Atrial Septal Defect–Associated Pulmonary Hypertension: Outcomes of Closure With a Fenestrated Device

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Unlike other intracardiac shunts, there is no known linear relationship between ostium secundum atrial septal defects (ASD) and development of pulmonary hypertension (PH). PH is observed in 8% to 10% of all ASD patients. Atrial septal defect–associated pulmonary hypertension (ASDAPH) is usually independent of the degree, duration of shunting, and defect size. Complete closure of ASD in these patients can be detrimental due to the potential risk for increase in pulmonary vascular resistance (PVR). Fenestrated closure allows for controlled residual shunt providing adequate cardiac output with a mechanism for decompression in the event of critical increase in PVR. After approval from institutional review boards and agencies protecting human subjects, 42 patients from 29 international centers underwent compassionate use of the Occlutech® Fenestrated Atrial Septal Defect (FASD) device. Physician implanters reported outcomes via electronic survey. Follow-up data were available for 25 patients (72% female, n=18) from 18 centers. Symptomatic improvement was observed in a majority of the patients with reduction in New York Heart Association class III symptoms from 68% at baseline to 8% at long-term follow-up. Mean oxygen saturation improved from 93% at baseline to 97% at long-term follow-up (P=0.0066). Reduction in right atrial pressure and mean pulmonary arterial pressure were also noted. During follow-up, one patient had spontaneous occlusion of the fenestration requiring emergency stenting. No other major complications were observed. FASD implantation improves outcomes in patients with ASDAPH; however, further studies are required in a large cohort of patients to determine timing of intervention, optimal fenestration size, and long-term prognosis.

Key Words—atrial septal defect, atrial septal defect–associated pulmonary hypertension, congenital heart disease, fenestrated defect closure, interventional cardiology, pulmonary arterial hypertension

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Atrial septal defect (ASD) is a common congenital heart defect (CHD) with a worldwide incidence of 1.64 per 1,000 live births.\textsuperscript{1} Ostium secundum ASD is the most prevalent type accounting for 70\% to 80\% of all cases. The absence of major clinical symptoms and physical findings can lead to delayed diagnosis; as such, ASDs are often diagnosed in adulthood.\textsuperscript{2} Approximately 8\% to 10\% of patients with ASD can develop pulmonary hypertension (PH), a severe chronic condition with progressive increase in right ventricular (RV) pressure and pulmonary vascular resistance (PVR), associated with secondary right heart failure, and high mortality rates.\textsuperscript{3} Patients with untreated CHD have higher risk for developing PH.\textsuperscript{3} CHDs characterized by high pulmonary pressures and/or high pulmonary flow states are more commonly known to develop reversal of shunting and cyanosis known as Eisenmenger’s syndrome.\textsuperscript{4} However, there is no known linear relationship between PH and ASDs and the pathogenic mechanism is unknown.\textsuperscript{5}

A subset of patients with ASD may develop severe PH at an earlier age. This atrial septal defect–associated pulmonary hypertension (ASDAPH) is more common in females accounting for around 85\% of all observed cases. ASDAPH often presents in the second or third decade of life and is typically independent of the degree of shunting.\textsuperscript{6,7} ASDAPH has better prognosis than idopathic PH possibly due to right-to-left shunting preventing cardiovascular collapse and syncope during critical increase of PVR.\textsuperscript{8} However, outcomes on medical therapy in ASDAPH are unreliable with worsening of symptoms in the majority of patients despite multiple drug therapy.\textsuperscript{8}

Although transcatheter or surgical repair of ASDs with normal mean pulmonary arterial pressure (MPAP) is considered safe and widely accepted procedure with negligible mortality, there are no specific techniques or established guidelines for the treatment of ASDAPH.\textsuperscript{9,10} Complete closure of the defect in ASDAPH by transcatheter or surgical approach may result in complications associated with sudden increase in PVR.\textsuperscript{11-14} The concept of fenestrated closure is to keep a residual shunt, which acts as a decompression mechanism, and maintains adequate cardiac output in the event of critical increase in PVR.\textsuperscript{13-15} Various improvisations in transcatheter and surgical closure have been attempted without significant success.\textsuperscript{16,17} This led to the development of a novel transcatheter closure device, the Occlutech\textsuperscript{®} Fenestrated Atrial Septal Defect (FASD) device.\textsuperscript{18,19} We report outcomes of FASD device implantation in ASDAPH.

**MATERIALS AND METHODS**

Patients with ASDAPH refractory to medical therapy underwent compassionate use of FASD device approved by institutional review boards and agencies protecting human subjects. The outcomes were reported via electronic survey by physician implanters from 18 centers. Patients with moderate to large ASDs with adequate margins were considered suitable for implantation.

**Device Deployment**

Pre-deployment transthoracic echocardiogram (TTE) and transesophageal echocardiogram (TEE) were performed to confirm the position of the defect and its characteristics. The Occlutech\textsuperscript{®} FASD device consists of a nitinol (51\% nickel, 49\% titanium) wire mesh (Figure 1). A flexible waist with a fenestration connects the 2 retention discs corresponding to the size of the ASD, which completely conforms to the atrial septum after deployment. Two very thin polyethylene terephthalate patches ensure faster sealing of the ASD while optimizing endothelialization for a sustainable atrial communication. The devices are available in 3 fenestration sizes with a proprietary delivery system based on the device diameter (Table 1).

![Figure 1: The Occlutech® Fenestrated Atrial Septal Defect device.](http://meridian.allenpress.com/aph/article-pdf/18/1/4/2456311/1933-088x-18_1_4.pdf)

| Fenestration size (mm) | Device                  | Delivery system (F) |
|------------------------|-------------------------|---------------------|
| 5                      | 25ASD15F, 25ASD18F, 25ASD21F | 11                  |
| 6                      | 25ASD24F, 25ASD27F, 25ASD30F | 12                  |
| 8                      | 25ASD33F, 25ASD36F, 25ASD40F | 14                  |
|                        | 25ASD44F, 25ASD48F       | 16                  |

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to confirm the anatomy and margins of the ASD. The appropriate device and fenestration size for individual patients were determined based on the pre-deployment cardiac catheterization data including balloon sizing. The patients underwent transcatheter FASD implantation via femoral vein access under fluoroscopic and TEE guidance (Figure 2).

**Patient Follow-up**
Follow-up visits were defined as short-term if they occurred between 1 and 6 months and long-term if they occurred between 6 and 12 months after the procedure. Clinical parameters (New York Heart Association [NYHA] functional class, 6-minute walk test [6MWT], and oxygen saturation), echocardiographic outcomes (device position, patency, and direction of shunting), and catheterization data were collected during follow-up.

**Statistical Analysis**
Data were analyzed using Statistical Analysis Software (Version 9.4, SAS Institute, Inc., Cary, NC). Numerical data were expressed as mean and standard deviation or median and range. A paired t-test and ANOVA were performed to compare pre- and post-deployment changes. A P value of <0.05 was considered to be significant for differences between parameters.

**RESULTS**

**Patient Characteristics**
Forty-two patients underwent transcatheter FASD implantation at 29 international centers. Complete pre- and post-procedural data were available for 25 patients from 18 centers (Table 2). The majority of patients were adults (84%, n=21/25) with median age of 60 years (range 5 to 80 years). Female preponderance was noted (72%, n=18/25). Based on NYHA class, patients were considered severely symptomatic if they had either NYHA class III or IV symptoms; 68% of patients had NYHA class III symptoms (n=17/25), the remaining 32% (n=8/25) had class II symptoms. 6MWT was used to assess the aerobic capacity and endurance of the patients. Severely symptomatic and bedridden patients (n=4) who could not complete the 6MWT were not included in the statistical analysis. The mean baseline 6MWT distance was 228±183 meters for 14 patients who completed the test. The mean oxygen saturation prior to implantation was 93±4% (n=25) ranging from 88% to 100%.

On pre-deployment TTE and TEE, patients were noted to have ASDs ranging in sizes from 15 to 40 mm. It was observed that 63% of patients (n=12/19) had bidirectional or right-to-left shunting across the fenestration. Baseline mean left atrial (LA) and right atrial (RA) pressures were noted to be 13±4 mm Hg and 12±3 mm Hg, respectively. The MPAP was 46±19 mm Hg. All patients were receiving anti-PH medications, with 20% on dual drug therapy (Table 2). All patients were on anticoagulants and antiplatelet agents alone or combination therapy for variable duration (range 6 weeks to 6 months) depending on the individual institutional protocols.

**Deployment**
All patients (n=25) underwent successful implantation on the first attempt. No deployment-related complications were reported in any of the patients. The im-

**Figure 2:** Post-deployment 3D TEE shows a well-seated FASD device in the interatrial septum with a patent fenestration.

| Table 2. Patient Demographics |
|--------------------------------|
| Age (years) | 51 (5-80) |
| Gender (female) | 18/25 (72.0%) |
| Height (cm) | 155±19 |
| Weight (kg) | 70±24 |
| Systemic systolic pressure (mm Hg) | 122±16 |
| Systemic diastolic pressure (mm Hg) | 68±9 |
| Mean arterial pressure (mm Hg) | 86±9 |
| Procedure time (minutes) | 76±44 |
| Fluoroscopy time (minutes) | 13±11 |

**PH medical therapy**
- None | 1/25 (4.0%) |
- Dual | 5/25 (20.0%) |
- Triple | 19/25 (76.0%) |

| Implanted AFR device fenestration size |
|---------------------------------------|
| 5 mm | 5/23 (21.7%) |
| 6 mm | 13/23 (56.5%) |
| 8 mm | 5/23 (21.7%) |
planted FASD devices had fenestration sizes of 5 mm (n=5), 6 mm (n=13), and 8 mm (n=5) with device size ranging from 15 to 40 mm. The mean procedural time was 76 minutes with mean fluoroscopy time of 13 minutes (range 6 to 34 minutes). Immediate post-deployment TEE confirmed optimal device position and patency of the fenestration in all the patients.

**Follow-up**

Of the 25 patients included in the study, 23 completed short-term follow-up, and 12 completed long-term follow-up (Table 3). Significant symptomatic improvement was observed in the majority of patients following FASD deployment. NYHA class III symptoms were observed in 13% of patients (n=3/23) at short-term follow-up and in 8% (n=1/12) at long-term follow-up as compared to 68% patients prior to FASD device implantation. One patient who had atrial fibrillation and was being medically treated experienced symptomatic improvement and was found to be in sinus rhythm on follow-up electrocardiogram. The mean oxygen saturation improved from baseline of 93±4% to 96±3% (n=22, P<0.0001) at short-term follow-up and 97±2% at long-term follow-up (n=12, P=0.0066). There was significant improvement in the endurance level of patients evident from improvement in 6MWT distance from 228±183 meters to 351±179 meters at short-term follow-up (n=12, P=0.0081), and 457±83 meters at the long-term follow-up (n=5). Follow-up TTE demonstrated optimal position and patent fenestration in all patients at short-term follow-up. The TTE also demonstrated RV remodeling and it was observed that 78% patients (n=7/9) had left-to-right shunting across the fenestration with obvious decrease in RV size.

As cardiac catheterizations were performed in only 5 patients at long-term follow-up, the data were not suitable for statistical analysis and comparison with baseline data. Instead the immediate post-deployment catheterization data (n=17) were used for comparison and calculation of the P value. Cardiac catheterization showed expected, but trivial decrease in RA pressure from 12±3 to 10±3 mm Hg (n=17, P=0.0502) immediately after deployment, and a substantial decline to 8±4 mm Hg (n=5) at long-term follow-up. The LA pressure remained unchanged at 14±4 mm Hg (n=16, P=0.9252) in the immediate post-deployment assessment and decreased from 13±4 to 11±1 mm Hg at long-term follow-up (n=4). A significant decrease in mPAP was noted from 46±19 to 42±17 mm Hg immediately after deployment (n=17, P=0.0004) and to 42±22 mm Hg at long-term follow-up (n=5). At short-term follow-up, all patients continued anti-PH medication therapy. At long-term follow-up one patient had the fenestration closed with an ASD occluder device in view of symptomatic improvement, normalized mPAP, and hemodynamics. Post-implantation symptomatic improvement and normalization of the rhythm was observed in one patient with atrial fibrillation.

**Complications**

All patients had uneventful device deployment with no deployment-related complications. One patient developed pseudoaneurysm with an arteriovenous fistula at the catheterization site requiring thrombin injections. No device-related complications were observed except in one patient on aspirin therapy who was observed to have worsening of symptoms on follow-up and had the 24 mm FASD device with 6 mm fenestration. On subsequent evaluation, TTE revealed complete occlusion of the fenestration. The patient underwent emergency re-catheterization and stenting of the fenestration resulting in improved hemodynamics.

**DISCUSSION**

PH often develops in CHD as a result of prolonged left-to-right shunting and may ultimately be associated with a fixed increase in PVR, which may render some patients inoperable.3 The risk of developing Eisenmenger’s syndrome is

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**Table 3. Change in Parameters Following FASD Implantation**

| Parameters                  | Baseline (mean ± SD) | Short-term follow-up (mean ± SD) | Long-term follow-up (mean ± SD) | P value |
|-----------------------------|----------------------|---------------------------------|---------------------------------|---------|
| **6MWT distance (m)**       | 228±183 (n=14)       | 351±179 (n=12)                  | 457±83 (n=5)                    | 0.0081  |
| **NYHA functional class**   |                      |                                 |                                 |         |
| I                           | 0 (0%)               | 7 (30.43%)                      | 8 (66.7%)                       |         |
| II                          | 8 (32.0%)            | 13 (56.5%)                      | 3 (25.0%)                       |         |
| III                         | 17 (68.0%)           | 3 (13.0%)                       | 1 (8.3%)                        |         |
| IV                          | 0 (0%)               | 0 (0%)                          | 0 (0%)                          |         |
| **RA pressure (mm Hg)**     | 12±3 (n=24)          | 10±3 (n=17)                     | 8±4 (n=5)                       | 0.0502  |
| **LA pressure (mm Hg)**     | 13±4 (n=23)          | 14±4 (n=16)                     | 11±1 (n=4)                      | 0.9252  |
| **mPAP (mm Hg)**            | 46±19 (n=24)         | 42±17 (n=17)                    | 42±22 (n=5)                     | 0.0004  |

†Due to low n and high standard deviations for short- and long-term, only descriptive statistics are given. ‡Represents cardiac catheterization data from the immediate post-deployment period.
50% in patients with a large ventricular septal defect, 90% in unrepaired atrioventricular septal defect, and almost 100% in truncus arteriosus. The timing and severity of the development of PH in CHD is directly related to the size and severity of the defect and is influenced by both the pressure and volume overload in the pulmonary arteries.3 However, the pathogenesis of PH with volume overload in ASD is not well explained by this mechanism in all the cases. Although, chronic volume overload in large or moderate-sized ASD is thought to cause irreversible changes to the pulmonary vasculature, most patients with only chronic volume overload do not develop PH. It has also been observed that in some patients with ASD, the progression rate of PH is much faster, and PH develops in childhood.

In our study, we observed there was wide variability in the size of ASDs ranging from 15 to 40 mm without direct correlation between the development of PH and the size of the ASD. Also, it was noted that there was no correlation between the duration of shunting and development of PH, as 16% of patients (n=4) in our study developed PH before the age of 18 years (Figure 3). This suggests a multifactorial cause or etiology for the development of PH in some patients with ASD, with a likely role for unknown genetic factors especially with female preponderance.7 The treatment of ASDAPH is controversial as treatment of PH in these patients with newer disease-modifying drugs leads to lowering of PVR resulting in increased pulmonary blood flow, which itself can lead to development of fixed PVR. The complete closure of ASDs in patients with PH using transcatheter or surgical approach is debatable as there is the potential for a deterioration of the pulmonary vascular disease with an increase in PVR and secondary signs of right heart failure without the positive effect of right-to-left shunting.10

Multiple improvisations have been attempted in surgical treatment of these patients.13-14 In 1959, Charles P. Bailey first demonstrated the use of flap valves made of a compressed ring of polyvinyl sponge for the treatment of atrial and ventricular septal shunts in patients with severe PH. Seven of the 8 patients treated using flap valve survived.13 Cho YH et al reported successful surgical correction of 16 patients of secundum ASD with severe PH using a fenestrated patch.14 Although the outcome of the study was satisfactory, surgical treatment has been associated with significant morbidity and mortality. There is possibility of bleeding and arrhythmia in patients with risk for development of moderate to severe PH. Patients who develop PH immediately or several months or years after ASD closure have poorer prognosis.12

Interventional ASD closure in patients with normal pulmonary arterial pressures or mild to moderate PH is now widely practiced and has replaced surgical ASD closure in many centers. Partial defect closure with a fenestrated device limits the left-to-right shunt in general and allows a right-to-left shunt as a decompression mechanism during episodes of transient rises of PVR while maintaining cardiac output. As such, a fenestrated ASD closure is the only available interventional treatment option in patients with ASD and moderate to severe PH. Due to unavailability of devices with fenestrations various modifications were tried in the existing devices. As reported by Kretschmar et al, two children were treated using the standard Amplatzer devices with fenestrations made using sheath dilators and balloon stretching. These fenestrations closed spontaneously 4 months after implantation.15 Skinner et al reported closure of large ASD with severe PH using an Amplatzer device with artificially created fenestration and maintaining the patency of the created defect using a 5 mm bare metal coronary stent. In this case the patency was maintained for 39 months after the procedure.16 Creating handmade fenestrations can be risky: large fenestrations can lead to excessive shunting, desaturation, and death whereas small-sized fenestrations are at risk for spontaneous occlusion. The difficulties in creating and maintaining a fixed size, stable fenestration in the devices for ASD closure in ASDAPH led to development of the FASD device.17,18 Successful use of FASD device in children with ASDAPH has previously been reported by Gonzalez-Barlatay et al.19

Figure 3: Pre- and post-deployment changes in the mean pulmonary artery pressure in individual patients with ASDAPH.
The implantation of the FASD device is technically identical to standard ASD closure devices. In our study, FASD implantation in all 25 patients was performed successfully at first attempt without any deployment-related complications. TTE demonstrated fenestration patency in all patients on post-implantation day 1. During follow-up, patients did not have any major complications like cardiac perforation, pericardial effusion, infective endocarditis, device erosion, embolization, and thrombus formation. No center reported any occurrence of new arrhythmias or deaths after device implantation. Spontaneous closure can occur in any fenestrated device; however, the frequency of fenestration occlusion was lower with the FASD device when compared to devices with handmade fenestrations. The occlusion of fenestration observed in one of our patients was after 6 months of implantation, which is a longer period of time than that reported with handmade fenestrations.

Symptomatic improvement was noted in most patients with improvement in NYHA functional class and improved quality of life. The symptomatic improvement may be explained by decreased left-to-right shunting, which results in normalization of RA size and pressure. The decreased RA size and pressure in the post-procedural period may help in reducing the precipitating factors for arrhythmia and make refractory arrhythmias amenable to medical treatment as was observed in one of our patients with atrial fibrillation. The decreased right-sided pressures can lead to RV remodeling, which results in improved hemodynamics. Moreover, the presence of the patent fenestration acts as a popoff valve and allows for shunting of blood from right to left in case of rise in PVR in the immediate post-deployment period and helps maintain cardiac output and oxygenation.

Limitations of Study
The retrospective nature of the study, lack of comparison to a standard treatment modality, follow-up protocol, and the limited number of patients were major limitations of our study. The variation in the techniques and protocols for management of these patients based on the clinical expertise of the physician implanters, the facilities available at different centers, and the severity of comorbidities may have also influenced outcomes in this series.

CONCLUSION
ASD closure in patients with severe PH using the FASD device was beneficial for patients without any major device-related complications. The fenestrated device restricts left-to-right shunting but allows for decompression of the right heart during pulmonary hypertensive crisis. Meticulous care toward patient selection, adequate defect sizing, as well as device and fenestration size is required for optimal outcomes. Further prospective studies are required in a large cohort of patients to determine the timing of intervention, appropriate fenestration size, and long-term benefits of the FASD device in ASDAPH.

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