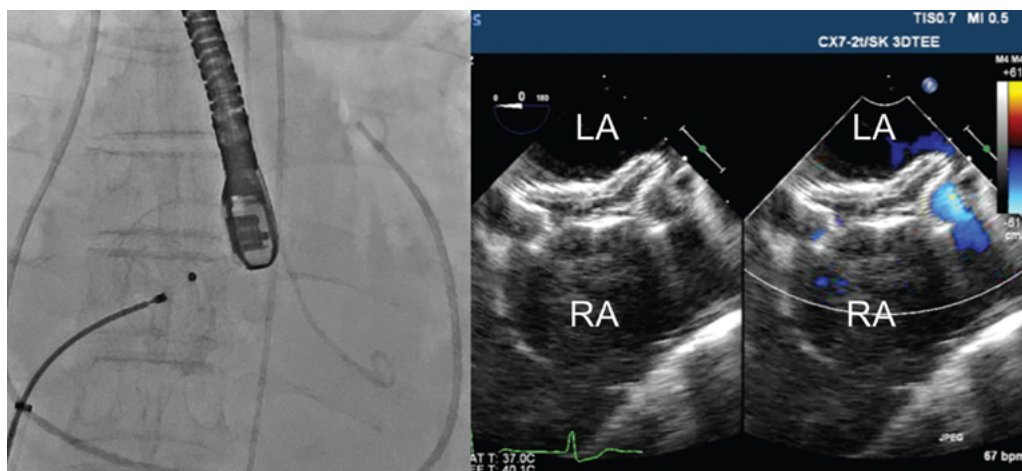
## Discussion

We successfully treated a patient with a giant ASD along with extreme pulmonary hypertension and massive pericardial effusion percutaneously with an ASO. Therefore, this case shows that appropriate and timely pre- and postoperative medical therapy combined with ASD closure can be effective.

In patients with significant ASD, left ventricular preload is reduced and a reduction of LV volume is observed over time [4]. In these cases, closure of an ASD with a large left-to-right shunt can increase left ventricular inflow, leading to left ventricular heart failure [5]. To avoid this catastrophic condition, two diuretics and an inotropic agent were administered intravenously for



**Figure 4.** Preoperative medication with two diuretics and an inotropic agent. Although right atrium pressure was decreased, pulmonary artery pressure was not changed by the treatment. ASO = Amplatzer septal occluder; m = mean; PA = pulmonary artery; PAR = pulmonary artery resistance; PCW = pulmonary capillary wedge; Qp/Qs = pulmonary-to-systemic blood flow ratio; RA = right atrium; s/d/m: systolic/diastolic/mean.

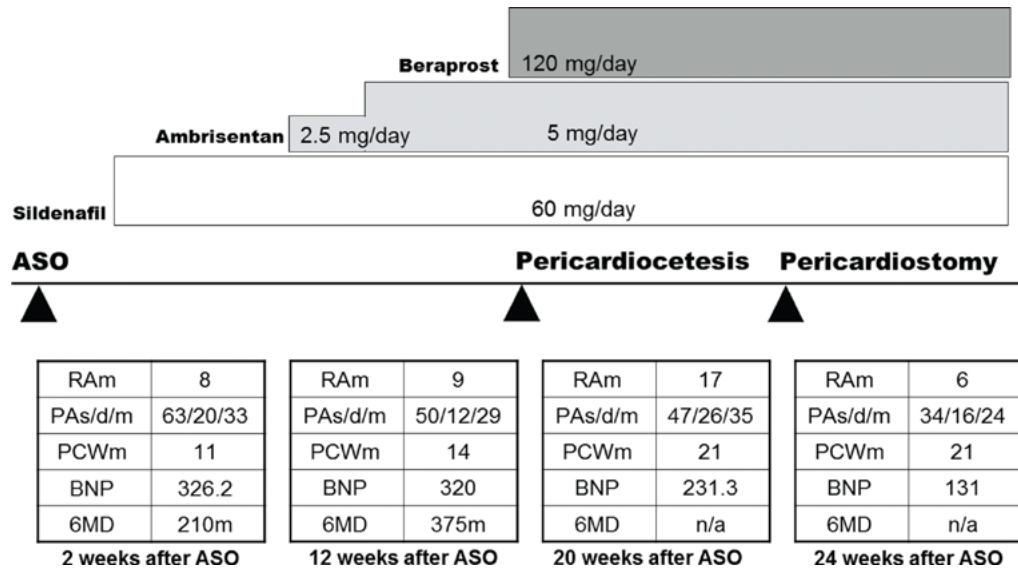


**Figure 5.** Angiographic (left panel) and TEE (right panel) findings during implantation of an Amplatzer septal occluder.

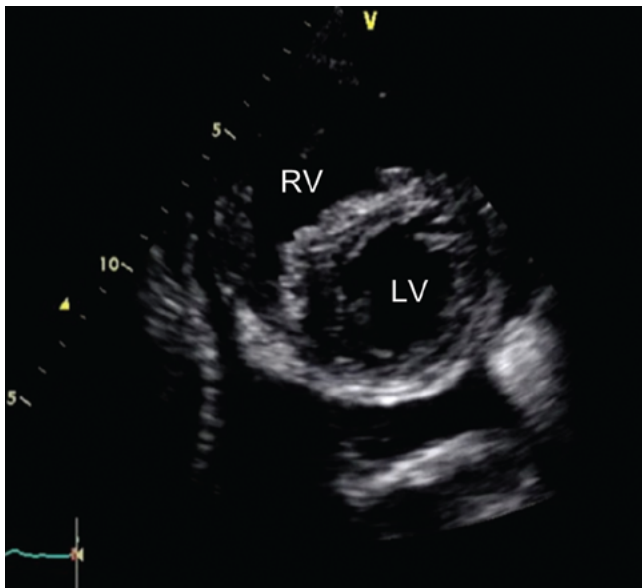
1 month before the operation. Although severe pulmonary hypertension was observed in this case, pulmonary artery dilators were not used before the procedure contrary to current recommendations for surgical treatment [6]. This is because a possible increase in left-to-right shunt due to a reduction in pulmonary artery resistance might increase left ventricular inflow, leading to left ventricular heart failure due to masked left ventricular restriction [7] at the time of ASD closure. In this case, ASD closure with an ASO was safely performed without

any hemodynamic deterioration.

As the massive left-to-right shunt was completely eliminated by ASO implantation, pulmonary artery pressure was expected to decrease after the procedure, but this did not occur. Therefore, potent pulmonary artery dilators such as a PDE5 inhibitor, endothelin receptor blocker, and PGI<sub>2</sub> analog were started incrementally after the procedure. The present case is considered to be high risk because the PVR was greater than 5 Woods units m<sup>2</sup>, and pulmonary artery pres-



**Figure 6.** Medications and examinations after ASO implantation. 6MD = 6-min walk distance; ASO = Amplatzer septal occluder; m = mean; PA = pulmonary artery; PCW = pulmonary capillary wedge; RA = right atrium; s/d/m = systolic/diastolic/mean.



**Figure 7.** Short-axis view of transthoracic echocardiography after drainage of the pericardial effusion 6 months after atrial septal defect closure. The left ventricular shape became round, and the dimension of the left ventricle increased.

sure was greater than 70% of systemic pressure [8]. The pulmonary hypertension-specific medications that we administered are reportedly effective even in high-risk patients during long-term follow-up [8]. However, in our particular case, probably due to the

presence of chronic massive pericardial effusion, the reduction in pulmonary arterial pressure fell short of our expectations. Because pericardiocentesis had no effect on pericardial effusion, pericardiostomy and drainage was performed according to previous reports [9]. Finally, the surgical procedure eliminated the effusion, which was followed by a decrease in pulmonary artery pressure to a nearly normal level and the disappearance of symptoms.

## Conclusions

We report a case of giant ASD with massive pericardial effusion treated by ASO followed by the administration of pulmonary arterial dilators. Potent pulmonary hypertension drugs and percutaneous closure devices make high-risk ASD treatment possible, even in elderly and frail patients.

## Conflict of Interest

The authors have no conflict of interest relevant to this publication.

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