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Ventricular septal defect (VSD) is present in 20% [1] of all patients with congenital heart disease. It was first described in 1879 by Roger and until now a small VSD is referred to as "maladie de Roger" [2].

**Anatomical Consideration:**
The anatomic location of the defect is important to define as it plays an important role in assessing feasibility for device closure of the defect. There are numerous classifications describing the VSD based on its anatomic location within the septum. The Kirklin classification describes the location of the VSD in relation to the ventricular septum when the defect is viewed from the right ventricle. A Type I Kirklin defect is the subarterial defect, which is located at the superior-anterior part of the ventricular septum. This defect is more frequently seen in the East Asian population and in about 5% of all the defects in USA. A Type II Kirklin defect is the perimembranous VSD and is located in the middle of the upper portion of the ventricular septum in close proximity to the aortic valve. It is present in 75% of the patients with a VSD and has a high incidence of spontaneous closure. The Type III Kirklin defect is present in the posterior portion of the ventricular septum in close proximity to the tricuspid valve giving it an inlet extension. This defect is present in 5% of the patients and is termed as the perimembranous VSD with inlet extension. The Type IV Kirklin defect is present in the muscular portion of the septum. It is present in 10-15% of the patients and is most commonly present at the apical portion of the ventricular septum. The muscular defects can be single or multiple giving rise to the Swiss Cheese VSD.

Ventricular septal defect is a congenital defect but can rarely be iatrogenic. The iatrogenic defects are seen in patients following traumatic injury to the chest and in 0.2% of the patients following ventricular rupture in myocardial infarction.

**Physiology:**
The clinical picture of these patients is dependent on the volume of left to right shunting. This in turn depends on the size of the defect and the ratio of the pulmonary vascular resistance to the systemic resistance. At birth the pulmonary vascular resistance is elevated and the amount of left to right shunting is restricted. As the pulmonary vascular resistance falls the left to right shunting across the VSD increases resulting in increased pulmonary venous return to the left atrium and left ventricle resulting in congestive heart failure. Patients with a large muscular or perimembranous VSD will present with signs and symptoms of congestive heart failure and failure to thrive.

Large VSDs are unlikely to close spontaneously as opposed to small to moderate sized VSDs which in the large majority of patients may undergo spontaneous closure. Patients with a moderate to large VSD are at risk for pulmonary artery hypertension, left ventricular volume overload, arrhythmias, double chambered right ventricle and aortic regurgitation. Therefore, in symptomatic patients or in those with left ventricular volume overload closure of the VSD should be recommended. A VSD untreated can eventually result in pulmonary artery hypertension resulting in cyanosis and this pathology was first described by

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Eisenmenger in 1898 [3]. All VSDs can be repaired surgically with minimal morbidity and mortality with the exception of the apical or Swiss cheese defects [4-6]. In patients with the apical muscular VSD and the Swiss cheese defect the morbidity is higher and transcatheter closure is a feasible alternative.

In 1952 Muller [7] described pulmonary artery band placement as a means to restrict the left to right shunt at the level of the VSD. This was followed by Lillehei [8] in 1955 who first described the successful surgical closure of the VSD.

In 1987 Lock [9] pioneered the technique for Rashkind device closure of the VSD and thereby started the era for transcatheter closure of this defect. Since then there have been numerous reports for transcatheter closure of the ventricular septal defect with numerous devices including the Rashkind device, the clamshell device, the buttoned device, the Starflex, the Amplatzer muscular VSD device, the Amplatzer Membranous device and the Nit Occlud device. Transcatheter closure of VSD can be safely done in patients with the muscular VSD and in selected patients with a perimembranous VSD where at least a 2 mm aortic rim is present.

MUSCULAR VSD:
Surgical closure of the muscular septal defects [4-6] of the Swiss cheese type and those that are at the apical portion of the ventricular septum are associated with significant morbidity and mortality. Surgical closure of the apical muscular VSD is through a right ventricular incision and this results in a ventricular scar which can have long term sequela. In this subgroup of patients the transcatheter approach is the preferred approach with a comparable success rate and decreased morbidity and mortality.

Prior to device closure these patients will have a Chest X-Ray, ECG and a transthoracic echocardiogram. The echocardiogram will delineate the position, size and the rims of the VSD and the volume overload status of the left atrium and ventricle. In addition the ventricular septum is also assessed for the presence of additional VSDs. The long axis view will demonstrate the anterior ventricular septal defects. In the short axis view at the level of the mitral valve the anterior defects are present between 12 and 1 O’clock position, mid muscular defects are present between 9 and 12 O’clock position and the inlet defects are present between 7 and 9 O’clock position. The apical four-chamber view at the level of the atrioventricular valves will demonstrate the apical, mid muscular and inlet defects. If the transducer is angled more anteriorly the subaortic and the anterior VSDs are defined. A good transthoracic echocardiogram is essential for appropriate patient selection for transcatheter closure of the defect.

Devices:
Currently there are two devices available for closure of the muscular VSD. The Cardioseal device (NMT, Boston, MA) is FDA approved for patients who are a high surgical risk. This device was originally designed for closure of the atrial septal defect. The second device is the Amplatzer Muscular VSD occluder (AGA Medical, Golden Valley, MN). Clinical trials with this device have been completed and the device is awaiting United States FDA approval. The device (Fig 1) is made of 0.004”-0.005” Nitinol wire. It has a right and left retention discs with a 7 mm connecting waist. The retention discs are 8 mm wider than the waist. There is a screw on the right-sided disc to which the delivery cable is connected. The devices are available from 4-16 mm diameter (diameter of the waist) in 2 mm increments. The device can be delivered through a 6-9 Fr delivery sheath. The delivery system is similar to the delivery system of the Amplatzer Septal Occluder for ASD closure and consists of a sheath, dilator, loader, delivery cable and a pin vise. The delivery cable is placed through the loader and screwed to the device. The device is then pulled back into the loader under saline and the loader is flushed with saline. Amin [10] and his colleagues were the first to report the use of this device.

![Cardioseal device](image1.png) ![Amplatzer Muscular VSD occluder](image2.png)
Percutaneous Closure Protocol:
The procedure (Fig 2) is done under general anesthesia and TEE guidance (Figure II). TEE is done to assess the number of defects, the location of the defect and its relationship to adjoining structures such as the chordae, papillary muscles and the AV valves. In all patients right internal jugular, femoral venous and arterial access is obtained. The right internal jugular access is especially helpful in patients with a mid, posterior and apical VSD. The anterior muscular defects are closed through the femoral vein. The patient is heparinized for the procedure and the activated clotting time (ACT) is kept above 200 seconds throughout the procedure. A routine right and left heart catheterization is done. The steps for muscular VSD closure are illustrated in Figure III. A left ventricular angiogram is performed in the hepatoclavicular view (35°LAO/35°Cr) to delineate the location, size and number of ventricular septal defects. A Judkins right coronary artery catheter or a cobra catheter are utilized to cross the VSD from the left ventricular side.

Figure 1: The Amplatzer muscular VSD device shown in different profiles.

Figure 2: TEE images of muscular ventricular septal defect closure with an Amplatzer muscular VSD device. A. Apical 4-chamber view of a muscular VSD; B. Color Doppler flow across the VSD; C. The muscular VSD in the short axis view; D. Color Doppler flow across the muscular VSD in the short axis view; E. Wire across the VSD; F. Sheath across the VSD; G. The left ventricular disc is deployed in the left ventricle; H. The right ventricular disc is deployed in the right ventricle; I. The device is released from the delivery cable; J. There is no residual shunt across the VSD by color Doppler. RA – Right atrium; RV – Right ventricle; LA – left atrium; LV – left ventricle.

Figure 3: Fluoroscopic and angiographic steps in the closure of a muscular VSD.
The VSD can be crossed from the right ventricle in patients who have a large mid muscular or an apical VSD. At all times care must be taken to ensure that the catheter is free of the trabeculae and the chordae. Once the VSD is crossed a 0.035 " J exchange length wire is positioned in the pulmonary artery. This wire is snared from the right internal jugular and exteriorized to form an arteriovenous loop. An appropriate sized delivery sheath is then advanced from the right internal jugular vein over the wire to the apex of the left ventricle and the exchange wire is removed. A device 1-2 mm larger than the VSD diameter at end-diastole is selected and advanced through the sheath. The left sided retention disc is delivered in the mid left ventricle cavity and the entire assembly (sheath- device, cable) is pulled back to the septum. The waist is then delivered by retracting the sheath over the cable. Further retraction of the sheath over the cable will deploy the right ventricular disc. If the VSD is single, angiography in the left ventricle is sufficient to delineate the position of the device and monitor the steps. However, if multiple VSDs are present, both angiography and TEE should be used to verify the device position. Once correct device position is confirmed, the device is released from the delivery cable using the pin vise. The patient is given cefazolin during the procedure and SBE prophylaxis is recommended for 6 months following complete closure of the defect.

PeriVentricular Closure Protocol:
The periVentricular approach is the preferred approach in small infants and in those patients with complex congenital heart disease undergoing surgery for associated cardiac defects. The procedure can be done either in the operating room (patients with complex cardiac defects) or in the catheterization laboratory (patients with single or two VSDs) under TEE guidance. After the chest and the pericardium are opened the free wall of the right ventricle is punctured in the direction of the VSD away from the moderator band with an 18- gauge needle. A 0.035" short wire (Terumo, angled tip) is positioned in the left ventricle and an appropriate sized sheath is advanced to the left ventricle over this wire. A device 1-2 mm larger than the size of the VSD is delivered under echocardiographic guidance as described above. The advantage of the periVentricular approach is that the patient may not need to be placed on cardiopulmonary bypass. On occasions, after closure of the first VSD as described above, the second VSD may not be easily crossable from the right ventricle free wall. In such cases, percutaneous approach under fluoroscopic guidance is done. The wire crosses the VSD from the left ventricle side and the wire is advanced to the pulmonary artery. At that point, snaring can be done via a sheath placed from the right ventricle free wall to the main pulmonary artery. Once the wire is snared, a wire loop is formed where the wire entered from the femoral artery, left ventricle, VSD and out the right ventricle free wall. Then over the wire, a short delivery sheath is advanced into the mid left ventricle cavity. The remainder of the steps are similar to the conventional periVentricular approach.

PERIMEMBRANOUS VENTRICULAR SEPTAL DEFECT

Closure of the perimembranous VSD was first reported using the button device and the Rashkind device that were not originally designed for the perimembranous VSD. In addition trials are underway using the Amplatzer Membranous VSD device(AGA Medical Corporation, Golden Valley, MN) which has been specifically designed for the perimembranous ventricular septal defect.

In all patients transthoracic echocardiography is done to delineate the defect. The parasternal long axis view will demonstrate the defect. In this view tilting the probe towards the right ventricular outflow tract
will outline the supracristal defect. In the short axis view at the level of the aortic valve, the perimembranous defects are seen between 7 and 9 O’clock position, the membranous defects are seen between 9 and 12 O’clock position and the supracristal defects are seen between 12 and 1 O’clock position. In addition there needs to be a 2 mm rim on the aortic side for effective and safe closure. Device closure is contraindicated in the supracristal VSD (Type I Kirklin defect) and patients with an inlet extension (Type III Kirklin defect) of the VSD need to be carefully evaluated for feasibility of closure.

**Amplatzer Device and delivery system:**
The Amplatzer Membranous VSD occluder is a self expanding, self-centering and retrievable double disc device (Fig IV). It is made of 0.003”-0.005” Nitinol wire. The device has a 1.5 mm long waist and a right and left retention discs. The right-sided disc is symmetrical and is 2 mm larger than the diameter of the waist. The left sided disc is asymmetrical with the aortic end being 0.5 mm larger than the waist and the ventricular end being 5.5 mm larger than the waist.

The ventricular end of the left sided disc has a platinum marker. This marker is important to guide correct device deployment. There is a screw on the right-sided disc with a flat part. This flat part is important when attaching the device to the delivery cable and the pusher catheter. The device is available in sizes ranging from 4-18 mm in diameter (diameter of the waist) in 2 mm increments and can be delivered through a 6-9 Fr delivery sheath.

The delivery system consists of a delivery cable, sheath and dilator, pusher, loader and a pin vise. The pusher catheter has a metal capsule at its end. The capsule has a flat part that corresponds to the flat part in the screw of the device. These two flat parts should align with each other. The delivery cable is pushed through the pusher catheter and the device is loaded in the customary fashion. The cable is then pulled back and the flat part of screw is opposed to the flat part of the capsule. This is an important step to allow the left sided disc to be delivered in the correct orientation (the platinum marker towards the patient’s feet). The device and the pusher catheter as a single unit are pulled back into the loader catheter under saline solution.

![Figure 4: Amplatzer Membranous VSD device. Legends: A- Delivery sheath; B- Pusher cable; C- Metal capsule of pusher cable; D- Delivery cable; 1 to 5 is the right sided disc; 2 is the waist; 3 to 4 is the left sided disc with a platinum marker at 3.](image)

**Closure Protocol:**
The procedure can be done under conscious sedation and transthoracic echocardiographic (TTE) or intracardiac echocardiographic (ICE) guidance. However, many prefer to perform the procedure under general anaesthesia and TEE guidance. A TEE is done to assess the location, rims, and size of the VSD and to guide the placement of the device (Figure V). Access is obtained from the right femoral vein and artery. Once access is obtained the patient is heparinized to keep the ACT above 200 seconds. A routine right and left heart catheterization is performed to assess the hemodynamics and to calculate the Qp/Qs ratio. The steps for perimembranous VSD closure are illustrated in Figure VI. An angiogram is done in the 50-60° LAO/15-20° cranial projection to define the position of the VSD. With the help of a Judkins right coronary...
artery catheter the VSD is crossed using a 0.035” Terumo angled glide guide wire. The wire is advanced and it usually goes to the main pulmonary artery (in occasions it goes to the superior vena cava).

The catheter is then advanced over this wire to the position in the main pulmonary artery. The glide wire is removed and a 0.035” exchange length Noodle wire (AGA Medical Corporation, Golden Valley, MN) is advanced to the pulmonary artery where it is snared and exteriorized through the right femoral vein using a goose neck snare (ev3, Plymouth, MN). An appropriate sized delivery sheath is then advanced to the apex of the left ventricle. A device which is 1-2 mm larger than the size of the VSD is selected. The device is screwed to the delivery cable. It is important to align the flat part of the microscrew of the device with the flat part of the capsule located at the end of the pusher catheter.

**Figure 5:** TEE images of perimembranous VSD closure with an Amplatzer Membranous VSD device. **A-D.** 2-D and color images of a perimembranous VSD; **E.** Wire across the perimembranous VSD; **F.** Sheath through the perimembranous VSD; **G-I.** Steps of device deployment; **J-L.** The device is released with no residual shunt. RA – Right atrium; RV – Right ventricle; LA – left atrium; LV – left ventricle.

**Figure 6:** **A.** LV angiogram demonstrating the perimembranous VSD; **B-G.** Steps of device deployment; **H-I.** Release of the device with LV angiogram showing no residual shunt; **J.** Aortogram with no aortic insufficiency.
This will ensure correct orientation of the device with the platinum marker towards the patient’s feet. After that the pin vise is tightened to the cable at the end of hub of the pusher catheter to prevent premature dislodgement of the device. The device is then advanced through the sheath and the left ventricular disc is delivered midway between the anterior mitral valve leaflet and the left ventricular outflow tract to prevent entanglement of the device with the mitral valve. The entire system is pulled back to the septum and the platinum marker at this time should be pointing towards the patient’s feet for proper orientation of the device. The waist is then delivered followed by the right ventricular disc. Position of the device is confirmed by echocardiography (TEE, TTE or ICE) and by angiography in the left ventricle.

Once good position is confirmed and the aortic valve is interrogated for any impingement of the device on the leaflets, the pin vise is loosened and the pusher catheter is retracted over the cable to disengage the flat parts (capsule and microscrew of device) from each other. Then the device is released by counter clockwise rotation of the pin vise. A repeat angiogram in the left ventricle is done to assess position of the device and any residual shunt. An ascending aortogram can be done to assess the relationship of the device with the aortic valve. However, this can be best assessed by echocardiography. All patients receive cefazolin for the procedure and SBE prophylaxis is recommended for 6 months following complete closure of the VSD.

**DEVICES AND RESULTS:**

**Muscular VSD Closure with the Rashkind, Clamshell and StarFlex device**

Closure of the muscular VSD was first reported by Lock [9] using the Rashkind device. Due to the high rate of residual shunt and larger delivery system, the device was modified initially to the Clamshell device and later due to high rate of frame fracture, it was modified to the Cardioseal and the Starflex devices. Knauth and colleagues [11] reported their results on 170 patient where the VSD was closed with the Clamshell and StarFlex devices. The device was successfully placed in 168/170 patients and 40% of the patients received multiple devices. In 7.7% (13/168) the device had to be explanted because of malposition, embolization or the presence of a significant residual shunt. This device has been cumbersome to use and was not designed specifically for the ventricular septum.

**Closure of the VSD intraoperatively**

Fishberger and his colleagues [12] described the use of the Rashkind device intraoperatively in a select group of patients who were felt to be unfavorable candidates for percutaneous closure of the VSD. The results were less than satisfactory with only 5/9 patients surviving the procedure. Chaturvedi et al [13] reported the successful intraoperative closure of the VSD in 4 patients. One patient died post operatively from low output failure. All other patients had complete closure of the VSD by 5 months. Murzi and his colleagues [14] reported closure of the muscular ventricular septal defect in 5 patients. There was one post operative death in a patient with a moderate to large VSD from intractable right ventricular failure. In one patient who had closure of a perimembranous VSD with a prosthetic patch complete heart block occurred and the patient underwent a permanent pacemaker placement.

**Closure of the perimembranous defect with the Rashkind device**

Kalra and his colleagues [15] reported closure of the VSD in 30 patients (28 perimembranous) with the Rashkind device in 27/28 patients. In the patients undergoing device placement there was a trace residual shunt in 30% of the patients.

**AMPLATZER MUSCULAR VSD DEVICE**

Closure of the VSD by the Amplatzer muscular and perimembranous closure device was first reported by Amin [10] and his colleagues who created a surgical muscular VSD in 5 dogs and used 5 Yucatan pigs with naturally occurring perimembranous VSD. All animals had intraoperative closure of the VSDs. There was complete closure of all 5 muscular VSDs. In the perimembranous group 3/5 had complete closure at 3 mos and one animal had aortic insufficiency.
Hijazi [16] and his colleagues reported the transcatheter technique of muscular VSD closure in 8 patients using the Amplatzer VSD occluder. All 8 patients underwent successful closure of the VSD with immediate complete closure occurring in 2 patients and by 6 months all patients had complete closure of the ventricular septal defect. There were no major complications of the procedure.

Arora [17] and her colleagues reported the closure of the muscular VSD in 50 patients. In 48/50 patients closure was done using the Amplatzer Muscular VSD device. There were no residual shunts and transient complete heart block occurred in one patient. At a follow-up of 2-90 months there was no device embolization, infective endocarditis, hemolysis or late conduction defects.

The immediate and midterm results for the US registry [18] reported the results in 75 patients in whom 83 procedures were done by the percutaneous (70/75) or perventricular approach (6/75). In this group 44% of the patients had multiple ventricular septal defects and multiple devices were implanted in 21% of the procedures. The complete closure rate at 6 months was 92.3%. Major complications occurred in 11% of the patients and there were 2 procedure related death.

**IATROGENIC VSD**

Ventricular septal rupture occurs in 0.2% of the patients post myocardial infarction and remains associated with a high morbidity and mortality rates. Transcatheter closure [19-22] is the preferred approach in this hemodynamically unstable group of patients. In a paper by Holzer and his colleagues [19], there were 18 patients who underwent closure between 2000-2003. The procedure was done in the acute phase in 27% of the patients. The device was deployed in 16/18 patients. 11/16 patients were alive at a median of 332 days with 8 patients having a none to small residual shunt and 2 patients having a moderate shunt.

In another paper by Holzer and his colleagues [20], two patients with an iatrogenic VSD occurred after aortic valve replacement with a prosthetic aortic valve underwent successful closure using the membranous VSD occluder [20].

**PERIMEMBRANOUS VSD**

The AGA membranous device is specifically designed for closure of the membranous VSD as opposed to other devices which were designed to close other heart defects.

Hijazi [23] and his colleagues described the first 6 patients to undergo closure of the VSD located in the membranous portion of the ventricular septum. The patients ranged in weight from 15-45 Kg with a median weight of 29 Kg. All patients had immediate complete closure. The only complication was trace aortic regurgitation in one patient.

Thanopoulos [24] and his colleagues described the closure of the muscular and perimembranous VSD with excellent results. There were 13 patients with a perimembranous VSD who underwent closure. The only major complication was device embolization in one patient secondary to catheter manipulation. The device was successfully retrieved and the VSD was successfully closed with another Amplatzer perimembranous VSD device. There was an 84% complete closure rate immediately following the procedure and at 1 month the complete closure rate was 92.3%.

Bass and his colleagues [25] also reported on their experience using this device in 27 patients; twenty-five had successful attempt with 23/25 of the patients having complete closure within one week from the procedure. No major complications were reported.

Pawelec [26] and her colleagues described the closure of the perimembranous VSD using the Amplatzer perimembranous VSD device in 9 patients. At a mean follow-up of 11.5mos there was a significant decrease in the left ventricular end-diastolic diameter. There were no complications noted.

**COMPLICATIONS:**
The complications associated with device placement are infrequent. Device embolization and migration is
rare with the Amplatzer device since the device is repositionable and retrievable. Embolization can occur to the pulmonary artery or the systemic circulation. In the large majority of patients the device can be snared and removed. Arrhythmias are transient and complete heart block is rarely seen. Currently, the incidence of complete heart block after device closure of perimembranous VSD is around 2%. Long-term follow-up and careful evaluation for long-term arrhythmias and heart block is important. Air embolism is a potential complication and careful attention to detail is essential. Hemolysis is rare in patients with complete closure of the defect. We believe that pre soaking the device with the patients own blood may enhance complete closure and hence decrease in hemolysis. Valvular regurgitation is minimized by assessing the position of the VSD in relationship to adjoining structures by TEE or TTE prior to device release.

Closure of the VSD by the transcatheter route is the preferred approach in a select group of patients. Complications with the Amplatzer device are rare in the hands of a skilled operator. Long-term follow-up of device patients and comparison with contemporary surgical group is essential before routine use of devices is recommended.

Bibliography


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