Congenital Heart Disease

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ARTICLE

The ASD that Wouldn't Go Away: An Unusual Case of ASD Device Failure in a Patient with Marfan Syndrome

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ABSTRACT

Marfan syndrome patients have connective tissue abnormalities that predispose them to intracardiac defects and postoperative complications. We present a case of late onset ASD device failure secondary to device movement within the atrial septum in a girl with Marfan syndrome. This case study suggests that further studies are necessary to determine the optimal device and approach for ASD repair in this patient cohort.

KEYWORDS

Congenital heart disease; atrial septal defect; device closure; connective tissue disease; marfan syndrome; transcatheter intervention

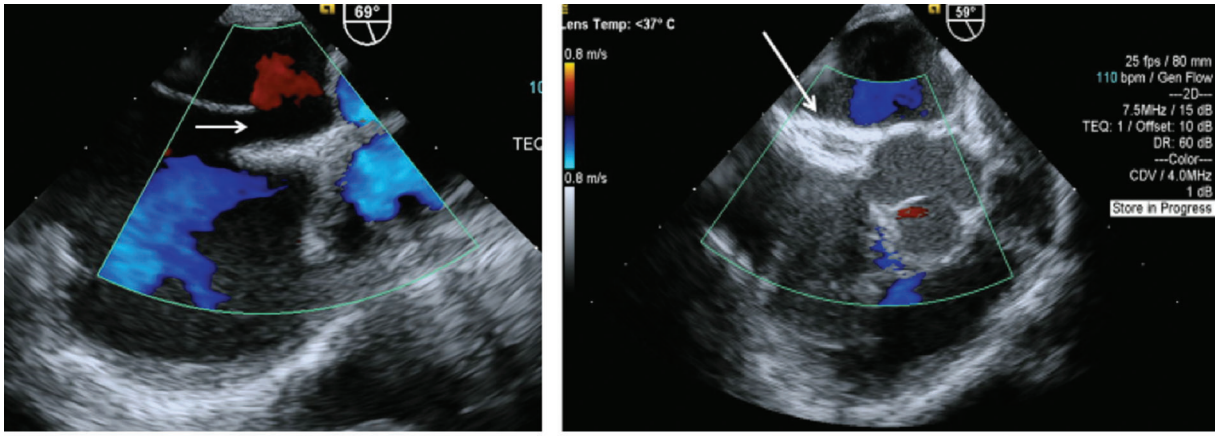
1 Introduction

Patients diagnosed with Marfan syndrome have an increased prevalence of structural cardiac lesions compared to the general populace, including, but not limited to, aortic root dilation, pulmonary artery dilation, mitral regurgitation, coronary artery aneurysm, and atrial septal defects (ASDs) [1,2]. Additionally, pediatric patients suffering from Marfan syndrome have higher rates of postoperative complications after cardiac procedures compared to the general populace [3,4]. We describe a pediatric patient with Marfan syndrome and small to moderate secundum ASD who underwent successful transcatheter ASD device closure with a GORE CARDIOFORM Septal Occluder (GSO), which is a non-self-centering device. Eleven months later, the device was found to have detached from the superior and anterior region of the septum requiring surgical removal of device and ASD patch closure.

2 Case Report

A five-year old female was diagnosed with Marfan syndrome at the age of 2 years and 10 months after fulfilling the Ghent criteria with aortic root dilation and history of ectopia lentis. She was also noted to have multiple characteristic physical exam findings and height >95th% for age. Subsequent genetic panel revealed a known pathogenic mutation in FBN1. Her father, who was incarcerated at the time, was known by foster parents to be of height 6'6"; little other family history is known.

A small to moderate ostium secundum atrial septal defect (ASD) with left to right shunting was noted on initial transthoracic echocardiogram (TTE). There was no evidence of mitral valve prolapse or mitral regurgitation and her aortic root was mildly dilated at 2.30 cm (z-score 2.9). She developed progressive right ventricular dilation and was referred for transcatheter ASD device closure at 5 years of age. Transesophageal echocardiogram (TEE) was utilized to guide the closure which was performed under general anesthesia. Right and left heart hemodynamics were normal with a small step-up in saturations from the left to right shunt. The defect measured 7 mm with inferior and superior vena caval rims of 10 mm each, a mildly deficient retro-aortic rim of 4.5 mm, total septal length of 29 mm, and mild misalignment of atrial septum (Fig. 1a). Due to concerns about potential erosion in the setting of a connective tissue disorder and underlying aortic root dilation with a mildly deficient retro-aortic rim, a non-self-centering 25 mm GORE CARDIOFORM Septal Occluder was successfully placed without complication (Fig. 1b). Of note, balloon sizing was not performed prior to device placement.



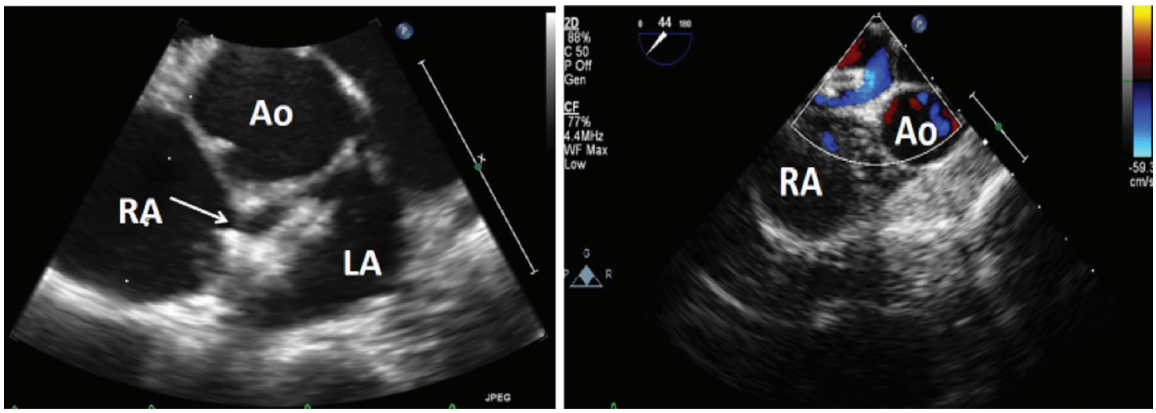
(a) (b)

Figure 1: Transesophageal echocardiogram demonstrating malaligned septum with secundum atrial septal defect (arrow) measuring 7 mm (a). Transesophageal echocardiogram showing device closure with 25 mm GORE CARDIOFORM Septal Occluder (arrow) (b). Note device position relative to aorta and absence of shunting by color flow imaging

Echocardiogram the following day confirmed proper placement of the device and no residual shunt. A repeat study five months later also demonstrated proper placement. Eleven months after device closure, TTE demonstrated that the superior portion of the device was no longer attached to the superior vena cava and a new 7 mm shunt was noted there (Fig. 2a). Transesophageal echocardiogram confirmed that the superior and anterior edges of the device had separated from the superior vena cava and retro-aortic rims with a small to moderate residual shunt around the edges of the device (Fig. 2b). Due to concern for further device movement, she was referred for surgical removal and ASD closure.

Intraoperative inspection of the interatrial septum revealed the GSO was well incorporated along its inferior rim with complete endothelialization of the entire device (Fig. 3). There was a significant residual defect along the superior edge just under the superior vena cava orifice, where the right atrial disc of the device had prolapsed into the left atrium. The device was carefully freed from its endocardial attachments and removed. In addition, there was an area of scarring on the posterior wall of the left atrium, likely representing an area of contact with the portion of the device that prolapsed through the defect. Autologous pericardium was harvested and used to close the ASD. Recovery was uncomplicated with post-repair TEE revealing no residual defect and she was discharged home on post-operative day two.

Four months after surgery, the patient remained well and TTE demonstrated normalized right heart size without residual defect.



(a) (b)

Figure 2: Transthoracic echocardiogram parasternal short axis view 11 months after 25 mm GORE CARDIOFORM Septal Occluder implantation (a). Note that the device has prolapsed into the left atrium and a residual defect (arrow) is now apparent. Transesophageal echocardiogram at time of surgical device removal and ASD patch closure (b). Note misalignment of device with septum and aortic root and residual left to right shunting as demonstrated by blue flow. Ao = aorta; ASD = atrial septal defect; LA = left atrium; RA = right atrium

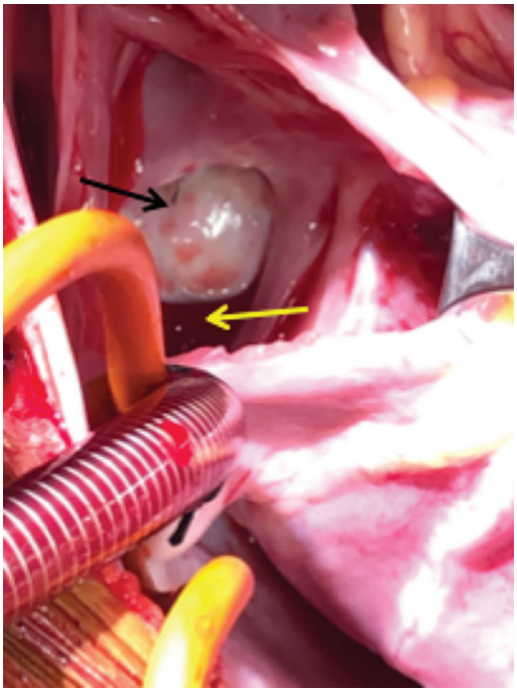


Figure 3: Intraoperative view of atrial septum from right atrial view. Note complete endothelialization of the device (black arrow), which is displacement into the left atrium, as well as the residual atrial septal defect (yellow arrow)

3 Discussion

We present a case of late onset ASD device movement within the atrial septum in a girl with Marfan syndrome. Although initially well positioned within the defect, over time a portion of the device detached from the septum leading to shunting and concerns for stability, which prompted surgical removal and patch closure of the ASD. To the best of our knowledge and after review of the literature, this is the first reported case of late device movement with the GORE CARDIOFORM Septal Occluder in a patient with Marfan syndrome. We hypothesize that while the device was initially well placed as evidenced by the complete endothelialization noted at the time of surgical removal, the underlying tissue characteristics and septal malalignment eventually led to the superior portion of the device detaching from the septum.

In 2001, the Amplatzer Atrial Septal Occluder (ASO) was approved by the FDA for trans-catheter repair of ASDs and has since been used widely for this purpose. While extremely effective, the device carries a small risk of potential erosion (0.1% to 0.3%) that remains poorly understood [5]. Reported in the literature are two such cases involving Marfan syndrome patients, one of a pediatric patient with perforation of the occluder through the left atrial wall and the other of an adult male experiencing cardiac tamponade due to perforation secondary to device erosion [6,7]. The pediatric patient, a nine-year old boy with mild aortic root enlargement, mitral regurgitation and prolapse, and secundum ASD received a 24 mm ASO. Transesophageal echocardiogram at the time of closure noted a smaller retroaortic rim (3–4 mm) but sufficient margins for placement of the device. The aortic root continued to dilate and two years later he underwent elective aortic root repair. Intraoperative inspection demonstrated penetration of the left disc of the ASO into the non-coronary sinus of the aortic root with subsequent aneurysmatic ballooning which was repaired at the time of surgery [6].

Marfan syndrome patients have connective tissue abnormalities that predispose them to intracardiac defects as well as *de novo* and postoperative aortic dissection and rupture [1–4]. This substrate could be more susceptible to breaches in tissue integrity and may represent a hostile environment for more rigid ASD devices. Additionally, devices within the atrial septum move throughout the cardiac cycle, potentially creating more shear forces on the underlying abnormal tissue.6 In our case, the septal malalignment may have prevented the device from naturally conforming within the plane of the defect, thereby exerting continuous external force on the device and tissue which eventually gave way.

While we considered using an ASO for this defect, we opted to use a GSO given the above case reports. The GSO received FDA approval for closure of small to moderate secundum ASDs up to 17 mm in diameter in 2015. While other ASD closure devices have a defined waist embedded with fabric to promote closure, the GSO used is a non-self-centering device and relies on device-to-defect ratio to provide tissue apposition and promote endothelialization of the device. Because of the devices' ability to move within the defect, it can shift as it conforms to the septal anatomy. We theorize that the newly approved GORE CARDIOFORM ASD Occluder may be a better option for these patients as this device is self-centering with an adaptable waist that conforms to each unique atrial septal anatomy, thereby potentially increasing device stability and reducing device free movement within the septal wall in patients with Marfan syndrome. Although we did not perform balloon sizing in this case, we would strongly recommend this practice in those patients who may have increased tissue mobility, as this information may have led to a different device selection.

4 Conclusion

Tissue abnormalities in Marfan syndrome patients may predispose to device failure after transcatheter ASD closure. Further studies are necessary to determine optimal device and approach for this patient cohort, including whether utilizing pre-placement balloon sizing and/or self-centering devices may minimize the risk of migration events as seen in our patient.

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Conflicts of Interest: Dr. Gordon is a proctor for WL Gore & Associates and Abbott.

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