

An Aneurysmal Interatrial Septum with a Patent Foramen Ovale and Multiple Fenestrations Closed by A Single Occluder Device

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Abstract- The patent foramen ovale (PFO) usually is a very small potential opening in the atrial septum. Under the conditions of normal hemodynamics with higher left atrial than right atrial pressures, the septum primum is forced against the foramen by the higher left atrial pressure and there is no actual persistent opening through the foramen. However, with any, even transient, increase in right atrial pressure this flap or “valve” can be pushed away from the septum and forced open. This results in the shunting of blood and anything else in the right atrium from the right atrium to the left atrium. Often the “valve” of the foramen becomes redundant and develops an “aneurysm” of the atrial septum. A large, redundant septum primum can have several additional openings or “fenestrations” in it. The PFO is now can be treated by interventional percutaneous therapy. This case represents a 24-year-old male with an aneurysmal interatrial septum and patent foramen ovale associated with multiple fenestrations. The defects were closed by a single *Amplatzer® septal occluder*.

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Introduction

The patent foramen ovale (PFO) usually is a very small potential opening in the atrial septum where the septum primum has not completely sealed the embryonic reopening of the ostium secundum in the septum secundum. In a PFO, the septum primum tissue, which constitutes the “valve of the foramen” is sufficient in size to cover the entire ostium secundum, but at the same time has not sealed the opening on the left side of the septum.

Under the conditions of normal hemodynamics with higher left atrial than right atrial pressures, the septum primum is forced against the foramen by the higher left atrial pressure and there is no actual persistent opening through the foramen. However, with any, even transient, increase in right atrial pressure this flap or “valve” can be pushed away from the septum and forced open. This results in the shunting of blood and anything else in the right atrium from the right atrium to the left atrium. For example, if a clot forms in the right atrium, its passage to the left atrium through the PFO can lead to the

embolic events in the systemic circulation, such as coronary, cerebral, or any other arteries in the body. This phenomenon is known as the paradoxical emboli.

Often the “valve” of the foramen becomes redundant and develops an “aneurysm” of the atrial septum. A large, redundant septum primum can have several additional openings or “fenestrations” in it.

Case Report

A 24-year-old Mediterranean male presented to the emergency department with typical angina chest pain. He had no history of previous cardiac problem. At presentation his electrocardiogram showed right bundle branch block associated with ST elevation in V1-V4. Serum levels of cardiac biomarkers were requested which showed elevated troponin I level. Patient was admitted and underwent percutaneous coronary angiography which demonstrated normal coronary arteries without any lesion or stenosis. The diagnosis of paradoxical emboli was made for the patient with assuming the right ventricle as the source of emboli and

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the interatrial septum defect as the predisposing factor. Echocardiographic evaluation revealed mild to moderate left ventricular enlargement with moderate systolic dysfunction (LVEF: 35%). Left ventricular diastolic function was normal. Wall motion abnormality was noted at anterior circulation with large left ventricular apical organized clot. Aortic valve was bicuspid, with no stenosis but mild insufficiency. Pulmonary artery pressure was in the normal range. Interatrial septum seemed aneurysmal (Figure 1).

In contrast study passage of bubbles into the left atrium was conspicuous. For further evaluation transesophageal echocardiography (TEE) was performed.



Figure 1. Transesophageal echocardiography. Aneurysmal interatrial septum is shown with arrow. Arrowheads indicate fenestrations.

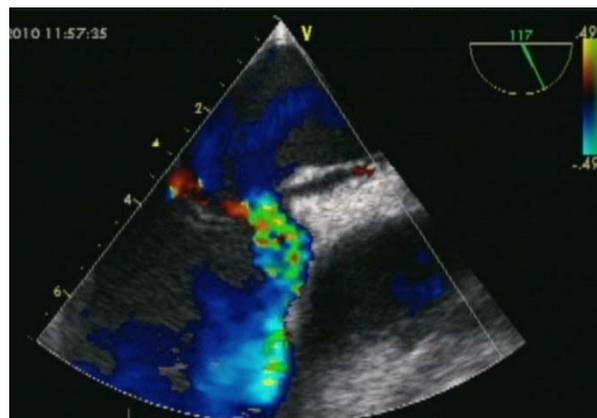


Figure 2. Transesophageal echocardiography shows fenestrations of interatrial septum. Note the turbulent flow across the fenestrations.

In TEE a fenestrated septum was seen with three separate holes, a patent foramen oval with an elongated flap valve and two smaller-sized fenestrations (Figures 2, 3).

The size of the patent foramen oval and the fenestrations were 6, 3 and 2mm, respectively. The distance between the PFO and the first fenestration was about 2mm and the second defect lied 9mm distant from the first one. The defect was closed by a single *Amplatzer® septal occluder* (Figures 4, 5). The size of right and left atrial discs of the device were 35 and 25mm, respectively.

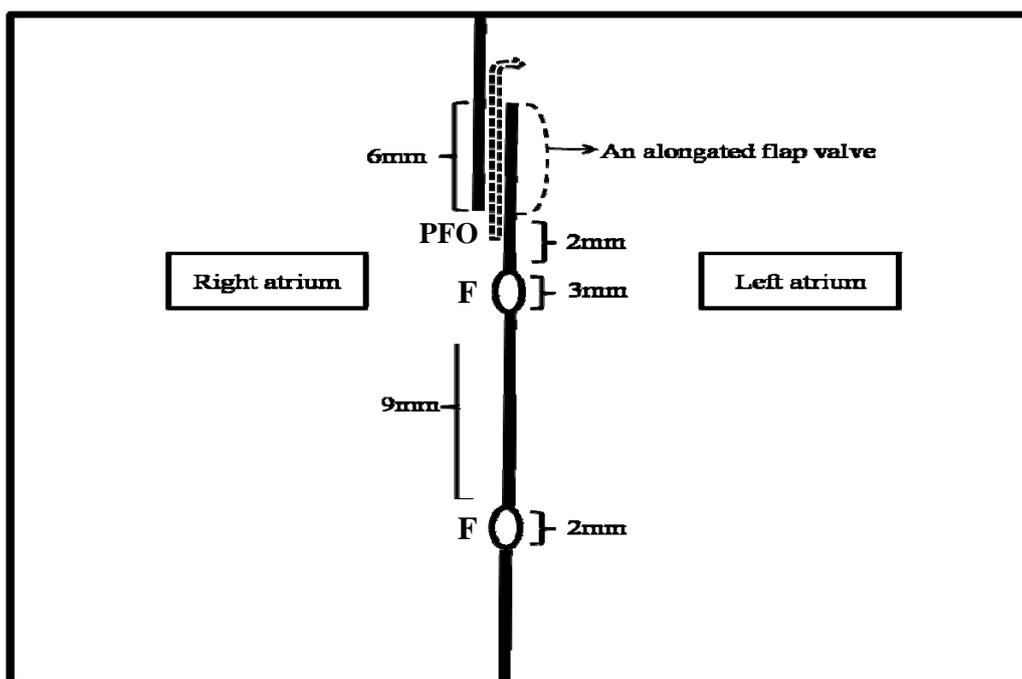


Figure 3. A schematic view of the patient's interatrial septum. The dashed arrow shows passage of blood from the right atrium to the left atrium through the patent foramen ovale. F; fenestration, PFO; patent foramen ovale.

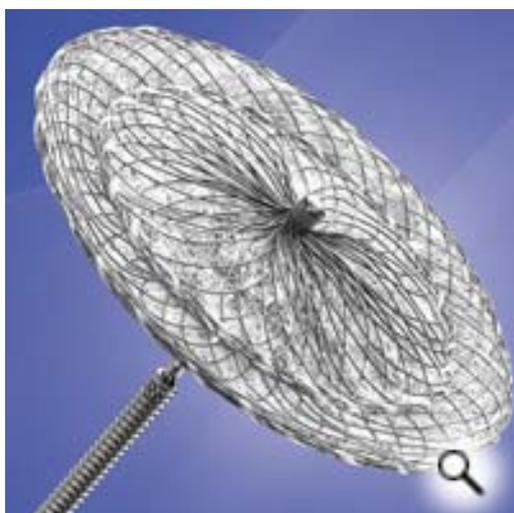


Figure 4. AMPLATZER® PFO Occluder. Each Occluder is made of a Nitinol wire mesh that is shaped into two flat discs and a middle, or "waist" to pull the discs up to the septum wall. Polyester fabric inserts help close the hole and provide a foundation for growth of tissue over the occluder after placement.

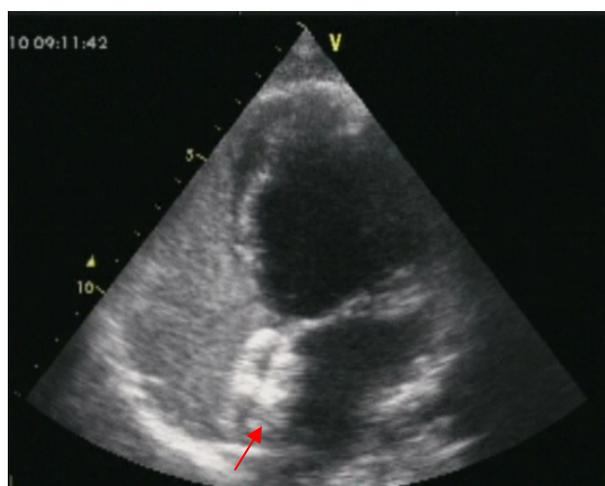


Figure 5. 4-chamber apical view of 2D transthoracic echocardiography after PFO closure. PFO occluder is shown (arrow). No passage of bubbles to the right atrium is seen on contrast study with agitated saline.

Discussion

Patent foramen ovale (PFO) is a common congenital cardiac lesion that persists into adulthood (1, 2). In most of the cases it may be asymptomatic and needs no further evaluation. However, it is a risk factor for several

serious clinical syndromes, including paradoxical systemic embolism, such as decompression sickness (3-5), ischemic stroke (6), myocardial infarction (7) and complications of pulmonary thromboembolism (8). As a matter of fact, in these situations attempts should be done to close this interatrial septum defect

The PFO is now can be treated by interventional percutaneous therapy (9), and numerous new technologies are either available or under development for this lesion closure. Percutaneous PFO closure, first performed in 1989, is now possible with any of 5-7 different devices depending upon availability during various phases of investigational development (Figure 1).

The initial and intermediate-term results of transcatheter closure of ASDs are satisfactory with most devices. However, device deformities, residual shunt, distal migration, and failure of deployment have been reported, mostly with older versions of these devices. Serious complications, including perforation of atria, atrioventricular valve damage, and mortalities, have been reported with larger devices, which are often needed for fenestrated atrial septal defects (F-ASD). Although the indications for the procedure are the same as for surgical closure, the selection criteria are stricter in terms of defect size and surrounding rim tissue. The presence of a sufficient rim of atrial tissue (at least 5 mm) surrounding the defect is considered one of the most important criteria for catheter closure with insufficient rims.

Amplatzer® septal occluder (ASO)

The Amplatzer® septal occluder (ASO) is a self-expanding double-disc centring device constructed of nitinol wire mesh which conforms to septal wall. The two discs are linked together by a short connecting waist corresponding to the size of the ASD. The discs and waist are filled with polyester fabric sewn with polyester thread. The nitinol and polyester construction of the PFO occluder allows rapid closure success and enhanced tissue ingrowth. The ASOs are available in four sizes to properly fit the occluder to PFO size. The right atrial disc sizes include 18, 25, 30 and 35 mm and the left atrial disc sizes are 18, 18, 30 and 25mm, respectively. The delivery system is not too large, and the device can be delivered through an 8 to 9 Fr long sheath. The ASO can be used for closing even very large defects, is self-

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centering, retrievable, and repositionable. The placement of the device can be done relatively simple with recapturable delivery. The residual shunt and complication rates are very low.

Although the increasing experience allows patients with unusual septal anatomic configurations or physiopathologic characteristics to be evaluated for percutaneous closure, large-diameter defects, multiple defects or fenestrated atrial septal defects and interatrial aneurysm with fenestrations may still pose a challenge to device closure in a significant proportion of patients.

Reported Solutions for F-ASDs

Percutaneous closure of fenestrated atrial septal defects using occluder devices has become a popular, effective, and safe method of treatment of in spite of years of successful surgical experience. The suggested techniques for percutaneous closure of F-ASDs can be categorized into three methods:

The first is percutaneous placement of two Amplatzer® septal occluder devices. The smaller ASD device which is deployed first may be partially overlapped by the larger. The drawback of this technique is that there are no data on endothelialization of two devices when overlapping. The second method is placement of a single device after performing a balloon atrial septostomy. This seems very hazardous and creates a larger hole which is not regularly elliptical in shape, and thus balloon sizing becomes unpredictable (over- and under-sizing). The third method was reported by Szkutnik *et al.* They believed in a fenestrated ASD that the distance of two defects is about 7mm; it is enough to close only the larger opening. They believed although residual leaks maybe observed but “they will resolve by the time”.

In this case, there were three defects with conspicuous distance from each which posed a challenge to close all defects with a single occluder device. As described before, the total area of the interatrial septum which should be covered by the occluder was a 22mm defect, creating a sufficient rim should be added to this measurement. In fact, the device might have closed only one defect with the other two remaining open because the convexity of the aneurysmal membrane might not have allowed the retention skirt to cover other defects. On the other hand, the mobility and shape of the septum seemed to exclude the possibility to deploy two devices or three devices. They would have overlapped each other and likely would have created a deformed

interatrial septum. Interestingly, despite the presence of mentioned problems we succeed to close all of them with a single device without performing any further procedure such as septostomy.

Described challenging aspects of this intervention encouraged us to introduce the performed procedure in this case as a novel technique that may have only a few similar reported interventions.

In summary, this case represents a further expansion of technologic attempts to improve the results and simplify the procedure of device closure of patent foramen oval in the presence of a multiple fenestrated interatrial septum.

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