

# Case Report

## Percutaneous Correction of Two Congenital Heart Lesions: Atrial Septal Defect and Pulmonary Valve Stenosis

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### Abstract

Atrial septal defects (ASD) are the most common congenital heart lesion detected in adults and are uncommonly associated with pulmonary valve stenosis (PVS). Due to counteracting effects on the pulmonary circulation, this combination of congenital lesions may delay clinical symptoms until later in life, but increases the likelihood of rapid decompensation. In this case report, we describe the presentation of a 43 year old female patient with both ASD and PVS. We then describe the correction of both lesions via transcatheter procedure.

### Case Presentation

A 43 year old woman was evaluated at our institution for progressive exertional dyspnea and general fatigue over the past year. Additionally, she had a transient ischemic attack (TIA); presenting as 12 hours of right-sided numbness and weakness within the previous month and was treated with daily acetylsalicylic acid (ASA). She was also hypothyroid and on thyroxine replacement. On physical examination she was normotensive with no peripheral edema or cyanosis. All peripheral pulses were palpable and symmetrical. There was no jugular venous distention. Precordial examination revealed fixed wide splitting of the second heart sound, suggesting an underlying atrial septal defect (ASD) (Table. 1). She also had a loud systolic ejection murmur heard best at the upper left sternal border with a systolic thrill and pulmonary ejection click, suggesting pulmonary valve stenosis (PVS) (Table. 2). There was no clinical evidence of heart failure.

A twelve-lead electrocardiogram demonstrated normal sinus rhythm with no evidence of atrial abnormality, conduction defects or right ventricular hypertrophy. A subsequent echocardiogram confirmed

secundum-type ASD and significant PVS. Cardiac catheterization was performed and revealed a peak-to-peak right ventricle to pulmonary artery pressure gradient of 55 mmHg (Table. 3). There was significant post-stenotic dilatation of both pulmonary arteries (Fig. 1). Pulmonary valve leaflet fusion was noted, resulting in eccentric systolic doming of the leaflets, suggesting dysplastic congenital etiology. A left ventriculogram demonstrated an ejection fraction of 60%. Coronary arteriography revealed a right dominant distribution with no angiographic evidence of disease in the major epicardial vessels.

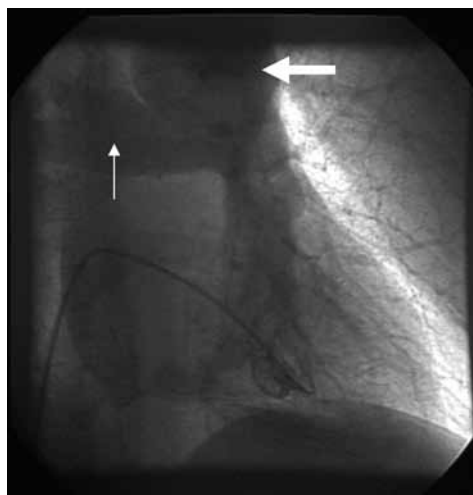


Figure 1. Initial right ventriculogram (right anterior oblique view) demonstrates marked post-stenotic dilatation of left pulmonary artery (large arrow) and mild dilatation of right pulmonary artery (small arrow).

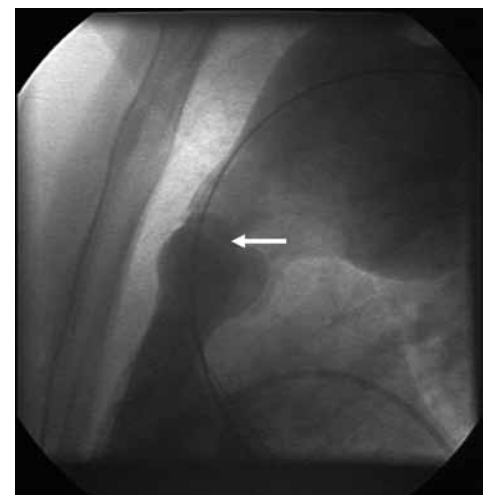


Figure 2. Right ventriculogram (lateral view) demonstrates marked post-stenotic dilatation of left pulmonary artery and doming of valve leaflets (arrow).

Table 1. Common presentation of an atrial septal defect<sup>4</sup>

Symptoms	Physical examination findings
Exertional dyspnea	Right, ventricular heave at LLSB
Fatigue	Wide, fixed split S2
Recurrent LRTIs	Systolic ejection murmur at ULSB
Palpitations	Midsystolic murmur at LLSB

Table 2. Common presentation of pulmonary valve stenosis<sup>4</sup>

Symptoms	Physical examination findings
None	Pulmonic ejection click following S1
Exertional dyspnea	Thrill at ULSB
Exercise intolerance	Systolic ejection murmur at ULSB
	Widely split S2 with soft P2 component
	Prominent <i>a</i> wave in JVP (severe cases)
	Right ventricular heave at sternum (severe cases)

Management options discussed with the patient included:

1. Surgical ASD closure with pulmonary valve repair or replacement
2. Transcatheter ASD closure using an Amplatzer® septal occluder (ASO) and balloon pulmonary valvuloplasty

After reviewing these treatment options, the patient preferred the transcatheter approach.

**Treatment**

The procedure was performed under general anesthesia with fluoroscopic and transesophageal echocardiographic (TEE) guidance. A right ventriculogram performed immediately prior to intervention confirmed pulmonary valve leaflet fusion and was used to assess the size of the pulmonary valve annulus (Fig. 2). Pulmonary valvuloplasty was then performed using the double-balloon technique. Two 10 mm balloons were fully inflated and deflated side-by-side across the valve annulus, leaving no residual balloon waist (annular constriction) at full inflation (Fig. 3). The post-procedure pressure gradient across the pulmonary valve was 3 mmHg, indicating an excellent hemodynamic result (Table. 3).

The stop-flow diameter of the ASD was estimated by inflating a sizing balloon across the opening and measuring the balloon waist with reference to the catheter diameter (Fig. 4). Cessation of flow across the ASD after balloon inflation was confirmed via TEE.

Based on the stop-flow diameter, a 14 mm ASO was selected as the closure device. The two discs of the ASO were deployed via catheter on either side of the ASD (Fig. 5). The device was designed to be securely anchored in place by push-pull action on the delivery cable. The ASO was finally released from the cable and TEE demonstrated successful ASD closure. The total fluoroscopic exposure time was 13.7 minutes and 45 cc of ISO-370 radiographic contrast were injected. The patient was slow to recover from general anesthesia but felt well by the next morning. She was in normal sinus rhythm and received antibiotic prophylaxis. She was also switched from ASA to warfarin for six months with a target INR of 2-3. A follow-up chest x-ray showed satisfactory placement of the septal occluder. Since the procedure, she has been followed by our adult congenital heart clinic. At nine years follow-up, she has demonstrated no significant complications from the procedure and has had no recurrence of her initial presenting symptoms.

**Discussion**

Atrial septal defects (ASD) are the most common adult congenital heart disease, comprising 10% of all congenital heart lesions detected at birth, and up to 40% detected in adults 40 years of age or older<sup>1</sup>. Overall prevalence is approximately 0.73 per 1000 live births. Secundum-type ASD is the most common variant, accounting for over 70% of all ASD, with a male:female ratio of 1:2<sup>2,3</sup>. In typical embryological development, a common atrial chamber is divided by growth and overlap of the septum primum and septum secundum. This overlap, along with perforations in both septa, provides a one-way valve enabling the right-left atrial shunt necessary for fetal circulation (foramen ovale). The septa normally fuse after birth when greater left-sided pressure pushes the foramen ovale closed, eliminating the shunt<sup>4</sup> (Fig. 6). An ASD often occurs due to inadequate growth of the septum secundum, abnormally large foramen ovale or excessive absorption of the septum primum. These lesions typically cause a left-to-right shunt due to higher left-sided pressures, resulting in right ventricular volume overload and

Table 3. Severity of pulmonary valve stenosis<sup>12</sup>

Peak-to-peak transvalvular pressure gradient (mmHg)	Category
<25	Trivial
25-49	Mild
50-79	Moderate
>80	Severe

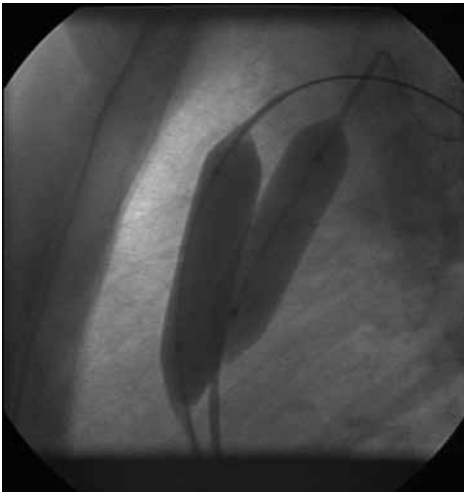


Figure 3. Successful valvuloplasty using double balloon technique. Lack of visible waist at valve annulus indicates successful dilatation.

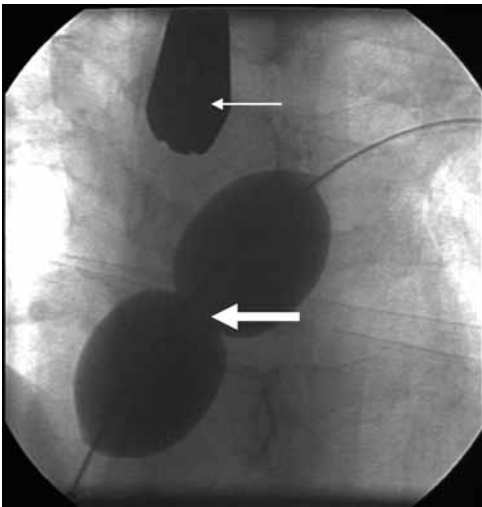


Figure 4. Estimation of ASD stop-flow diameter based on residual balloon waist diameter (large arrow). TEE probe is present (small arrow).

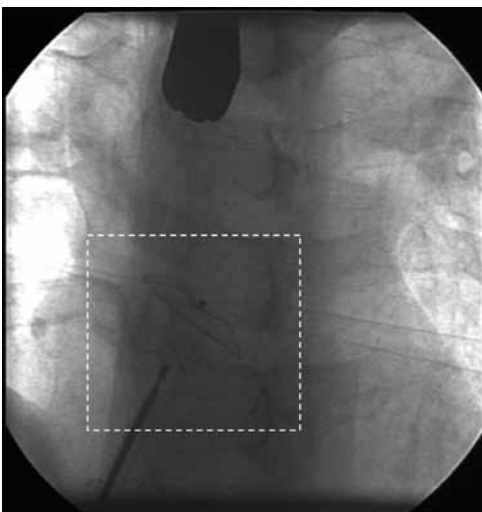


Figure 5. Anterior-posterior view indicates successful placement of ASO (box).

progressive pulmonary hypertension. Patients often present with exertional dyspnea or palpitations secondary to atrial arrhythmias (Table. 1), although with a stable left-to-right shunt, many remain asymptomatic until adulthood. However, an increase in shunt fraction may occur with the development of left ventricular diastolic dysfunction with advancing age and systemic arterial hypertension, intensifying symptoms. Adult patients may develop complications such as paroxysmal or chronic atrial fibrillation. There is also significant potential for paradoxical emboli via the ASD. These two factors increase the risk of thromboembolic episodes including stroke<sup>2</sup>, which may explain the TIA in our patient's history.

ASDs are uncommonly associated with significant pulmonary valve stenosis (PVS)<sup>5</sup>. The etiology of PVS is usually congenital, and most often involves fusion of the valve commissures. Prevalence of PVS is approximately 0.46 per 1000 live births, and is isolated to the pulmonary valve in 90% of cases<sup>3,4</sup>. Patients typically present with exercise intolerance and exertional dyspnea (Table. 2). The presence of PVS in the setting of an ASD may actually reduce or extinguish the left-to-right shunt fraction. In this case, the right-sided outflow obstruction leads to right ventricular hypertrophy and reduced compliance, thus raising diastolic pressure, which results in right atrial pressure elevation. This may help balance the pressure differential between the atria, thus slightly reducing right-sided volume overload. The outflow obstruction would also be expected to limit pulmonary hypertension in these patients<sup>5</sup>. However, given that right ventricular hypertrophy may be increased, right-sided congestive heart failure is more likely. Additionally, the high velocity jet across the stenotic pulmonary valve may result in significant post-stenotic dilatation of main and branch pulmonary arteries, as observed in our patient.

Individuals with large left-to-right shunts are at increased risk of developing Eisenmenger's Syndrome, in which chronic volume-overload of pulmonary circulation causes pulmonary vascular damage, pulmonary arterial hypertension and diminished right ventricular compliance. Implications of these complications include right atrial pressure elevation and reversal of shunt direction<sup>4</sup>. This causes cyanosis and results in significant morbidity, including diffuse organ damage and the potential for paradoxical emboli<sup>4</sup>. Patients with Eisenmenger physiology require careful review of pulmonary vasodilator reserve to determine eligibility for surgical procedures. Significantly elevated pulmonary vascular resistance with lack of vasodilator reserve indicates a high risk of right ventricular failure

following closure of an ASD, and greater potential for peri-procedure morbidity and mortality<sup>6</sup>.

Patients with combination ASD and PVS may present with early cyanosis and shunt reversal due to right ventricular hypertension as a result of the right-sided obstruction<sup>4</sup>. Despite shunt reversal, these patients may actually be protected from pulmonary hypertension. This is due to limited pulmonary flow secondary to the obstruction across the right ventricular outflow tract. Correction of PVS with ASD closure in these patients leads to a favorable outcome with correction of cyanosis and improvement of symptoms<sup>5</sup>.

Correction of secundum-type ASD may employ either a surgical or transcatheter approach. Surgical closure has been practiced with a high success rate for over 50 years, while transcatheter closure is a somewhat newer technique. In surgical closure, sternotomy is performed and the defect may be closed by direct suture or by pericardial or synthetic patch. In the transcatheter approach, the femoral vein is catheterized and an occlusive device is passed along the catheter and deployed across the defect<sup>1</sup>. Typically, the device comprises two collapsible discs for placement on either side of the atrial septum<sup>7</sup>. The transcatheter approach has several advantages, including avoidance of sternotomy and cardiopulmonary bypass. Recovery times are also shorter, and intensive care stay can normally be avoided<sup>1</sup>. The overall complication rate is lower than that for surgical intervention. Specific complications are typically less serious, consisting mainly of arrhythmias, but including hemodynamic instability, pneumothorax and embolization of the closure device<sup>1,8</sup>. Recent studies indicate comparable efficacy in surgical and transcatheter ASD closure based on short-term results, but there are currently no long-term follow-up studies to confirm the overall advantage of the transcatheter approach<sup>1</sup>. Additionally, transcatheter closure has been associated with post-operative atrial systolic dysfunction, thought to be due to mechanical obstruction from the closure device<sup>9</sup>.

PVS is typically corrected with percutaneous balloon valvuloplasty, which has been practiced for over 25 years<sup>7</sup> and is the current gold standard treatment<sup>5,10</sup>. In this technique, a polyethylene balloon is placed via catheter across the stenotic valve and then inflated, dilating the valve annulus. In cases where a single balloon is unable to provide sufficient dilatation, a double-balloon technique may be employed, in which two balloons are inflated side by side using two separate catheters<sup>10</sup>. If balloon valvuloplasty is unsuccessful or contraindicated, surgical valvotomy may be performed.

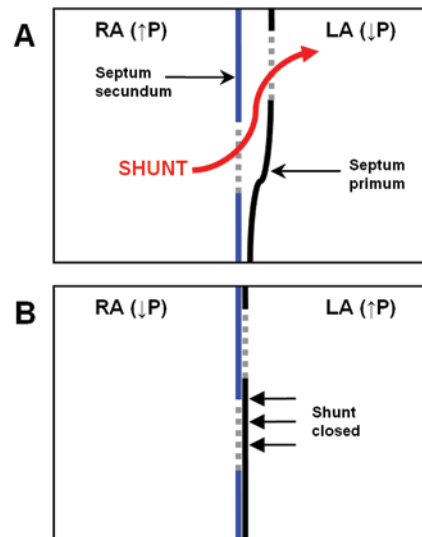


Figure 6. A) Normal right-left atrial shunt (foramen ovale) in fetal circulation maintained by higher right-sided pressure; B) Increase in left-sided pressure after birth forces closure of foramen ovale.

These procedures have comparable short- and long-term outcomes in adults<sup>11</sup>.

### Conclusion

This case illustrates the successful percutaneous correction of two congenital heart lesions. Patients with secundum-type ASDs typically become symptomatic in adulthood, as right-sided volume overload and pulmonary hypertension damage the cardiovascular system to the point of clinical significance. In the uncommon case of ASD associated with PVS, the onset of clinical symptoms is determined by the natural history of the predominant lesion, but may be delayed in comparison to ASD alone. Uncorrected, combined ASD and PVS may lead to shunt reversal and peripheral cyanosis even in the absence of pulmonary hypertension. Prompt correction leads to reversal of cyanosis and relief of symptoms. Percutaneous transcatheter management options may provide equivalent results to surgical correction with shorter hospital stay and avoidance of sternotomy and cardiopulmonary bypass.

### References

- Suchon E, Peiculewicz M, Tracz W, Przewlocki T, Sadowski J, Podolec P. Transcatheter closure as an alternative and equivalent method to the surgical treatment of atrial septal defect in adults: Comparison of early and late results. *Med Sci Monit*, 2009;15(12):CR612-617.
- Gessner IH, "Atrial Septal Defect, Ostium Secundum," *eMedicine Pediatrics: Cardiac Disease and Critical Care Medicine*. Revised November 10, 2008. <<http://emedicine.medscape.com/article/890991-overview>> (April 25, 2010).
- Samanek M, Slavik Z, Zborilova B, Hrobonova V, Voriskova M,

Skovranek J. Prevalence, treatment, and outcome of heart disease in live-born children: a prospective analysis of 91,823 live-born children. *Pediatr Cardiol*, 1989;10(4):205-211.

4. Lilly LS. *Pathophysiology of Heart Disease*. Baltimore: Lippincott Williams & Wilkins, 2007:380-389.
5. Medina A, de Lezo JS, Delgado A, Caballero E, Segura J, Romero M. Combined percutaneous atrial septal defect occlusion and pulmonary balloon valvuloplasty in adult patients. *Tex Heart Inst J*, 2000;27:216-7.

6. Schwerzmann M, Zafar M, McLaughlin PR, Chamberlain DW, Webb G, Granton J. Atrial septal defect closure in a patient with "irreversible" pulmonary hypertensive arteriopathy. *Int J Cardiol*, 2006;110(1):104-107.
7. Becker M, Frings D, Schroder J, Ocklenburg C, Muhler E, Hoffmann R, Franke A, Lepper W. Impact of occluder device type on success of percutaneous closure of atrial septal defects – A medium-term follow-up study. *J Interv Cardiol*, 2009;22(6):503-510.
8. Du ZD, Hijazi ZM, Kleinman CS, Silverman NH, Larntz K, Amplatz Investigators. Comparison between transcatheter and surgical closure of secundum atrial septal defect in children and adults: Results of a multicenter nonrandomized trial. *J Am Coll Cardiol*, 2002;39(11):1836-1844.
9. Boyd AC, Cooper M, Thomas L. Segmental atrial function following percutaneous closure of atrial septum using occluder device. *J Am Soc Echocardiogr*, 2009;22(5):508-516.
10. Park JH, Yoon YS, Yeon KM, Han MC, Kim CW, Byung HO, Yung WL. Percutaneous pulmonary valvuloplasty with a double-balloon technique. *Radiology*, 1987;164(3):715-718.
11. Chen CR, Cheng TO, Huang T, Zhou YL, Chen JY, Huang YG, Li HJ. Percutaneous balloon valvuloplasty for pulmonic stenosis in adolescents and adults. *N Engl J Med*, 1996;335:21-25.
12. "CCS Consensus Conference 2001 update: Recommendations for the Management of Adults with Congenital Heart Disease." The Nevil Thomas Adult Congenital Heart Library. <<http://www.achd-online.com/consensus/sec7.html>> (March 5, 2010).

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