Update on Transcatheter Device Closure of Congenital Septal Defects

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Abstract

Purpose of Review The goal of this paper is to review currently available devices for closure of atrial septal defects (ASDs) and ventricular septal defects (VSDs).

Recent Findings Favorable results from the ASSURED trial resulted in FDA approval for the most recently developed device for transcatheter ASD closure in the United States. Further studies are required to assist in the development or approval of safe devices for transcatheter perimembranous VSD closure in pediatric patients.

Summary Device closure is the less invasive and preferred management option for many ASDs, with multiple studies demonstrating lower complication rates, shorter hospital stays, and lower mortality than surgical repair. Complex ASDs that make device closure more difficult include large defects, rim deficiencies, fenestrated defects, multiple defects, and the presence of pulmonary arterial hypertension. Device closure has also become an accepted alternative to surgery for some types of ventricular septal defects VSDs, though challenges and limitations remain. Future innovations including novel devices and techniques are needed to further expand on the types of defects that can be safely closed via transcatheter approach.

Keywords Transcatheter intervention · Device closure · Atrial septal defect · Ventricular septal defect

Introduction: Atrial Septal Defects

History and Rise of Transcatheter ASD Closure

Atrial septal defects (ASDs) are one of the most common types of congenital heart disease (CHD), comprising 15% of all CHD with an estimated prevalence of 1.4 per 1000 live births [1]. The majority of defects are ostium secundum ASDs, which occur due to a deficiency of tissue in the area of the fossa ovalis [2]. Other types of ASDs include ostium primum (as part of an atrioventricular septal defect), superior or inferior sinus venosus, and coronary sinus defects (Fig. 1). Although most patients are asymptomatic during childhood and adolescence, hemodynamically significant left-to-right shunting over time can cause right heart dilation and failure, exercise intolerance, atrial arrhythmias, and pulmonary arterial hypertension (PAH) in adulthood [2].

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For patients that meet indication for ASD repair to prevent such sequela, treatment includes surgical and transcatheter options. The first catheter-based intervention for ASD closure was successfully performed by King and Mills, pioneering a new alternative to open heart surgery for ASD management and paving the way for innovation in transcatheter interventions [3].
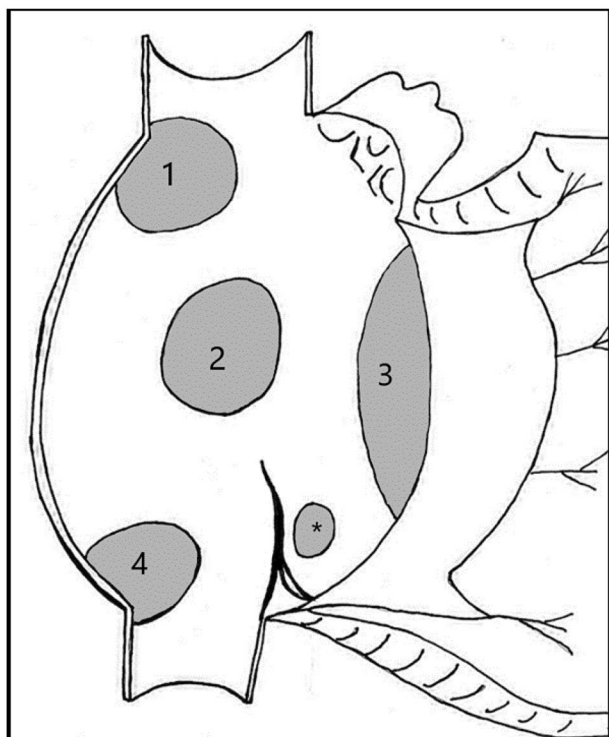
Transcatheter device closure is a less invasive option to surgery and now the preferred method for treatment of secundum ASDs at most institutions, with overall success rates > 95% [4, 5]. Transcatheter ASD closure is safe, with multiple studies demonstrating lower complication rates, shorter hospital stays, and even lower mortality than surgical repair [4, 6]. This shift towards catheter-based interventions became possible due to innovation and significant advancement in the types of devices and delivery techniques over the last few decades. There are now currently four devices from two companies that are available in the United States and approved by the U.S. Food and Drug Administration (FDA) for secundum ASD closure: Amplatzer Septal Occluder, Amplatzer Cribriform Occluder, Gore Cardioform Septal Occluder, and Gore Cardioform ASD Occluder. The primary focus of this review is to discuss currently available devices for transcatheter closure of secundum ASDs and

Fig. 1 Relative anatomic locations of defects within the atrial septum are depicted as follows (1) superior sinus venosus defect, (2) secundum atrial septum defect, (3) primum atrial septal defect, (4) inferior sinus venosus defect. *Os of the coronary sinus*

review techniques used to close complex secundum ASDs. A summary of device features is found in Table 1.

Devices

The Amplatzer Septal Occluder (ASO) (Fig. 2A) was the first device approved by the FDA for ASD closure in 2001. It is a self-expandable and self-centering double-disc occlusion device made of Nitinol mesh designed for the discs to sit in the right and left atria and the waist to rest in the defect. It can close defects up to 38 mm in diameter [5]. Multiple studies have demonstrated long-term safety and efficacy of the ASO. A prospective study including 1000 patients at 50 different centers demonstrated successful closure in 97.9% of patients at 2-year follow-up years. The average static ASD diameter was 14.5 mm, and the average stop flow diameter was 17.6 mm. There was a deficient retro-aortic rim (<5 mm) in 11.5% of cases. The investigated safety and adverse events included additional endpoints of hemodynamic compromise and device erosion. Hemodynamic compromise was rare, occurring 0.7% (7 events in 6 patients). Device erosion as the cause of hemodynamic compromise was also rare, occurring in 3 patients (0.3%) [5]. Erosion through the aortic root or the atrial wall has been associated with deficient

septal rims (especially aortic and superior rims), larger defects relative to the septum and patient size, and oversized and larger devices [5, 7]. Although a serious late complication, device erosion remains rare (from 0.04 to 0.3%) and continues to be monitored [5, 7–9]. Hopefully improving patient selection and newer device options for larger defects will reduce this risk in the future, as well as continued surveillance to help identify risk factors. Despite this complication, advantages of the ASO device include a lower profile delivery sheath compared to the Gore devices and a large variety of techniques described for complex defects.

The Amplatzer Cribriform Occluder (Cribriform ASO) (Fig. 2B) is a non-self-centering device with a thin central waist and large, equally sized atrial discs designed to close fenestrated ASDs with multiple defects. It is available in 18–35 mm (with the size corresponding to the atrial disc diameter). A small, retrospective study has demonstrated that it can be used safely and effectively in pediatric patients to close fenestrated atrial defect defects. Optimal device size is > 1.5 times the fenestrated septal length, but less than the total septal length. Device placement is most effective when a small, central fenestration is crossed and provides for maximal coverage of fenestrations and lower incidence of significant residual shunting [10]. Other strategies are available for closing fenestrated defects using multiple ASO devices to close larger fenestrations that are remote from one another, and other devices discussed separately can also be used [11].

The Gore Cardioform Septal Occluder (GSO) (Fig. 3A) is a non-self-centering double-disc device without a central waist composed of nitinol wire frame covered with polytetrafluoroethylene (ePTFE) that is soft and compliant. It is available in three sizes (20 mm, 25 mm, and 30 mm) with the size describing the diameter of both the left and right atrial discs. It is approved for both patent foramen ovale and secundum ASD closure. Safety and efficacy were demonstrated in a multicenter US trial which prospectively followed 400 patients at 21 sites. The median patient age was 6.9 years, and median defect diameter was 9.7 mm with a stop flow diameter of 12 mm and maximum diameter of 17 mm. Device implantation was performed in 93.5% of patients, and clinical device closure was successful in 98.8% of patients. There were no device embolization, erosions, or reinterventions at 6 months. Overall, it performed well in single and multi-fenestrated defects, and with deficient aortic rim defects, with 23.8% of closures performed in fenestrated defects and 41.8% of closures having deficient aortic rims (<5.0 mm). The size of the device should ideally be at least 1.8 times the stop flow diameter, limiting its use to defects 17 mm or less in diameter. Although it can be stretched to larger defects in some situations, other devices should be considered for larger defects [12]. Wire frame fractures did occur in 6–7% of device closures, though they have not been associated with any clinical sequela [12, 13].

Table 1 Summary of device features for ASD closure devices

| Device	Maximum defect Size Amenable for Closure	Material	Self-centering	Device Size (defect range/ maximum defect size)	Right atria disc size	Left atria disc size	Sheath Size without Wire	Sheath Size with 0.035" Wire	Closure Success at 6-12 months	Embolization Rate	Significant complication (risk factors)
Amplatzer ASO	4-38mm (size = stop flow diameter or 1 mm larger)	Nitinol mesh with polyester material	Yes	4; 5; 6; 7; 8; 9; 10; 11; 12; 13; 14; 15; 16; 17; 18; 19; 20; 22; 24; 26; 28; 30; 32; 34; 36; 38	12; 13; 14; 15; 16; 17; 18; 21; 22; 23; 24; 25; 26; 27; 28; 29; 30; 32; 34; 36; 38; 40; 42; 44; 46; 48	16; 17; 18; 19; 20; 21; 22; 25; 26; 27; 28; 29; 30; 31; 32; 33; 34; 36; 38; 40; 42; 44; 46; 50; 52; 54	6; 6; 6; 6; 6; 6; 6; 7; 7; 7; 7; 7; 7; 7; 8; 8; 9; 9; 9; 10; 10; 10; 12; 12; 12; 12	-	97.2%-98%	0.2%-1.1%	Device erosion (possible risk factors include deficient aortic rims, large defects and oversizing the device)
Amplatzer Cribriform	9-17.5 (size 2x total distance of defects)	Nitinol mesh with polyster material	No	18 (9); 25 (12.5); 30 (15); 35 (17.5)	18; 25; 30; 35	18; 25; 30; 35	8; 8; 8; 9	-	92%	0%	
Gore Cardioform Septal Occluder (GSO)	11-17 (size 1.75x stop flow balloon sizing)	Nitinol wire with ePTFE membrane	No	20 (11); 25 (14); 30 (17)	20; 25; 30	20; 25; 30	10; 10; 10	12; 12; 12	98.8%	0%	Wire frame fracture
Gore Cardioform ASD Occluder	8-35 (stop flow diameter)	Nitinol wire with ePTFE membrane	Yes	27 (8-15); 32 (13-20); 37 (18-25); 44 (23-30); 48 (28-35)	27; 32; 37; 44; 48	27; 32; 37; 44; 48	10; 10; 11; 12; 14	12; 12; 13; 14; 16	100%	1.6%	Wire frame fracture (oversizing the device is a possible risk factor)

Fig. 2 Available Amplatzer devices include (**A**) Amplatzer Septal Occluder, (**B**) Amplatzer Cribriform Multi-Fenestrated Septal Occluder. (Amplatzer is a trademark of Abbott or its related companies. Reproduced with permission of Abbott, © 2023. All rights reserved)
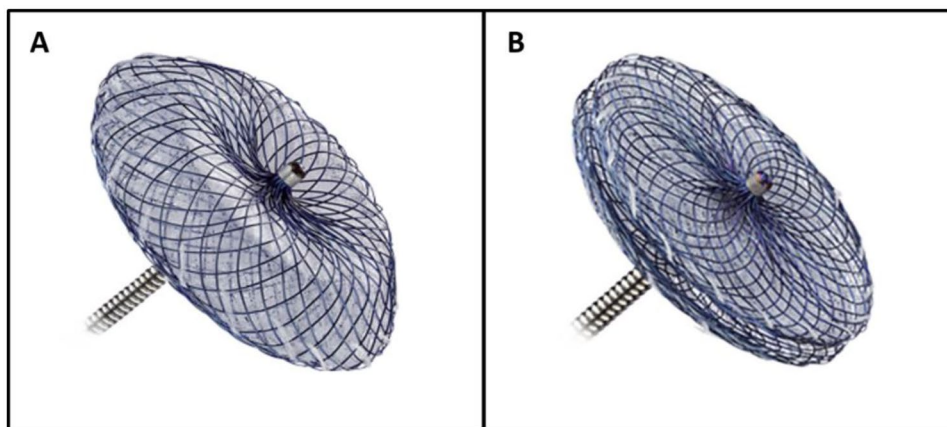
The Gore Cardioform ASD Occluder (GCA) (Fig. 3B), FDA approved in 2019, is the most recently approved device for transcatheter closure of secundum ASD and was designed to be a hybrid of previously approved devices. The GCA is composed of a nitinol wire frame covered with ePTFE, similar to the GSO, but with a self-centering waist. Unlike other devices, the left atrial disc and intra-disc occluder components deploy as a 3D cone that is pulled back into the defect before the right atrial disc is opened. It is available with five different disc diameters and can close defects up to 35 mm in diameter. The ASSSURED trial, a prospective, multicenter study including 22 US sites and 125 patients, demonstrated device safety and clinical efficacy [14•]. The cohort was primarily pediatric patients, with 72% of patients < 18 years old and a median age of 12.3 years. The median stop flow diameter was 17 mm, with nearly half of the defects measuring > 18 mm (43%) and 57% of defects having a deficient aortic rim. Importantly, nearly 1/3 of patients had both diameter > 18 mm and deficient aortic rim. 96% of patients had successful device placement, with 100% closure success at 6 and 12 months follow up (4% had clinically insignificant residual shunts and no patient had significant shunts). The most significant adverse events were device embolization (2 cases, 1.6% of patients) and new arrhythmias. New atrial tachyarrhythmias occurred in approximately 5% of patients, with some requiring medical treatment. Three patients required device removal within 24 h of implantation (two for device embolization and one for refractory supraventricular tachycardia). Wire frame fractures were an expected complication of the GCA given prior experience of the GSO, but occurred more frequently than anticipated, occurring in 36% of cases. Although there were no clinical sequela at 6-month follow-up, ongoing surveillance of wire frame fractures is required to assure there are no long-term sequelae of this issue such as perforation, device impingement on other structures, device instability, or thrombosis. Overall, the GCA was able to successfully treat a broad range of defects including patients with both deficient aortic rims and large defects. The biggest advantage of the Gore devices is that the long-term risk of device erosion, as seen with the ASO, has not been reported with either the GSO or GCA devices to this point [14•].

Fig. 3 Available Gore devices include (**A**) Gore Cardioform Septal Occluder, (**B**) Gore Cardioform ASD Occluder (© 2017 W. L. Gore & Associates, Inc. Used with permission)
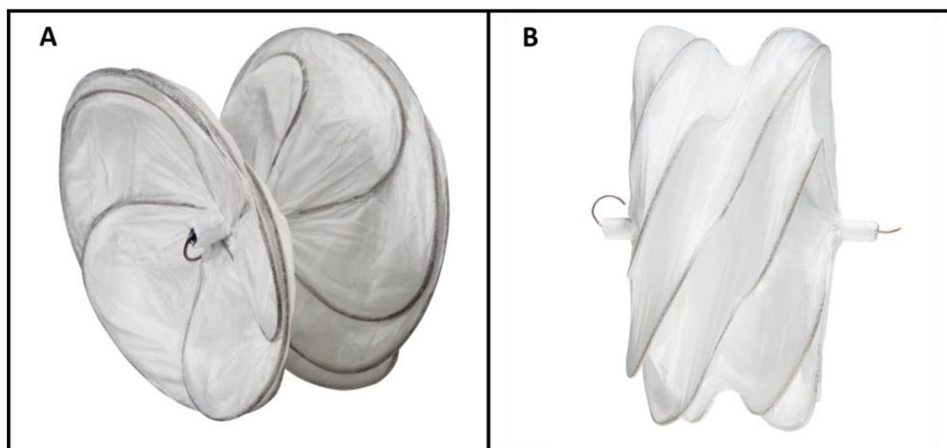
Approach to Complicated ASDs

Complex atrial septal defects that make transcatheter device closure more difficult include large defects, rim deficiencies, fenestrated defects, multiple defects, and the presence of pulmonary arterial hypertension.

Some patients with unrepaired ASDs, especially older patients, may also have pulmonary arterial hypertension. Leaving large defects unrepaired can lead to progression of pulmonary vascular disease and ultimately Eisenmenger syndrome, yet complete closure of an ASD in patients with severe PAH carries significant risk and is contraindicated [2]. Partial closure of ASDs in patients with moderate to severe PAH has been described by performing fenestrated device closure using Amplatzer devices [15]. The authors manually created fenestrations about 1/4 the diameter of the device by manually removing part of the patch in ASO devices and in some cases added an additional fenestrated of 4–5 mm to ensure adequate flow. They placed fenestrated devices in 51 patients without any periprocedural pulmonary hypertensive crisis. Overall, the strategy of combining fenestrated device closure with post-intervention pulmonary vasodilator therapy was safe and effective with many patients having an improvement in New York Heart Association functional class and pulmonary artery pressures at follow-up. For patients with persistent severe PAH, ballooning or stenting of the fenestration can be performed [15]. Although only currently available in the USA under compassionate use, the Occlutech Atrial Flow Regulator negates the need for post-market modification of devices and may reduce risk of spontaneous closure. Deployed similarly to other ASD devices, the Occlutech Atrial Flow Regulator is a Nitinol-framed device with 4, 6, 8, and 10 mm fenestrations located centrally with discs ranging from 16 to 30 mm and sheath sizes 10–12 French [16].

Using additional sheaths to manipulate the angle of deployment is one method to close large defects with complex anatomy or deficient rims. The delivery system of the Gore Cardioform ASD Occluder (GCA) may not allow for ideal alignment of the device with the atrial septum during closure of larger ASDs or ASDs with deficient retro-aortic rim. A long, curved Mullins sheath can be rotated clockwise in the IVC to assume a posterior position, allowing for improved angulation and better alignment of the GCA device to the atrial septum. The 12–16 Fr 64cm Mullins sheath along with the GCA delivery catheter has been successfully used to facilitate GCA device closure, especially in larger defects > 32 mm, and may be considered a first-line method for device deployment in large ASDs [17]. Additional long sheath combinations have been described for assisted deployment of the GSO and ASO devices – including the Hausdorf sheath (GSO), the St. Jude SL2 transseptal sheath (ASO) and steerable sheaths

(ASO). Although not a long sheath, an additional technique for improving septal alignment during device deployment is the suture-assisted technique [18]. A long Prolene, Vicryl, Polypropylene or silk suture can be looped through the guide wire loop in the GCA delivery sheath and tension placed on the suture during deployment to flex the sheath and create an ideal delivery angle.

Although specific to the ASO device, numerous implantation techniques have been described to aid successful closure – especially in the case of large defect or defects with rim deficiency. One such technique is deployment from the right upper pulmonary vein. This method describes placing the delivery sheath in the right upper pulmonary vein combined with a rapid ASO device deployment as the sheath is withdrawn from the pulmonary vein. The technique was found to improve alignment of the left atrial disk along the plane of the atrial septum and improve successful device closure rates when deployment from the left upper pulmonary vein was unsuccessful [19]. Balloon-assisted techniques can also facilitate closure of large ASDs. Large ASDs often have insufficient rims, which do not allow the left atrial disc to anchor, and the floppiness of the atrial septum can also be an issue. In this technique described by Dalvi et al., a sizing balloon was inflated in the right atrium and pushed against the atrial septum. The left atrial disk was pulled back from the ostium of the right upper pulmonary vein toward the septum with the balloon supporting the septum and left atrial disk. The waist and right atrial disk were then released with the balloon still in position against the septum. The balloon is then removed and then the waist and right atrial disk are pushed toward the left disk to flatten the device. This was successfully completed with the ASO device in 14 patients without complications [20]. Numerous other techniques exist for the ASO including the "Greek maneuver", "tulip-bud deployment", and dilator/catheter assist. The "Greek maneuver" involves full uncovering of the left atrial disc and 2/3 uncovering of the right atrial disc to align the left atrial disc with the septum prior to complete deployment [21]. In "tulip-bud deployment", the left atrial disc is partially uncovered to form a "tulip-bud" in alignment with the atrial septum followed by a rapid uncovering and deployment of the device [22]. Finally, in the dilator assist technique, with access in the contralateral femoral vein, a long sheath is advanced over a wire in the left upper pulmonary vein and remains in place through atrial disc deployment to stabilize the device.

Newer delivery systems have also been developed to aid in closure of ASDs with complex septal anatomy and deficient rims. The Amplatzer Trevisio intravascular delivery system has a highly flexible delivery wire allowing for optimized alignment with the atrial septum, reduced-tension deployment, and improved deployment control. A case-series described successful transcatheter closure using

the Trevisio system in 9 pediatric patients with anatomically complex ASDs with no complications. Six patients had absent aortic rims including one with an aneurysmal septum, two patients had complex residual atrial septal defects in setting of complex congenital heart disease, and one had interrupted IVC requiring access from the internal jugular vein for device deployment [23].

Future Directions

Future innovations in catheter-based atrial septal closures include device closure of sinus venosus and coronary sinus defects, as well as the use of biodegradable devices. Superior sinus venosus defects are located at the junction of the right atrium (RA) and superior vena cava (SVC) and characterized by a deficient wall between the SVC and right upper pulmonary vein usually resulting in partial anomalous pulmonary venous return [2]. While there are no FDA-approved devices for this indication, successful transcatheter closure of these defects has been performed using covered stents deployed in the SCV-RA junction to close the ASD and redirect right pulmonary vein blood flow into the left atrium. Using advanced cross-sectional imaging and 3D models as part of pre-procedural planning, one institution has been able to perform this procedure successfully in 25 patients [24]. While described in younger patients, the majority of implants have tended to be in older adults. With further experience and longer follow-up studies, it may become a more widely accepted alternative to surgical closure at other institutions, but long-term sequelae including arrhythmia burden are not yet clear. Coronary sinus defects are rare defects involving a connection between the coronary sinus and left atrium due to an absent or deficient coronary sinus septum. Multiple case reports have described successful transcatheter closure of coronary sinus ASDs using ASO devices in adults and a covered stent in an infant [25, 26]. While sinus venosus and coronary sinus defects are less common than secundum defects, they represent examples of lesions previously managed exclusively by surgical correction.

The long-term presence of metal devices can lead to some rare long-term sequela - erosion, arrhythmias, thrombosis - and can be problematic if patients need future procedures that involve crossing the atrial septum. Innovative bioresorbable devices are being developed, which may provide an alternative in preventing such issues. One such example, the Carag bioresorbable septal occluder, has no metal framework and is composed of poly lactic-co-glycolic acid. After endothelialization, the device begins resorbing at 6 months and completely resorbs by 2 years. It is currently being evaluated in clinical trials in Europe and may provide a promising future for transcatheter device closure.

Novel approaches to the management of lesions previously managed by surgery alone and bioresorbable devices represent innovations that continue to expand the options for catheter-based interventions.

Ventricular Septal Defects

History and Rise of Transcatheter VSD Closure

Ventricular septal defects (VSDs) account for up to 40% of all congenital heart defects [27]. Prevalence in newborns is as high as 5% with the use color flow Doppler echocardiogram; however, reports of prevalence vary given that many small VSDs may close shortly after birth with another source reporting a prevalence as low as 4 per 1000 patients [28, 29]. VSDs can occur in isolation or in association with other types of congenital heart disease and are described based on their location in the ventricular septum, with the most common terminology being perimembranous, muscular, inlet, and outlet septal defects [30]. Large VSDs often lead to pulmonary overcirculation in infants with symptoms of tachypnea and failure to thrive and ultimately lead to pulmonary vascular disease and Eisenmenger syndrome if not closed. Small or moderate sized, pressure restrictive VSDs may not cause symptoms during childhood, but can lead to long-term sequela including left heart dilation, pulmonary hypertension, arrhythmias, aortic insufficiency, double-chambered right ventricle, or endocarditis. VSD closure should be performed in children or adults when there is evidence of a hemodynamically significant shunt. Indications for closure include left heart dilation, failure to thrive, Qp:Qs > 1.5:1, history of endocarditis, and aortic valve insufficiency [31, 32]. Although surgery continues to be a safe and effective means of management for all types of VSDs, over the past few decades, transcatheter device closure has become an accepted and less invasive alternative in many muscular and some perimembranous VSDs.

The first transcatheter device closure of a ventricular septal defect was performed in 1987. Six patients ranging from infants to older adults with either perimembranous or muscular VSDs who were not surgical candidates underwent device closure with a Rashkind double umbrella device [33]. Other early reports emerged describing successful transcatheter closure for selected patients with membranous or muscular VSDs using the Rashkind device [34, 35]. By the early 2000s, Amplatzer devices became available for both membranous and muscular VSD device closure.

Transcatheter device closure for membranous VSDs has posed significant challenges due to the close proximity of the defect to the aortic and tricuspid valves and risk of conduction disturbances. Initial studies of the Amplatzer Membranous VSD Occluder were promising, showing technical success and encouraging closure rates. The device was designed as a self-expandable double disc device composed of nitinol

wire mesh. US phase 1 trial results demonstrated successful closure rates of 91% initially and 96% at 6 months with rare adverse outcomes [36]. However, follow-up studies demonstrated rates of complete heart block that were unacceptably high, up to 22%, occurring even months after device placement due to impingement on the conduction system [37, 38]. The Amplatzer Membranous VSD Occluder is not approved for clinical use in the USA due to this complication.

More recently, further development of this technology and implantation techniques have improved success rates and safety for transcatheter VSD device closure, especially in muscular defects that can be difficult to locate and close surgically. Additionally, softer devices are being used off-label for closure of membranous VSDs with lower reported rates of heart block.

Current FDA-Approved Devices

Muscular VSDs frequently decrease in size or close spontaneously over time and do not require intervention. However, larger or multiple muscular defects can be hemodynamically significant and necessitate closure. Muscular defects can be challenging to repair with surgery, due to poor visualization of the defect through the tricuspid or semilunar valves or location at the apex. Surgical management sometimes requires a ventriculotomy or temporary palliation with PA banding in infancy, or closure can be achieved with a transcatheter or hybrid approach [39]. Indications for transcatheter muscular VSD closure includes patients with hemodynamically significant muscular VSDs that weigh > 5 kg, while contraindications include pulmonary vascular resistance > 7 Wood U/m2, contraindications to anti-platelet therapy, or anatomic concerns such as inadequate rims (< 4 mm between the defect and valves) [40].

The Amplatzer Muscular VSD Occluder (Abbott), approved in 2007, is currently the only FDA-approved device with an indication for VSD closure in the US (Fig. 4). It is a self-expandable device made of a nitinol mesh frame with two flat disks linked by a central connecting waist and is available from 4 to 18 mm in 2-mm increments. Each disk also has Dacron patch material within the Nitinol scaffold to enhance thrombosis and occlusion [41]. Multiple clinical trials and reports have demonstrated the safety and efficacy of this device for closure of muscular VSDs [41–44]. The IDE (investigational device exemption) trial was a prospective, multi-center study enrolling 75 patients and demonstrating successful device implantation in > 85% of patients. Inclusion criteria were the presence of a muscular VSD that was either hemodynamically significant or with a history of resolved endocarditis. There was complete closure in 47% of cases immediately following the case, which improved to 92.3% at 1-year follow-up. The median VSD size was 7 mm, ranging from 3 to 16 mm, and successful closure was achieved in anterior, apical, mid-muscular, and posterior VSDs. There were major complications in 10.7% of cases and two deaths resulting from the procedure. Major complications included device embolization (2.7%), hypotension and/or cardiac arrest requiring inotropes (12%), and conduction abnormalities (20%), though most were temporary with only two cases of persistent right bundle branch [41]. Other known device-related complications, including vascular injury, endocarditis, significant residual shunts, and hemolysis, were rare [41–44]. Overall, smaller patients (< 5 kg) have an increased risk of procedural-related complications and residual shunts, and device closure should be performed in these patients carefully and with clear indications [41].

Both antegrade and retrograde approaches can be used for transcatheter device closure. The antegrade approach involves crossing the defect from the left ventricle, snaring the wire in the superior vena cava or pulmonary artery, and externalizing it through the venous access site, creating an arteriovenous rail for subsequent antegrade device delivery (Fig. 5). A retrograde approach involves advancing the delivery system through the defect from the left ventricular side and has been increasingly used to close smaller defects off-label with the use of newer devices that require lower profile

Fig. 4 Example of Amplatzer devices commonly used for ventricular septal defect closure include **A** Amplatzer Muscular Ventricular Septal Defect Occluder, on-label FDA approved use, **B** Amplatzer Ductal Occluder – II (ADO-II), off-label use. (Amplatzer is a trademark of Abbott or its related companies. Reproduced with permission of Abbott, © 2023. All rights reserved)
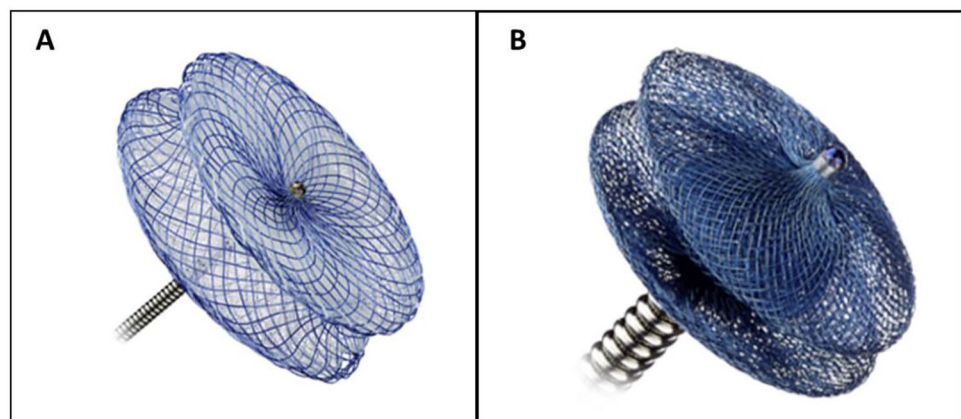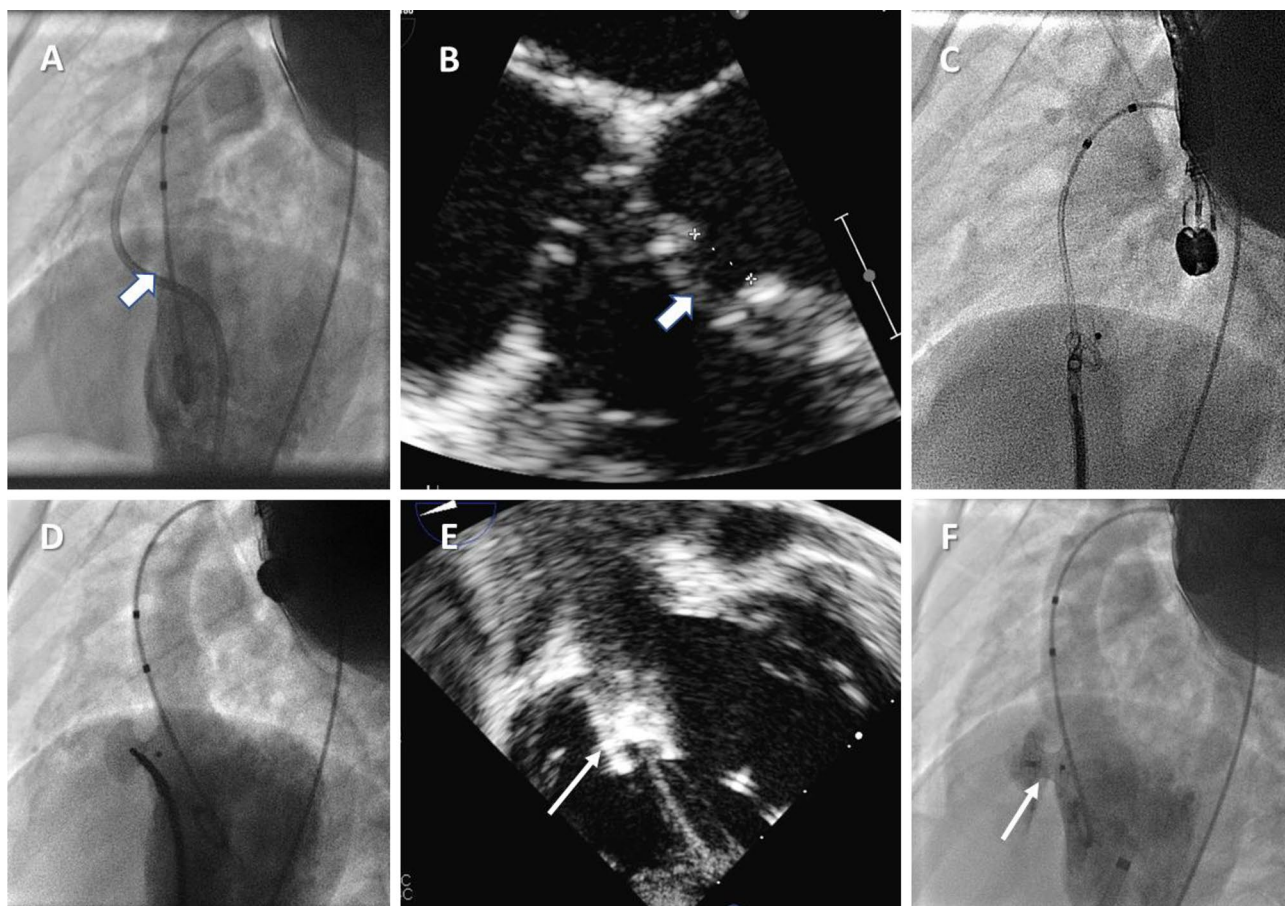
Fig. 5 Percutaneous transcatheter muscular VSD closure: The muscular VSD (*thick arrow*) is assessed and measured by left ventriculogram (**A**) and trans-esophageal echocardiogram (TEE) (**B**). The defect is crossed retrograde with a wire from the left ventricle and snared in the pulmonary artery to create an arterio-venous loop with the wire from femoral artery to femoral vein. A 45 degree Abbott Amplatzer Torque Vue sheath is advanced from femoral vein into left ventricle and a 6mm Abbott Amplatzer Muscular VSD Occluder Device is deployed across the VSD and released from the delivery cable once positioning is confirmed by angiography and TEE (**C–D**). After release, the device is in stable position by TEE and angiography (*thin arrow*) with trivial shunt through but not around the device (**E–F**)

delivery systems. It allows for safe closure of the defect from the arterial side and removes the need for an arteriovenous rail, which can lead to hemodynamic compromise in smaller infants [45].

Hybrid Approach

Transcatheter closure for small infants can be limited due to the inability to manipulate large sheaths in a small patient, and surgical repair can be difficult due to poor visualization. A hybrid perventricular approach allows for device closure of larger defects in smaller infants through a full or partial sternotomy. The hybrid procedure can be performed in the operating room under TEE guidance without cardiopulmonary bypass and without the need for fluoroscopy in most cases. Perventricular puncture of the RV free wall allows for more direct access to the defect and allows for

shorter sheaths and larger devices to be used. A wire can be advanced directly across the defect into the left ventricle for device deployment under echocardiographic or fluoroscopic guidance. Hybrid approaches can also be combined with other surgical procedures such as PA band removal. This hybrid perventricular approach to close muscular VSDs has been associated with high success rates (90–97%) and few adverse events and allows for the avoidance of cardiopulmonary bypass and a ventriculotomy incision [46–48].

Other Devices and Future Directions

Other devices have been used off-label to close perimembranous and muscular VSDs. The Amplatzer Ductal Occluder (ADO) I is an asymmetrical device that has primarily been used to close membranous VSDs associated with an aneurysm. Its design allows for the device retention disc to be implanted

entirely within the aneurysm away from the conduction system of the ventricular septum, thereby minimizing the risk of complete heart block. Case series report successful closure rates > 90% and only two cases of temporary heart block, with no patients requiring permanent pacemakers [49–51]. It is usually delivered from an antegrade approach. The ADO II device is a symmetrical, double-disc device with a low-profile delivery sheath that allows for an antegrade or retrograde approach. It has been used successfully to close both perimembranous and muscular VSDs, with successful closure rates > 85% and rare cases of complete heart block requiring surgical device removal [51–53, 54•] (Fig. 4).

Outside of the USA, other devices are available for muscular and membranous VSD device closure. The PFM Nit-Occlud VSD coil device consists of a nitinol coil with Dacron fibers and an asymmetric design with larger left-sided cone and smaller right-sided cone. It was designed for membranous and muscular VSD closure and was approved by the European Union in 2010. In a multi-center European registry, it was implanted successfully in 92% of patients, with 97% closure rate at 1 year. There were rare transient conduction abnormalities, but no long-term complete heart block [55]. Long-term efficacy and safety of the device for perimembranous VSDs with associated aneurysm tissue have been demonstrated, with a success rate of up to 96.6%. Adverse events included rare cases of device embolization, endocarditis, tricuspid valve regurgitation, residual shunts, hemolysis, and self-limited conduction abnormalities with no long-term complete heart block [56]. Overall, this flexible coil device appears to have fewer rhythm complications than previous membranous VSD closure devices, but does have an initial higher rate of residual shunts and hemolysis that usually resolves on its own. Different versions of the Cera VSD Occluders (Lifetech, China) were able to close membranous, muscular, and post-infarction VSDs. These devices have a self-expandable nitinol frame and are available in both symmetrical and asymmetrical designs. A prospective, multi-center study for device closure of membranous VSDs demonstrated successful implantation in 91% of cases, with 2% having trivial and 7% having moderate residual shunts at subsequent follow-up. There was one case of complete heart block requiring a pacemaker [57]. Other available devices include Occlutech membranous and muscular VSD occluders, which are soft, self-expanding nitinol-based devices; however, there is limited literature available describing their efficacy and safety.

Conclusions

Transcatheter device closure of septal defects has been shown to be a safe and effective treatment option using devices available and approved within the USA. Continued development and improvement of devices will help to expand the safe use of transcatheter device closure for not only secundum ASDs and muscular VSDs, but also septal defects in other locations.

Compliance with Ethical Standards

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